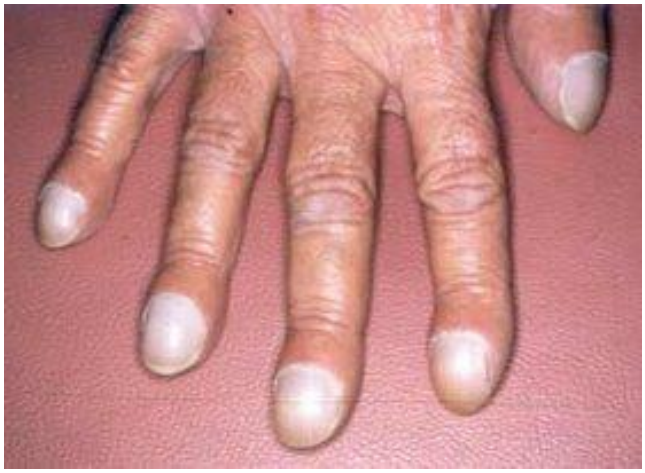
Secrets of Paces

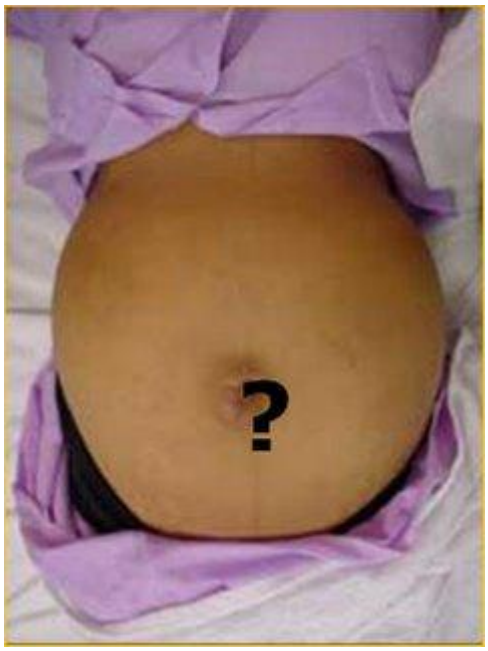
Index **Page**

Clubbing



Clubbing is a common sign in clinical practice. It gives clue to the underlying disease.When clubbing is present, please demontrate nail bed fluctuation, palpate wrist for tenderness seen in HPOA, look for nicotine staining of nail and central cyanosis.

I am sure everyone could give a good differential for clubbing.
CVS: cyanotic heart disease, IE
Lung: bronchiectasis, fibrosing alveolitis, lung cancer
Abdomen: chronic liver disease, IBD



In PACES, you will be asked the differential diagnosis for your patient. Some of the common combination of the signs would be:

1. Liver + Spleen
* CML
* Myelofibrosis
* Lymphoproliferative
* Cirrhosis + portal hypertension (CLD signs)
* Hereditary hemolytic anemia (Jaundice, pallor, hyperpigmented, short stature, young age)

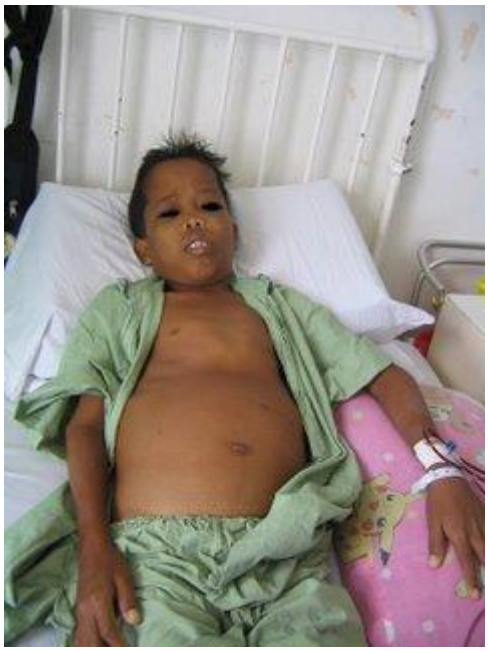
2. Liver + Spleen + LNs
* CLL
* Lymphoma
* CML in blastic crisis

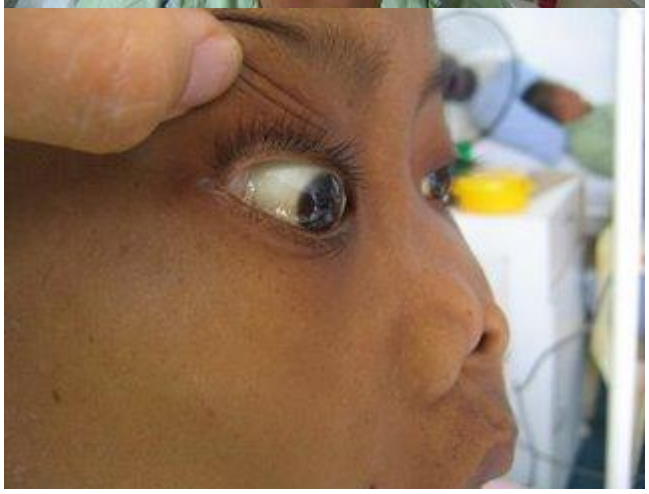
3. Spleen + LNs
* CLL/ lymphoma
* Infections eg Infectious mononucleosis
* Felty syndrome (RA hands!)

4. Isolated Spleen
* CML/ Myelofibrosis
* Lymphoma
* Polycythemia rubra vera (plethora)
* Chronic malaria (fever, jaundice)
* IE, typhoid
* Cirrhosis with portal hypertension (CLD signs)

5. Isolated Liver
* Cirrhosis (CLD)
* Mitotic lesion (primary or secondary)
* CCF (tender liver, edematous)
* Acute hepatitis (tender, fever, jaundice, no CLD)
* Liver abscess (tender, fever, jaundice, no CLD)

Thalassaemia





This 20 year old young man was referred from our paediatric colleagues for further

follow up and management. Please examine his abdominal system.

I would like to complete my examination by examining his genitalia, doing a per rectum examination.

This thin young man is pale, jaundiced, hyperpigmented and short for his age. He has frontal bossing, prominent maxillaries and chipmunk teeth which are maloccluded. There is also sparse axillary and pubic hair
The abdomen is distended and there is a left lower quadrant scar. There is hepatomegaly, extending 4 finger breadths beneath the costal margin. The liver has smooth edges and non tender on palpation. The kidneys are not ballotable. There is no shifting dullness to suggest the presence of ascites.

I did not find any lymph nodes in him. There were no features to suggest chronic liver failure. I also note he does not have any abdominal puncture wounds to suggest the use of desferral

In summary, this is short young man with jaundice, pallor, hepatomegaly, a previous splenectomy done and has thalaessaemic facies and possible underlying hypogonadism and haematochromatosis. He has chronic hemolytic aneamia. He is transfusion dependent.

I would like to offer the differential of a haemoglobinopathy. The most common haemoglobinopathy in this part of the world is Thalaessaemia.

Questions

1.What investigations would you like to do?

I would like to confirm the underlying cause of hemolytic anemia by doing serum electrophoresis. I would like assess the complications of treatment and patient's disease by doing LFT, coagulation profile, hormonal assay, hepatitis serology and echocardiogram. I will assess his functional status using FBC.

2.How would you like to manage this young man?

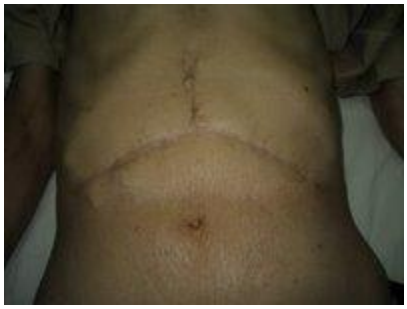
a. Treatment of Disease- hypertransfusion, Splenectomy, Pneumococal vaccination, bone marrow transplant in some centres
b. Treatment of its complications-hormone replacement therapy, iron chelation

therapy, control heart failure

c. Patient and family education, genetic counseling is also important so that family members can be screened and treated early.

> **Scars in Thalassemia** may be due to previous cholecystectomy or splenectomy.
> **Laparascopic scars** may give you a small scars (Look for it carefully!!)

A Tale of Scar: Mercedes Benz scar

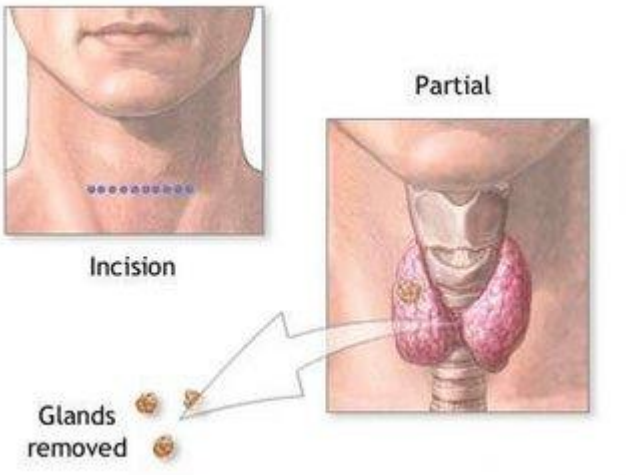


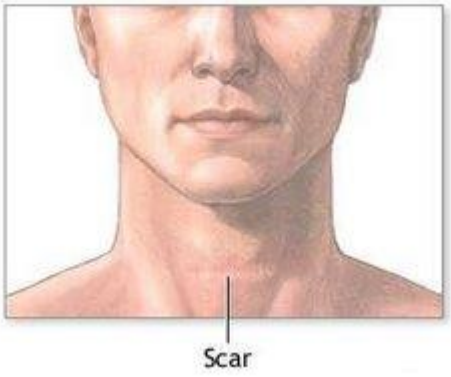
My friend got this in his exam recently in London.

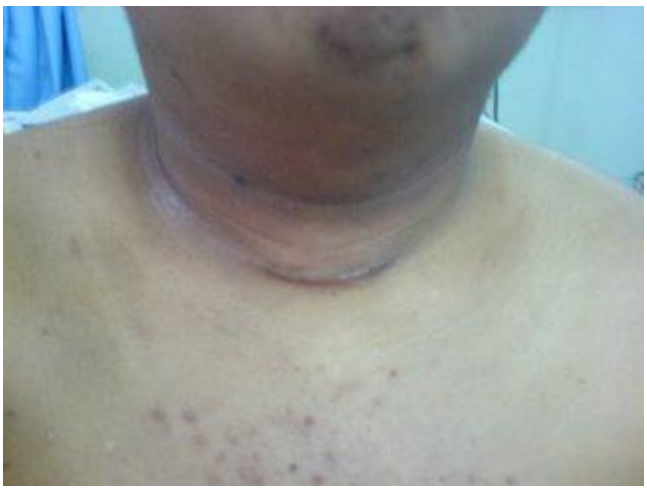
He was asked : what are the likely cause of this scar ?

Liver transplantation as evidenced by presence of mercedes benz scar. This is useful in carrying out total gastrectomy, operations for renovascular hypertension, total oesophagectomy, liver transplantation, extensive hepatic resections, and bilateral adrenalectomy.

A Neck Scar In Abdomen Station







Parathyroidectomy scar is smaller than thyroidectomy scar.

It is located higher than post-tracheostomy scar (sternal notch).

"Abdomen station: Examiner introduced a middle aged thin lady having tiredness and SOB."

I introduced myself, positioned and stood at the foot side. She had sallow complexion, a midline / right iliac fossa and right lumber scar on the abdomen. Examination of hands was unremarkable. On left arm there was a AV fistula with palpable vibrations/hum. Above this was a small scar. She had pale conjunctiva. A transverse about 3cm scar on the thyroid. On abdominal palpation there was palpable and ballotable mass 15cm x 6cm with irregular surface in left lumber region. No other masses were palpable in the abdomen and there was no venous hum on auscultation. No sacral or pedal oedema and there were no lymph nodes anywhere. I expected a transplant in LIF scar but there was none.

Examiner asked what is the diagnosis after I presented the case. I said 'These findings are suggestive of APKD and patient has right nephrectomy. She has ESRF and is on renal replacement therapy'

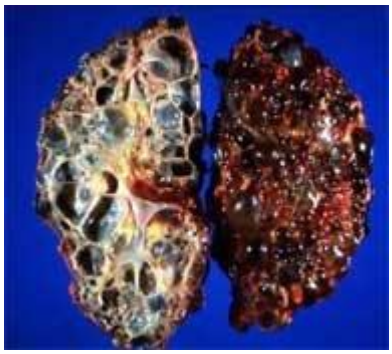
Only then, examiner reminded me of thyroid scar and asked is it related to her present condition. I was stressed since the scar was much smaller than usual thyroidectomy scar. Then I suddenly realised that this patient had 'parathyroidectomy' and a small scar on the left arm was due to one of the parathyroid transplant in the arm.

Take home messages:

1. Neck scar in abdomen station = suspected parathyroidectomy scar

2. Be prepared to recognise the reasons behind the scars in abdomen station

Renal enlargement (PCKD)



Patient with ***bilateral*** renal enlargement are commonly shown in PACES. The causes would be:
1. Polycystic kidney disease (very common and almost 100% in PACES)
2. Bilateral hydronephrosis (rare in PACES)
3. Amyloidosis (rare)

Having diagnosed a mass as a kidney, you must be able to support this with your physical findings:
1. Ballotable (ask patient to *inspirate* and *hold* the breath)
2. Resonant to percussion
3. Examining hand can get between swelling and costal margin
4. Slight movement downwards on inspiration

Unilateral renal enlargement is less common in PACES.
1. Nephrectomy in polycystic kidney (look for scar!)
2. Renal cysts
3. Carcinoma
4. Hydronephrosis
5. Hypertrophy of a single functioning kidney

**Remember, should you encounter a kidney case in PACES, 50% of them are polycystic kidney and the other 50% would be renal transplant!
**Dont mistake hepatosplenomegaly as bilateral kidney mass and vice versa.

Renal transplant

Iliac fossa scar with a mass equivalent to transplanted kidney

1. **Causes**
   * Most of the cases shown are polycystic kidney disease
   * Other causes include DM (check for dermopathy and diabetic ulcers), GN and etc

   2. Complications
   a. Side effects of medications
   ** Azathioprine - myelosuppression with bruises, pallor, jaundice
   ** Cyclosporin - hand tremor, hypertrichosis, gum hyperplasia, high BP
   ** Prednisolone - Cushing's syndrome, pallor secondary to UGIB
   b. Hepatitis B/C with CLD signs

   3. Functional capacity
   * Signs of graft rejection - tender transplanted kidney
   * Signs of infections - they have 1001 reasons to get infections
   * Uremic/hepatic encephalopathy - unlikely to appear in PACES!

Renal transplant

Mr XYZ has ESRF and has had a renal transplant in the RIF which is working well. He has gum hypetrophy which evidence of previous Cyclosporin use and warty fingers which is evidence of previous Azathioprine use. He also has a buffalo hump and centripetal deposition of adipose tissue which is evidence of long term steroidal use.He has previous scars from Tenckhoff catheters for peritoneal dialysis. He has also had haemodialysis using the permacath and from abrachiocephalic fistula which is no longer working, He has no ascites and his abdomen is non tender

I would like to comlplete my examination by checking Temp chart, BP, urine dip for glycosuria, fundoscopy

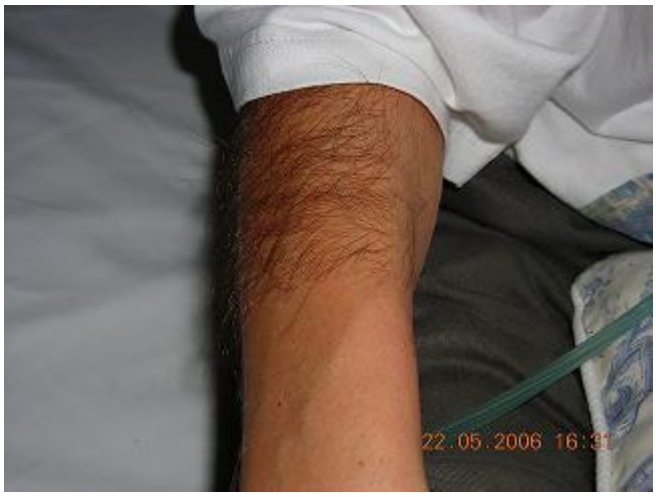
Q: What do you think is the cause of renal failure?
A:Being of Asian descent Diabetes is highest on my list but could also be due to hypertension, glomerulonephritis, adult polycystic kidney dse, nephrocalcinosis.

Q: You mentioned about all scars except one on the left abdo , what was it for?
A: May be some unrelated abdominal surgery, or complications from CAPD or left renal surgery.

Q: Can you tell us the indications for nephrectomy in APCKD?
A: Very large kidneys, recurrent abdominal pain, recurrent haemorrhage into cysts and malignant change.
I later found that this patient had APCKD and had undergone left nephrectomy with RIF transplant.

A Hirsute Man With Sallow Appearance (Right transplanted kidney):

This gentleman has lethargy of 6/12 duration. Please examine his abdomen.



I would complete my examination on this gentleman by checking his blood pressure, looking at his temperature chart and dipstick the urine to look for evidence of haematuria and proteinuria. I would also like to do a per rectal examination to exclude malaena as a possible cause for his pallor.

This gentleman is lying comfortable on one pillow. He is on supplemental O2 administered via nasal prong running at 2L/min. He has a sallow complexion. There is no peripheral stigmata of chronic liver disease. He has no flapping tremour. He has hirsutism as evidenced by coarse hairs over both of his arms as well as on the face. He looks pale however there is no jaundice. There is gum hypertrophy.

On inspection of the abdomen, there is a transverse surgical scar noted at the right lumbar region extending to the infraumbilical region which is well healed. The umbilicus is centrally located and inverted. Palpation of the abdomen has revealed a mass situated at the right iliac fossa which measures around 5x8cm in size, is firm in consistency, non-tender and it produces a dull note upon percussion. There is no other mass felt in the abdomen. There is no ascites and the bowel sounds are normal. There is no signs of fluid overload.
Therefore, I would like to draw conclusion that this gentleman has a right transplanted kidney complicated by signs suggestive of cyclosporin side effects, currently no signs of fluid overload and not in uraemic encephalopathy. The possible etiologies for his renal failure I would like to offer include diabetic nephropathy, hypertensive nephropathy and other primary glomerulonephritis.

During periphery examination, I noted an AVF. So, that gave me a clue this was a renal patient and the case probably a APCKD or transplanted kidney. There was no stigmata of CLD. Abdominal examination reviewed a RIF mass with an overlying mass, a RHC mass with bilateral balottable kidneys. So, confidently, I said, "This patient has a right transplanted kidney. He has ESRF secondary to APCKD".
Examiner asked me, "What do you think the RHC mass is?". I wasn't sure. I "hentam", most probably a right polycystic kidney. Examiner wasn't satisfy and asked "what else?". I said, it could be a liver. Why? Because, this patient was hemodialysis-dependent previously, so he could have contracted infective hepatitis due to Hep B or C. "What else?". By that time, the bell rang. Didn't get into the management part.

Comment:
In retrospectively, I thought, that could be a polycystic liver!!

You must try to score clear pass for abdomen station. It's a gift because there are only 3 main diagnoses that you need to consider i.e. ***liver problems, kidney problems or hematological problems.***

The abdomen case that I had in UK was a case of kidney transplant. The theme for that case was the patient complaint of lethargy for 2/52. Clinically, she was not Cushingoid. She was pale and there was no AVF. Abdomen examination showed multiple abdominal scars and one of them was a RIF surgical scar. There was a mass under the RIF scar. There was no fistula noted over the abdomen.
I was confused by the multiple abdominal scars and the history of lethargy. I concluded that the possible diagnosis would include Crohn's ds or kidney transplant. The examiner challeged me and asked which one was the most possible diagnosis. I chose the wrong one.
Retrospectively, I missed an ulcer over the right foot. At the end of the exam, I met the lady and she told me she was a case of ESRF secondary to diabetic nephropathy with kidney transplant done 6 years ago. She was not Cushingoid because she was only on low dose of steroid. The abdominal scars were due to previous CAPD and she was not on HD before.

This case taught me to think simple for abdomen case. Just focus on the three main diagnoses and look for the signs that support or eliminate each one of them. It is very unlikely to be a case of Crohn's disease because they are too ill to appear in PACES!

Cardiology

MedicAlert

Look for this MedicAlert bracelet in every patient in PACES. It might be your lifesaver on your PACES day!

Believe me, the 2nd picture show the exact diagnoses for the patient!
"Complex Congenital Cyanotic Heart disease"!

Coarctation of the aorta

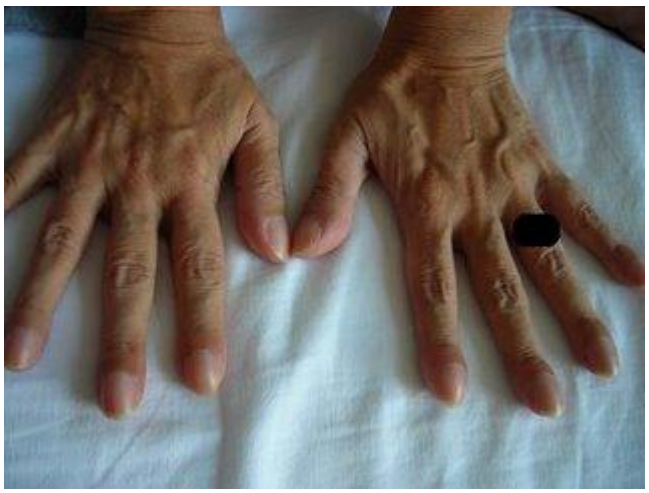
Mr K is admitted with uncontrolled hypertension. Please examine the cardiovascular system.

I would complete my examination by measuring his blood pressure on both sides. Examiner:"The blood pressure is 170/100mmHg on both sides."

The principal findings on this young man are radiofemoral delay with absent lower limbs distal pulses. I could not feel the posterior tibial and dorsalis pedis pulses on both sides, though the popliteal pulses were feeble. There were no puncture marks or haematoma on the groins to suggest a recent intraarterial procedure. Examination of the precordium revealed no abnormalities, in particular, there is no murmur to suggest an intracardiac shunt or aortic root abnormalities. There was no scar seen. There was no sign of heart failure.

With the aforementioned findings, I think this young man has coarctation of aorta complicated by uncontrolled hypertension. It would be most pertinent to look for signs of target organ damage urgently, ie preliminarily funduscopy, urinalysis and serum creatinine. In view of his age, I would also consider the differential diagnosis of Takayasu arteritis.

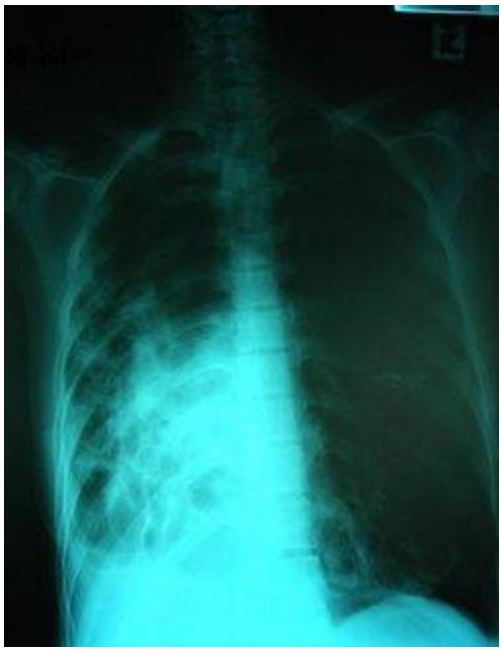
Kartagener’s syndrome



The unifying diagnosis I would like to consider for this patient is Kartagener syndrome. My principle findings on cardiovascular examination is that she has dextrocardia. The apex beat is at the right 5th intercostal space. There is no murmur to suggest an intracardiac shunt. Clinically, she is not in cardiac failure and she is in sinus rhythm.

She has obvious finger clubbing and on auscultation of her chest, there are coarse crepitations at the bases which diminish following a cough. I would be keen to examine the abdomen to look for associated situs inversus.

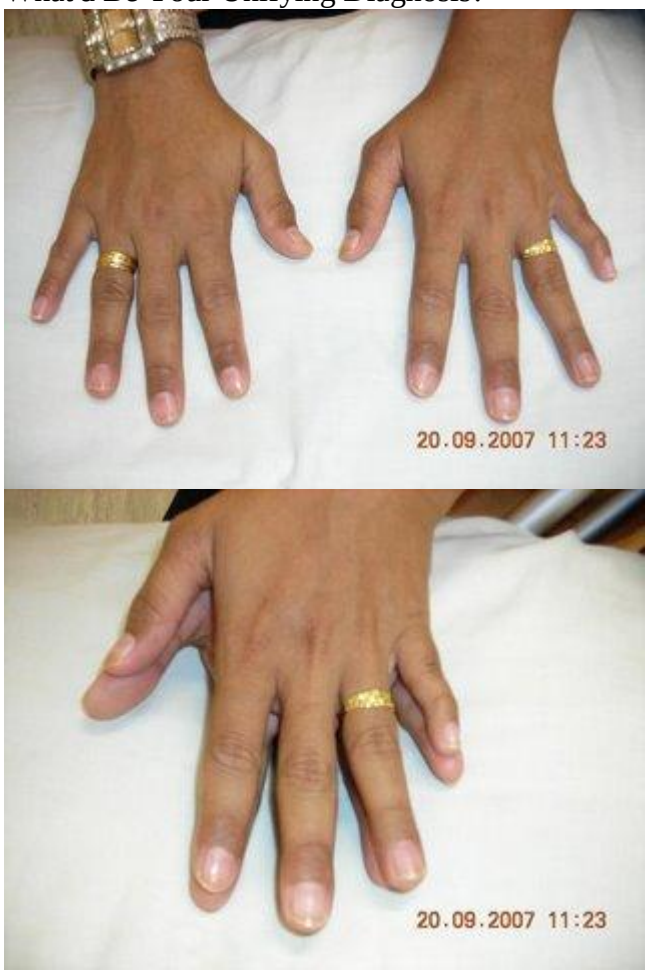
How would you like to investigate this patient?
I would do a plain chest radiograph to confirm dextrocardia and to look for features of bronchiectasis.
(Examiner:"Here you are.")



The cardiac silhouette falls on the right side and there are ample findings which are in keeping with bronchiectasis. There are cystic changes resembling a bunch of grapes on the lower zones. Tramline opacities are noted especially on the left base. However, the gastric bubble appears to be on the left side hence I think this patient doesn't have situs inversus to complete the syndrome.

Holt-Oram syndrome

What'd Be Your Unifying Diagnosis?



It's been a while since our last 'casual' life blogs.

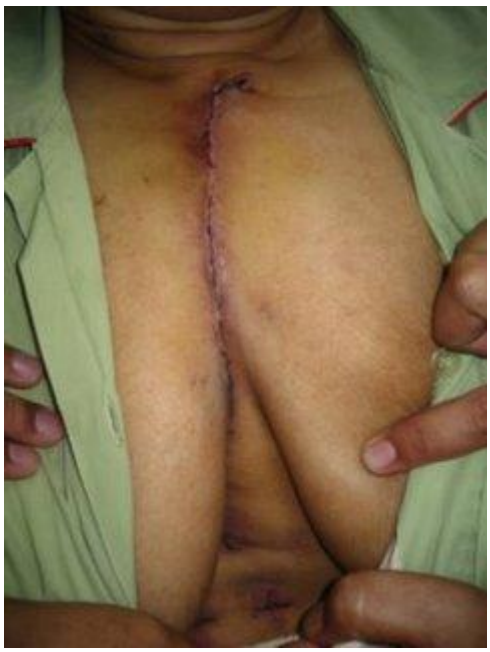
I saw this lady during my combined antenatal and medical follow-up.

She had a congenital cardiac lesion which is corrected. Clinically she is pink on air and there is no finger clubbing.

What would you expect the lesion to be and what would be your diagnosis?
It's Holt-Oram syndrome.

She doesn't have a murmur as her ASD was closed.

Midline Sternotomy Scar: mitral valve replacement



Examine the Cardiovascular system

This pt has a pulse rate of 80/min which is irregularly irregular. There is no radio-radial delay or collapsing pulse. There is no clubbing or peripheral stigmata of infective endocarditis. She is pale. There is no jaundice, cyanosis. JVP is not elevated and there is no pedal oedema of the legs.

On examination of the precordium, there is a midline sternotomy scar.
Apex beat is displaced, 5th intercostals space 2 cm lateral to MCL. No parasternal heave or thrills.
Metallic click of the 1st HS, normal 2nd HS
Lungs clear

I would like to complete my examination by checking the fundus for Roth spot or fundal haemorrhage, look for signs of overwarfarinization, examine the CNS for evidence of CVA, look for splenomegaly, urine dipstick for haematuria, check BP, all peripheral pulses

My diagnosis is that this pt has Mitral valve replacement and is in atrial fibrillation and is anaemic. There are no signs to suggest infective endocarditis, pulmonary

hypertension, overwarfarinization or heart failure. There is also no evidence of valve leakage.

Questions:

1. What are the causes of anaemia ?
*Bleeding due to anticoagulation
*Haemolysis
*Secondary to IE
2. Advantages of Porcine heart valve
*No need anticoagulation
3. Disadvantage of Porcine heart valve
*Degeneration with time – may need reoperation (after about 7 years)
*Calcification
An elderly gentleman had already been properly exposed and positioned propped up at 45o.
"Please examine the cardiovascular system and tell us your findings."
Again, a rather simple stem. Nothing fancy.
I quickly noticed a midline sternotomy scar on the patient. I gathered my clinical experience and in my mind, I had these possibilities outlined:

1. Patient had a valve replacement done, either metallic or bioprothetic - *this is by far the commonest scenario encountered in PACES with sternotomy scar*
2. Possible valve repair
3. Patient had a CABG done - ?evidence of venous harvest
4. Patient had sternotomy for excision of cardiac tumours or masses eg, atrial myxoma, resistant IE etc or closure of large septal defects
5. Sternotomy for non-cardiac surgery eg, thymectomy, repair of other midline structures etc. - *this is highly unlikely in PACES station 3!*

I proceeded to pick up his hands and started examining the peripheries. The only positive sign was of an irregularly irregular pulse at about 60-70bpm. There were no stigmata of IE and in particular, I could not see any scar of previous venous harvest both in the upper and lower limbs.
He was pink on air and there was no jaundice. The venous pressure is not elevated and, as mentioned earlier, there was a midline sternotomy scar which was well healed.

There was no scar elsewhere at the precordium.
At this juncture, I gathered that the positive signs were:

1. Rate-controlled AF
2. Midline scar

I told myself that I should be looking hard for a mitral valve pathology hence needing surgical intervention, with the distant possibility of a previous large ASD now closed.

On examination of the precordium, the apex beat is not displaced. There was no thrill. The were no signs of raised pulmonary pressure. The first heart sound was loud and the second heart sound was well heard. There were no other added sounds. There was no signs of cardiac failure and no signs to suggest overwarfarinisation.

"So what do you think?"

"I think this gentleman has had a mitral valve replacement done, complicated by atrial fibrillation currently rate-controlled. There was no signs of cardiac failure. There were no metallic sounds heard on auscultation and hence it cannot be a metallic valve. It is possible that this gentleman has had a bioprosthetic mitral valve implanted."

"Are you sure? What would be the other possibilites?"

"It is still possible that he had a mitral valve repair done previously."

"Which one do you think is more likely?"

"I think a valve replacement would be more likely as the presence of atrial fibrillation would indicate the chronicity of the mitral valve pathology, which is, mitral stenosis in this case in view of the undisplaced apex beat. A valve repair is usually being employed to ameliorate a less severe or chronic valve pathology."

"Ok then, you say that it is a bioprothesis. Would you still want to give this patient warfarin?"

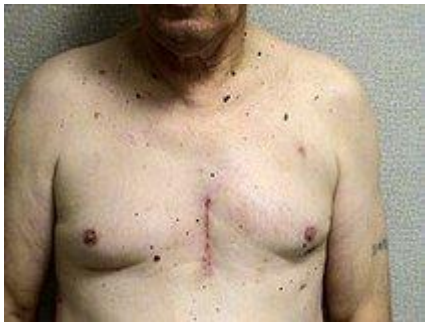
"Yes. I would like to anticoagulate this patient for the indication of atrial fibrillation."

"Why is that so?"

