431

Radiology Team

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Lecture 3: Radiology of Hematopoietic Disorders

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◆Important ◆Doctor's notes ◆Team's notes

From Slides | Extra Images

Radiology of Hematopoietic Disorders

Classification:-

- Hematopoietic disorders are classified into disorders of:-

CELLS	RBCs	Anemia	Sickle cell
			Thalassemia
		Polycythemia	
	WBCs	Lymphoma	
		Myeloma	
	PLATELETs	Bleeding / clotting	
PLASMA		Bleeding / clotting	

The lecture will only discuss these

- Anemia:- there will be some changes that can be seen on radiological images.
  1) Reactive increase in RED BONE MARROW (1st response) [Expanded bone marrow in bones including bones of hands, feet, limbs, skull. Decreased T1 MRI signal in vertebral body bone marrow than adjacent discs] this is called INTRA-MEDULLARY HEMATOPOIESIS. (Normally in Adults, bone marrow is usually in axial skeleton *spine, skull and to a less extent in the pelvis and proximal long ends of the bone like humerus and femur*.)
  2) New marrow areas in potential organs (if the anemia is still not corrected), which normally don't produce blood cells in adult life. [Liver, Spleen, Lymph nodes, Thymus, Paraspinal areas with possible extension into spinal canal outside the Dura, Kidneys, Meninges, and Skin] These can also be a site for RBC production. This is called EXTRA-MEDULLARY HEMATOPOIESIS.
  3) Transfusions, and iron overload [Increased CT density (brightness) and changes in MRI signal of liver & spleen]. (When the patient is severely anemic and needs transfusions.)

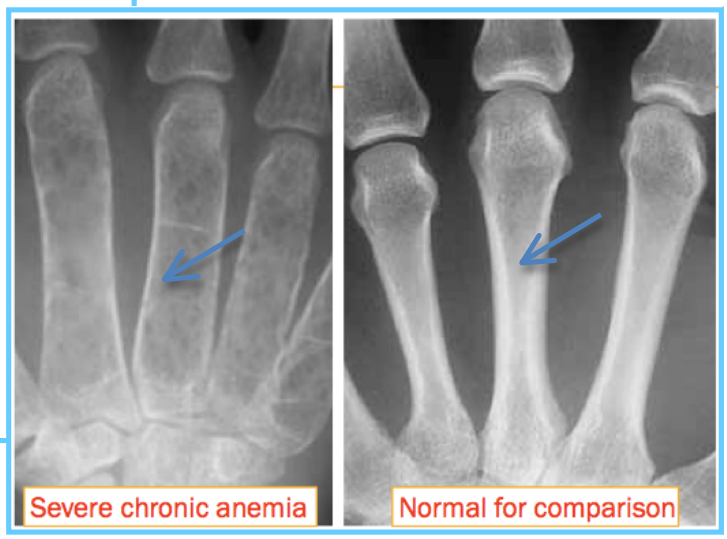
  [N.B. These signs are NOT specific to any kind of Anemia]

- Sickle Cell Anemia:- (has some additional signs.)
  1) Increased risk of infections. (Pneumonia and osteomyelitis)
  2) Increased risk of infarction. (Spleen, Kidneys, Brain, Bones, any organ can get affected). (due to the sickling of the RBCs inside the vessels, so the vessel will get occluded and ischemic)

Thalassemia

Decreased bone density with coarse trabeculae

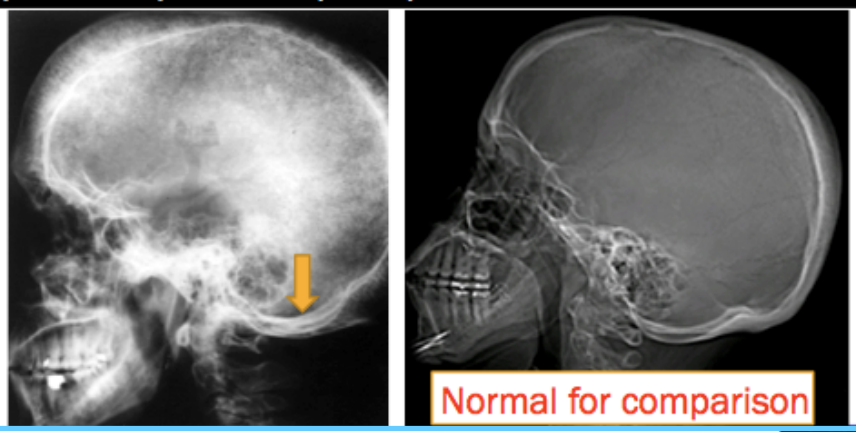
Wide medullary cavity with thin cortex

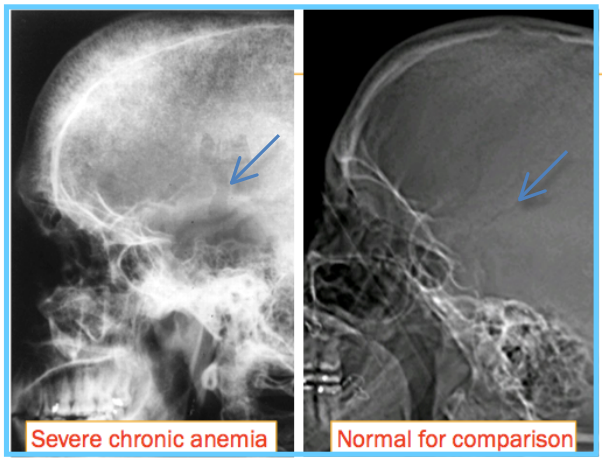


Abnormal:- When bone marrow becomes hyperplastic and increases in amount → The bones are expanded and the diameter is increased, and there's cortical thinning.
(Sign of anemia, not specific to Thalassemia.)