"To reduce the risk of thromboembolism. Warfarin is shown to be superior than aspirin alone in preventing thromboembolism, for example stroke, which causes significant morbidity and mortality."

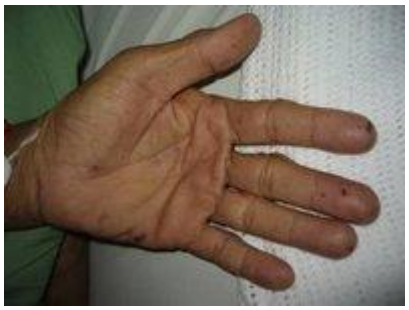
"Do you know the percentage of thromboembolic risk reduction by giving patients warfarin?"

"Sternotomy scar"



When one see this midline sternotomy scar, it carries 2 possibilities i.e. CABG with underlying IHD or prosthetic valve replacement.
For CABG, look for presence of functional MR and presence of pacemaker for underlying arrhythmias.
Please bring your ear near to the chest to listen for prosthetic click with an unaided ear then ascertain which valve is replaced.

Infective endocarditis



What is the sign shown ?

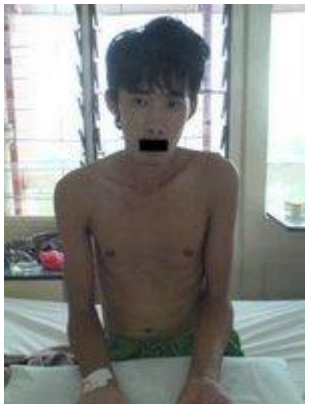
What is the diagnosis ?

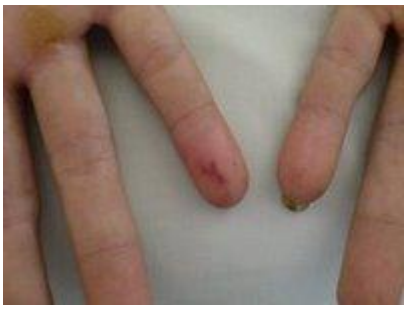
What is the possible underlying valve lesion ?

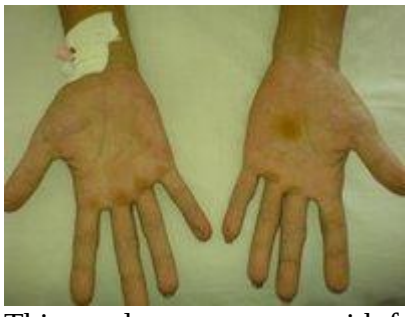
He has Janeway lesion, although you do see it at the pulp, however it is non-tender and non-palpable.

He happens to be an IVDU and therefore has a TR murmur. Also has MR and thromboembolic phenomenon of stroke.

Infective endocarditis Update







This gentleman presents with fever for 3/52. Please Ex his cardiovascular system

I would like to complete my examination by checking

1) fundus – Roth spot
2) Dipstick the urine for hematuria
3) Temperature chart
4) Abd- spleenomegaly

This gentleman looks thin and he has a brannula at his right wrist. He has stigmata of IE as evidenced by presence of Osler nodes and Janeway lesion. There is no finger clubbing or splinter haemorrhage. There are also IV puncture marks over the cubital fossa.
PR is 90 bpm regularly regular. There is no radial-radial delay and the pulse is not collapsing.
He is pink and not jaundice.
Jugular venous pressure is not elevated.
Apex beat is not displaced and there is no thrill or Left parasternal heave.
There is a PSM best heard at the apex and non radiating

This gentleman has IE currently not in failure. He probably is an IVDU in view of the presence of IV puncture marks.

Investigations:

1) FBC- WCC, normochromic normocytic anaemia
2) ESR
3) Blood C&S- 3x fr different sites over 1 hour time span
4) ECHO

Treatment

1) Antibiotics- IV C. Pen + Genta x 2/52
IV Cloxa + Genta x 2/52 until review C&S
2) Surgery if
a. Persistent Blood C&S/ relapse
b. Abscess
c. Fungal
d. Damaged valve
e. 2nd / 3rd degree heart block secondary to aortic valve vegetation
f. New aneurysm of the sinus of Valsalva

IE prophylaxis for Dental Procedure(Circulation May 8 2007)

1) Prosthetic heart valve

2) Previous IE

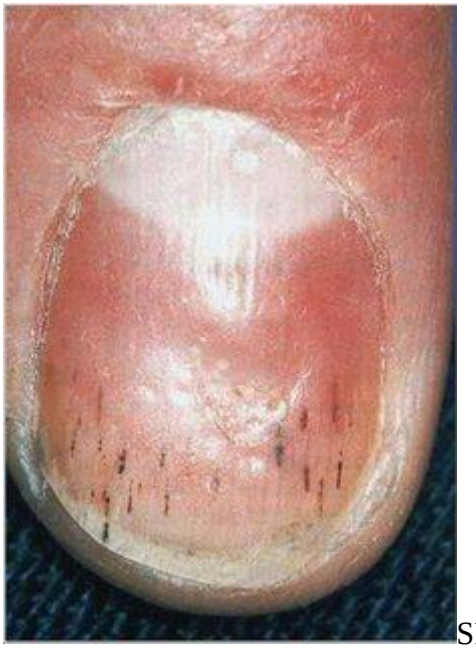
3) Congenital heart disease

4) Cardiac transplant patients who develop valvulopathy

give oral amoxy 2g, 30 min before dental procedure

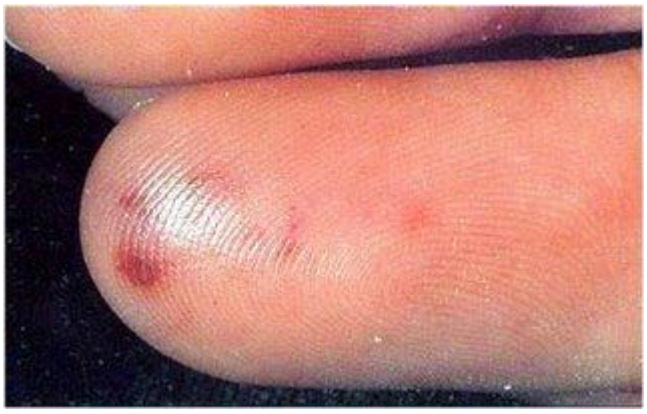
Prophylactic antibiotic for IE not recommended for GU/ GI tract procedure

IE signs



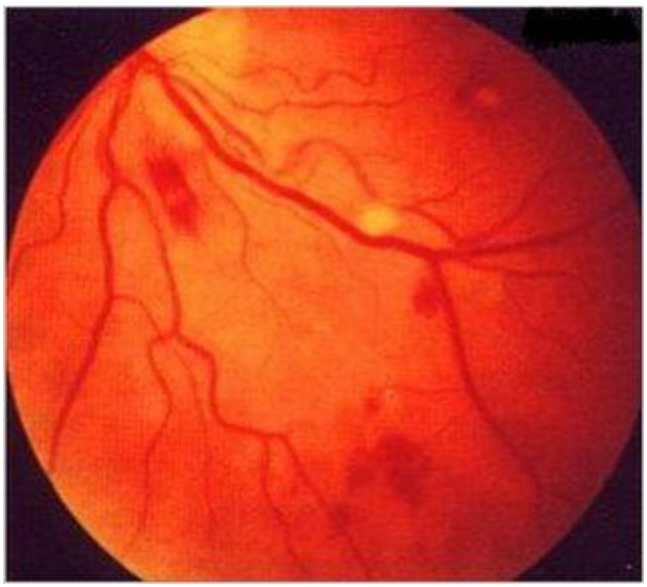
SPLINCTER HAEMORRHAGE

subungual linear “vasculitic” lesions



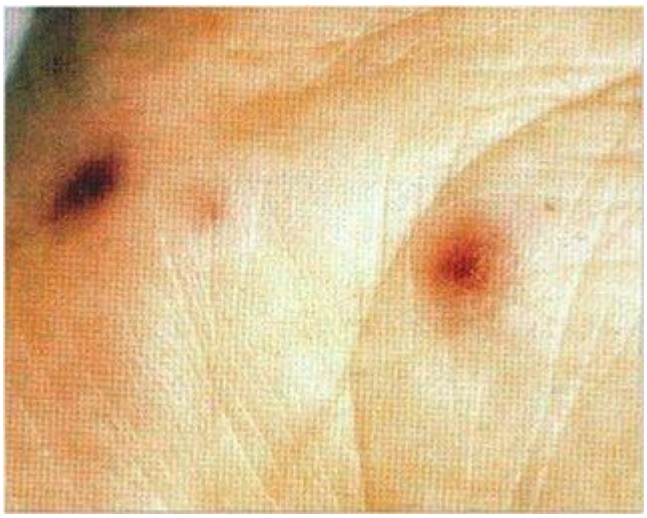
OSLER’S NODES

red, raised tender nodules on the pulp of the fingers (or toes), or on the thenar or hypothenar eminences



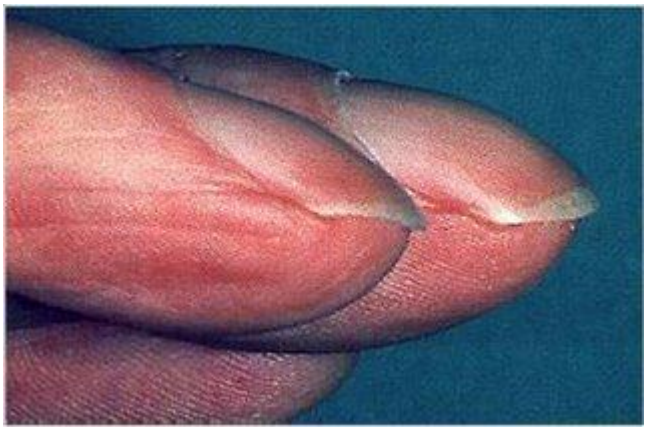
ROTH'S SPOT

intraretinal haemorrhages with white centers representing fibrin thrombus occluding a rupture blood vessel



JANEWAY'S LESION

Non-tender, erythematous maculopapular lesions, containing bacteria, which occur very rarely on the palms and pulps of the fingers



CLUBBING

"Does this patient need IE prophylaxis?".

1. MUST on IE prophylaxis

- Prosthetic heart valves
- IE patients
- Complex cyanotic congenital heart diseases

2. SHOULD on IE prophylaxis

- Congenital cardiac malformations like VSD (with the exception of isolated secundum atrial septal defects)
- CRHD like MS, MR, AS, AR
- Hypertrophic cardiomyopathy
- Mitral valve prolapse

Secondly, they will ask what procedures need IE prophylaxis? For PACES, just remember all dental procedures need IE prophylaxis. (In fact, NOT all of them need one).

Other procedures will includes:

1. Respiratory

- Tonsillectomy

- Rigid bronchoscopy

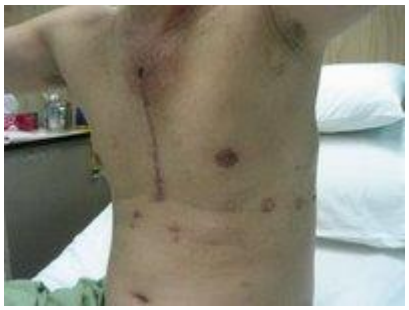
2. GI

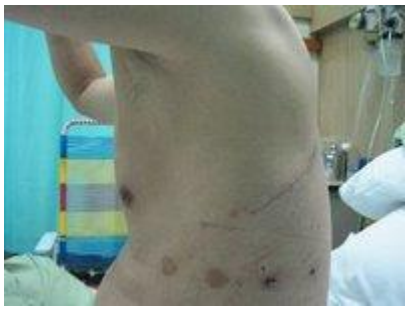
- ERCP
- Operation that involve intestinal mucosa
- Biliary surgery
- Esophageal stricture dilatation
- Sclerotherapy for esophageal varices

3. Genitourinary

- Cystoscopy
- Prostate surgery like TURP
- Urethral dilatation

Post-operative pericardial effusion





A consultoid cardiologist took me for a short case 1 hour ago.

This man had a clear cut mitral valve replacement ! However the question was what was the other scar due to ??

Left sided pericardial window and the smaller scars for the drains.
It's open pericardial drainage by thoracotomy approach. This patient's MVR was complicated by post-op pericardial effusion.

A Tearful Cyanotic Boy

I was paged by my ED colleague at 1800H to see an emotionally labile 15 year-old boy who presented with history of recurrent haemoptysis. He is a known congenital heart disease patient who has defaulted our paediatric cardiology colleagues f/u years ago. He is in functional class II-III prior to presentation.

Record:

I would complete my examination by taking his blood pressure and to examine the abdomen, in particular, to look for a pulsatile liver.
Fingers and toes are clubbed. Cyanosed both peripherally and centrally, this boy has a regular pulse of 85 bpm, normal in volume and not collapsing in nature. Pulses are present and equal on both sides. No peripheral stigmata of infective endocarditis.

The venous pressure is elevated with a prominent v wave. The precordium is hyperdynamic with a visible pulsation noted on the right side of the chest. This is dextrocardia. The apex beat is situated at the right 5th ICS MCL. There is presence of parasternal heave with palpable P2 however there is no thrill. Auscultation revealed normal 1st and 2nd heart sounds. There is an early diastolic murmur best heard at the right 3rd ICS with patient in full inspiration, grade 3/6, and is non-radiating. There is another pansystolic murmur best heard at the right lower sternal edge, grade 2/6, and is non-radiating as well. There is loud P2 with variable splitting of the 2nd heart sound.
Lungs are clear to auscultation and there is no ankle oedema.
I would like to draw conclusion that this patient has dextrocardia with Eisenmenger syndrome most probably due to a congenital septal defect; now complicated by pulmonary hypertension, not in cardiac failure and there is no stigmata to suggest infective endocarditis. He has pulmonary regurgitation and tricuspid regurgitation which are in keeping with severe pulmonary hypertension.

Verdict:

It turned out that this patient has a complex congenital heart lesion comprising of:

1. Dextrocardia
2. VSD

3. ASD
4. Double outlet right ventricle
5. Severe pulm HT

Pitfalls:

Further history revealed that this child has been diagnosed with the above condition by our paediatric cardiology colleagues when he was 7. His mother relayed to me that the family were utterly upset by a open comment made a doctor while he was teaching his juniors that this patient would not be able to survive beyond 12. And hence they defaulted, pretty much out of frustration.

I think it is pertinent that we should be very tactful while breaking this news to the family.

Remember the 5T's of cyanotic congenital heart diseases?

1. Tetralogy of Fallot (TOF)
2. Transposition of the great vessels (TGV)
3. Tricuspid valve atresia
4. Total anomalous pulmonary venous return
5. Truncus arteriosus

Any of the acyanotic congenital lesions ie VSD, ASD and PDA can present with cyanosis when they develop Eisenmenger syndrome later in life.

Hence, in PACES MRCP, remember:

-Young patients (<20) with cyanosis + clubbing ->> think of TOF (commonest) and other primary cyanotic conditions
-Older patients (>20) with cyanosis + clubbing ->> think of acyanotic conditions with Eisenmenger syndrome (commonest be VSD)

Mx of Valvular Heart Lesions

The *ultimate* question for valvular lesion is "How would you manage the patient?"
The general principles are:
1. *ALL* patients need IE prophylaxis.
2. Medical treatment in general is meant for symptomatic relief except nifedipine in AR.
3. Pregnant issues.
4. *ALL* stenotic lesions need surgery when symptomatic.
5. Regurgitant lesions may need surgery prior to symptomatic eg LV dysfunction or dilatation.

Valve lesion	Medical	Surgical Indications	Intervention
MR	*Consider diuretic for symptomatic relief. *Use anti-coagulation and rate control in AF.	Severe MR and one of the following: *Symptoms *LV dilatation (end systolic size>4.5 cm) *LVEF<60%	*Mitral valve repair *Mitral valve replacement
MS		*Moderate or severe MS (mitral valve area ≤1.5 cm2) and symptoms *Pulmonary HPT or hemoptysis *Recurrent embolic event despite good INR	*PTMC *Mitral valve repair *Mitral valve replacement

AR	*Afterload reduction (nifedipine is most studied agent)	Severe AR and one of the following: *Symptoms *LV dilatation (end-systolic size >5.5 cm) *LV dysfunction (LVEF <50%)	Aortic valve replacement
AS	None	*ALL symptomatic *Valvular gradient >50mmHg *Aortic valve area ≤0.6 cm2 *Asymp with LV dysfunction	Aortic valve replacement

Severity of Valve Lesions

Aortic stenosis

Narrow pulse pressure

Soft S2

Narrow or reverse S2

Systolic thrill and heaving apex beat

S4

Cardiac failure

Aortic regurgitation

Wide pulse pressure

Soft S2

Duration of decrescendo murmur

Presence of LV S3

Austin Flint murmur

LVF

Mitral regurgitation

Larger LV (displaced apex)

S3

Mitral stenosis

Narrow distance S2 and opening snap

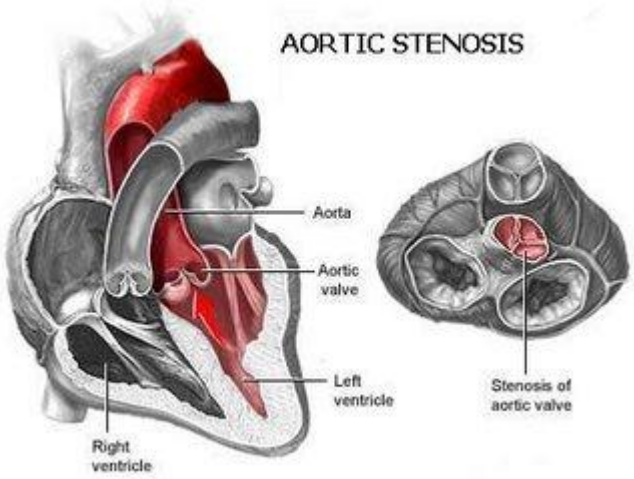
Longer diastolic murmur

Pulm HPT

Mitral stenosis

A middle-aged lady presented with reduced effort tolerance, I was asked to examine her cardio-vascular system. I started by examining the peripheries. I then noted patient had irregularly irregular pulse. What came to my mind were two differential diagnosis, either Mitral valve disease or ASD. Her apex beat was not displaced and was tapping in nature (very classical). There was parasternal heave. S1 and S2 were loud and there was a MDM over the apex. No other abnormal sound was heard. Examiner asked me what was the findings and I started presenting all the positive signs from the peripheries. I concluded as: "This patient has pure mitral stenosis, secondary to chronic rheumatic heart disease, complicated by atrial fibrillation and pulmonary hypertension; clinically she is not in cardiac failure and there is no stigmata of infective endocarditis". The examiner asked me whether there was any other murmur heard and I said no. Examiner did not probe further, so I presumed I was correct. They only questioned me on how to assess severity clinically.

Aortic Stenosis



Record :
I would end my exmination by checking the BP for narrow pulse pressure, fundoscopy for Roth spot, abd for splenomegaly, urinalysis for proteinuria and haematuria, all peripheral pulses.

This lady has a PR of 95/min, regular, slow rising, no radio-radial delay and no collapsing pulse. There is no clubbing or stigmata of infective endocarditis. There is no pallor, jaundice or cyanosis. JVP is not elevated. There is no pedal edema of the legs. There is a right mastectomy scar. The apex beat is not displaced, L 5th ICS at MCL, heaving in character. There is no parasternal heave or thrill felt. Normal 1st and 2nd HS. There is a Grade 3 ESM at the aortic radiating to the neck. Lungs - clear

My **diagnosis** is that this lady has aortic stenosis most likely due to CRHD with no evidence of PH, IE or cardiac failure.

eMRCPian : What other possible aetiology ?
Ans : Degenerative valve or Bicuspid valve

eMRCPian : What are the signs of severe AS ?
Ans : Narrow pulse pressure, Soft S2, Narrow split S2, S4, systolic thrill and heaving apex beat, cardiac failure

eMRCPian : How would you investgate this pt ?
Ans :

CXR - cardiac enlargement, calcification of valve

ECG - LVH, LAD

Echo - confirm dx, assess severity, assess LV function

eMRCPian : How do u MX this pt ?

Ans :

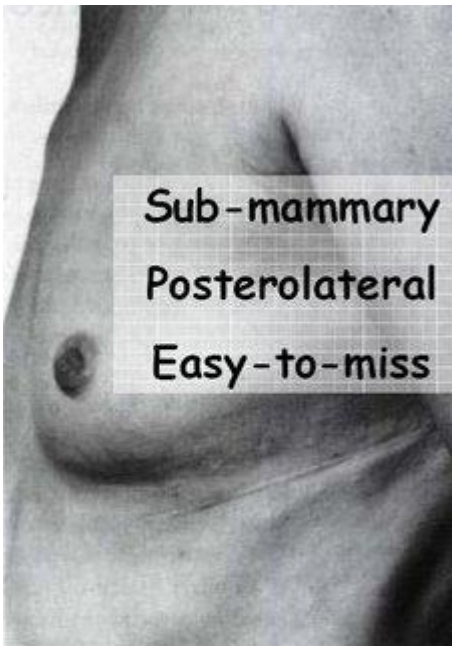
Divide into symptomatic and asymptomatic

Symptomatic - AV replacement

Asymptomatic - follow up pt, assess severity

AVR for asymptomatic - valvular gradient > 50 mmHg, valve area < 0.6 cm2

Tell-Tale Valvotomy Scar



I can't stress enough the importance of looking for scar.

But, some scars are easily missed in exam! Valvotomy scar is one of them.

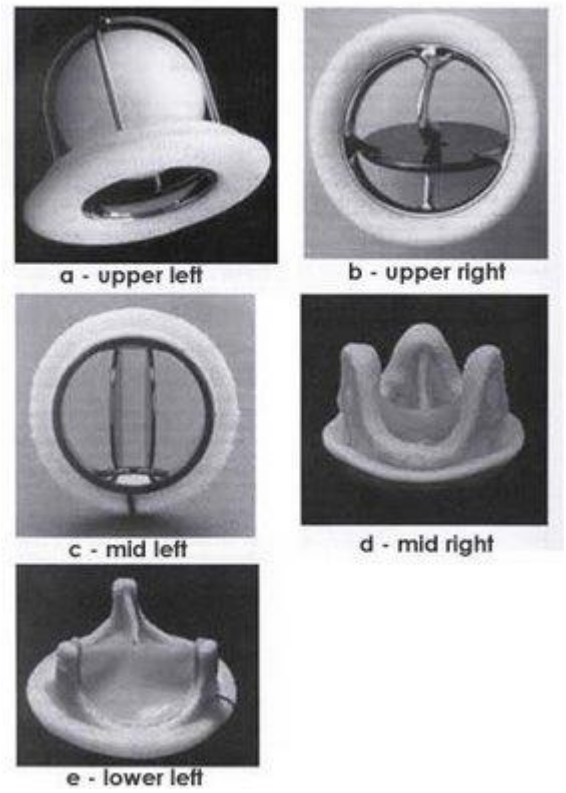
What should you suspect before putting your stethoscope on the precordium?

Answers:

MS

MR complications of valvotomy

METALLIC Prosthetic Valve



5 types of prosthetic heart valves:

A. Starr-Edwards mitral caged ball valve.
B. Medtronic Hall tilting disk valve.
C. St. Jude bileaflet valve.

D. Hancock porcine valve.
E. Carpentier-Edwards bovine pericardial valve.

(NOT as short case)

Don't confuse yourself with the types of prosthetic valves. For PACES setting, you only need to know about:

1. Which valve is replaced?

- Mitral valve: metallic S1, loudest at mitral area
- Aortic valve: metallic S2, loudest at aortic area

2. Any complications?

- evidence of valce leakage
- IE
- Thromboembolism eg embolic stroke
- Bleeding due to overwarfarination
- Hemolytic anemia

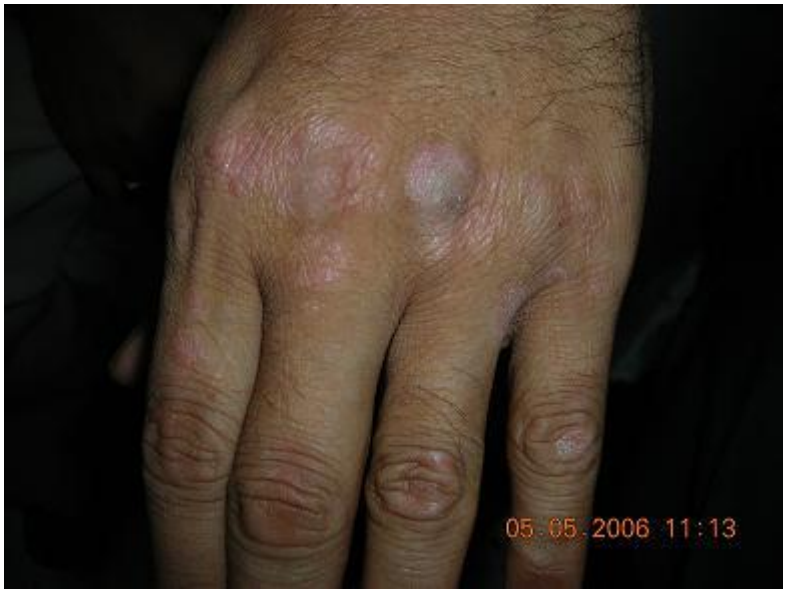
Pitfalls

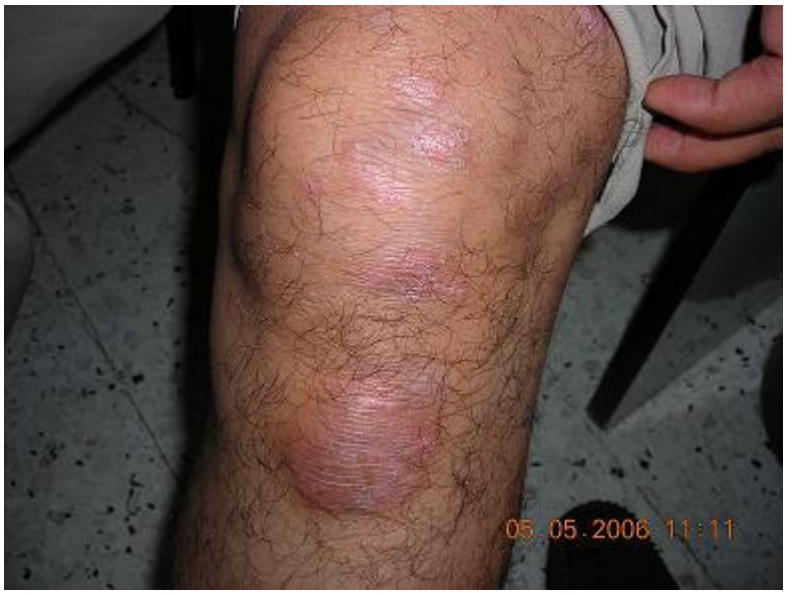
Fail to recognize bivalve replacement

Mistaken systolic flow murmur as murmur of valve dysfunction

-*ONLY* 2 important murmur you need to remember for valve leakage i.e. AR and MR

Dermatology

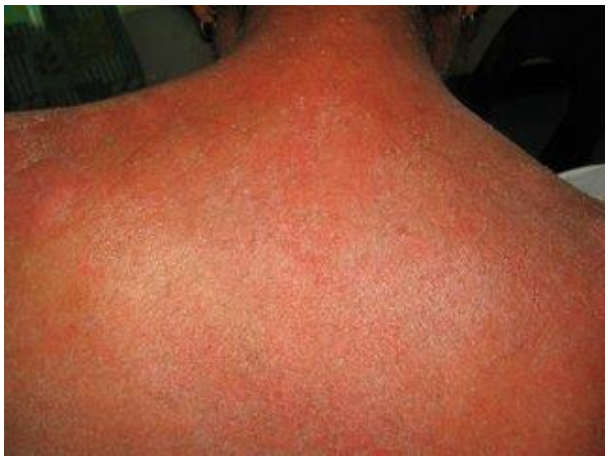


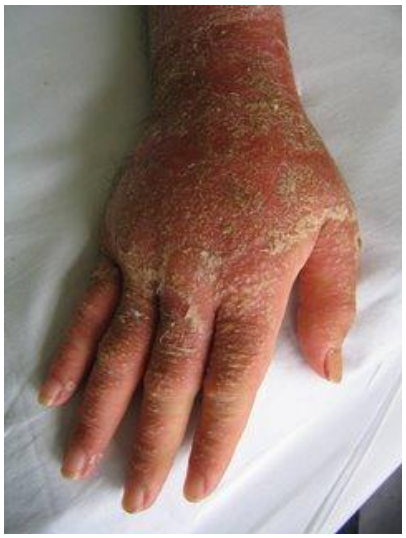


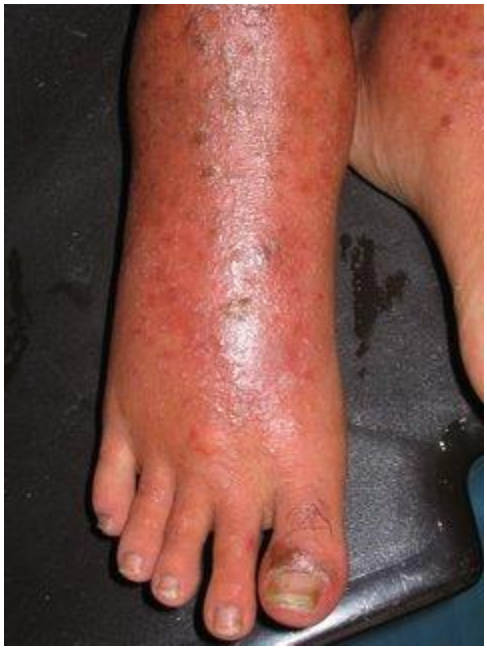
This patient has psoriasis without joint involvement.

There is pitting of the fingernails and thickening of the nail plates. Onycholysis is noted in some of the fingers. However, there is no yellowish discolouration of the nails and there is no joint involvement. There are patches of silvery scaly, salmon-pink and well-circumscribed circular plaques noted at the elbows, knees and dorsal aspects of the fingers. The scalp is spared and there is no lesion elsewhere.

Pustular Psoriasis







Stem

"This young man presented with this condition after being given some treatment for a his plaque psoriasis . Please examine his skin"

Suggested presentation

This febrile unwell-looking young man has generalised widespread creamy white pustules with an erythematous base. Some of the pustules have erupted, resulting in red oozing erosions . The palms and soles are also involved. He also has generalised onycholysis with discolouration and misshaped toes and fingernails.The patient has tenderness and warmth over both knee joints.

The diagnosis is pustular psoriasis complicated by gout. The likely cause for this presentation is massive intradermal steriod injection.

(Patient in this picture had pre-existing plaque psoriasis, so did his father and paternal grandfather. He was given Intradermal methylprednisolone 200mg daily instead of the recommended 10 mg per month. Not shown in the pictures, he had evidence of plaques on his elbows, hairlines and behind the ears)

Questions that may be asked

1. What differentiates this from skin infections?

The pustules are sterile on culture and the patient's white cell count may be normal

2. What is the explanation for the patient's knee pain?

Psoriasis is a condition where there is increased cell turnover so there is hyperuricaemia, which results in crystal deposition in knee joins, giving rise to gout.

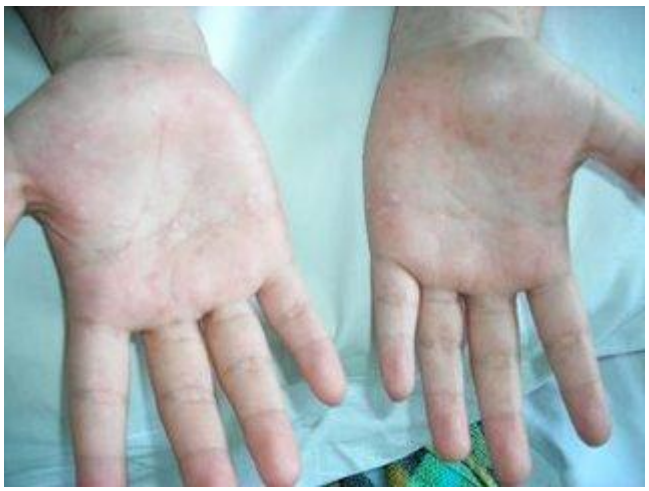
3. How would you manage this patient?

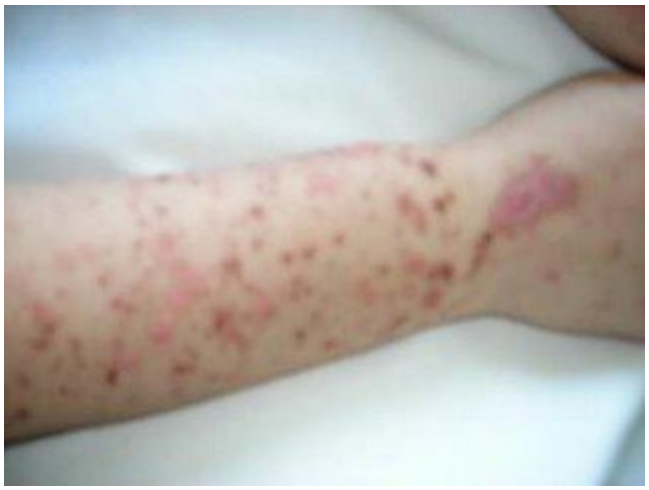
The patient should be managed like a exfoliative erythrodermic patient, paying close attention to barrier nursing, fluid replacement, prompt treatment of infections, adequate nutrition

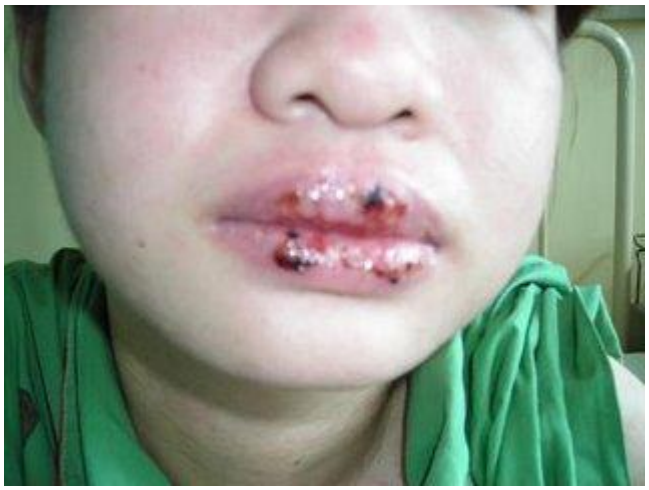
4. Name me the definitive treatment for pustular psoriasis.

Acitretin, an oral retinoid.

Steven Johnson syndrome







This series of photos belongs to a lady who presents with a rash.

There are multiple erythematous well circumscribed maculapapular lesions noted bilaterally on the palms and extensor and flexor compartments of forearms bilaterally.

The lesions on the forearms are deemed to be at different stages of development.
There are 2 patches of confluence on the distal right and left forearms.
The lesions on the patient's palms could not be assessed in details.

In addition to the lesions on the hand, there is an ulcerating lesions with signs of dried hemorrhages on the patient's upper and lower lips.There is no lesions noted on the patient's face.
I can only guess for this case. Could this be a case of Steven Johnson?

This are outlines of management of steven johnson syndrome given by a dermatologist from Ipoh GH:

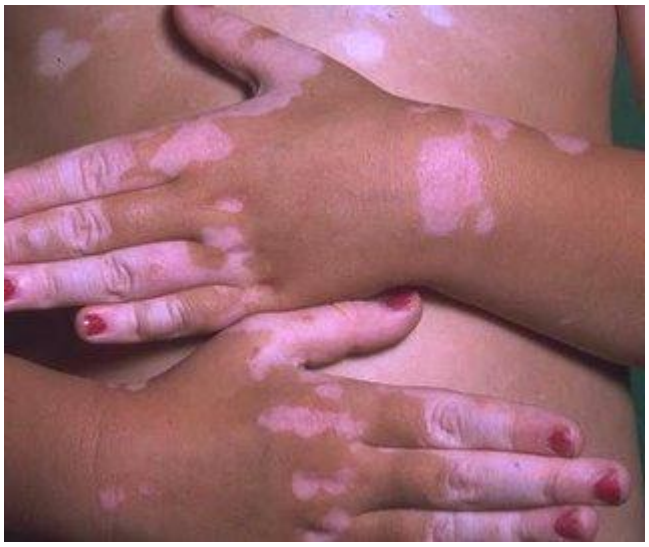
1) Vital Sign monitoring
2) Withhold Offending drug
3) Adequate hydration
4) Strict I/O chart
5) Assess disease severity daily
6) Oral lesion: NaHCO3 gurgle 4hrly
Syrup nystatin 6hrly
7) Crusted lip lesion: use liquid parafin soak with gauze apply to lips for 10min 4hrly to soften the crust, then slowly remove it
8) Body and lim lesions: KMno4 wash to raw area 2-3x per day, normal saline bath bd then apply gerila cream to raw area after KMno4, aqueous cream 2hrly

Basic dermatology:

1. Macules

** Well circumscribed change in colour of the skin. It is a discrete flat lesion.

** Examples: vitiligo, viral exanthems, cafe-au-lait spots.

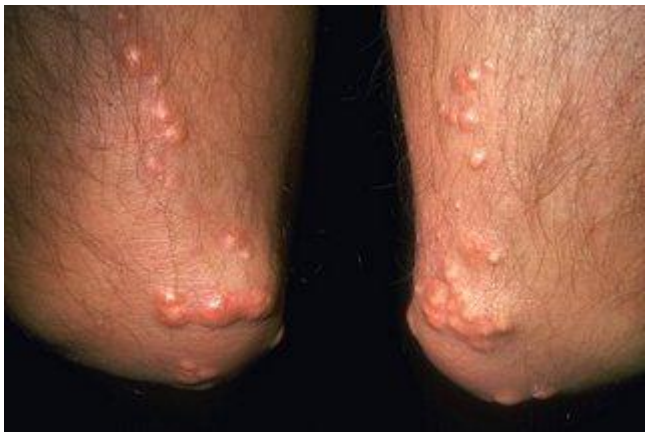


2. Papules and Nodules

** Well circumscribed, solid elevated lesions.

** <1cm in diameter are papules, >1cm in diameter are nodules.

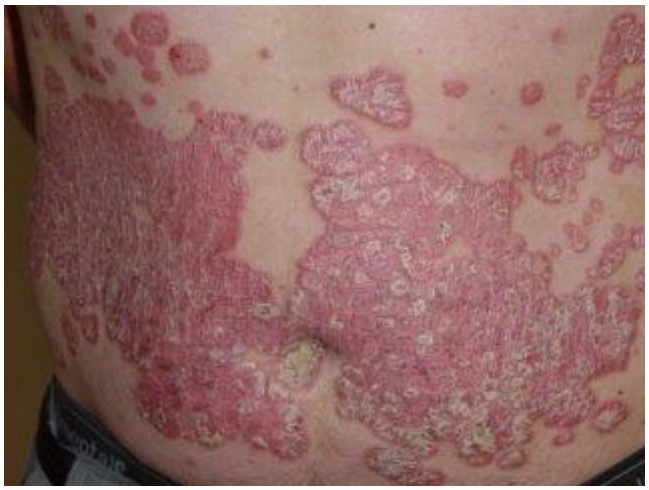
** Examples: xanthomas, molluscum contagiosum, amyloidosis.



3. Plaques

** A flat topped palpable disc shaped lesion.

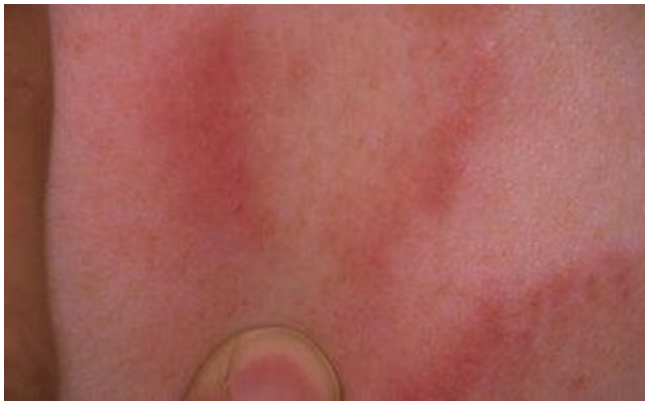
** Classical examples are psoriasis, lupus erythematous.



4. Wheal

** Rounded, erythematous, flat elevation which is transient vascular response with dermal oedema.

** Examples: urticaria.



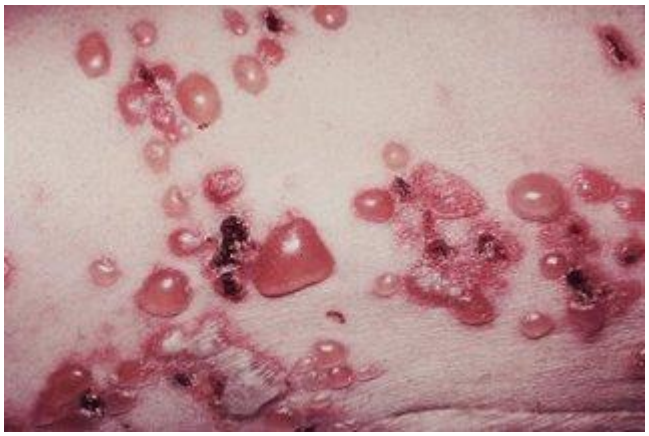
Wheal is compressible

5. Vesicles and Bullae

** These terms describe different sizes of blister i.e. circumscribed elevation containing fluid.

** Vesicles(<5mm),>5mm)

** Examples: herpes simplex, pemphigus, pemphigoid.

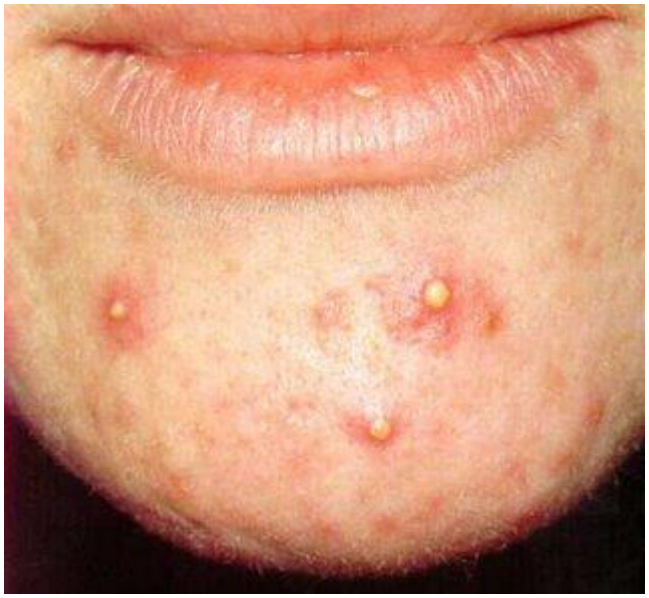


Pemphigoid

6. Pustules

** A visible accumulation of free pus.

** Examples: pustular psoriasis, acne

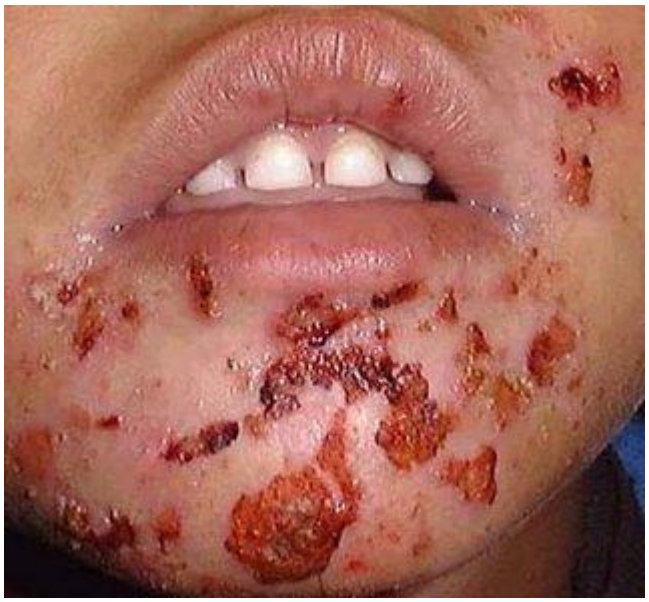


Acne vulgaris

7. Crust

** An accumulation of dried exudate, serum, blood. It is commonly known as scab.

** Examples: impetigo, ecthyma.



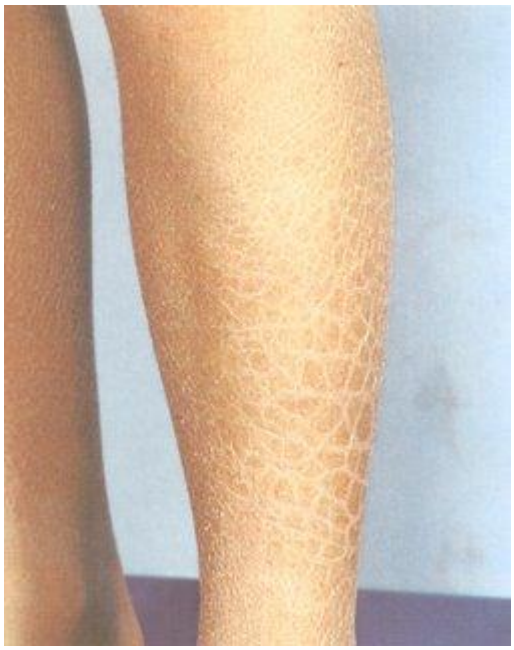
Impetigo

8. Scale

** Flaking of the skin surface due to increased loss from the stratum corneum(SC).

** It implies an abnormality in SC formation or damage to the epidermis.

** Examples: psoriasis, ichthyosis, tinea infections

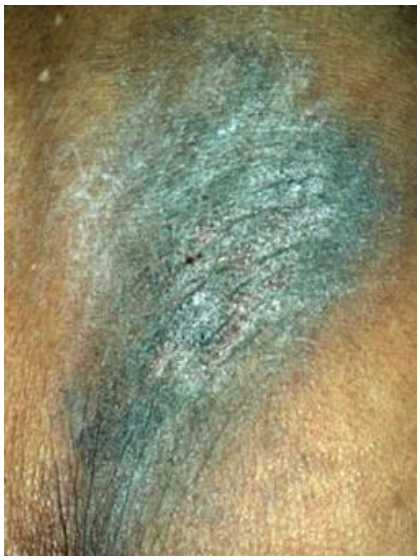


Ichthyosis vulgaris

9. Lichenification

** Areas of increased epidermal thickness secondary to chronic rubbing.

** Examples: eczema, lichen simplex chronicus

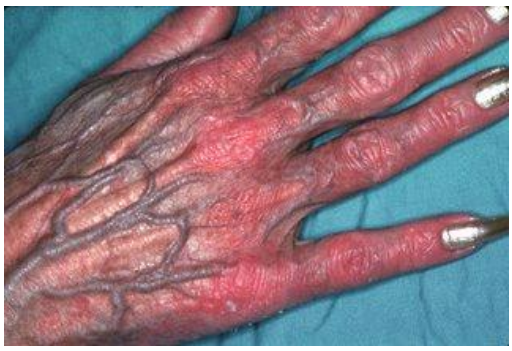


10. Atrophy

** Diminution of some or all layers of skin.

** In epidermal atrophy, the epidermis is thin, transparent and the vessels are prominent.

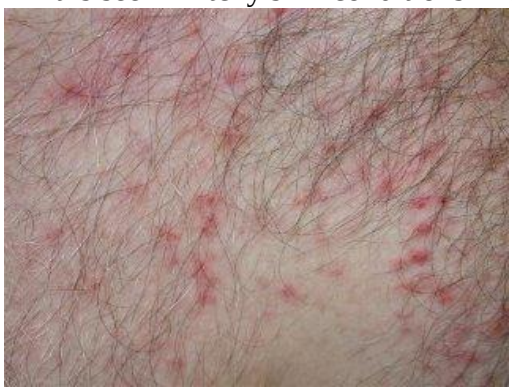
** Examples: Steroids induced atrophy



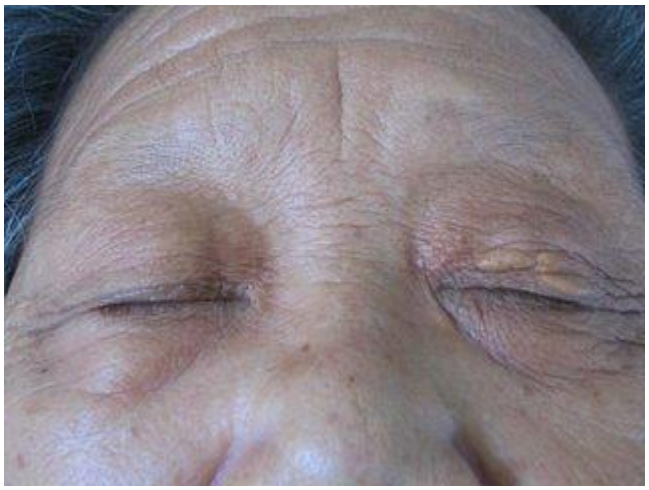
11. Excoriations

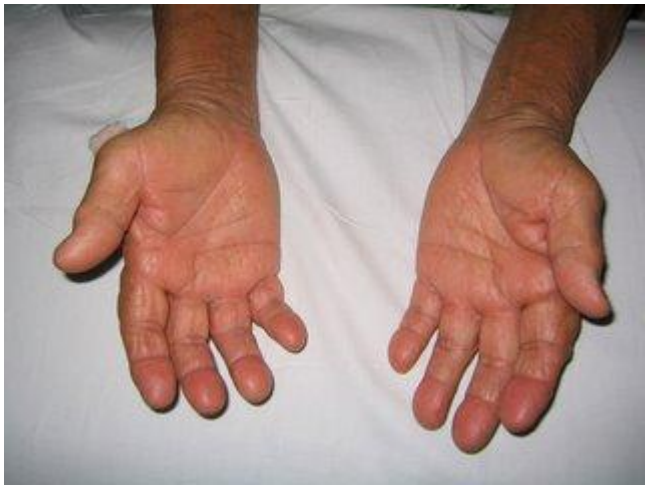
** A shallow abrasion due to scratching

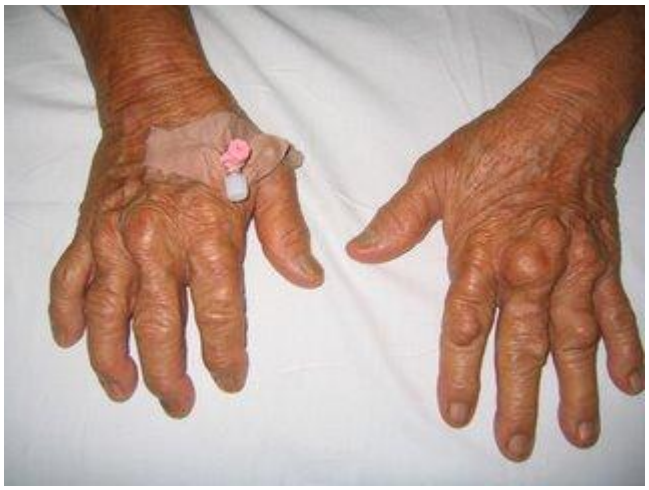
** It is seen in itchy skin conditions like eczema, scabies, urticaria.

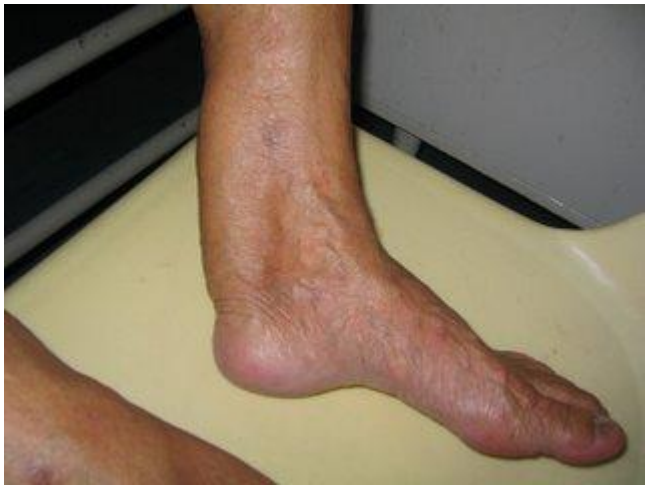


Familial hypercholesterolaemia









Examine this 80 year old lady's hands and comment

(Hint, she should have died long ago, and yet's she's fit as a fiddle. Her mother had the same condition and died her 80s)

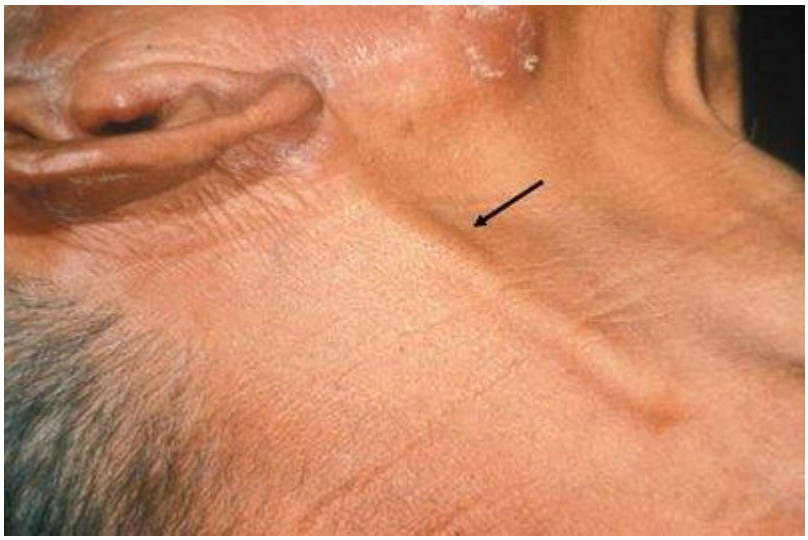
This lady has familial hypercholesterolaemia,autosomal dominant inheritance.pitures shows

1.xanthlasmata palpabrarum

2.tendon xanthomas

3.achillus tendon xanthoma.

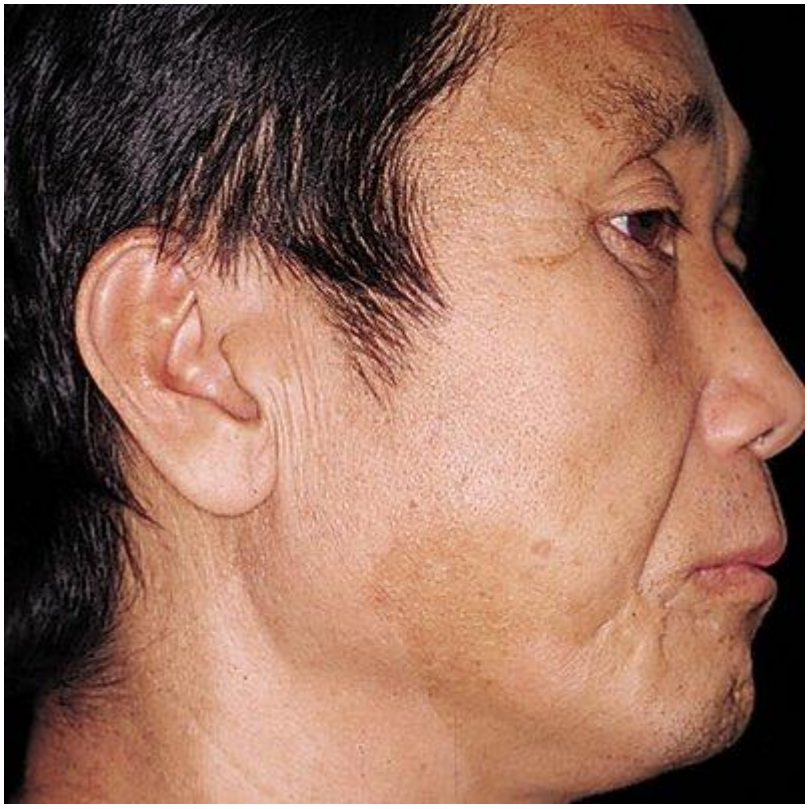
Hansen's Disease



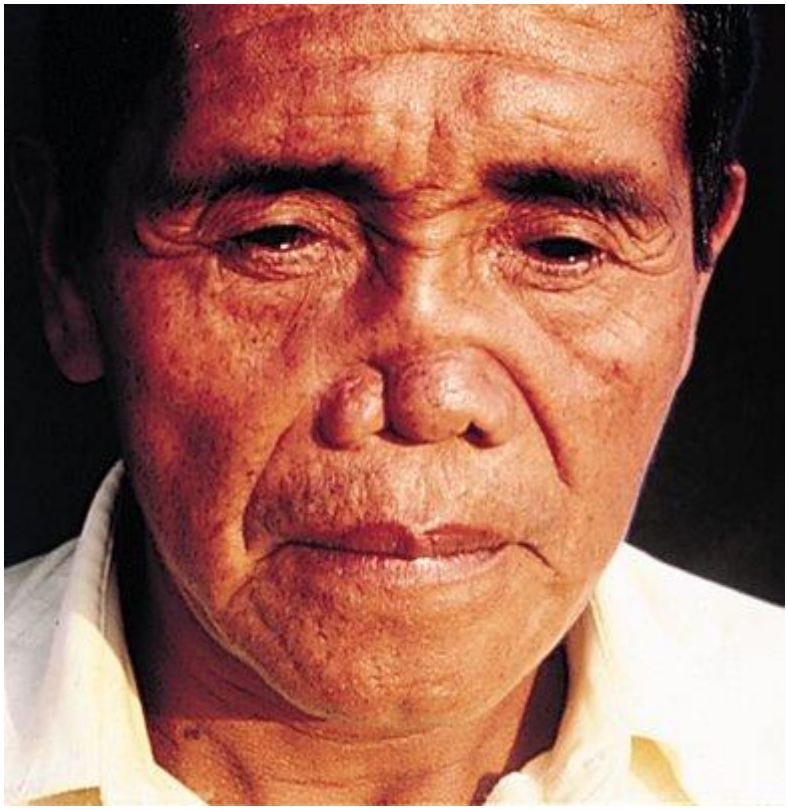
What is your diagnosis if this case appear in skin substation? It is not JVP nor Corrigan's sign! It is a thicken great auricular nerve.

Think of ***Hansen's disease (Leprosy)*** in this case. You may see this case in Malaysia PACES center.

The next step is to search for a skin lesion with sensory loss. Ask for contact history



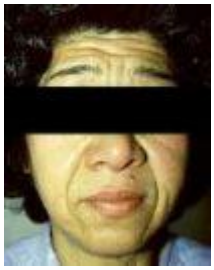
Hypopigmented macules in confluence (patches) of tuberculoid leprosy.

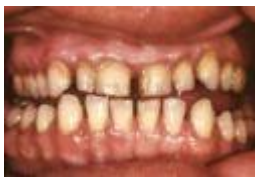


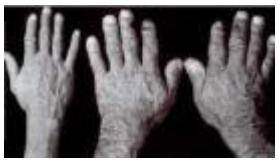
"Leonine facies" of diffuse lepromatous leprosy.

Endocrinology

Acromegaly







" please examine the patient's hands".

By one look at the patient, we are happy to conclude that the patient has acromegaly, the next thing in my mind is what the examiner want me to pick up from the patient's hands with acromegaly.

Possible things to pick up:
1) Carpal Tunnel syndrome
2) Osteoarthritis
3) Pure hand feature of acromegaly

We may easily get a 4 mark if we follow the stem, go accordingly to point out the features that support the diagnosis.
Steps to follow in order for acromegaly patient in the very moment of 2 min before we use up 3 min for presentation.
1) Hand : large spade like hand, doughy hand, sweaty palm, tinel's sign
2) Armpit: especially to look for skin tag (most candidate miss it, and we may only get 3 if we miss it)
3) Face: Prominent supraorbital ridge, prognatism, widely-space teeth, macroglossia, increase in skin creases/ wrinkles
4) Visual field assessment -bitemporal hemianopia

5) Lower limb: large foot, osteoarthritis of knee, thicken heel pad
6) complete by mentioning: checking BP, urine for glycosuria, examine for galactoria

Present as such order to make it systematic but the opening sentence must be based on the stem, in this case, "examine the hand"
Eg:
This patient has features of acromegaly. I say so because she has a large doughy, spade like hand. Her palm appeared sweaty. There is evidence of carpal tunnel syndrome as the tinel sign is positive. I found a skin tag over her right axillary area. Her voice appears to be coarse, she has prominent supraorbital ridge, and prognatism. She has macroglossia with widely-space teeth. Her foot appears to be large. Her heel pad is thickened. There is sign of osteoarthritis over her knee joint. There is no evidence of bitemporal hemianopia.

In conclusion, Mdm X has acromegaly. The most likely cause is a macroadenoma. There is sign of active disease in view of presence of sweaty palm, and skin tag. I would like to further assess her cardiovascular system.

These are the questions the examiner asked me:

1) How do you confirm the diagnosis
Failure of suppression of GH level to less than 2ng/ml after oral glucose confirm the diagnosis
2) What further test you want to do?
Localization of pituitary tumour by requesting a MRI of brain
3) What are the treatment?
Optimization of cardiovascular risk by strict BP and sugar control, refer eye for Visual field perimetry, definitive treatment: transphenoidal hypophysectomy, medical: somatostatin analogue: Octreotide, GH receptor antagonist: Pegvisomant

Further possible question:
1) What other source of GH if MRI shows no macroadenoma?
Ectopic growth hormone production, small cell CA lung
2) How do you monitor treatment response?
Insulin like growth factor

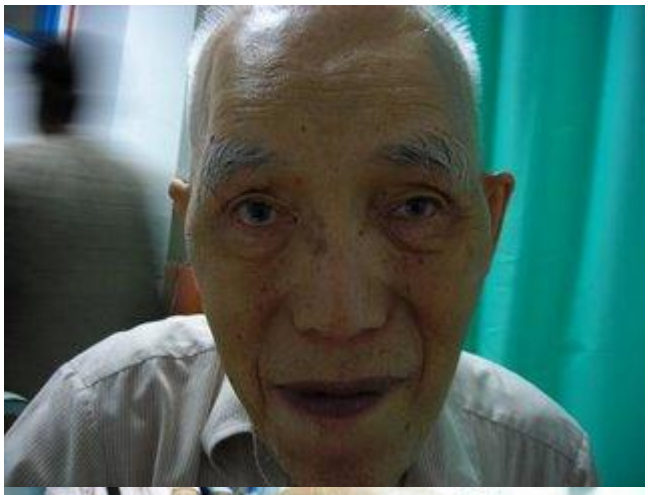
3) Do you know of any endocrine adenoma association with acromegaly?

MEN type 1 -parathyroid hyperplasia, pituitary adenoma, pancreatic tumor

Need to examine calcium level

Acromegaly

Look at this patient and proceed.



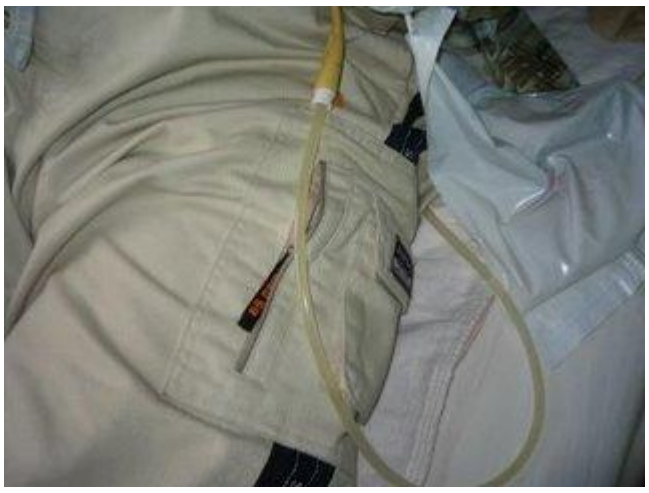
Suggested presentation:

I would be very much interested in knowing his blood pressure and I would complete

my examination by examining the visual field and dipstick his urine for glycosuria. *(Examiner: "His blood pressure is 110/80mmHg and there is no visual field abnormalities")*

This patient has acromegaly as suggested by his coarse facies and spade-like fingers. There are prominent supraorbital ridges and the nose and mouth are both relatively big with evidence of overbiting of the lower jaw. On inspection of his oral cavity, there is macroglossia with interdental separation. Tinel's sign is negative. There is no axillary skin tags and excessive sweating. Coupled with a normal blood pressure and normal visual field, I think the disease activity is currently quiscent.

NB: As demonstrated in this case, candidates should not be taken aback by information offered by the examiners midway during your presentation. Instead, you should be keen to expect responses from examiners since that information is important in formulating the complete diagnosis.



This patient has acromegaly and the disease is inactive, currently complicated by bilateral carpal tunnel syndrome and urinary tract infection

Examination of the hands reveals large spade like hand. It is however not sweaty or warm. Positive Tinel sign suggestive of bilateral carpal tunnel syndrome.
There is no proximal muscle weakness
There is prominent supraorbital ridge with prognathism and large nose and large tongue. There is also interdental separation.
There is no visual field defect and no goiter.
There is heel pad thickening

I would also like to check the BP, urine dipstick for glycosuria, organomegaly and the old photograph of this pt.

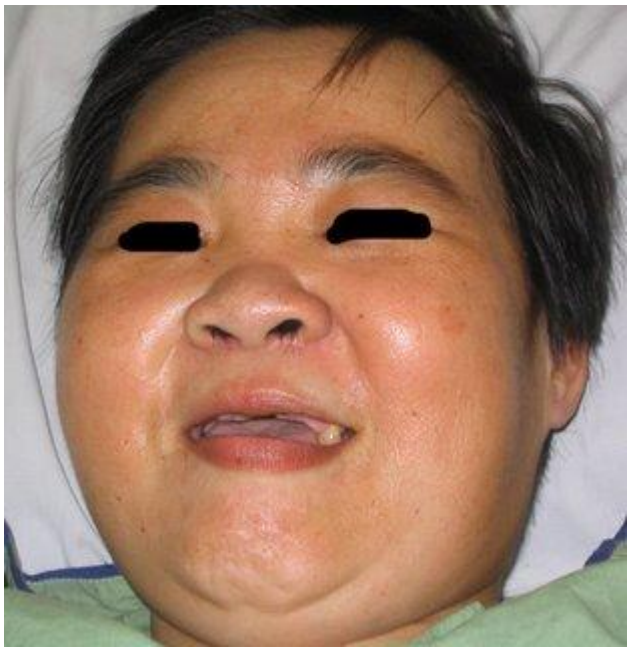
Acromegaly is a gift in endocrine substation. You should not miss the striking features of acromegaly.
**Face-prominent supraorbital ridges,large nose and tongue, increased interdental separation, prognatism.

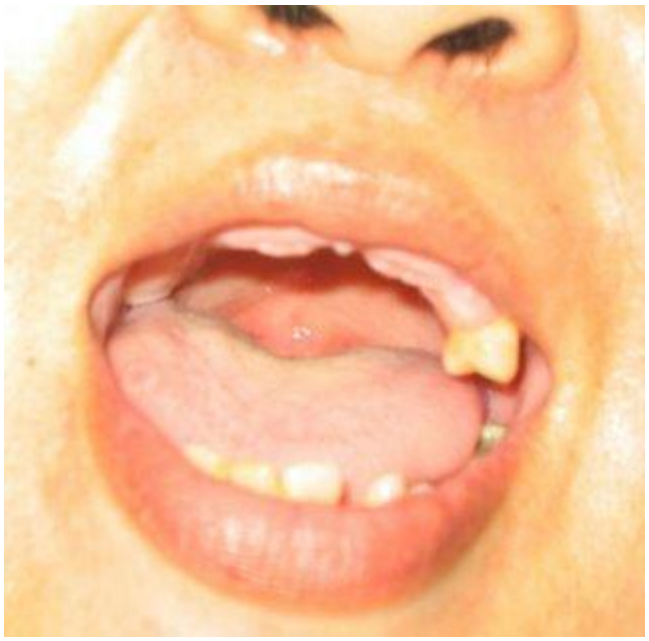
Then, proceed to look for:
1) Neck for goiter
2) Visual field for bitemporal hemianopia
3) Hand- carpal tunnel syndrome, spade-like hand, thick , doughy hand with thick hand and sweaty palms
4) Skin- look for skin tag over the axilla.
5) Leg- leg odema (may indicate heart failure)

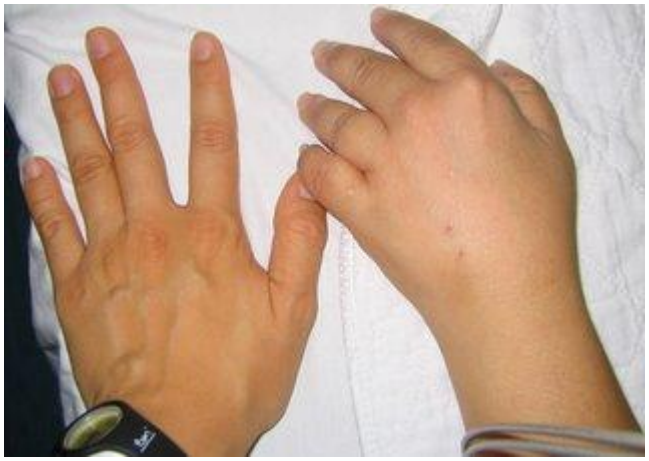
Lastly, tell examiners that, you would
1) check abdomen for hepato-splenomegaly and look for testicular atrophy
2) Check BP and urine for sugar
3) Check joint for athropathy
4) Ask for patient's old photo or IC for comparison with current facies features

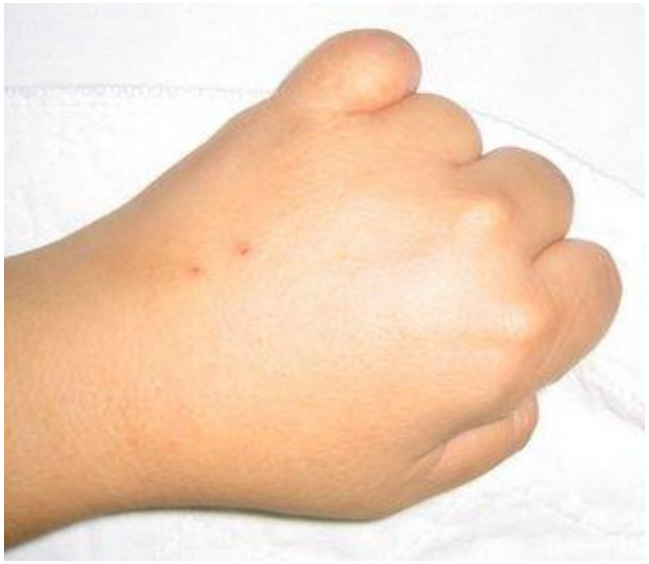
Tips: As a MRCPian, you are expected to assess the disease activity. Please pay attention to check for sweaty palm, skin tags, visual field and BP.

Pseudohypoparathyroidism









The stem

This 30year old lady was noted to be short by the polyclinic MO. Would you like to examine her?

Suggested presentation

This young woman is overweight, short, with rounded facies, missing teeth, short fingers and a short neck. She also has shortening of both fourth metacarpals.
The combination of these features suggest the diagnosis of pseudohypoparathyroidism type 1a
I would like to elicit Chvostek' and Trosseau's sign as she may have hypocalcaemia.

Questions that may be asked

What is the basic abnormality in Type Ia patients?

Ans. There is target organ resistance to the action of parathyroid hormone. The defect occurs proximal to the formation of the second messenger , cAMP

How will you treat this patient?

Acute symptomatic hypocalcaemia can be corrected with infused calcium Gluconate with cardiac monitoring to prevent hypercalcaemia. For long term treatment, use of calcium supplements and activated Vitamin D to keep calcium levels within the normal range. (Activated Vit D eg calcitriol to increase absorption of Calcium in guts and increased retention in kidneys)

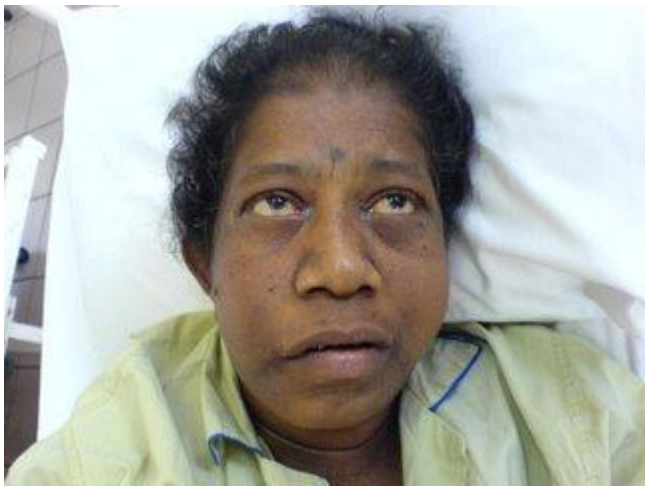
What biochemical test can you do in this patient

Ellsworth Howard Test. In a normal person, infusion of parathyroid hormone will cause a rise in urinary phosptate and cAMP excretion. In Type I A patients, there is no increase in urinary cAMP or phosphate.

What are other causes of short stature you know of?

Genetic for example familial, achondroplasia, Turners and Noonans and Downs syndrome
Nutritional or general illness during childhood eg low birth weight, congenital heart disease or renal disease

Lady with hoarse voice



Thickened and coarse facial features, perorbital puffiness

A spot diagnosis in endocrine substation in station 5.

General inspection

** Hoarse voice, response and movements are slow

** Overweight

** Myxoedematous facies as shown in the picture

Check eye for jaundice(?chronic active hepatitis)and pallor (?hemolytic anemia).

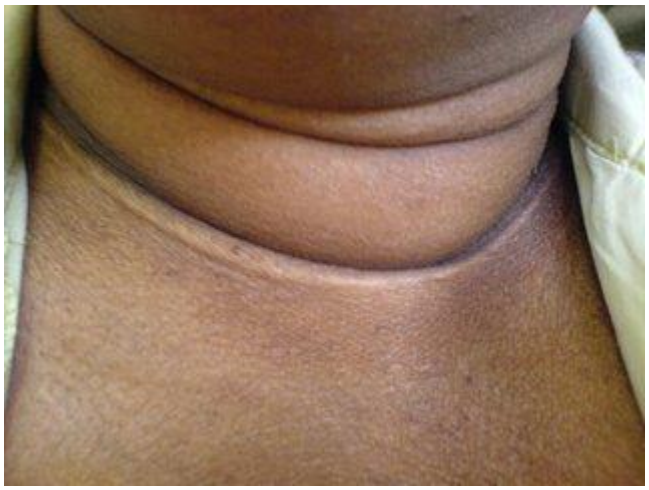
Don't forget to look for **Grave's disease eye signs**.

Rub the hair with your fingers to feel for coarse hair.

Check the mouth for macroglossia.

Check the neck for goitre and scar

Don't miss a scar in this case! In PACES, 99.9% of hypothroidism is due to previous total thyroidectomy.



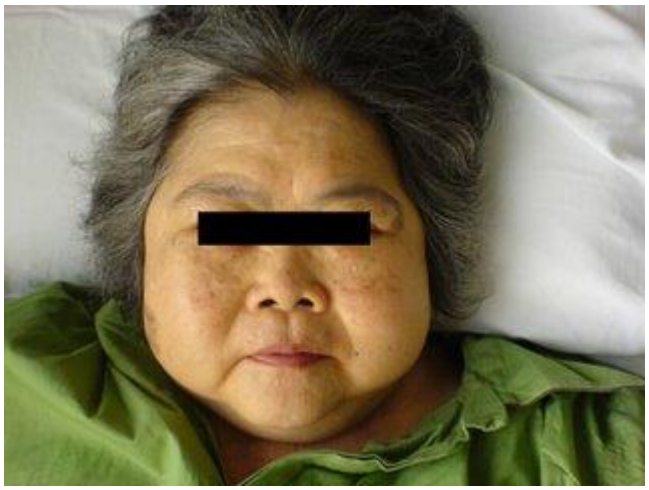
Thyroidectomy scar!

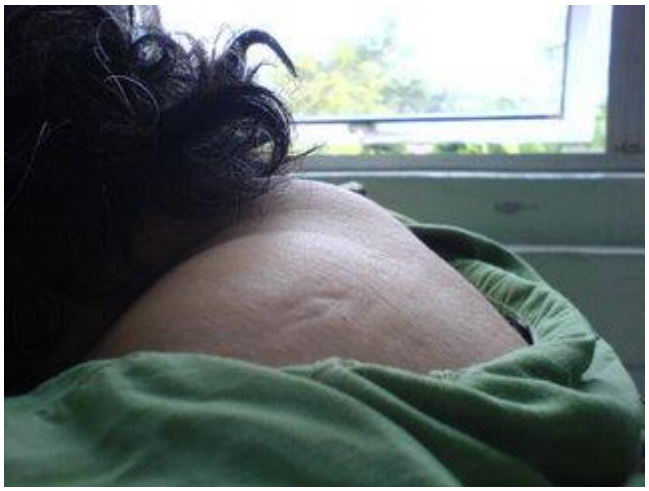
Then, **proceed to the hand** to count the pulse rate i.e. bradycardia and look for acropachy in Grave's ds.

Test for proximal myopathy over shoulder.
Check for pretibial myxoedema and pedal edema in **lower limbs**.
Lastly, position the patient in a chair to test the **ankle jerk** for slow relaxation phase.
Remember, this is the most important sign that indicate the patient is clinically hypothyroid!
End your examination by saying that you would complete it by checking CVS, Lung and cerebellar signs.

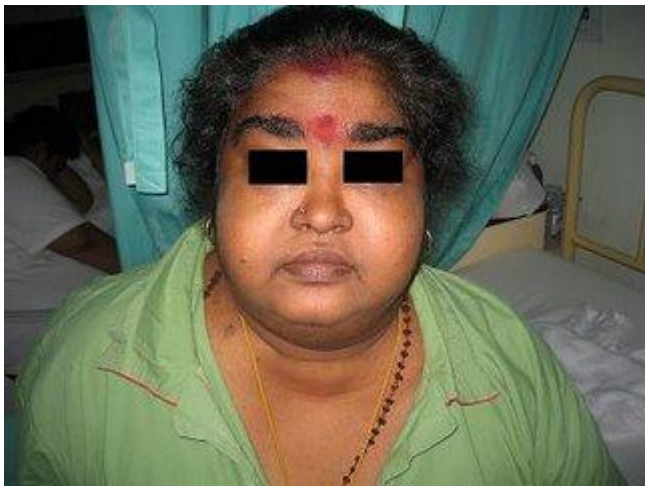
Learning points:
** Don't miss a scar in hypothyroid patient!
** Always look for Grave's ds signs in hypothroid patient. Remember, the treatment may cause hypothroidism!

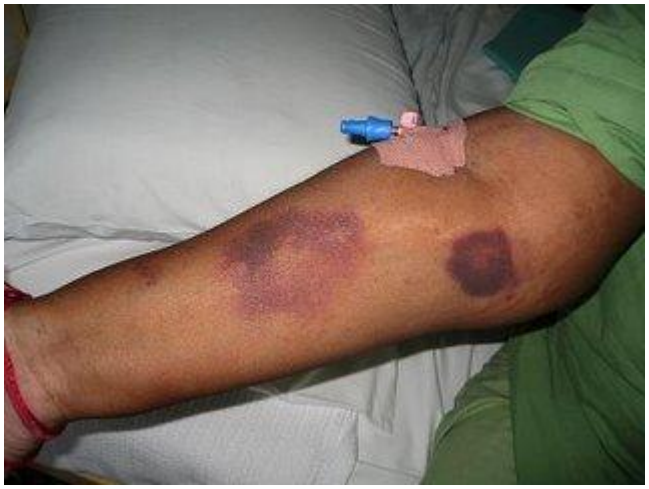
Cushing's syndrome





Cushing's syndrome is another popular spot diagnoses in PACES. The candidate is usually asked to "Look at the patient's face".





I would be keen to examine this lady's blood pressure and dipstick the urine for evidence of overt glycosuria and proteinuria. I also would like to complete my examination by checking the visual fields and performing a funduscopy to look for evidence of retinopathy and papilloedema. A history of steroid usage is of prudent importance in this patient.

This lady has Cushing's syndrome as evidenced by the presence of moon-face associated with hirsutism and acne. There are buffalo hump and truncal obesity as well. There is thinning of the skin with localised bruises especially over the cubital fossae of both arms. However there is no purplish striae noted on the abdomen and inner thighs. There is no oral thrush. Patient has proximal weakness. The back is straight with no features of kyphoscoliosis.

Questions:

1. What are the causes of Cushing's syndrome?

Outline: Pituitary-driven, adrenal and ectopic or iatrogenic sources.

2. How would you investigate to determine the cause?

I would screen the patient by doing a 24hr urinary free cortisol first and overnight dexamethasone suppression test. A high dose dexamethasone suppression test is then needed to localise the source of excess cortisol. An elevated ACTH level would isolate the source to either from pituitary or ectopic source.

3. What are the other tests to determine the source of ACTH (to distinguish primary from ectopic)?

CRH and inferior petrosal sinus sampling. An MR of the pituitary gland is helpful as well.

4. How would you manage Cushing's syndrome?

Outline: If possible, the underlying source of excess cortisol should be removed. In cases where the source cannot be removed, I would manage the complications which may arise from Cushing's syndrome.

5. What is Nelson's syndrome?

(A reminder: In real exam situation, you are unlikely to get so far. If you do, then there are 2 extreme possibilities, ie, either you score tremendously..or you failed badly!)

There are 3 main steps in dealing with this case:

First, be able to recognise all the *striking features* of Cushing's syndrome.

** Typical moon-face with plethora, hirsutism and acne.

** Truncal obesity with interscapular and supraclavicular fad pads.

** Purple striae over the abdomen, around the shoulders and breasts and thighs.

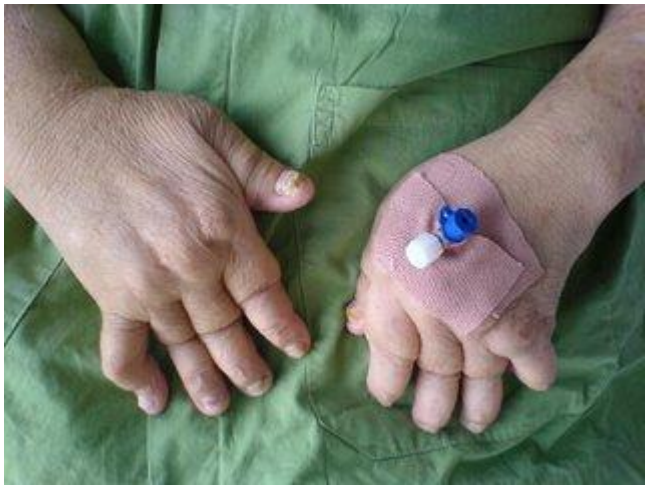
** Thin skin and easy brusing commonly found over limbs

** Proximal myopathy (shoulders & hips) and spinal tenderness (osteoporosis)

Then, you must always look out for the possible underlying diagnosis as it is not enough for a PACES candidate to get the diagnosis of Cushing’s syndrome only in MRCP.

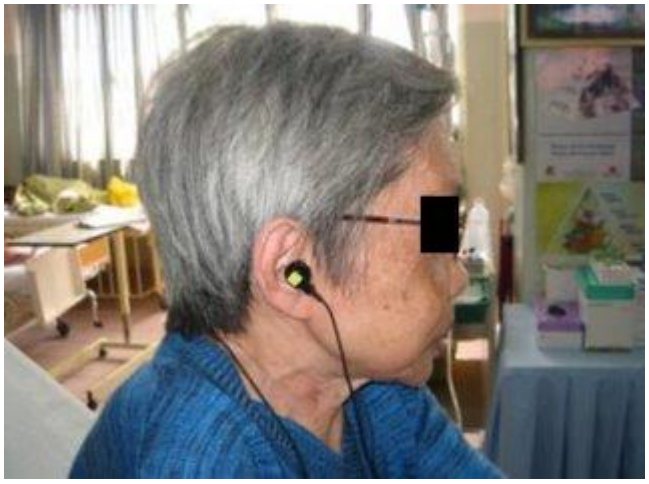
Lastly, please complete your examination by mentioning that you would like to measure the BP and test the urine for sugar.

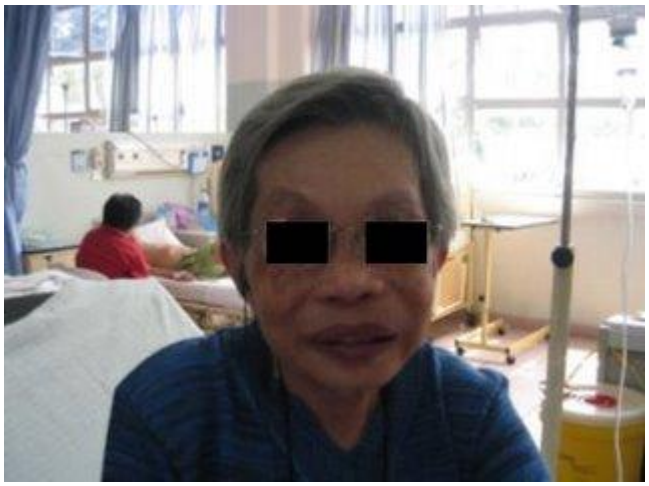
Points to remember: The commonest cause is still iatrogenic i.e. secondary to steroid. So, please look for RA hands, gouty arthritis, nephrotic syndrome and etc.

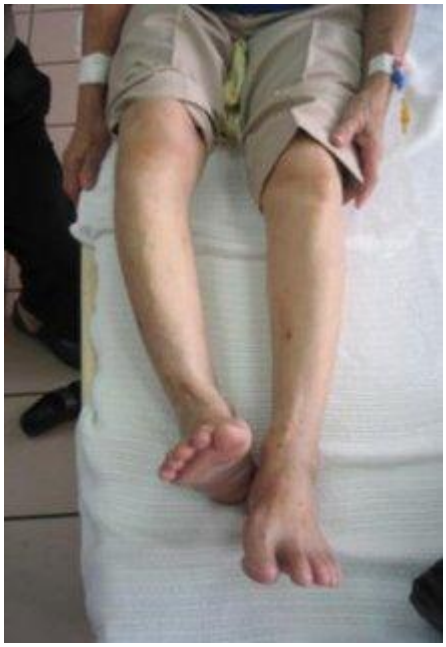


RA hands

Paget’s disease





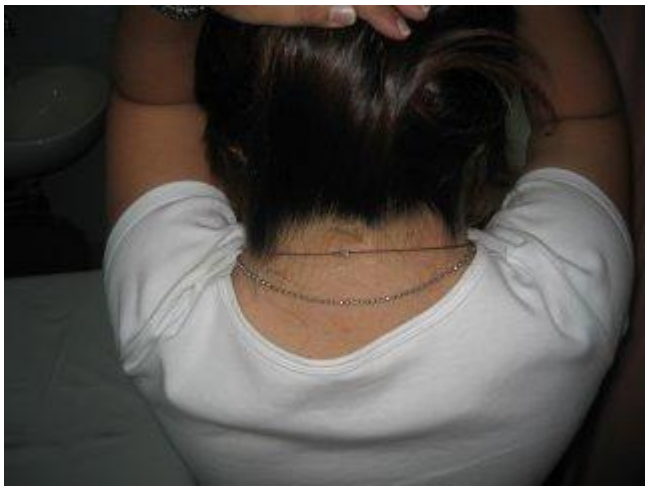


My diagnosis is that this elderly lady has Paget’s disease as evidence by enlargement

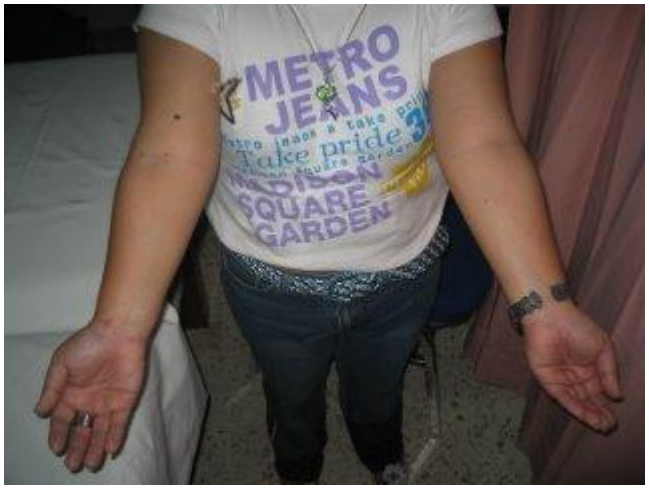
of the skull, bowing of the Right tibia which is warmer than the left one. She also has a hearing aid which suggests her condition is complicated by deafness. She is kyphotic.
I would like to look at the fundus for optic atrophy and angioid streaks, examining the CVS for heart failure. I would also do a urinalysis for evidence of hematuria (urolithiasis).
Investigations include ALP, urinary hydroxyproline which are elevated and a normal Ca and PO4.
**When is biphosphonates indicated for Paget's ds?

Turner’s syndrome

Short stature



Webbed neck and low set hairline



Wide carrying angle

Approach

Short stature

Hands

*Short 4th metacarpal

*Wide carrying angle (cubitus valgus)

Face

*Hypertelorism

*Epicanthic fold

*High arched palate

Neck

*Webbed neck

*Low hairline

Chest

*ASD scar

*Shield like chest

*Wide spaced nipple

Lymphoedema

Mental retardation is rare

Left sided heart lesions i.e. aortic stenosis , coarctation of aorta

Questions:

What renal abnormalities are associated with Turner's

Horseshoe kidney and hydronephrosis

Is pregnancy possible ?

Yes in the mosaic individuals with normal 46 XX cell line

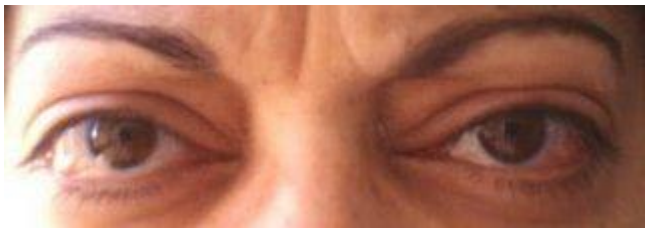
How would you manage such patients ?

Tretment is supportive and includes hormone replacement therapy on attaining puberty, primarily to prevent osteoporosis and induce sexual maturation or menses

Differential diagnosis

Noonan's syndrome

"Grave's disease"



PACES candidates should know everything about Grave's disease on their fingertips. It is very common. In fact, I got it twice for PACES exam and there was always a drink beside of the patient!

Usual themes:

1. Please examine this lady who presented with palpitation for 2/52
2. This lady presented with neck swelling. Please examine.
3. Please examine this patient's thyroid status.

My advice is to divide the examination into two objectives.
First, assess the thyroid status and signs to support Grave's disease. Second, examine the neck for diffuse goitre. The sequence of examination would depends on the theme given. For theme no. 1 & 3, I would examine the thyroid status then proceed with neck examination.

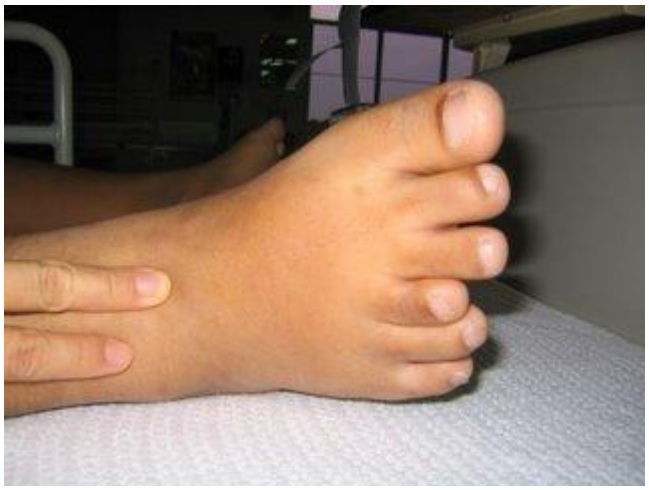
Thyroid status would include:

1. Hands: pulse, sweaty palms, tremor
2. Arms: proximal myopathy, hyperreflexia
3. Eyes: lig lag, lid retraction

Points that support Grave's disease would include:

1. Diffuse goitre
2. Thyroid acropachy, onycholysis, palmar erythema
3. Exophthalmos, chemosis, ophthalmoplegia
4. Pretibial myxoedema.

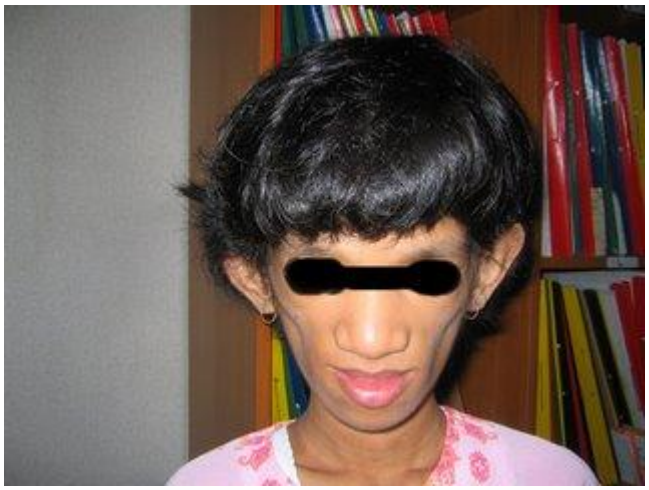
Laurence Moon Biedl syndrome

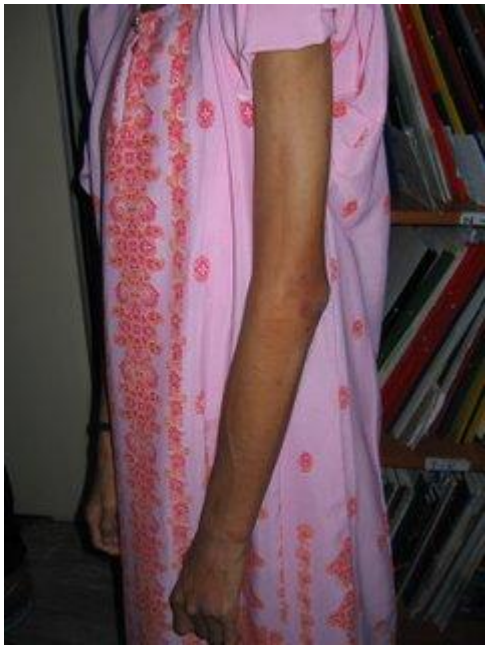


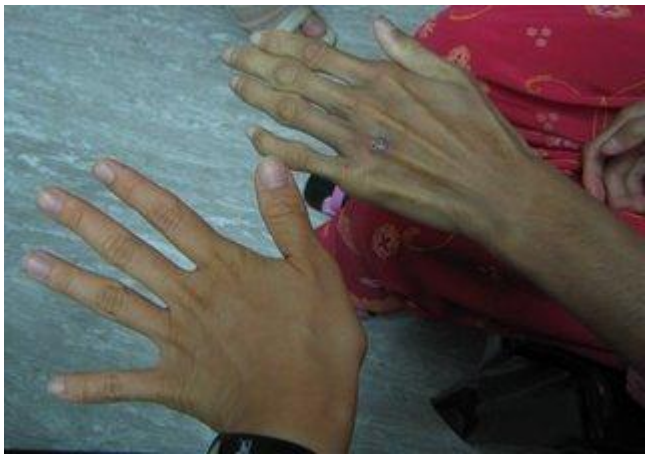
Please look at this mentally challenged young man's hand and feet and proceed.
It is a rare, recessively inherited disorder which affects approximately 1 in 100,000 babies born.

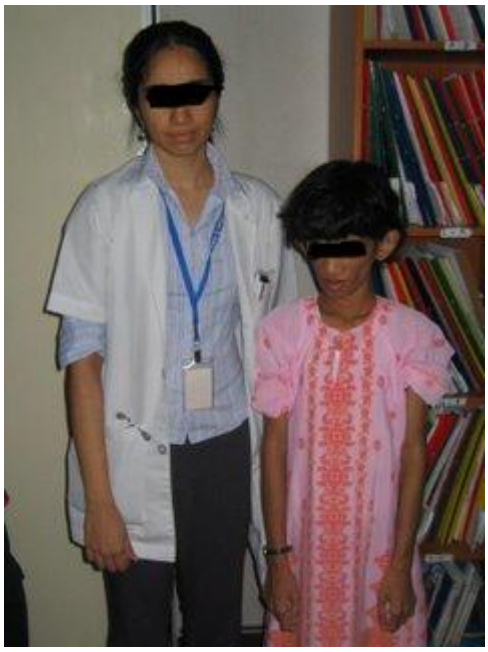
The main features include visual impairment due to retinitis pigmentosa. They are obese with extra fingers and/or toes (polydactyly) and/or partially fused digits (syndactyly). The male patient may have underdeveloped genitals. Unfortunately, they have
developmental delay, speech and co-ordination problems, and often, learning difficultie. Other health problems may occur, most frequently kidney (renal) abnormalities.

Lipodystrophy









"This 17 year old girl presented to endocrine clinic with a history of polyuria and polydypsia. Please look at her and proceed."

This young girl who is short for her age has masculine facial appearance caused by loss of fat in the face. She has prominent muscles and inframazillary dimples.She also has loss of fat (fat atrophy) in the arms and hands, giving her a muscular appearance.

The diagnosis is **partial lipodystrophy**.

As the opening statement says she has polyuria and polydyspia, I would like to do a urine dipstick for sugar as this condition is associated with diabetes

Questions you may be asked

(1) What other conditions may be associated with lipodystrophy apart from diabetes?

**HIV on antiretroviral therapy

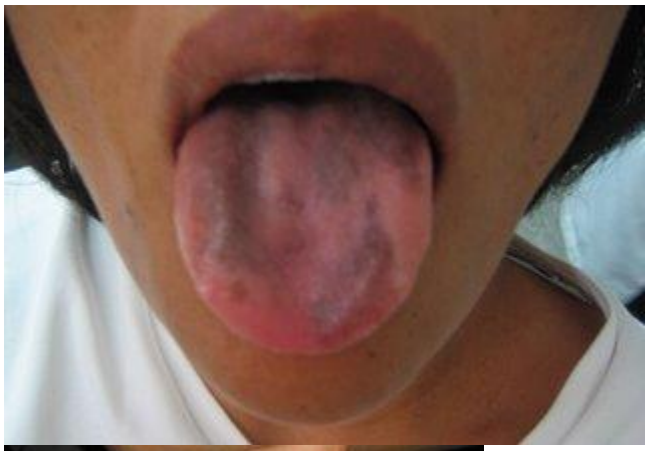
**Mesangiocapillary glomerulonephritis

**Localized scleorderma

**Morphea

**Chronic replasing panniculitis

Hyperpigmentation (Addison's disease)



Possiblity of opening statments

Look this lady who presented with weakness, loss of appetite and weight loss.

Suggested examination method

Hands-look at palmar creases for hyperpigmentation (Compare with your own palm)

Mouth-look at tongue, buccal mucosa for hyperpigmentation

Elbow: pressure points hyperpigmentation

Other sites: nipples, previous scars

Look for vitiligo

Tell examiner you would like to complete examination by:

checking her blood pressure (postural hypotension)

look for sparse axillary and puvic hair

Abdomen-?scars of previous bilateral adrenalectomy
Ask to check for visual field defect if you suspect Nelson's Syndrome.

Remember Nelson's syndrome is a enlarging pituitary tumour caused by previous bilateral adrenalectomy (done in the past for treatment of Cushing's Disease. Hyperpigmentation caused by high ACTH levels.)

Suggested presentation

"This young lady has generalized hyperpigmentation, which is more marked in the tongue and the palmar creases. The history suggest that she has Addison's disease. "

Questions you may be asked

(1) What are the causes of Addison's disease?

Idiopathic (80%)
TB (important to remember if you are taking exam in Singapore/Malaysia)
Metastasis
HIV infection

(2) How will you investigate this patient?

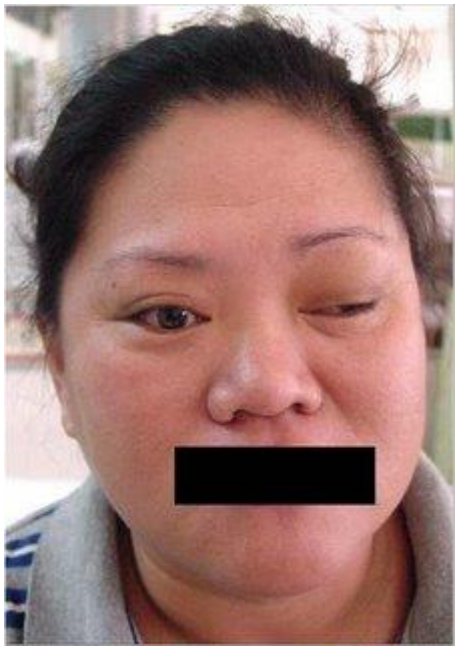
Biochemical tests to look for hypoNa, hyperK, metabolic acidosis
Blood glucose to look for hypoglycemia
I will do a screening test using the synacthen test to confirm the diagnosis.
I will also measure ACTH levels and cortisol levels.

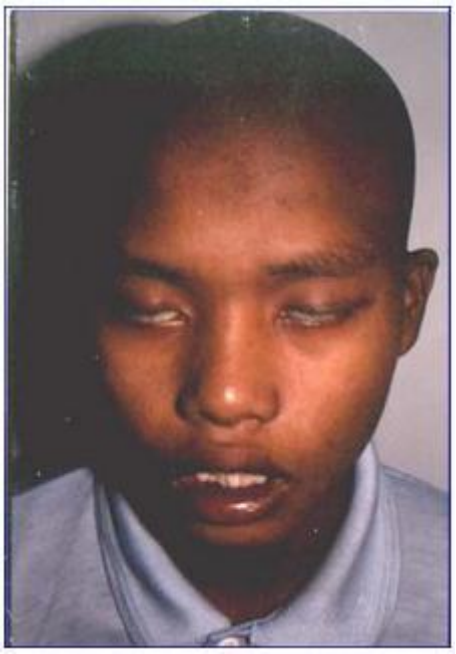
Suggested reading for this topic

Polyglandular syndrome type I and II
Interpreting the synacthen tests.

Neurology

Bilateral LMN 7th CN palsy



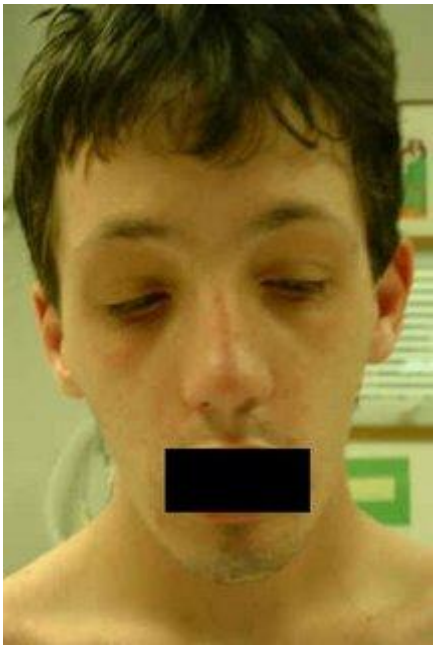


The complete 3rd nerve palsy syndrome:
1) Complete ptosis
2) The eye looks down and out
3) Dilated pupils unreactive to direct light and to accommodation

The 3rd feature is important in differentiating surgical vs medical lesions.

a. **SURGICAL lesions**
   Aneurysms, trauma, & uncal herniation involves the pupil by compression.

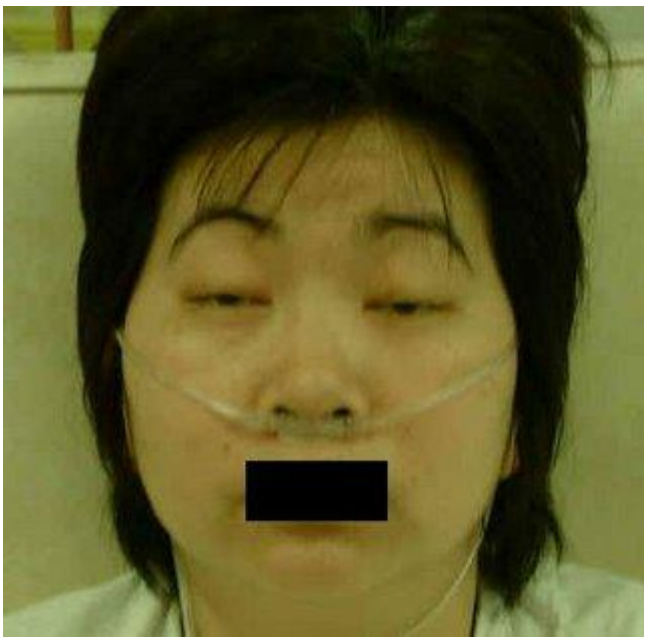
   b. MEDICAL lesions
   Hypertension, diabetes usually spare the pupil because the microangiopathy associated with these medical lesions involves the vasa nervorum, causing neural infarction of the trunk of the nerve, but sparing the superficial pupillary fibres.



Myopathic facies

Ptosis with normal pupils is worth for a special discussion. What is the differential diagnosis? The clue starts from the moment you shake the patient's hand!

1. Myotonic dystrophy
* Bilateral ptosis
* ***Myotonia with hatchet look***
**** Wasting*** of temporalis, facial muscles, masseter, sternomastoids



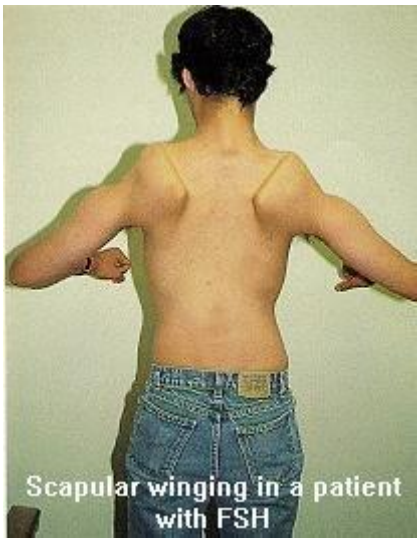
MG patient in respiratory distress

2. Myasthenia gravis
* Unilateral/bilateral ptosis
* ***Fatiguibility***

***** Variable strabismus/diplopia
* Bulbar symtoms (though not all)

3. Facioscapulohumeral dystrophy

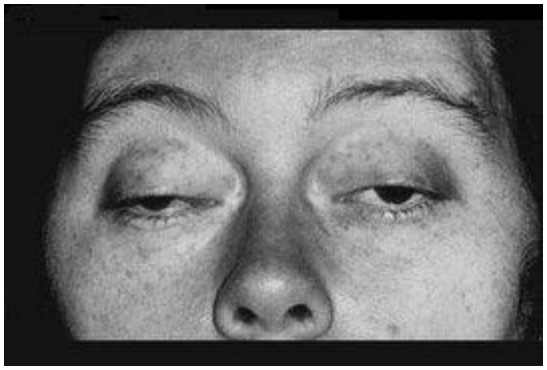
* Bilateral ptosis
* ***Wasting*** of facial muscles,sternomastoids and limb-girdle muscles.
* ***Winging of scapula***
* ***Foot drop*** - classical case has weakness of anterior tibialis

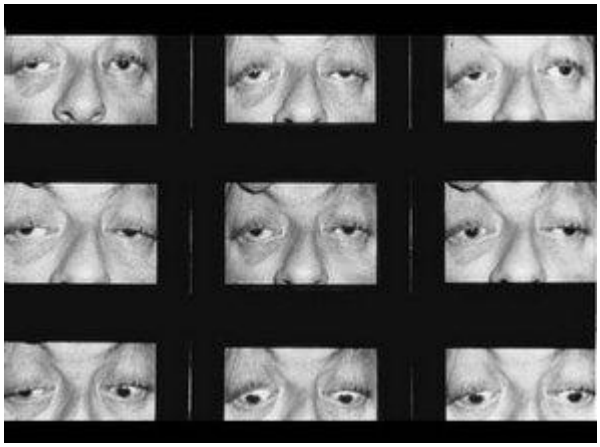


Foot drop

4. Chronic Progressive External Ophthalmoplegia(Kearns-Sayre Syndrome)

* Bilateral ptosis
* ***Symmetric*** ophthalmoplegia
* Look for retinitis pigmentosa





Bilateral ptosis with severely decreased motility in all directions of gaze

5. Oculopharyngeal muscular dystrophy
*Scenario may suggest history of ***dysphagia***
*Extraocular weakness
****Distal*** limb weakness

Horner's syndrome

Oculosympathetic pathway involved in Horner's syndrome. This 3-neurone pathway projects from the hypothalamus to the spinal cord i.e. T1, then to the superior cervical ganglion and finally to the pupil, the levator palperbrae and the sweat glands of the face.

A lesion at any site along the pathway can produce Horner's syndrome

1st-order neuron

- Arnold-Chiari malformation
- CVA/intrapontine haemorrhage

- Basal meningitis (syphilis)
- Intrinsic tumour - glioma
- Syringobulbia/ syringomyelia

2nd-order neuron

- Pancoast tumour
- Occlusion/dissection
- Tumour
- Trauma – surgical,birth
- Thyroid malignancy

3rd-order neuron

- Tumour
- Granuloma
- Herpes zoster
- NPC

Clinical features:

1. Partial ptosis
2. Miosis
3. Lack of sweating (stroke the face with pen or dorsum of your hand)
4. Enopthalmos (don't need to mention i.e. too subtle to observe)

Approach of examination

1. Confirm the diagnosis of Horner's syndrome
2. Search for the cause

Look for neck scar, wasting of muscles of hand

Examine

Neurological - lateral medullary syndrome (common)

Respiratory – Pancoast tumours (common)

Neck - lympadenopathy (mitotic lesion e.g. NPC)

1. Look at the level of the eyelid, look for asymmetry

- Unilateral/bilateral
- Partial/complete

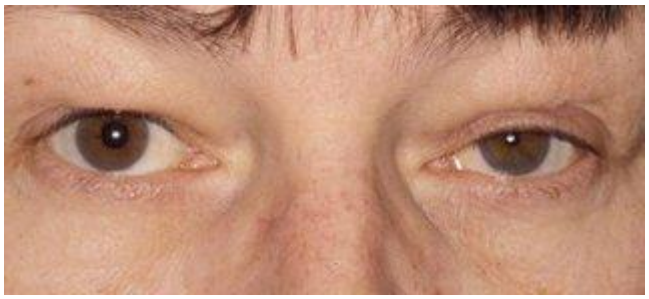
2. Then, proceed to check the pupil, look for anisocoria (unequal pupil)

- dilated in 3rd nerve palsy
- constricted in Horner's syndrome
- normal size pupil in MG, CPEO, dystrophy myotonica

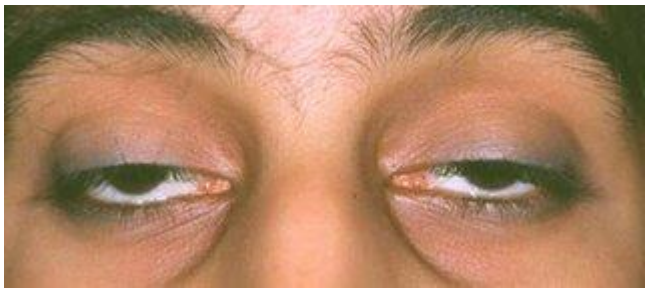
3. Look for eye position, any strabismus

- looking downward and outward in 3rd nerve palsy
- variable strabismus in MG

Right 3CN palsy

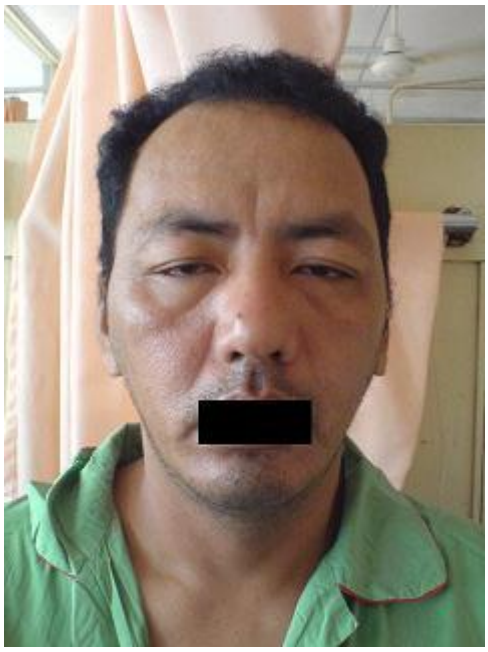


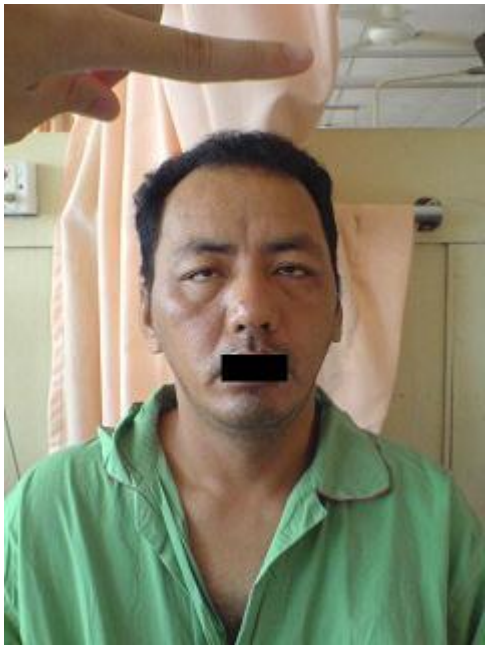
Left Horner's syndrome

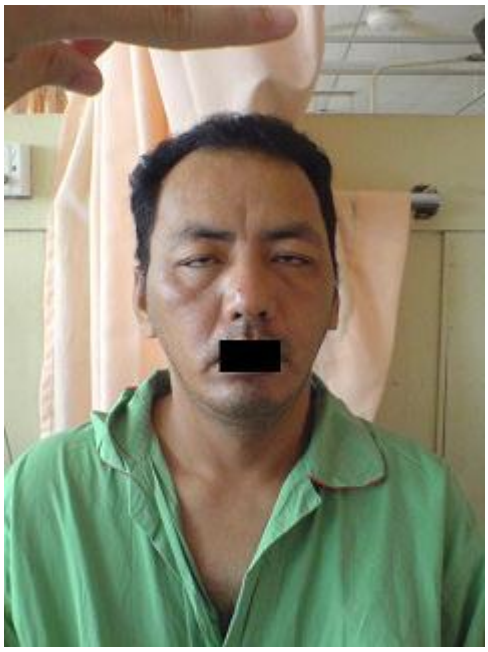


Myasthenia Gravis with bilateral partial ptosis

30 year old gentleman was told by his friends that he has abnormal eyes. Please examine the eyes.







What's the Diagnosis?

PTOSIS

*An unequal bilateral ptosis could be easily mistaken to be a unilateral ptosis.

Inspect

Dystrophia myotonica look (suspect during hand shake)

Eye looking downward and outward

Look at the pupils

-**dilated** in surgical 3rd nerve palsy

i. isolated: PCOM aneurysm(check hemiplegia, cerebellar, extrapyramidal signs)

ii. III, IV, V1, VI CN–superior orbital fissure synd(II involved)/cavernous sinus thrombosis

-**constricted** in Horner's syndrome (common: Pancoast tu, LMS)

i. V, IX, X (LMS)

ii. Neck (inspect for scars, thyroid; palpate for LN; auscultate for carotid aneurysm

iii. Percuss Upper zone- Pancoast

iv. UL - small muscle wasting, pronator drift (MS, syringomyelia)

sensation

cerebellar

-**normal** size pupil in **MG**, CPEO, dystrophy myotonica, Miller Fisher, medical 3rd nerve(DM, H/T, vasculitis)

i. Eyes

1. Fatigability- upward gaze
2. VARIABLE Strabismus and diplopia in MG(no particular CN opthalmoplegia). Medical 3rd nerve palsy?

ii. Face
1. V CN
2. VII CN
3. Bulbar (count 1-50)

iii. Muscle
1. Neck Flexion
2. Proximal myopathy
3. Reflex (ELS, Miller Fisher)

Look for
1) Treatment given: median sternotomy , subclavian line
2) Complication: tracheostomy, gastrostomy
3) Fundus (CPEO)
4) Ass A/I dis

Diagnosis of Myasthenia gravis:
FVC, Edrophonium test, Anti Acetylcholine receptor Ab, EMG, CXR, CT mediastinum
Treatment:
Long acting cholinesterase inhibitor
Thymectomy

Disociated sensory loss:

Damage to *either pathway in isolation* results in dissociated sensory loss. The damage usually occurs in the spinal cord.

1. Causes of spinotahlamic loss

- Syringomyelia
- Brown-Sequard syndrome - contralateral leg
- Lateral medullary syndrome - contralateral to other signs
- Anterior spinal artery thrombosis

2. Causes of doral column loss

- Subacute degeneration of the spinal cord
- Multiple sclerosis
- Brown-Sequard syndrome - the ipsilateral leg
- Tabes dorsalis
- Spinocerebellar degeneration e.g. Friedreich's ataxia

Young lady with walking stick (spinocerebellar ataxia)

Possibilities outlined in my mind:

1. Cerebellar syndrome - young lady with walking stick, must consider gait abnormalities. Differentials to consider: FA, SCA, MS, ?Wilson and other infiltrative disorders
2. Bulbar or pseudobulbar palsy - Differentials: MND, ?young stroke
3. Parkinsonism - unlikely PD in young lady. Differentials: drug use (neuroleptics), ?substance abuse

I proceeded to talk to the patient. To be honest, it didn't strike me to be of anything abnormal for a moment. Luckily, I stayed with the locals for 3/52 prior to the exam and hence could appreciate the subtle slurring and staccato'ing. It wasn't very clear even with phrases like "British Constituition" or "Hippopotamus"!

At this jucture, the examiners interrupted me: "So what do you think?"

I gathered my thoughts and quickly said: "I've noticed very subtle scanning speech and staccato speech. I would like to proceed to examine the cerebellar system fully."
"Please,do."
I proceeded to elicit every cerebellar sign that I could. The lady had obvious past-pointing, more on the right side and dysdiadochokinesia. However, truncal stability was relatively preserved. There was sustained nystagmus on looking to the right. She displayed broad-based gait with failure to perform tandem walk. The Rhomberg's sign was negative.

I was happy to conclude that she had ample of signs to suggest a cerebellar syndrome, with the possible underlying etiologies being MS (young lady and in temperate region), and hereditary ataxias such as SCA or FA. I would complete the examination by looking into her fundi for optic atrophy as well as to examine the lower limbs for long tract signs.

"If you are given an opportunity to ask one question, what would you ask?"

"I would elicit history of similar illness running in the family."

"Please do."

I proceeded to ask the nice lady with the question and I found out that her mother and the elder brother had the same condition.

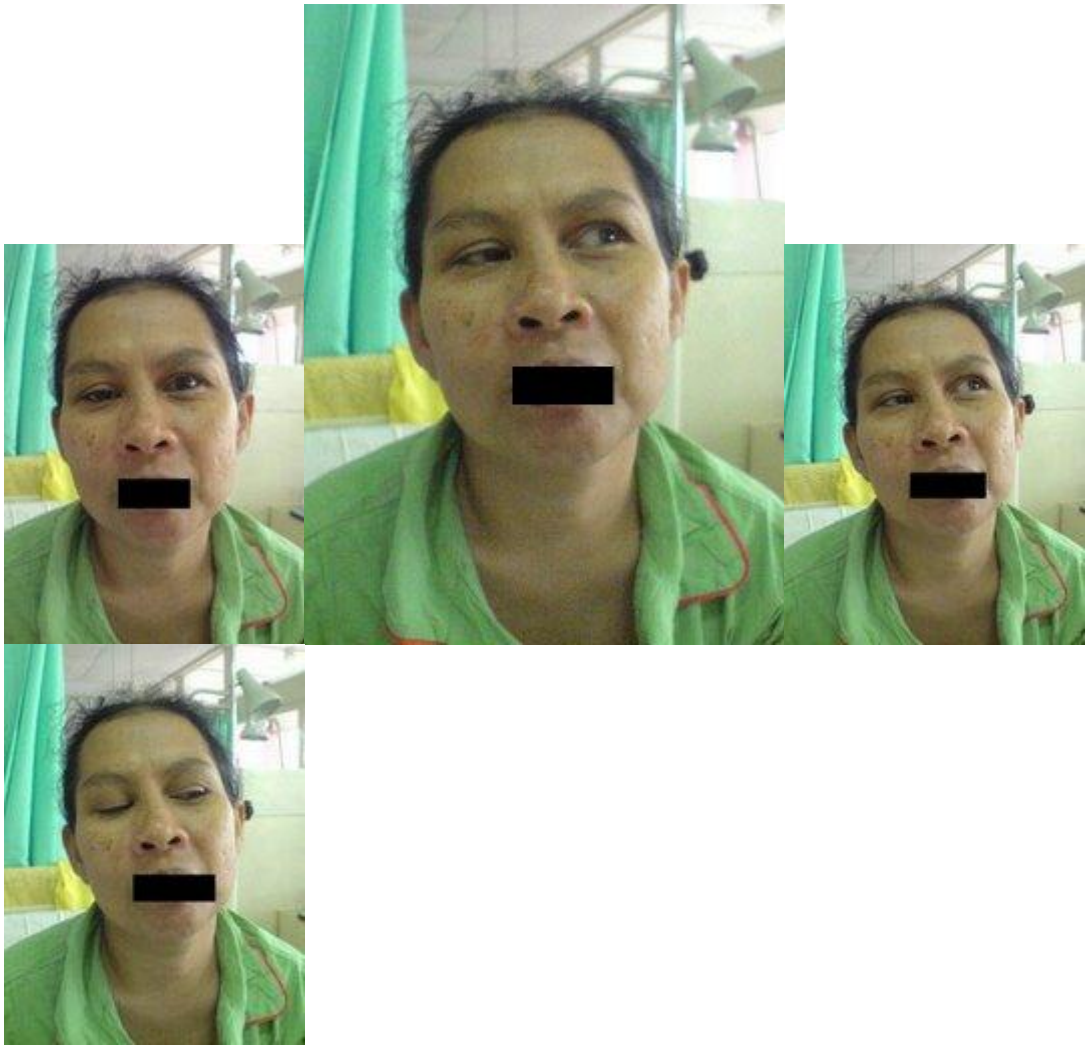
"So what do you think?"

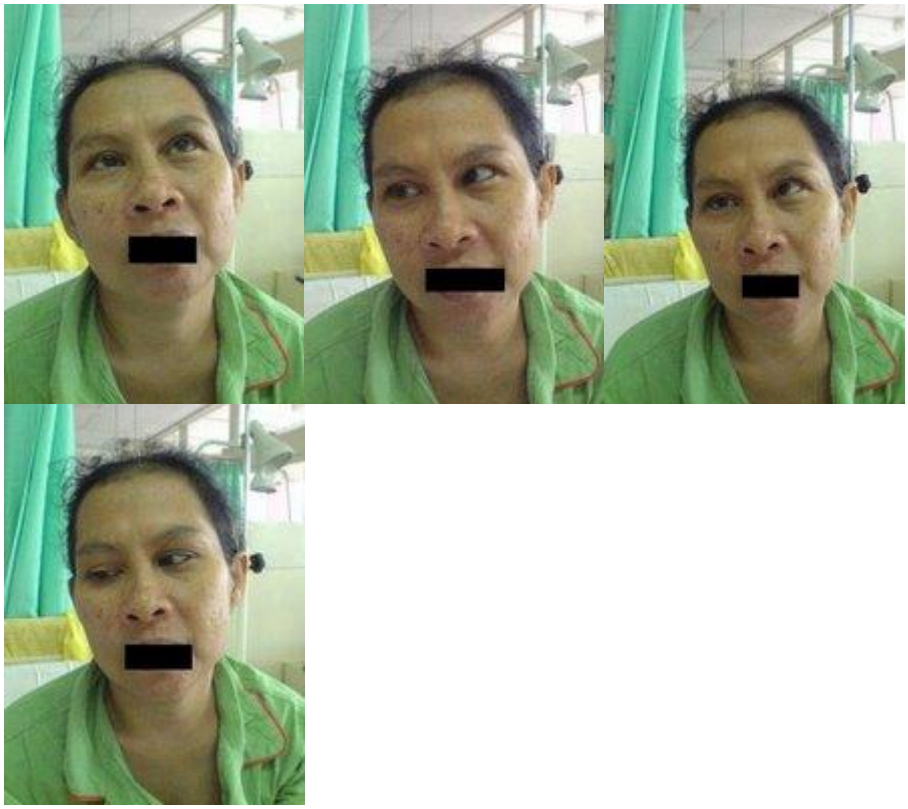
"The inheritance pattern is likely of autosomal dominant type, and hence it cannot be Friedrich ataxia. I would think this patient has **spinocerebellar ataxia** of autosomal dominant type."

The examiners nodded and smiled: They tossed me a few more questions on the other possible causes of cerebellar signs in a young lady before concluding the station.

Ophthalmoplegia

Puan Rahimah complains of giddiness. Please look at her and proceed.





Examiner: Dr. Pan, please present your findings.

Puan Rahimah's head is rotated to the right. She has right eye ptosis. There is no strabismus at rest. Both her pupils are spared. There is no fatigability. There is mild impairment of upwards and upwards lateral movements of the right eye. Intorsion is intact. She has diplopia upon looking at all direction except looking downwards . The outer image disappears upon closing the right eye. There is no nystagmus. The rest of the cranial nerves are intact.

I would like to complete my examination by examining the patient's BP (H/T), urine for sugar (DM), upper limbs and lower limbs for any pyramidal signs including the reflexes (CPEO, ELS) and cerebellar sign. I would also like to look into the fundus to look for any papilloedema. (/retinitis pigmentosa)

In summary, Puan Rahimah has complex diplopia and I would like to consider differential diagnosis of

1) Mononeuritis multiplex
2) MG (no fatigability)

3) Miller Fisher
4) CPEO, Ocular muscular dystrophy
5) Grave's (proptosis?)
6) Cavernous sinus thrombosis/ superior orbital fissure syndrome(if V CN is involved)

What other investigation would you like to do?

1) ESR to screen for vasculitis
2) MRI of the brain (infarct / SOL esp at the midbrain)

Causes of isolated III CN palsy

Medical:

1) DM/ H/T/ Vasculitis
2) Multiple sclerosis

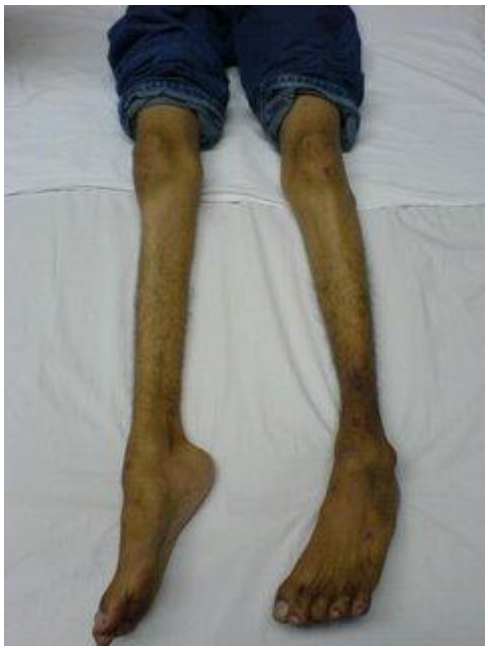
Surgical

1) Posterior communicating artery aneurysm/ Midbrain tu
2) Subacute meningitis

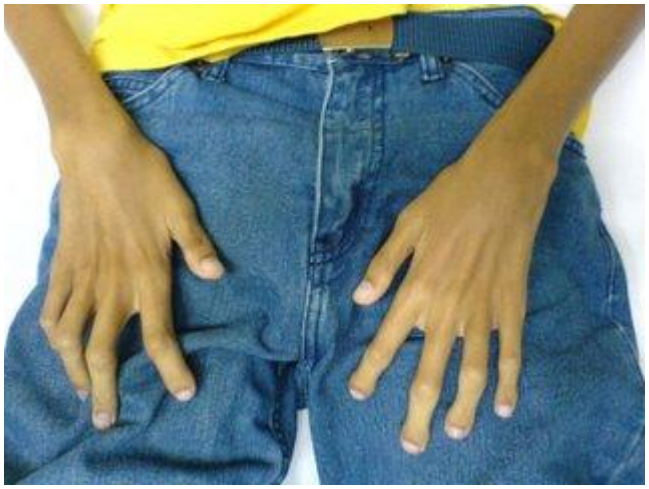
Causes of isolated VI CN palsy (look for involvement of VII CN)

1) SOL
2) DM/ H/T/ Vasculitis
3) Multiple sclerosis
4) Subacute meningitis

Charcot-Marie-Tooth disease



This patient came for an elective investigation in neurology clinic today PM.
Scenario: 20-year-old, male with difficulty in walking for 8 years. Examine the LL.
Examination reveals *LMN lesion*. There is wasting of the lower limbs muscle. There is no fasciculation and the tone is reduced. The weakness involves predominantly the distal muscles. The extensors of the toes and feet are weak. The knee and ankle jerks are absent and the plantar reflexes show no response. The sensation to pin prick and propioception are preserved.



What do you want to examine next?

a. There is wasting of the small muscles of the hands.

b. He had bilateral foot-drop and high stepping gait.

c. Ask for a family history of similar problem.
Yes, his elder brother also has the similar condition.

Discussion

The diffential diagnosis that should be considered here are:
1. Peripheral neuropathy i.e. motor neuropathy
**Hereditary as suggested by 8 years history of onset and the age of presentation at 12 yo i.e. Charcot-Marie-Tooth ds (HMSN)
**CIDP unlikely because of no sensory involvement

2. Motor neuron disease - Unlikely
** Wrong age of presentation for MND
** No signs to suggest UMN lesion (mixed UMN+LMN)
** No fasciculation (Hallmark for MND)

3. Muscular dystrophy - Unlikely
** Pseudohypertrophy but not severe wasting
** Predominantly proximal wasting

Conclusion

The diagnosis is Charcot-Marie-Tooth disease.

Further questions

What is the investigation that is planned for him in neurology clinic?

How many types of HMSN do you know?

What is the inheritance pattern for this disease?

I guess one would order a NCS to look for either velocity or the amplitude is affected. There are HMSN type 1, and 2, and the distal spinal muscular atrophy. Type 1 being the demyelinating form and type 2 axonal degeneration type

Inheritance is either autosomal dominant or recessive. Need to look into the eyes as well.

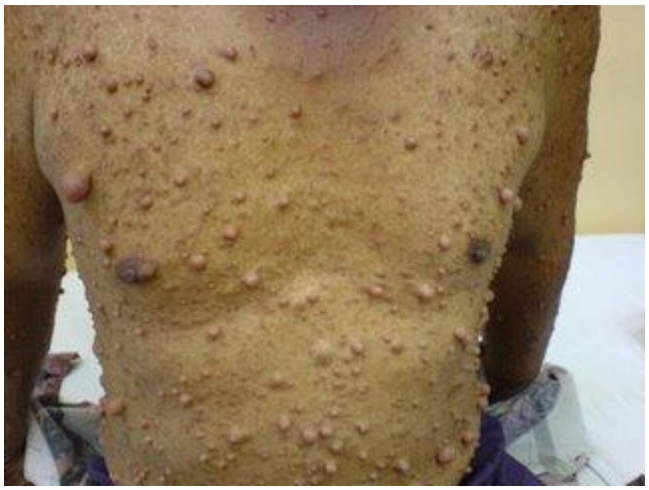
Brain Pacemaker

Deep brain stimulation (DBS) is one of a group of treatments involving surgical implantation of a medical device called a brain pacemaker, which sends electrical impulses to specific parts of the brain.
It is used for advanced Parkinson's disease resistant to medical treatments. Other indications include severe essential tremor, primary dystonia and epilepsy.
It is true that Parkinson's patients are now increasingly receiving DBS devices. So, when you see a scar at the left/right pectoral region in a parkinson patient, please think of :
1- DBS device.
2- Cardiac pacemaker for associated arrhythmia(coincidental findings).

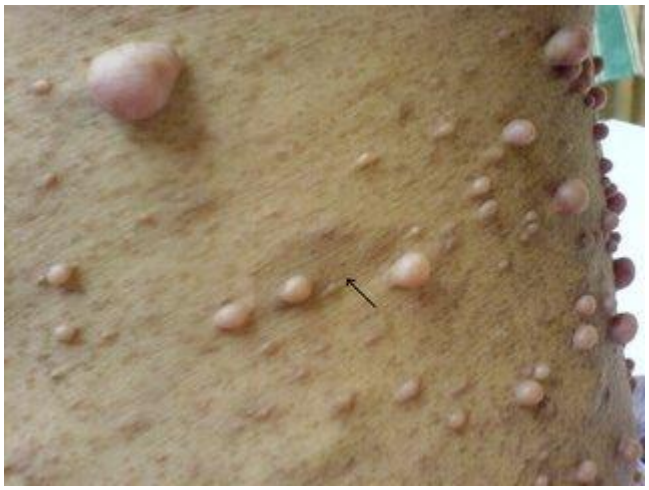
Spot diagnosis: Neurofibromatosis

One of the spot diagnosis in skin substation. As a PACES candidate, we need to demonstrate the various skin lesions (fibroma, plexiform neurofibroma, cafe-au-lait spots, axillary freckles) and also check for complications such as:

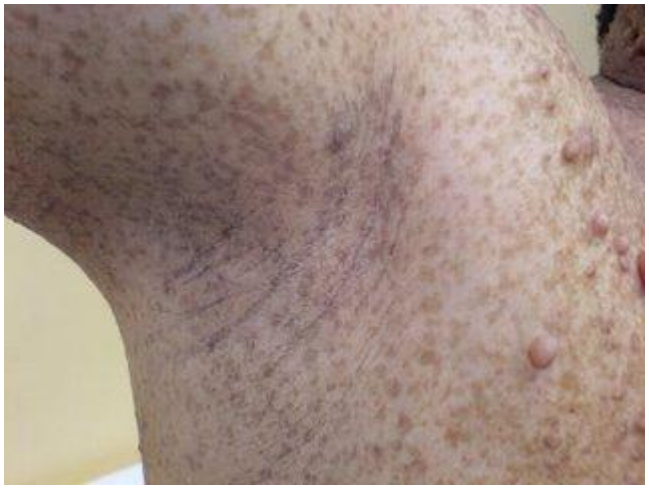
*Check hearing (acoustic neuroma)
*Visual acuity and fundi (optic glioma)
*Look for kyphoscoliosis
*Check BP for HPT in phaeochromocytoma



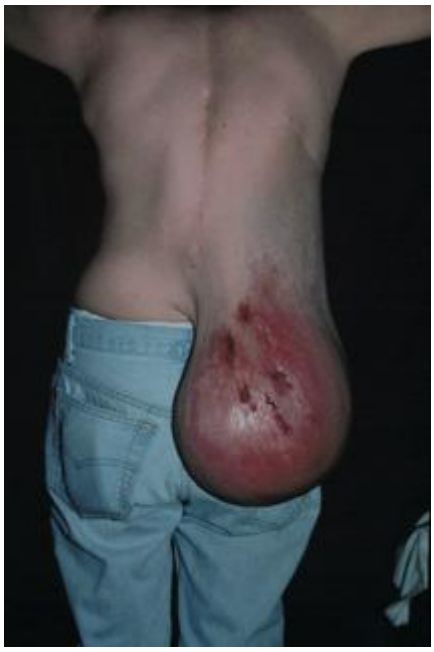
Multiple neurofibroma



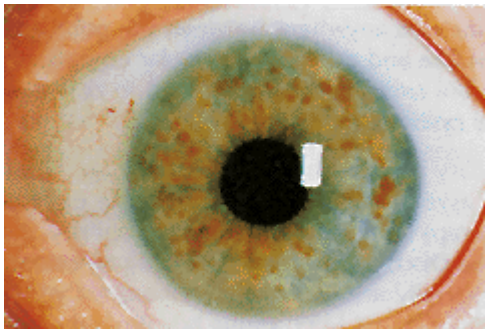
Cafe-au-lait spots



Axillary freckles



Plexiform neurofibroma



Lisch nodules in the iris

Spastic paraparesis:

It is a common case in neurology station. Most of the candidates would not have difficulties in eliciting the signs of lower limbs weakness with UMN signs such as hypertonia, hyperreflexia, ankle clonus and upgoing plantar responses. However, the struggle comes after presenting the findings i.e. formulation of differential diagnoses.

Don't panic! Just stick to the ABC of neurology by answering 2 questions.

1. Where is the lesion?

**UMN lesion signs tell you that the lesion lies above L1 as the pyramidal tracts end there.

**So, the lesion would be either at anterior horn cell, spinal cord (thoracolumbar/cervical) or cerebral level.

Anterior horn cell: fasciculations, no sensory loss, LMN signs like wasting of the small muscles of the hands, tongue fasciculations.
Spinal cord: sensory level, sphincter involvement, inversion of supinator/ biceps jerks.
Cerebral: intellectual impairment (in cerebral palsy), papilloedema (parasagittal meningioma)

2. What is the lesion?

** These depends on the scenario given in PACES. You need to correlate it to the *age and gender* of the patient and also the *onset of the disease*.

For example, a young lady with 2 weeks of weakness. You need to consider about demyelinating disorders, transverse myelitis (autoimmune eg. SLE), anterior spinal artery thrombosis (vasculitic eg SLE), infection eg abscess, trauma.

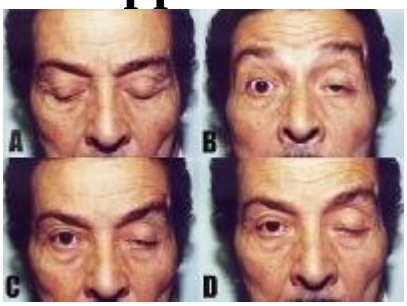
On the other hand, you should consider about the differential diagnoses of tumours, spondylosis with possible disc prolapse, chronic infection like Pott's ds for an old patient with months of weakness.

The moral of the story is you should generate a list of possible causes before facing

the music. Please remember to sing a song that match the music!

"I would like to complete my examination by checking the spine, anal tone and S2/3/4 sensation, ULs, CNs, cerebellar signs and fundoscopy."

An Approach To Bilateral Partial Ptosis (Part 1)



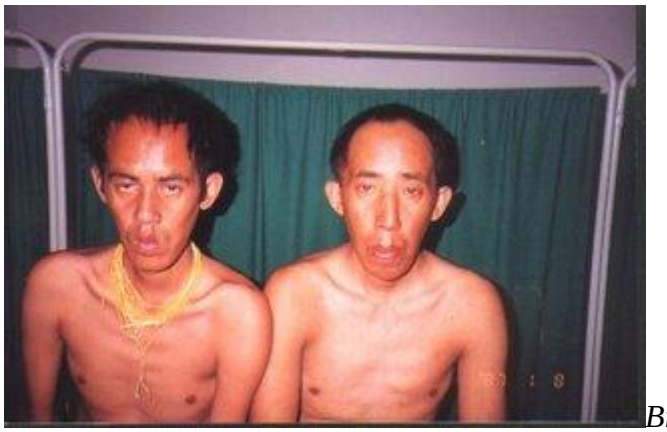
The usual stem of CNS station would be:
Examine this patient's cranial nerve.

It would be lucky if on inspection, there is obvious partial ptosis. The immediate next step would be thinking of the following 3 possibilities:

1. Is it bilateral Horner syndrome?
2. Is it bilateral 3rd CN palsy? (Rather unlikely to appear in real exam if it is due to central cause rather than peripheral)
3. Is it neuromuscular weakness?

Our Part 1 will concentrate on neuromuscular weakness
During CN examination, we have effectively ruled out Horner and CN III palsy. So, it is likely partial ptosis due to neuromuscular weakness. At this point, there should be at least 5 differential diagnosis in our mind:

1. Myasthenia Gravis/ LEMS
2. Gullain Barre Syndrome (Miller-Fisher)
3. Dystrophic Myotonia
4. Fasciomusculoscapular dystrophy
5. Oculopharyngeal Muscular Dystrophy

 *Brothers with Dystrophic Myotonia*

Therefore, at the end of CN examination, we need to do something extra to get the most likely diagnosis. Do as follow:

1. Check for fatiguability (eye lids, upper limbs, nasal speech)
2. Check for thymectomy scar (may be hidden by patient's dress, and it would be fatal for missing the scar)
3. Check for diplopia on extreme gaze
4. Check for myotonia, by percussing the thenar muscle, and ask patient to grip hands
5. Check lower limb reflex for areflexia in GBS
6. Check for winged scapula

During presentation, we need to mention about the possibility of medical emergency, in this situation, we should mention:

1. "I would like to complete my examination by requesting a bed side spirometry to assess for respiratory mucle weakness" -for suspected Myasthenia/LEMS/GBS
2. "I would like to examine upper and lower limb for ascending weakness and aflexia" -for GBS

High Stepping Gait

High steppage gait occurs due to foot drop where dorsiflexion of the foot cannot occur. The knee is flexed and raised to allow the foot to clear the ground. It occurs in L4, 5 root lesions, common peroneal lesions, peripheral neuropathy. Occasionally it may be due to lesions in the motor cortex.

EYES ARE THE WINDOW TO THE SOUL

This gentleman presented with **giddiness**. Please look and proceed

I would like to complete my Ex by checking the fundus for papilloedema or optic atrophy, corneal reflex and the gag reflex. I would also like to Ex the UL and LL for pyramidal signs and finally measuring the BP.

This gentleman has Left Internuclear Opthalmoplegia where by the Left eye is unable to adduct and the right eye has nystagmus with the fast phase towards the right side upon looking to the right. The rest of the cranial nerves are intact.

I would like to consider causes such as stroke or demyelinating disease.

Examiner : What are the other causes of INO?
Pan : Tumour, Drugs such as phenytoin and carbamazapine

Examiner : Where is the lesion?
Pan : Left Medial Longitudinal Fasiculus (connecting 3rd nerve nucleus to the opposite 6th nerve nucleus) at the brain stem

Examiner : How would you like to investigate this pt?
Pan : I would like to request for a MRI of the brain.

MRI came back as small left pontine infarct.

Gait disturbance:

The 1st part of the video clip clearly demonstrates positive Romberg's test. The patient is losing his balance upon closing his eyes. Please take note that the test is considered positive only when patient is about to fall. So, please guard the patient when perfoming this test.

The 2nd part shows *Stamping gait* - broad based, foot stamping on the floor.

Where is the lesion?
-It is seen in propioceptive disorders. The lesion is in the peripheral nerve or posterior column.

What is the lesion?
1.Peripheral neuropathy - DM,Alcoholism, Vit B12 deficiency etc
2.Tabes dorsalis
3.Subacute combined degeneration of the spinal cord (SACD)
4.Friedreich's ataxia

Dizziness:

The case mentioned is a 45-year-old Chinese man, working as a mechanic. I remembered he kept on complaining of dizziness from the moment he stepped into my clinic. To cut the story short, he had been having severe dizziness and imbalance gait for nearly 2 years. It was progressively worsen for the past 3 months before he sought treatment from medical doctor. Functionally, he was terribly disabled by those symptoms.

On examination, he was ataxic and hardly able to stand or walk steadily without any assistance. There was bilateral cerebellar signs as evidenced by presence of dysmetria, intentional tremor and failure of performing rapid alternate motion test.

There was obvious nystagmus demonstrated bilaterally. He had staccato speech. Further examination revealed that the lower limbs were hypertonic and hyperreflexic. Clonus was present with bilateral upgoing plantar responses. His sensation of pain and propioception was intact.

In conclusion, he has bilateral cerebellar signs with UMN signs to suggest corticospinal tract involvement.

The possible diagnosis that should be considered in this case:

1. Spinocerebellar ataxia (SCA)
2. Multiple sclerosis

How should you proceed to reach the diagnosis?
What is the investigation of choice at this stage?

Wallenberg's Syndrome

This Malay gentleman presented with 2 days history of dizziness. He cannot walk steadily without assistance.

Examination of the LL shows hemisensory loss up to neck region over RIGHT side. The muscle power is preserved. The propioception is intact. He is ataxic and there is tendency to fall to LEFT side. The heel shin test is impaired over LEFT side.

Further examination shows LEFT facial numbness with intact corneal reflex. There is signs consistent with LEFT Horner's syndrome i.e. partial ptosis and constricted pupil. The 9th and 10th CNs are intact. The eye movement is normal and there is no diplopia.

He has a LEFT lateral medullary syndrome.

Questions:

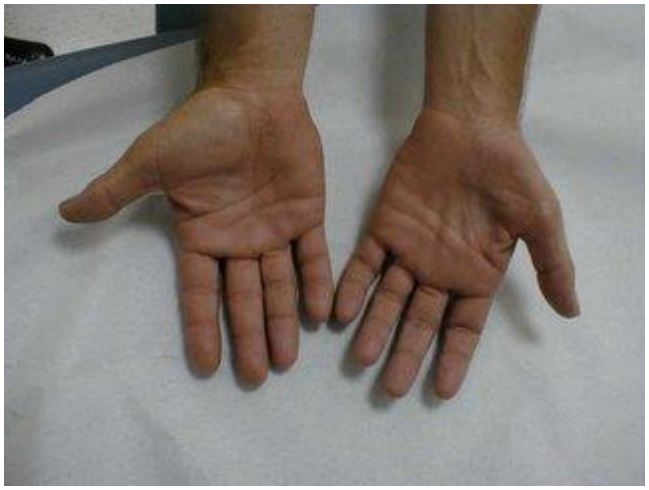
1. Where is the lesion?
2. What are the other clinical features?

Ipsilateral	Contralateral
Facial numbness (Vth) Diplopia (VIth) Nystagmus Ataxia (cerebellar) Horner's syndrome Ninth & tenth nerve lesions	Spinothalamic sensory loss Hemiparesis (mild, unusual)

Wasting Of The Small Muscles Of The Hands

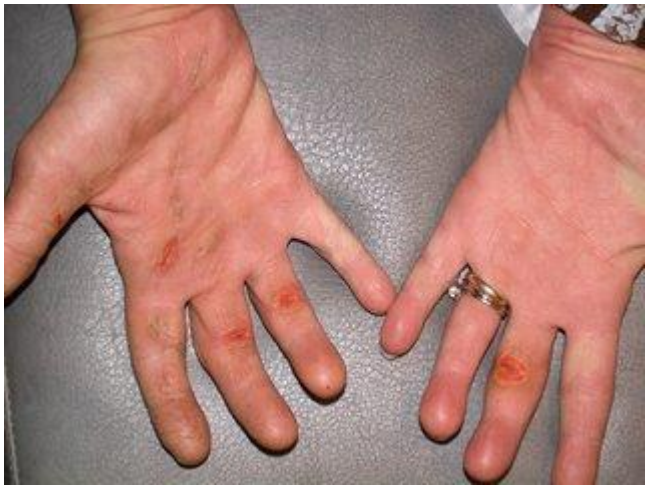
Examination of hands is very common in PACES setting. You may encounter the hands in locomotor substation or neurology station. There are not many differentials that you need to memorize but you need to have a mind mapping about the approach especially in neurology station.

1 st case is a 40 years old man with insidious distal weakness for > 12 years. 3 out of his 8 siblings are affected. He has wasting of small muscles of the hands resulting in guttering of the dorsum of the hands. There is generalized areflexia and glove pattern of sensory loss. He came for nerve conduction test which show reduced conduction velocity. The diagnosis is **Charcot-Marie-Tooth disease**.



2nd case is a middle aged man who had progressive weakness of hands for 10 months. He has wasting and weakness of small muscles of the hands. The hypothenar and

thenar eminences are wasted with left more severe than right. There is fasciculation seen and the reflexes are brisk. The sensation is normal. MRI cervical spine and NCS are normal. The diagnosis is **motor neuron disease (MND).**



3rd case is a 30-year-old lady with a **syringomyela** developed painless ulcers of the palms and fingers. Other possible skin signs are edema and hyperhydrosis due to interruption of central autonomic pathways.

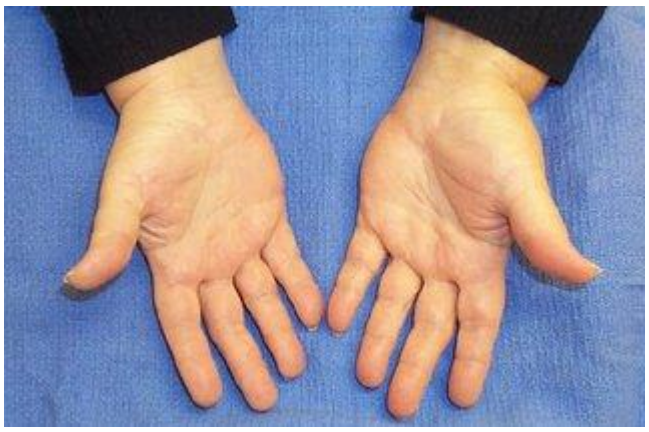
Confuse? Don't get panic. Just remember several points:

1. We should localize the lesion as LMN i.e. the lesion is at C8-T1 level including the cord, anterior horn cell, nerve roots and peripheral nerves.
2. NMJ is not included because it does not cause wasting. Muscular disorder is not included because it does not usually cause distal weakness.
3. Sensory examination is *VERY* important in localizing the lesion. For example, if there is sensory loss then **MND** is excluded from your differential lists.

Unilateral sensory loss: combined median & ulnar nerve palsy, C8-T1 nerve roots or brachial plexus lesion.

Bilateral sensory loss: glove (PN), C8-T1 cord lesion, dissociated sensory loss (syringomyelia).

Carpal tunnel syndrome:



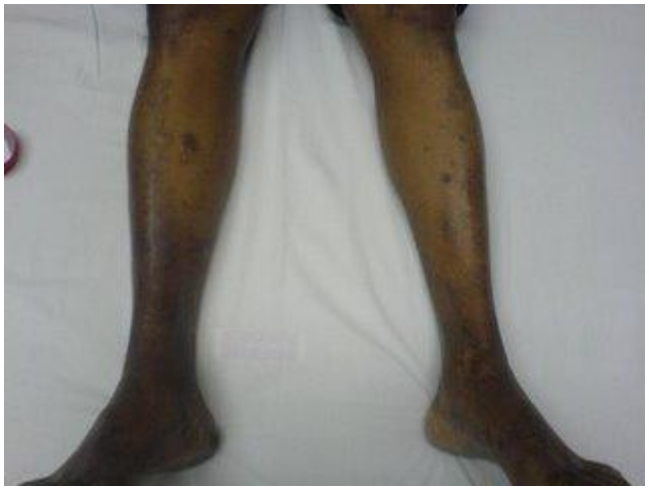
Here is a lady who came for nerve conduction study. She experience *Right* hand numbness that disturb her sleep.

Compare the thenar eminence of the *Right* hand with that of the *Left* hand and note that it is not as prominent on the *Left*. It is wasted. This is the result of median nerve entrapment at the flexor retinaculum, leading to carpal tunnel syndrome.
There is weakness of flexion, abduction and opposition of the *Right* thumb. The sensation also reduced over the lateral three and half fingers.
Then, proceed to elicit Tinel's sign which is positive in this case.
A positive Tinel’s sign for suspected carpal tunnel syndrome.

1. What is Phalen's maneuver?
2. What are the causes for carpal tunnel syndrome?

Peripheral Neuropathy

Here is an Indian gentleman who complaints of feeling shaky on walking for the past 3 years. The onset is insidious and his condition gradually worsen for the period mentioned.



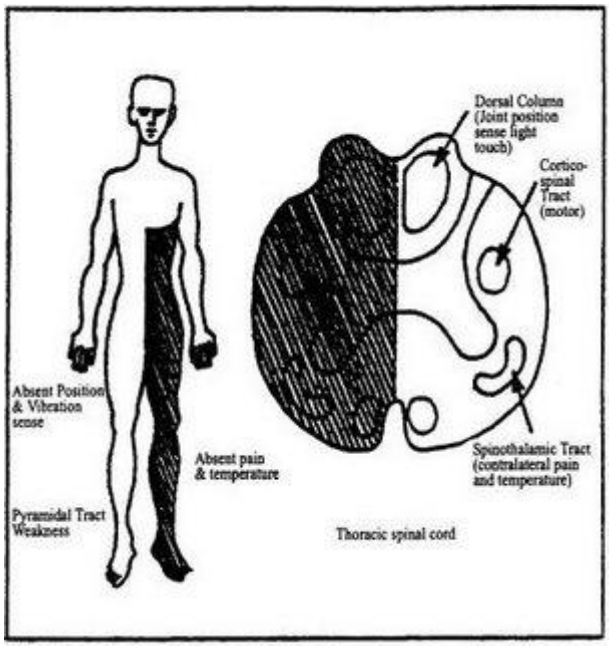
There are trophic changes noted over the lower limbs i.e. the skin is shiny, smooth and loss of hair. There are hyperpigmented skin lesions suggestive of diabetic dermopathy. There is no muscle wasting and the power is normal.
There is impairment of sensation to vibration sense, joint position and pinprick over a stocking distribution (from foot up to knee level). The reflexes are absent.

There is sensory ataxia as evidenced by positive Romberg's sign and stamping gait (broad based gait, foot stamping on the floor)

In conclusion, he has a peripheral neuropathy predominantly involving the sensory component. Functionally, he has sensory ataxia due to impaired joint position sense. The likely cause would be diabetes mellitus in view of the diabetic dermopathy.

I would like to complete my examination by:
1. Checking the upper limbs (sensory loss over a glove distribution)
2. Palpating the peripheral pulses (peripheral arterial disease due to the trophic skin changes)
3. Examine the fundus for diabetic retinopathy.
4. Checking the urine for sugar.

Brown Sequard Syndrome



I am now working in neurology unit. Hence, I will be showing you all more neurology cases. Recently, I came across a case of SLE who presented to us with subacute onset of right LL weakness. Clinically, she has monoparesis of right leg with reduced tone, the reflexes are reduced and extensor plantar response. There is loss of joint position and vibration senses on the right side. There is loss of pain senses on the left side up to the level of T7.

These features suggest a **diagnosis** of right hemisection of the spinal cord. An urgent MRI of spine is warranted to exclude space occupying lesion like abscess or tumour.

Our working diagnosis is tranverse myelitis related to SLE.
The differential diagnosis would include tumor (primary or metastatic), disk herniation, herniation of the spinal cord through a dural defect, epidural hematoma and vertebral artery dissection.

Proximal myopathy

The clinical diagnosis of proximal myopathy is usually straightforward. Proximal muscle wasting and weakness is easily demonstratable.

Tips:
**The weakness is bilateral and usually symmetrical.
**The sensory is always INTACT!

The *causes* that you need to consider are:

1. Muscular dystrophy

**Duchenne/ Becker muscular dystrophy (look for pseudohypertrophy of calf muscle
** **Fascioscapulohumeral dystrophy** (look for facial muscle weakness)
**Limb girdle muscular dystrophy

2. Inflammatory muscle disease

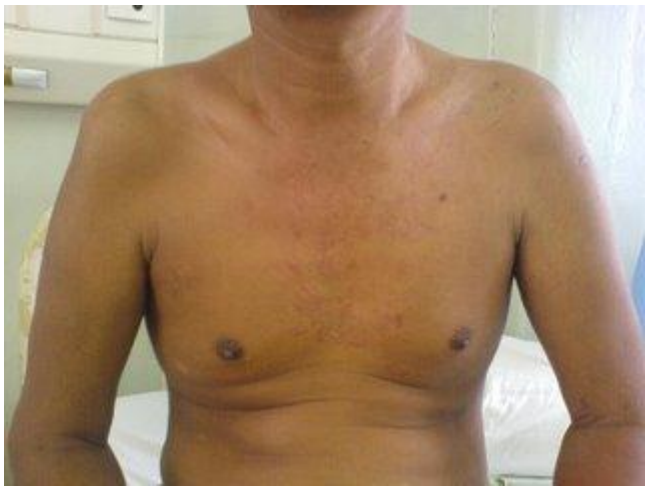
** Polymyositis (muscle may be tender)
** Dermatomyositis (Facial heliotrope rash and Gottron's sign)

3. Endocrinopathy

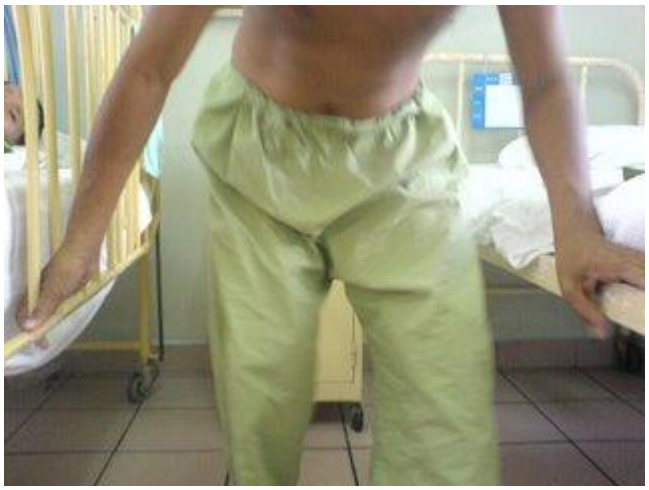
** Cushing's syndrome
** Thyrotoxicosis
** Acromegaly
**ALL will have obvious external features

4. Metabolic myopathies

** Hypo/hyperkalemia, Hypo/hypercalcemia



Wasting of deltoid muscle

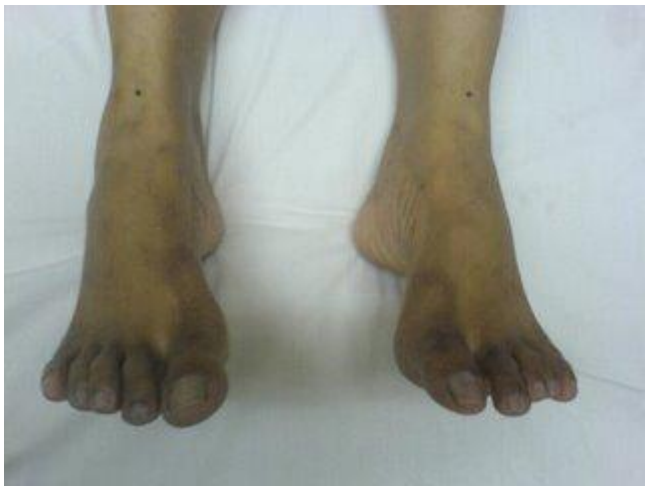


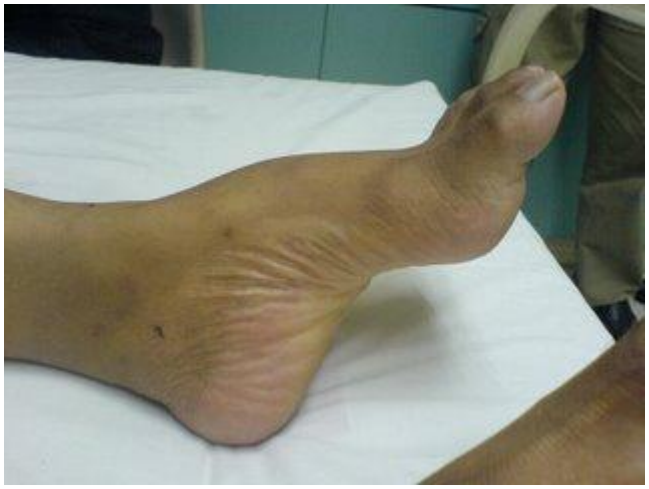
Difficulty in standing up from squatting

This patient was referred to neurology clinic for an investigation. He was then planned for an elective procedure.

What is the respective investigation and procedure?

Pes Cavus





Pes cavus or high arched foot is an important sign for PACES candidates. It carries a lot of weight in neurology station.

1) It means the underlying cause is long standing.
2) It means you need to strongly consider these diagnosis:
** **Charcot-Marie-Tooth ds**
** Friedreich ataxia
** Syringomyelia
** Others: poliomyelitis, cerebral palsy

Ophthalmology

There are several tips to get the correct diagnosis:

1. Prepare the patient by giving a clear instruction. Here is my suggestion.

> *Mr. X, I am going to come near you and examine your R/L eye with this light. It might be uncomfortable. Please bear with me. Now, look straight and keep your eyes still. Don't move your eyes but you can blink as you wish.*

2. Always check the macula area before presenting your findings.

> *Mr. X, please look into my light now.*

3. Think of 3 main diagnosis you need to know for PACES

> *Diabetic eye, **retinitis pigmentosa**, optic atrophy.*

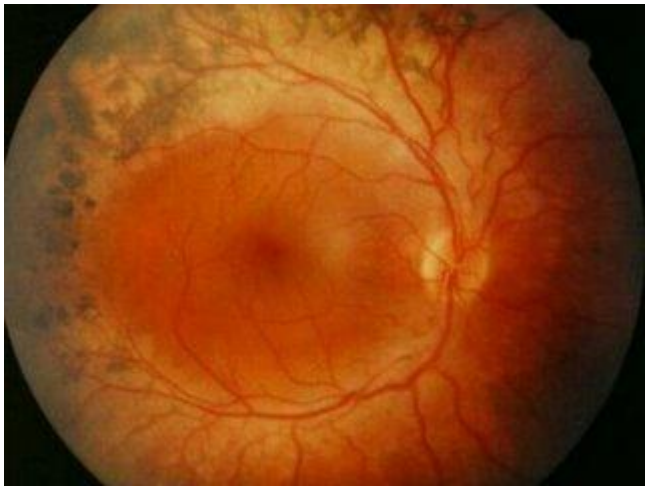
Yesterday, a consultant neurologist came to my hospital and took one of the PACES candidates for a mock.
This was what the patient had (picture above). It was Station 5 Fundoscopy.
1) What is your diagnosis ?
2) What else would you look for ?
3) What are the common causes ?
4) What would you look for in the other eye ?
The patient was a young lady.
What is your complete diagnosis ?

> The answer is that the patient had optic atrophy due to recurrent optic neuritis (in fact, the lady had 8 times optic neuritis but still has not fulfilled the criteria of MS based on the Mc Donald's criteria - MRI always normal)

The other important thing is to look for central scotoma and visual field(pituitary tumour eg craniopharyngioma causing optic nerve compression is a common PACES case)
When you look in the other eye, do not forget to look for papilloedema which will be present in Foster Kennedy syndrome(favourite question from Neurologists)
Do not also forget other causes of Optic atrophy such as AION, toxic(methanol, lead, tobacco, ethambutol), B1,B12 deficiency and consecutive optic atrophy from retinitis pigmentosa.
The 5 features of nerve injury includes
1) RAPD
2)Decreased visual acuity
3) Attenuated colour vision
4) Central/paracentral scotoma
5) Optic atrophy

3 common fundus PACES questions include retinitis pigmentosa, optic atrophy and diabetic retinopathy.

Retinitis Pigmentosa



One of the common case in eye station. Be sure that you can recognise this classic case i.e. widespread scattering of black pigment in a pattern resembling bone corpucles.

Don't forget to check for optic atrophy! The examiner will definitely ask you about the optic disc.

Then, proceed to check for visual field looking for tunnel vision.

Remember these few systemic disorders associated with RP.

1. Laurence-Moon-Bardet-Biedl syndrome

** Autosomal recessive

** Polydactyly (or check for amputation scar over the hands)

2. Refsum's disease

** Autosomal recessive

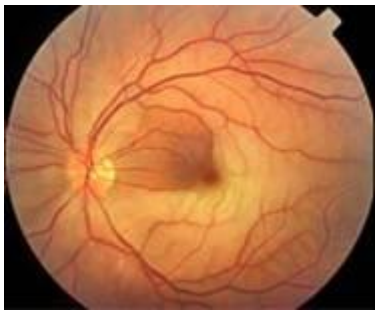
3. Hereditary ataxias

4. Familial neuropathies

5. Neuronal lipidoses

6. Kearns-Sayre syndrome

CRAO



What is the diagnosis ?

What are the causes ?

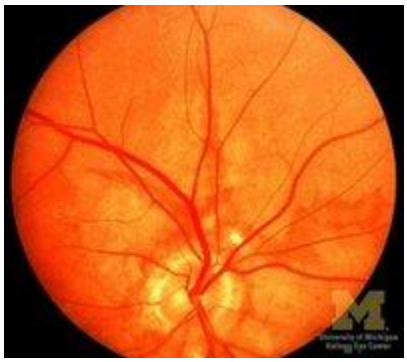
How to treat ?

Yes, the fundus is pale with cherry red spot at the macula, the arterioles are thin and scanty. Diagnosis of CRAO.
Clinically look for carotid bruit, AF, heart murmurs.

What is the diagnosis if this patient complaints of headache ? What are the 2 investigations you would offer ?

The patient must be having Giant Cell Arteritis, so I will proceed to check ESR and temporal artery biopsy. At the same time I would start high dose steroids to prevent visual loss in the other eye.

Pseudoxanthoma elasticum



What is the diagnosis? What else to look for?

The fundus examination showed angiod streak, I say this because of the presence of irregular red brown lines seen radiating from the optic disc. The lines end abruptly, and are larger in diameter compared to the vessels.

i will proceed to examine the right fundus. assess the visual acuity as the acuity can be affected in some cases, although this is usually asymptomatic.

Examine further for associated conditions.
1. Pseudoxanthoma Elasticum
- chicken plucked skin, mitral valve prolapse, young IHD/ Stroke.
2. Ehler Danlos Syndrome.
- lax skin, thin cigarette paper, hypermobile joints, GI bleed, Heart valve.
3. Paget's Ds
- large deformed skull, long bones bowing, Heart failure, nerve entrapment (facial nerve, 8th nerve, spinal cord) sarcoma, urinary stones.
4. Acromegaly - typical features in head, hands, skin tags, acanthosis nigricans, gynaecomastia, spine/ skeletal deformity, HPT, DM, Visual field

5. Lead Txicity - motor peripheral neuropathy, X-rays, Dental, history.

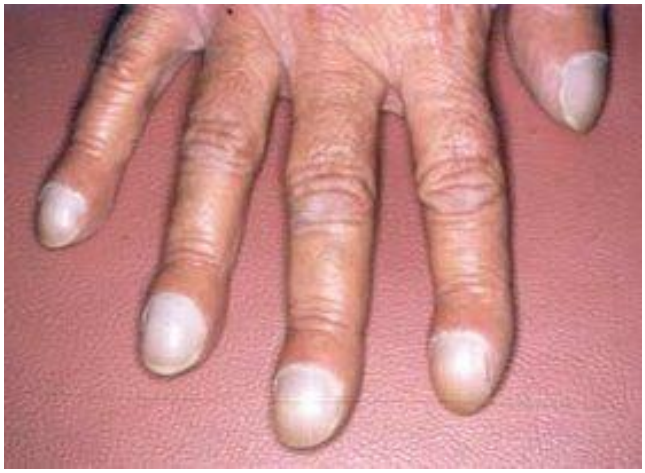
6. Sickle cell anaemia - African, pain, haemolysis.

Proliferative Diabetic Retinopathy

What is the diagnosis ?

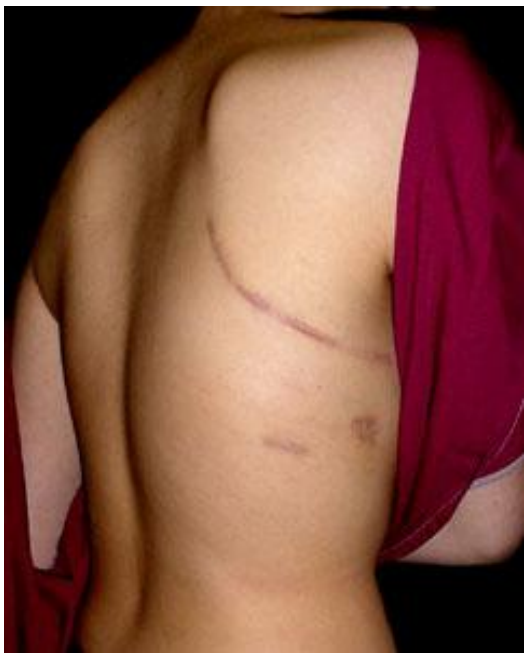
Proliferative Diabetic Retinopathy

Respiratory



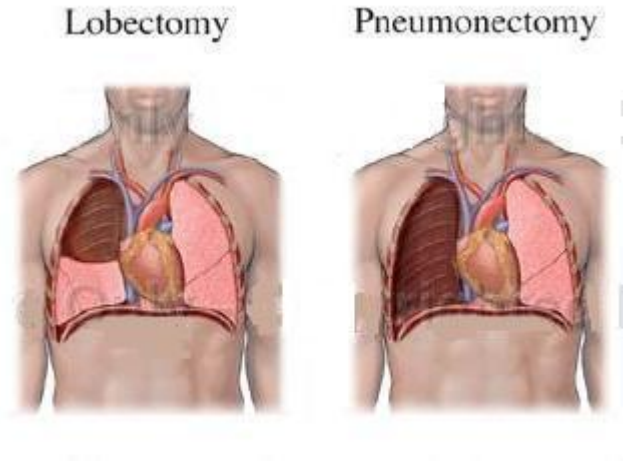
Nobody should miss this sign - *Clubbing*
Clubbing is a common sign in clinical practice. It gives clue to the underlying disease.When clubbing is present, please demontrate nail bed fluctuation, palpate wrist for tenderness seen in HPOA, look for nicotine staining of nail and central cyanosis.

I am sure everyone could give a good differential for clubbing.
CVS: cyanotic heart disease, IE
Lung: bronchiectasis, fibrosing alveolitis, lung cancer
Abdomen: chronic liver disease, IBD



A big thoracotomy scar, one could localize the lesion side by the presence of the scar. Then, you should pay attention to the air entry during auscultation.
a. If air entry is reduced, then think of lobectomy (d.t. mitotic lung lesion)and pneumonectomy.
b. If air entry is equal, then think of decortication and bullectomy.

Lobectomy vs Pneumonectomy



The similarities:

1. Both have thoracotomy scars.
2. Both have reduced chest expansion and reduced AE.

The differences:

1. The signs of lobectomy are **confined** to lobe which is removed. The signs are similar to *pleural effusion* except it is **not** stony dull.

2. The signs of pneumonectomy are **extensive** i.e. involve the whole lung. The side involved would be flatten. It is similar to *whole lung collapse*.

3. Normally, the tracheal is **central** in lobectomy except for upper lobe. The tracheal is almost **always** shifted in pneumonectomy.

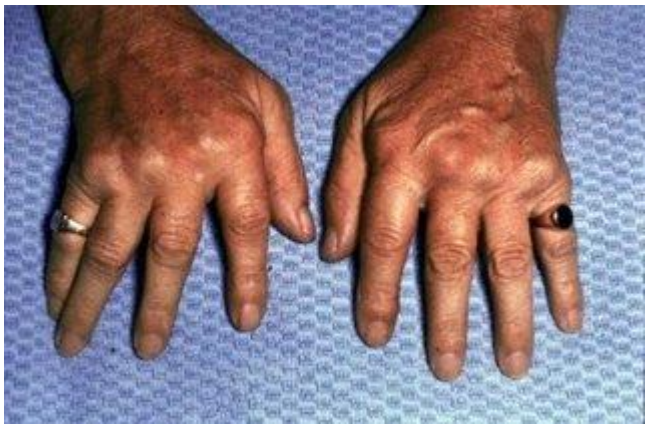
Fibrosing Alveolitis

The respiratory case that I had in UK was rheumatoid lung disease. I got no problem in detecting the clubbing and fine inspiratory crepitations bibasally. But, the outcome was 2/2!

Pitfall: I failed to identify the underlying cause for fibrosing alveolitis. I missed out the obvious sign i.e. Rheumatoid hand changes!

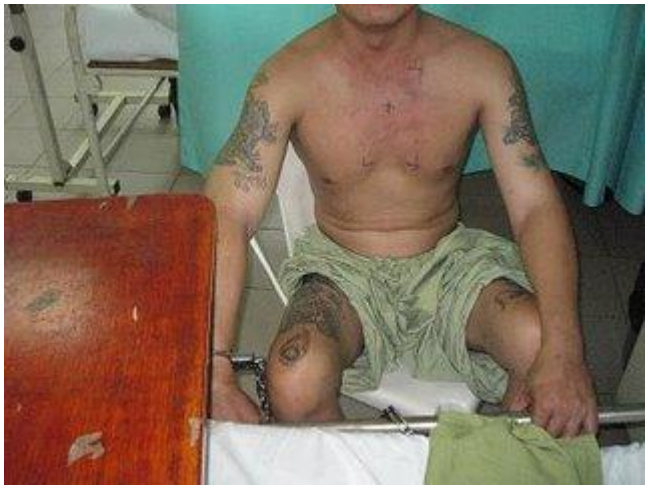
The moral of the story to learn is ALWAYS look for the underlying cause for fibroising alveolitis as cryptogenic fibrosing alveolitis is RARE for PACES!

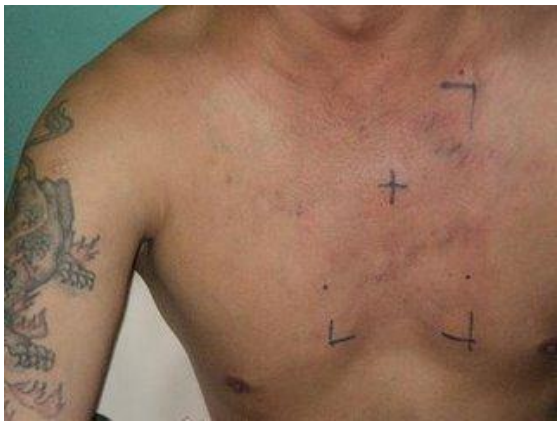
Face: malar rash (SLE)
telangiectasia, beak nose, microstomia (scleroderma)
facial pigmentation (amiodarone)
heliotrope rash (dermatomyositis)
Hand: RA, sclerodactyly



Never miss RA hands in respiratory station!

SVC Obstruction





There is prominent veins over this patient's anterior chest wall. They are tortuous and dilated. The neck veins are engorged with loss of pulsation. The face looks plethoric and suffused.
This is superior vena cava obstruction.
This patient has undergone recent radiotherapy for this medical emergency condition as evidenced by fresh radiation ink marks and skin erythema over the same area.

The patient:

He looks like a young bloke,I'll give probably in his late 20s to early 30s.
No surgical scars evident.
Patient is not cachexic.
No oxygen support line visible.
No IV line (for ?chemo) visible but the right hand is partially covered.
Tatoos could mean bloodborne infection e.g. Hep C, Hep B, HIV
HIV increases susceptibility of cancer.

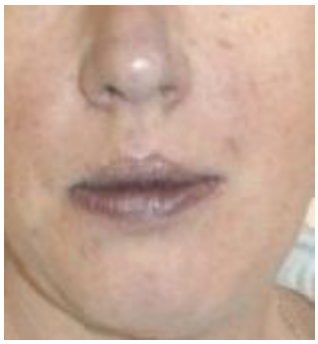
My top two differentials

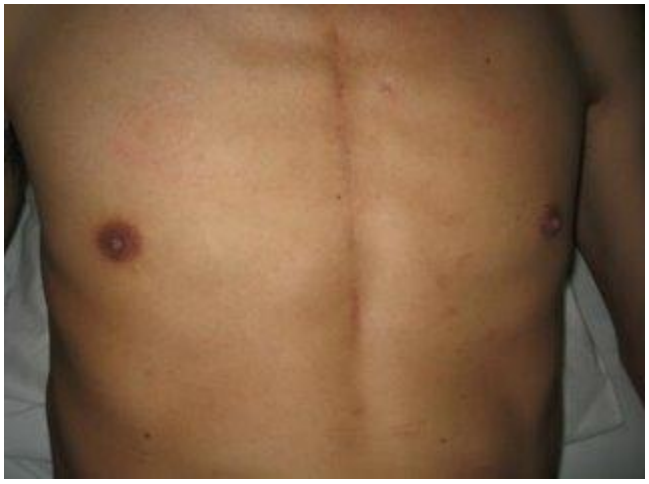
1. Small cell lung cancer (it causes most of SVC obstruction, unresectable, thus no surgical scar,strong association with smoking, he probably is a smoker to me:p)

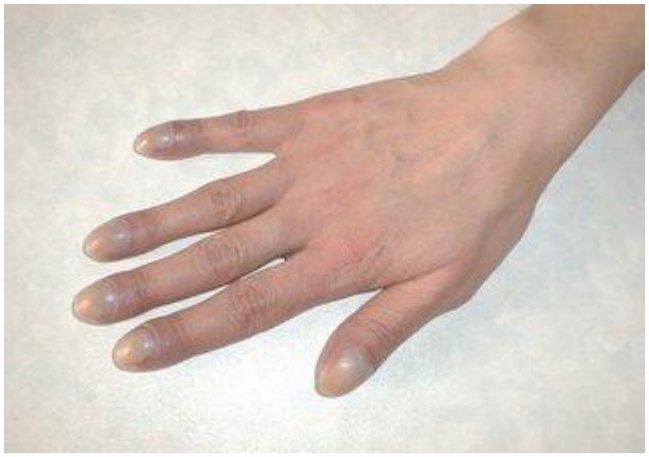
2. Hodgkin lymphoma (bi-peak distribution: young adults and elderly,insidious;I assume you would run the IV chemo here)

Others:
3. Metastastatic cancer. Tatoos could mean Hep C infection,Hep C could lead to hepatoma. (Against this : no visible organomegaly,?no jaundice, no visible scratch mark)

Central cyanosis with clubbing





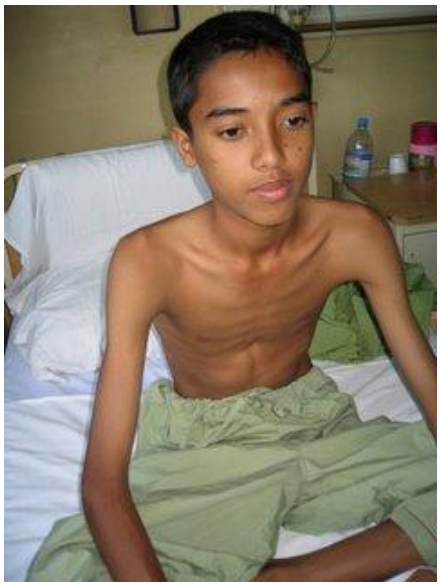


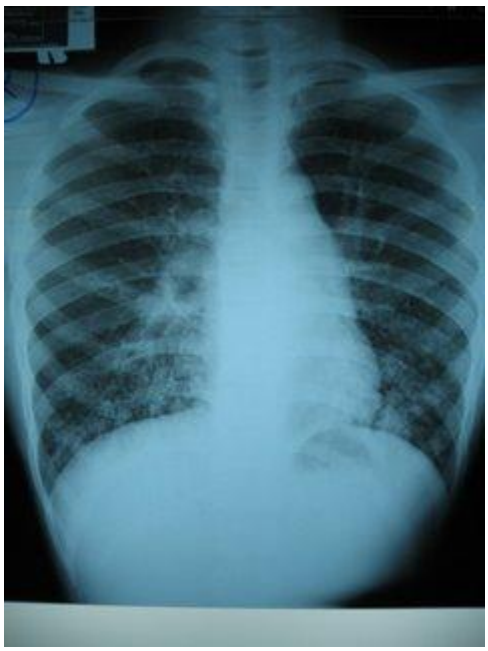
During yesterday's MRCP mock exam, Wuchereria had a tough time with one of the local examiner.

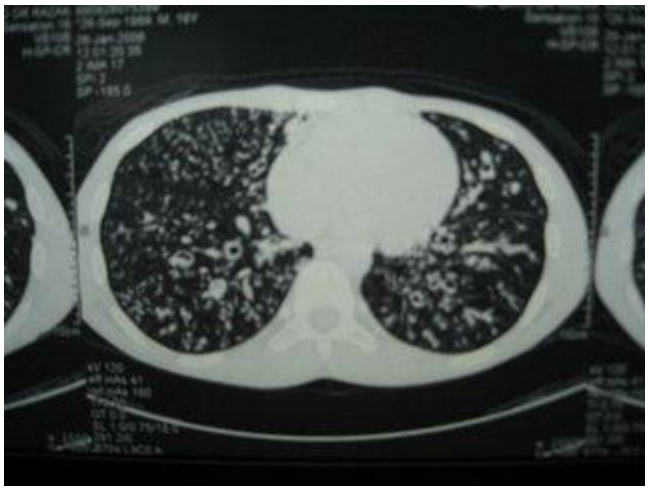
This patient has clubbing with cyanosis and bilateral fine crepitations over the bases with a scar as seen in the picture. He also has an area of consolidation over the R lower zone.

Differential diagnosis: Lung cancer, idiopathic pulmonary fibrosis, chronic suppurative lung disease or cystic fibrosis and previous lung surgery.

Crepitatins and clubbing







Record :

This young boy is tachypnoic at rest with a RR 24/min. There is clubbing noted, no HPOA or evidence of CO2 retention. There is no pallor, jaundice or cyanosis. JVP is not elevated. Trachea is central and apex beat not displaced. There is no cervical L/N enlargement. There is no pedal oedema of the legs. There are coarse inspiratory crepitations over bilateral lower zones.

I would end my examination by checking the sputum pot, temperature chart and other lymph nodes

My diagnosis is that this pt has bronchiectasis and is tachypnoeic. Possible cause include resp infection in childhood.

DD of crepitations and clubbing:

Cancer of lung, fibrosing alveolitis, lung abscess

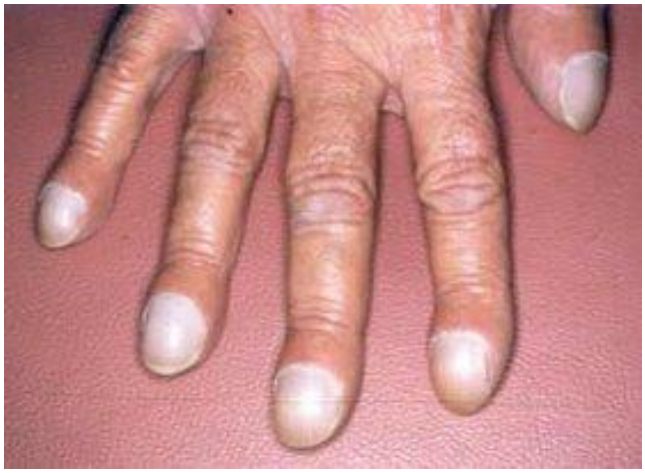
Investigations: CXR, HRCT, Sputum culture

Pleural effusion

A middle-aged lady presented with shortness of breath. Examine her respiratory system. First, I started from the periphery and went up to the head. No abnormality. Then, I went on to feel for the trachea. It was central. As I had discussed with my friend before, I proceeded to examine the back after telling the examiner. There, I noted a needle puncture mark at the left posterior wall. My instinct told me, this should be a case of pleural effusion. It was further confirmed pleural effusion after I elicited stony dullness percussion note with a reduced breath sound and reduce VR. Examiner went on to ask me about how I investigate and manage the patient.

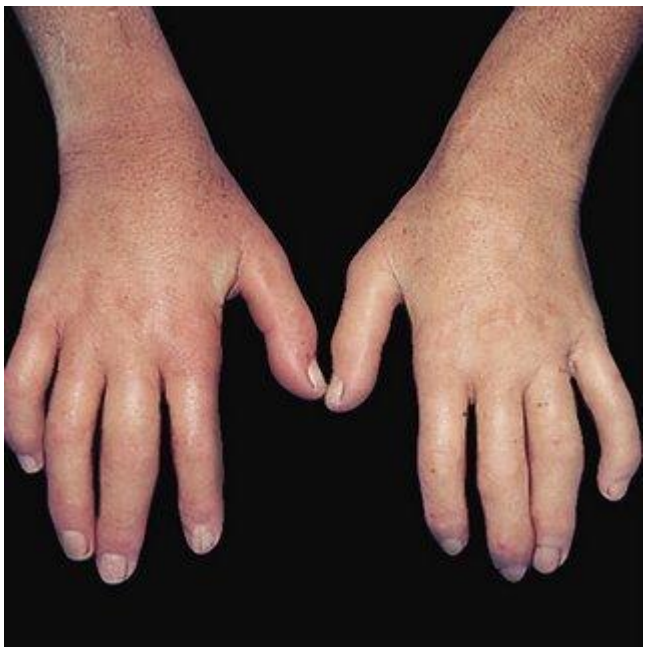
Comment:
This case was relatively straight forward and questions asked was quite direct. I made a terrible mistake here. My friend who sat in UMMC found a centrally placed trachea and he proceeded to examine from the back. After that, he mentioned to the examiner that he would like to complete examination by examining from the front and the examiner said, that was ok and stopped. So, presumed I got the similar scenario, I told the examiner the same thing and he asked me to proceed! I was panicky, there was only three minutes left! Instead, my mind went crazy, I examined the patient's breasts!!! (Thinking there might be a clue there).

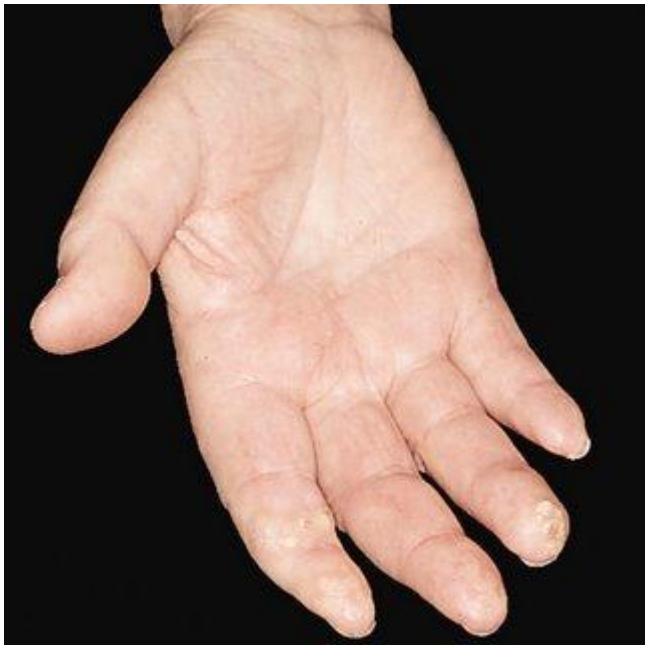
Rheumatology



Nobody should miss this sign - *Clubbing*
Clubbing is a common sign in clinical practice. It gives clue to the underlying disease.When clubbing is present, please demontrate nail bed fluctuation, palpate wrist for tenderness seen in HPOA, look for nicotine staining of nail and central cyanosis.

I am sure everyone could give a good differential for clubbing.
CVS: cyanotic heart disease, IE
Lung: bronchiectasis, fibrosing alveolitis, lung cancer
Abdomen: chronic liver disease, IBD





Scleroderma

What is the diagnosis ?

Any comments on this case ?

Raynaud's Phenomenon
Cyanosis at distal part of finger
Nail pulp atrophy
Vasculitic lesion
Areas of small focal necrosis and ulceration

Proceed

Hands - for arthropathy, function, CREST

Upper limbs - extend of skin involvement in Scleroderma, Proximal Myopathy.

Face - feature of scleroderma, SLE, Sjogren syndrome

Chest - Pulmonary Fibrosis

Heart - Loud P2, cardiomyopathy in scleroderma.

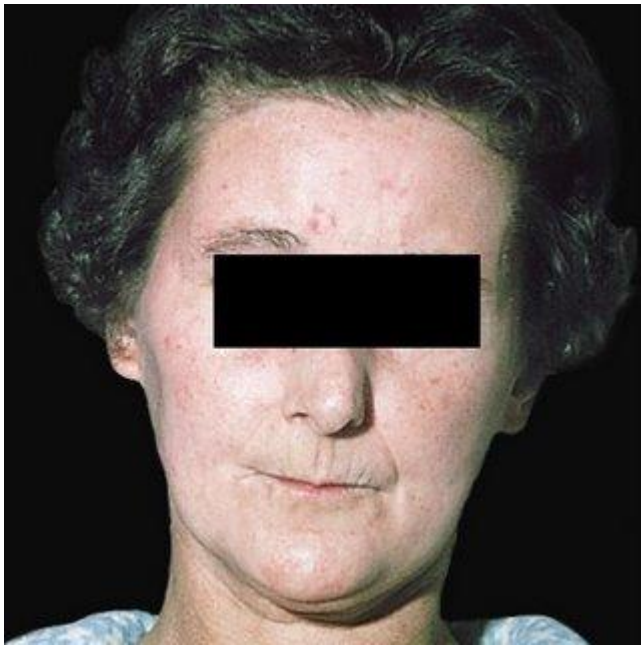
Abdomen - Hepatomegaly, suggestive of Hyperviscosity ?myeloproliferative, PRV etc

End - Drug Hx, Occupation Hx, Joint Pain, Dry eyes, Dry mouth
-look for features of collagen vasc disease

Secondary Raynauds -
Arterial Disease, Thoracic Oulet
Blood Disorder, Connective Tissue Disease, Drugs, Cold Exposure.

Scleroderma is a common short case in locomotor station. You are asked to examine the hands. Check for the characteristic skin features in the hands.

1. Thickening and tightening of the skin. It looks shinny.
2. Subcutaneous calcification over the finger tips or the extensor aspects of forearms or elbows.
3. Ulcerations over the bony eminences.
4. Areas of hypo- or hyper-pigmentations.
5. Look for polyarthropathy that may mimic RA. Assess the function of the hands.



Then, look at the face for microstomia, matted telangiectases over cheeks and lip margins.
If allowed to take history, ask about Raynaud's phenomenon and dysphagia.

Complications:

GI problems - affect any part of the GIT. thus may have GERD, dysphagia, early satiety, diarrhea, constipation

Respi - pulmonary fibrosis and pulmonary hypertension

CVS - cardiomyopathy, myocarditis, arrhytmia

Renal - renal crisis with severe hypertension....treatment of choice?

Cosmetic, depression, low self confidence etc

GI problems - affect any part of the GIT. thus may have GERD, dysphagia, early satiety, diarrhea, constipation

Respi - pulmonary fibrosis and pulmonary hypertension

CVS - cardiomyopathy, myocarditis, arrhytmia

Renal - renal crisis with severe hypertension....treatment of choice?

Cosmetic, depression, low self confidence etc

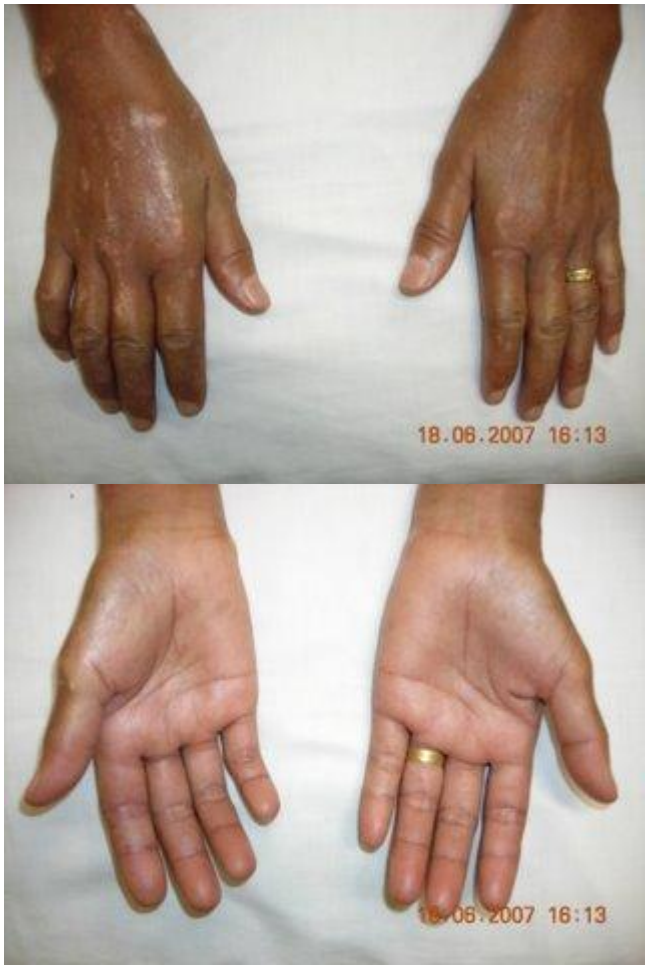
Don't forget about pericardial effusion (raised JVP, low pulse volume +- ascites) if examiner asks for cvs complication

Treatment:

ACE inhibitor for renal crisis.

And penicillamin for pulm fibrosis.

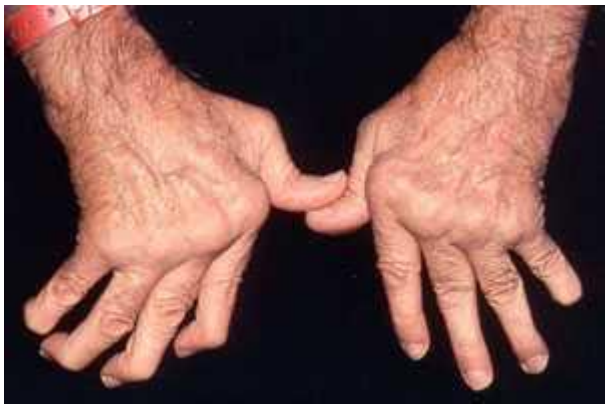
Systemic sclerosis (diffuse type)



Please examine this lady's hands and proceed accordingly.

1) What is your diagnosis?
2) What other systems would you like to examine?
3) How would you treat this lady?

Rheumatoid Arthritis



1. Inspection

Deformities
*Symmetrical deforming arthropathy
*Involving MCP/PIP joints
*Sparing DIP joints
*Swan neck
*Boutonniere's deformity
*Z deformity of the thumb
*Ulnar deviation

*Wasting of small muscles of hands

*Palmar Erythema

*Vasculitic nail changes

2. Palpation

*Warmth, redness, tenderness (active ??)
*Bony swellings (suggest subluxation)
*Synovial swellings

3. Percussion

*Carpal tunnel syndrome (Tinel sign)

4. Movement

*Grip
*Claw
*Flex/extend wrist

*Check ulnar/median nerve (abduct thumb, abduct fingers)

5. Functions

*Unbutton shirt
*Key holding
*Write

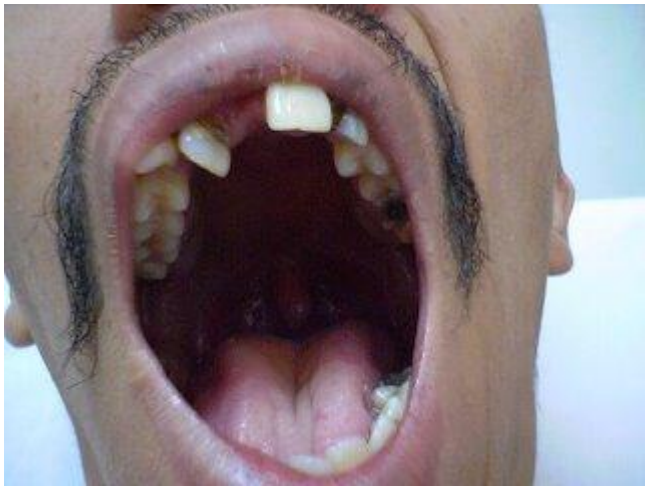
6. Check for Rh nodules

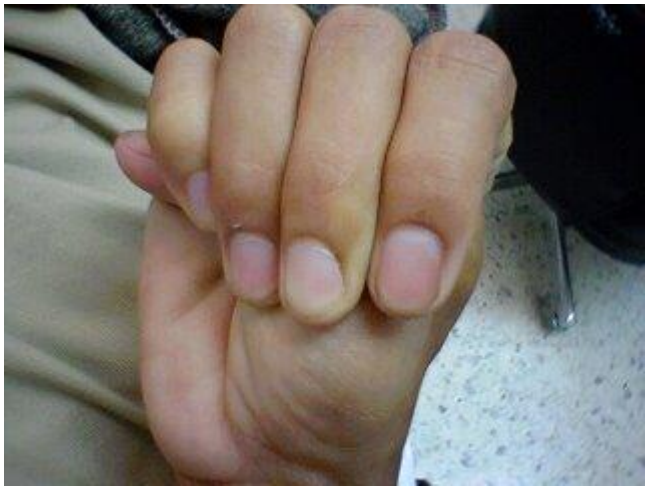
7. End examination

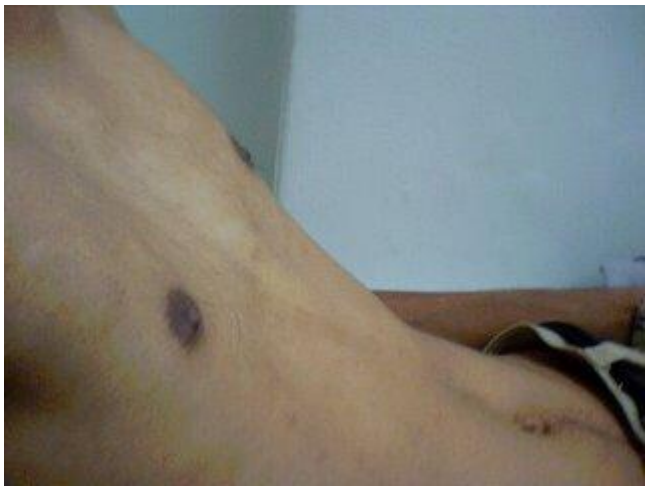
*Eyes – scleromalacia, scleritis, Sjogren's, keratoconjunctivitis sicca
*Lungs – pulm. Fibrosis, nodules
*Hepatosplenomegaly – amyloidosis, Felty
*Leg – Pyoderma gangrenosum
*Cervical – atlantoaxial dislocation

My diagnosis is Rheumatoid arthritis with limited hand function and the disease is currently not active

Marfan Syndrome







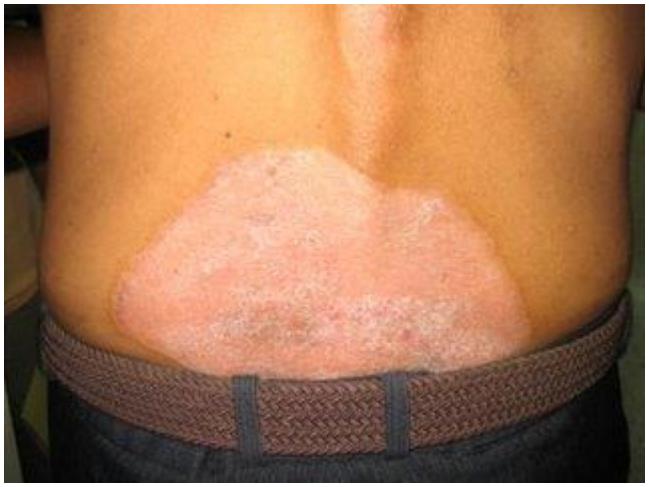
A popular spot diagnosis in MRCP station 5 or may be in CVS station. You notice this man has long fingers (arachnodactyly).Demonstrate to examiners and convince them that the patient's fingers are long and joints are hypermobile by demonstrating

thumb sign (ask patient to clench her thumb in her fist, the thumb should not exceed the ulnar side of the hand in normal subjects)

Then, look for other signs to suggest Marfan syndrome such as high arched palate (in this patient), upwards subluxation of the lens, kyphoscoliosis, and chest wall deformity i.e. pectus excavatum.

For the heart, remember you are likely to find either aortic regurgitation or mitral regurgitation.

Psoriasis With Axial Arthropathy



Well dermacated erythematous plaque with silvery scales.

I had been quite busy for the past few weeks as I just joined skin department and there are a lot of new things to learn. It is a whole new world for me to explore.
As we all know, the prevalence of psoriasis is about 2% of the population. It is the bread & butter of dermatology practice. Here is one of the new cases that presented with difficulty in extending his neck for the past few years. We nearly miss this sign until he stood up to thank us after the consultation - he is unable to extend his neck to look at us. In addition, he needs to turn the whole body to look to either side.

As the picture shown, the occiput is not able to make contact with the wall when heel and the back are against the wall. It indicates upper thoracic and cervical limitation.

Questions:

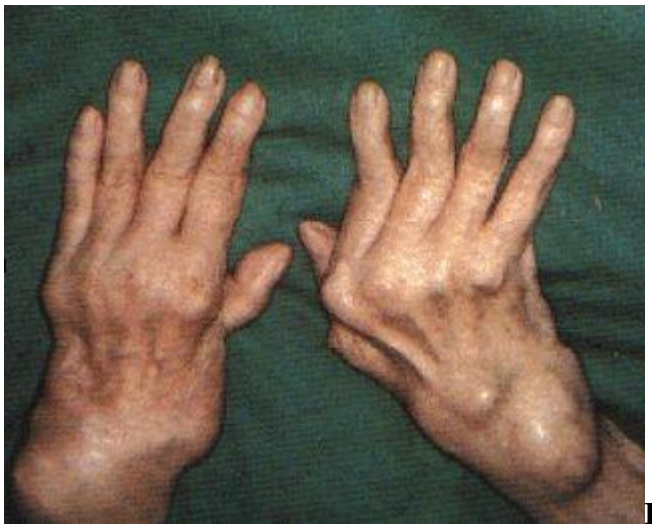
1. What are the types of psoriatic arthropathy?

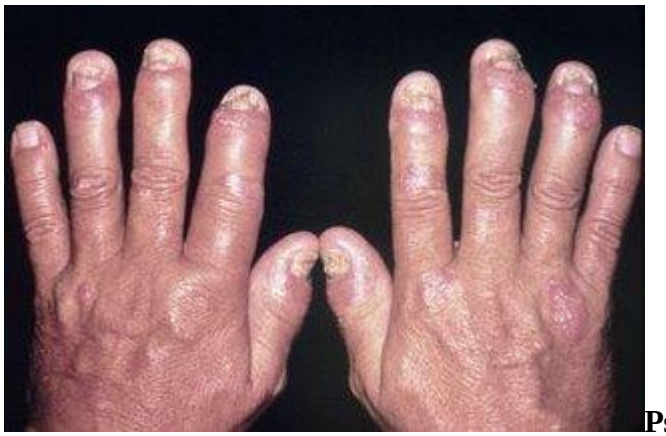
* Arthritis of DIP (Classical)
* RA like arthritis
* AS like arthritis (spondylitis and sacroiliitis)
* Mono- and asymmetrical oligoarthritis
* Arthritis mutilans

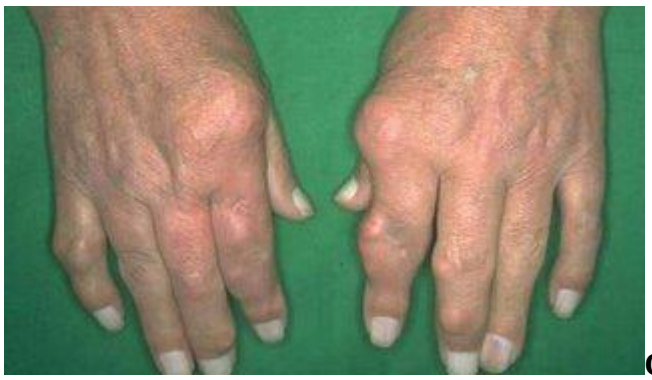
2. What is the most common psoriatic arthropathy?

* Mono- and asymmetrical oligoarthritis

There are not many locomotor cases that the examiners can produce for PACES. So, **please remember the following 3 classical locomotor cases**. Personally, I got RA during 1st attempt and psoriatic arthropathy during 2nd attempt

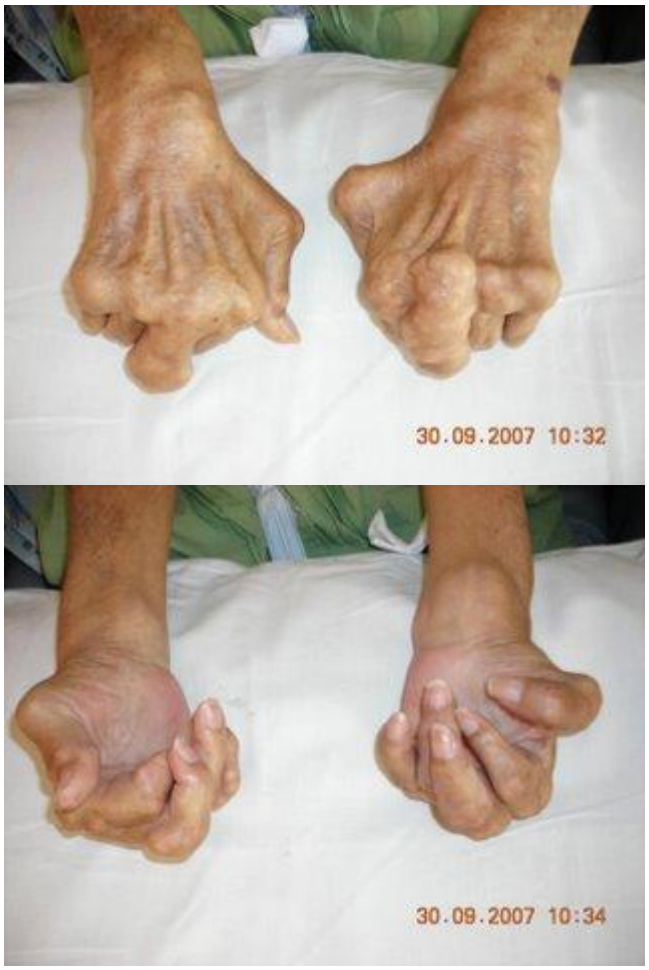
 **RA hands**: symmetrical deforming arthropathy with synovial thickening.

 **Psoriatic arthropathy**: asymmetrical arthropathy involving terminal IPJ with nail changes.

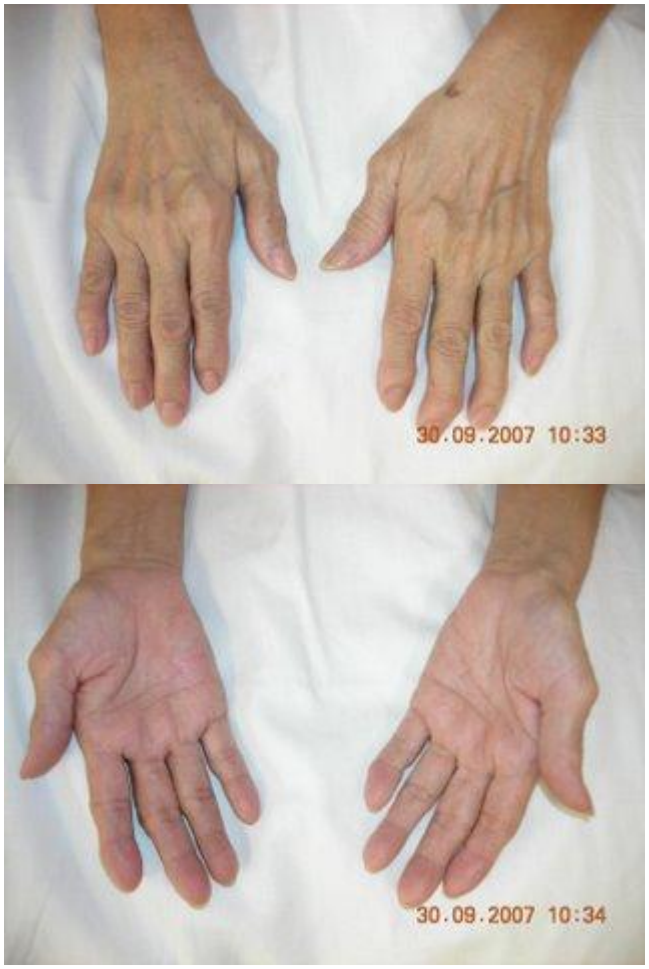
 **Chronic tophaceous gout**: asymmetrical deforming arthropathy with tophi formation.

Chronic tophaceous gout and OA.

Please examine this lady's hand and tell me your findings.



Case 2: This patient has difficulty in holding objects in her hands. Please examine her.



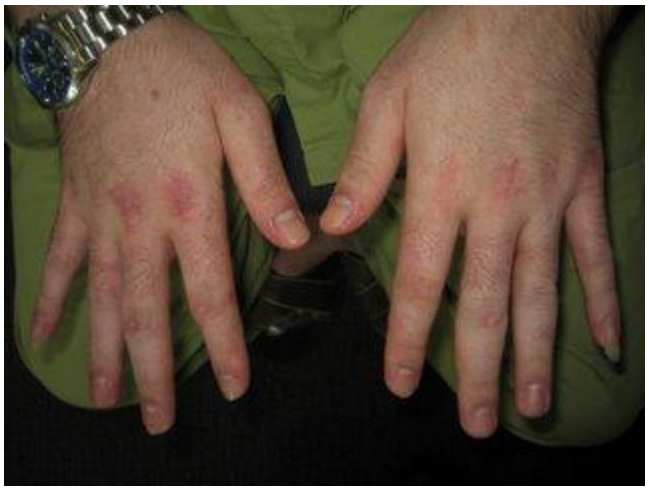
1rst patient has deforming polyarthropathy with multiple topaceous gout and reduced function of the hand and also wasting of the small muscles because of disuse atrophy specially of the introssie
2nd patient has bony swellings of the distal interphalngeal joints of the right hand and the little finger of thr right hand probably herbedens nodes.

Case 1: Chronic tophaceous gout
Case 2: Primary osteoarthritis

Differential diagnosis of OA:

Psoriatic arthropathy affecting DIPs
Non-classical presentation of RA
Chronic gout
HLA-B27 spondyloarthropathies but will have evidence of other disease process
Infective arthritis

Dermatomyositis



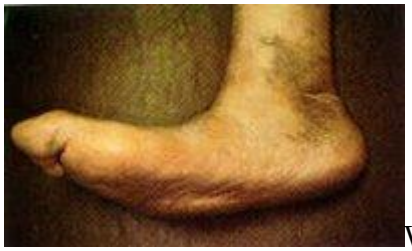
What is the diagnosis ?
This patient has epistaxis. What is it due to ?

This male patient (obviously from the watch) has rashes over the knuckles and interphalangeal joints. There is also vesicle formation and vasculitic lesion. The periungal areas are erymathous. This is dermatomyositis.
In view of the opening statement that the patient has epistaxis, I would like to investigate for an underlying Nasopharyngeal carcinoma.

Diagnosis:

Dermatomyositis presenting with Gottron's papule.
He also has nasopharygeal carcinoma.

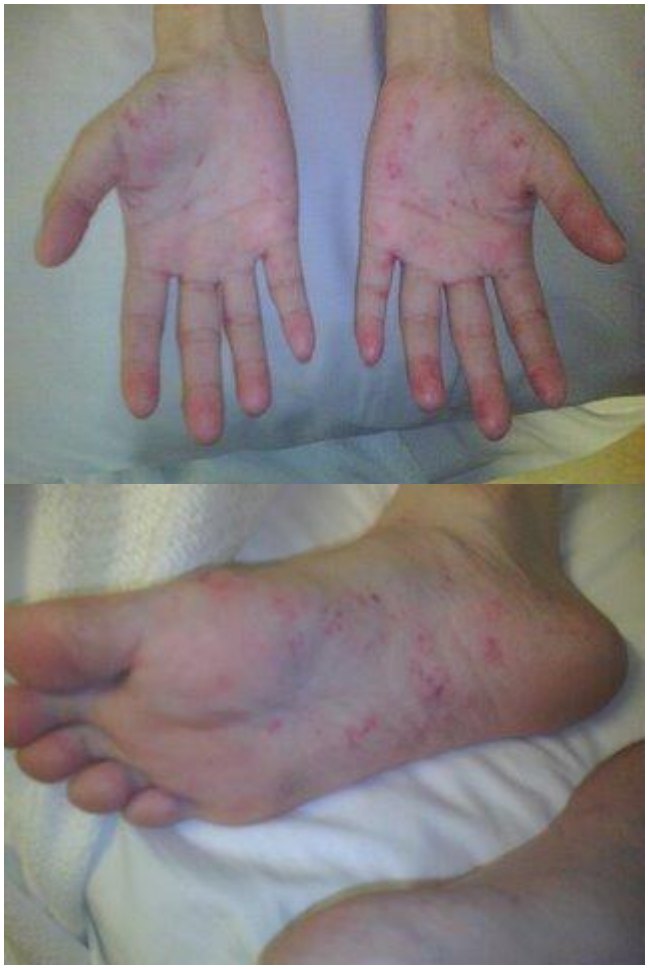
Charcot's joint



What is the diagnosis ?
How would you proceed to examine this patient ?
What is the cause ?

The answer is that this patient has Charcot joint. Need to think of DM as this involves the ankle. If it involves the knee, think of Tabes Dorsalis.

Mixed connective tissue disease



Look at this patient's extremities and proceed.

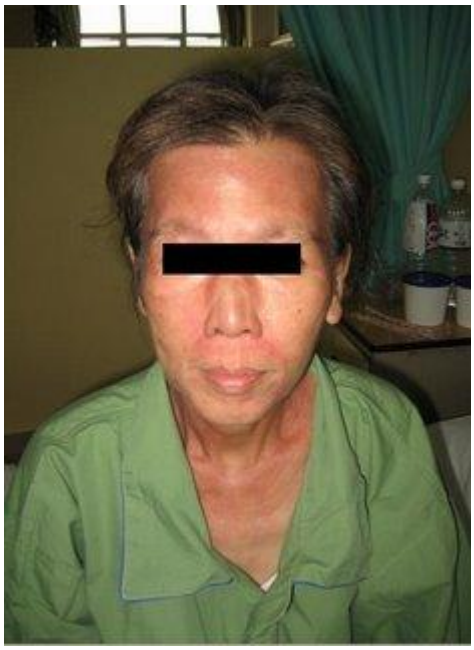
What are your findings? What are the possible differential diagnoses?

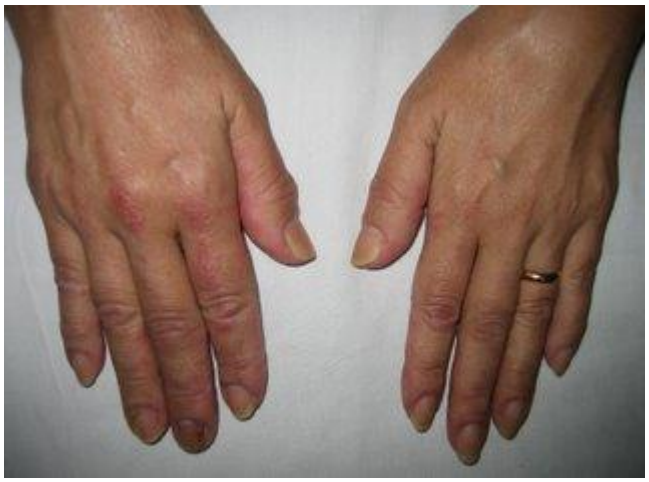
This lady in fact has MCTD with +ve anti-RNP. She also shows cutaneous stigmata of scleroderma upon palpation.

Panhypopit is right on the right track for the differentials.

Nope, it's not syphilitic rash for sure. And HHT won't have this confluent form of erythematous patches. Plus, it's the wrong place to look for HHT cutaneous signs!

Mixed connective tissue disease





Observe and comment the skin findings.

What are your differentials?

Think of 3 DD when you encounter facial rash possibly dt connective tissue disease:

1. SLE
2. Dermatomyositis
3. Mixed CT disease

Communication Skills

The 5 star list:

*Brain stem death

*Asking for postmortem

*Breaking bad news about terminal illness

*Breaking bad news about chronic illness

*Fitness to drive

*Cardiac rehabilitation

*Needlestick injury from HIV patient

*Resuscitation status in a terminally ill patient

*Screening for prostate cancer

*To ventilate or not to ventilate

*Withdrawing treatment

*Dealing with poor compliance

*Consent to participate in clinical trial

*Consent from a patient who does not have the capacity to give consent

*Managing a complaint after an adverse incident

These are the main topics that you **MUST** read and practice before your PACES day!

Asking For A Postmortem

Sympathy

Iâ€™m sorry to hear about the sudden death of your father and all the staff looking after him are saddened by the news

Explain the scenario

Explain that it is not absolutely certain what caused the death, give possibilities

Explain that one of the way of knowing exactly what may have caused the sudden deterioration is a postmortem

Check understanding

What do you understand regarding a postmortem ?

Intermnal examination of person just died, valuable information

Ask advance directive

Has your father ever told you regarding his objection towards postmortem ?

Benefits of postmortem

Valuable information about illness

Explain cause of death

Help Dr treat the same kind of illness

For research

Expain about postmortem

Limited vs Full postmortem

When, where and whom performing the postmortem ?

-by pathologist, in hospital mortuary

Reassurance

Reassure that the body would not be disfigured

Reassure that the funeral arrangements would not necessarily be delayed

As soon as possible

Anything suspicious, this organ will be retained to confirm diagnosis if no objection

Consent if anything retained

Results will be informed â€“ an appointment with consultant

Concerns

Support the decision

Inform brain death and withdrawal of treatment

Mr Smoki is a 65 y.o. gentleman admitted 3 days ago for acute exacerbation of COPD. He has been getting dyspneic for past few days, with fever and increased sputum production but has refused for admission. He was found collapsed by her daughter, but resuscitation team has managed to resuscitate him and was sent to hospital. He has history of recurrent admission for COPD. For the past 1 year, his COPD symptoms worsened, and he required 5 x admission for the past 1 year. He has been stopped smoking 3 months back due to progressive worsening dyspnea. He has been staying with his only daughter, and his wife has passed away due to myocardial infarct. His daughter Miss Norsmok has been quite supportive of her father so far, and has been spending her time all along during these hospital stay.

There has been no improvement in his father's condition throughout the ICU stay. He has spiking fever and it was complicated by septic shock and multi organ damage. His blood pressure has been low, an now require inotropic support. The ICU team has discussed poor prognosis to medical team. Brainstem test was done, and Mr Smoki was confirmed brain death. Therefore, withdrawal of treatment is imminent.

You are the senior medical officer in charge appointed to meet Miss Norsmok. Your task is to explain braindeath to her and discuss the issue of withdrawal of treatment.

1) Introduction

Dr: Im Dr X, im here to discuss with you about your father's condition. Do you have anyone coming along for the discussion?
Miss Norsmok: No, I came alone.

2) Explore how much she knows about her father

Dr: How do you feel about your father's condition? Did anyone tell you about your father's condition before?
Miss N: Nobody so far tell me in detail. But I was told that my father is critical. Doctor, did anything go wrong?

3) Explaining general condition

Dr: Im afraid we have bad news for you. Your father has been admitted with severe lung disease. We have to protect his lung into rest. His breathing is supported by our machine. It has been hard for your father to recover in view of his severe lung disease. Your father's condition was worsened by multiple organ failure due to severe infection. We have been trying our best but we failed to do so. (give a pause…)
Miss N: What do you mean? Fail?
Dr: Yes. The fact is hard to accept, but we have perform a test and have found that your father has brain death, in other word, your father has passed away. We are sorry.

Miss N: It can't be. He is still breathing, and he still have pulse..Are you sure my father has died?

Dr: Yes. Your father would not be able to breath by his own. It is the machine which help his breathing. And his pulse is due to medication, which temporally make his heart beat. Without artificial support, his heart will stop beating, and he would stop breathing. Therefore, your father has passed away.
Miss N: I can't believe it…(pause…)

4) Discuss withdrawal of treatment
Dr: You seems upset by the news. I could understand that. It is difficult for us to discuss this at this moment, but I need to tell you that we are going to withdraw treatment to your father. How do you feel about it?
Miss N: Are you going to give up on my father? Do you mean you want to let my father stop breathing?? Don't ever mention it!

(Do not be panic as most family would think that we are going to let the patient die. We need to confer to them that her thought is incorrect)

Dr: Miss Norsmoke, we never give up treating your father. It is however not appropriate to continue treatment in your father, as he already passed away. I'm sorry about this, and I know that you need time to accept.

Miss N: …What are you going to do Dr?

Dr: We are going to stop the medication that support the heart as well as stopping the machine. Your father would stop breathing and his heart beat would stop in a short

moment. There would be no suffering to your father as he has already passed away. (Mention the word "passed away" few times to remind the family about the death)

5) Explore reason if disagree

Miss N: No, I don't want you to do that. You cant stop the treatment.

Dr: It must be hard for the decision. But can I ask why do you think in such a way? Do you have any religious believe in this matter? Did your father ever express his will about how he would be treated before this?

Explore the reason and sort out any misconception is utmost important. Decision of the relative must be regarded important, although decision of withdrawing treatment lies in the hand of clinicians.

Miss N: No, there is no specific reason. I don't accept that my father be treated as such.

Dr: Withdrawing treatment may be unfamiliar to most people. I would address the issue to my consultant. In a short while, we would have another discussion with my consultant. Do not hesitate if you have any question regarding the problem. See you again.

Issue and Discussion:

Autonomy
Beneficence
Nonmaleficence
Distributive justice

Why withdraw treatment?

1) Withdrawal of treatment is an issue in intensive care medicine because it is now possible to maintain life for long periods without any hope of recovery.
2) It is often easier to withhold a treatment than to withdraw it once it has been instituted. Ethically, however, there is no difference between withdrawing a treatment that is felt to offer no benefit and withholding one that is not indicated.
3) About 70% of deaths in intensive care occur after withdrawal of treatment. This is not euthanasia. The cause of death remains the underlying disease process, and

treatment is withdrawn as it has become futile.
4) In general, treatment is withdrawn when death is felt to be inevitable despite continued treatment. This would typically be when dysfunction in three or more organ systems persists or worsens despite active treatment or in cases such as multiple organ failure in patients with failed bone marrow transplantation.

Patient autonomy

1) Autonomy is another of the basic precepts of ethical practice, but there are problems with its implementation in the intensive care unit. Most critically ill patients are not competent to participate in discussion because of sedation or their illness.

2) in the United Kingdom relatives do not have legal rights of decision making

3) Another difficult issue occurs when a patient may survive but with a poor quality of life

4) Relatives must be kept fully informed about the patient's condition, in particular regarding issues of limiting and withdrawing treatment.

5) Although decisions rest with the medical staff, it is unwise to limit or withdraw treatment without the agreement of the relatives

Problems

1) The referring team request continued futile therapy
This can usually be resolved by explaining the rationale and offering a second opinion from another intensive care consultant. If conflict still remains, treatment cannot be withdrawn. The family should not be informed of a decision to withdraw that is then rescinded because of interteam conflicts. It will reduce their faith in subsequent decisions and undermine confidence in the predicted outcome.

2) The patient's family requests continued futile therapy
Guilt usually plays a part in the family's request to continue treatment, although religious and cultural factors may also contribute. Agreement can usually be obtained by explaining the rationale again and offering a second opinion from within or outside the intensive care team. It is best not to withdraw treatment if there is conflict.

However, the final decision rests with the intensive care team. This underlines the need for good communication.

3) The family requests inappropriate discontinuation of therapy.
The rationale behind the therapy and the reasons why continuing treatment is thought appropriate should be explained. The duty of care is to the patient, not the family. Again, a second opinion can be offered.

4) The patient requests discontinuation of therapy.
Explain to the patient the rationale for the treatment and that, in the opinion of the intensive care team, a chance of recovery exists. It may be appropriate to offer a short term contract for treatment (for example, 48 hours then review). Ultimately, the competent patient has the right to refuse treatment even if that treatment is life saving.

Almost all the examiner I talked to put lot of stress on 'setting the agenda' for history taking & communication:

1. Your introduction with designation
2. Your role in management
3. Check the identiy of the patient & relative if said so in the scenerio
4. Set the agenda i.e why is this meeting
5. Will be willing to answer all questions
6. No hasty gesture i.e we have 10 minutes etc, if time finishes can say I feel a few issues needs to be addressed to will make another appointment within a few days.
7. Always, always- summarize

Remember these 5 core principles in medical ethics:

1. **Non-maleficence** - ie to do no-harm first! Remember this principle first. You will find that it's applicable in any scenarios, and it's indeed very true in our daily clinical practice.
2. **Beneficence** - ie to do good for patients
3. **Patient's autonomy** and right to **confidentiality**
4. **Truth-telling**
5. **Justice and sharing** - ie fairness in provision of care in face of limited resources

Hence, after spending 14 mins with the surrogate and 1 min of reflection, time for heads-up and face the music! Place your discussion with the examiners based on all these core principles. You won't go wrong. But to get a 4 would require much of your soft skills in tackling the surrogate's emotion and responses.

Here are some illustrations:

- A patient with advance statement for no-CPR came in with an umcomplicated MI. Your task is to explain with regards to his advance statement and its application to his current diagnosis.
  Principles: Autonomy vs beneficence

- Relatives of a patient with poor outlook came to you requesting ICU admission. Your ICU has 1 bed left. A young man with polytrauma is on his way to the hospital. Your task is to address this issue with the relatives.
  Principles: Autonomy (any advance statement?) vs justice and sharing of care

- You accidentally prescribed bactrim to a patient with a previous history of sulfur allergy. The patient developed Steven-Johnson syndrome. The father is keen to meet you for an explanation.
  Principles: Non-maleficence & truth-telling (apology)

- A young man is tested positive for HIV. He refused to tell his wife despite adequate explanation. Your task is to meet him and explain with regards to the risk of transmission of HIV to his sexual partner(s). **Principles: Autonomy (confidentiality) vs non-maleficence (public interests) and justice**