Normal:- Bones are broad at the edges and narrow in the middle.
Abnormal:- Expansion in the middle.

25-year-old man with β-thalassemia.
Lateral skull radiograph shows
expansion of diploic space with hair-on-end appearance
widened groove for middle meningeal artery
Spared occipital bone (arrow)





The bone marrow is between the inner cortex and the outer.

Abnormal:- The overall skull is very thickened, expanded with cortical thinning. There are also some areas that are more Lucent and areas that are darker → indicating bone marrow hyperplasia.

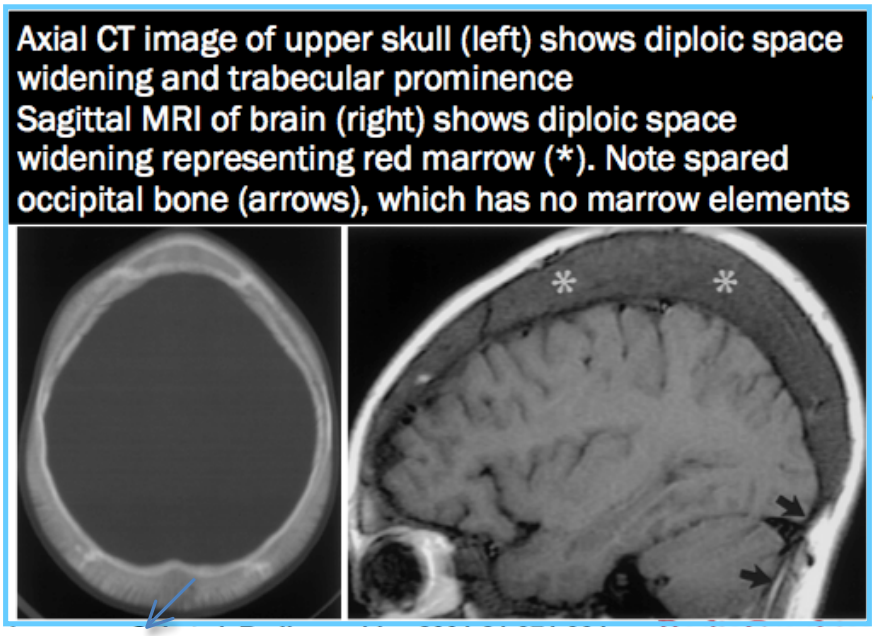
The occipital area has no bone marrow so it will be spared.

You can't see the bone marrow on x-ray only the expansion that's due to bone marrow hyperplasia.

Also bone marrow hyperplasia needs increase amount of blood supply so it will need more/larger blood vessels. You can see that the branch of the middle meningeal artery is enlarged.

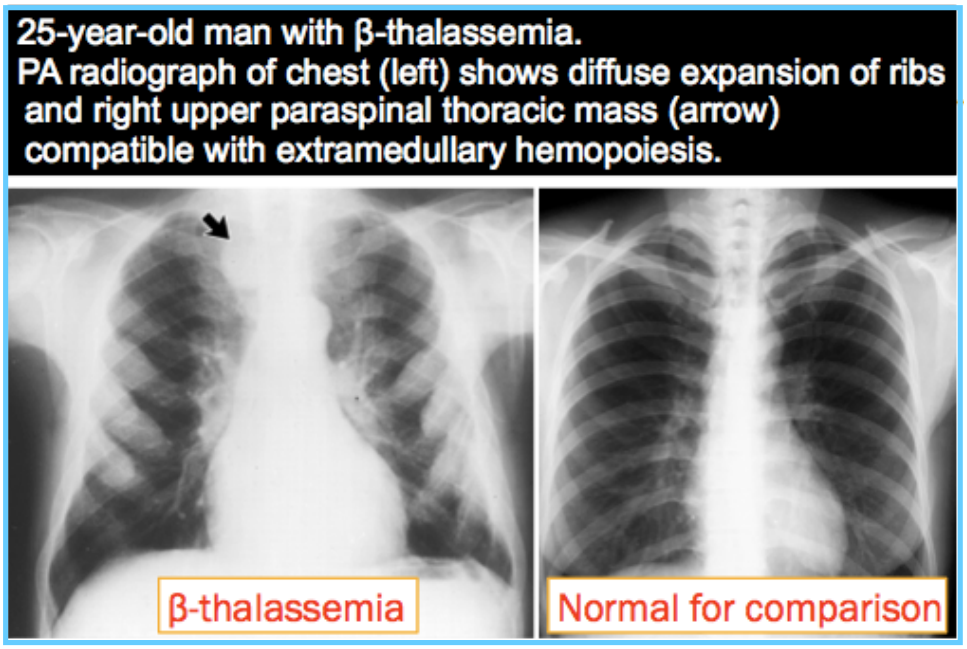
Not specific to thalassemia.

Both cases are intramedullary hematopoiesis.



When we do MRI we can actually see the bone marrow with the skull.

And the CT has basically the same appearance.



Abnormal:- The whole rib is expanded and especially the anterior end → Intramedullary hematopoiesis.

So basically the same sign of anemia; expansion of the bones.

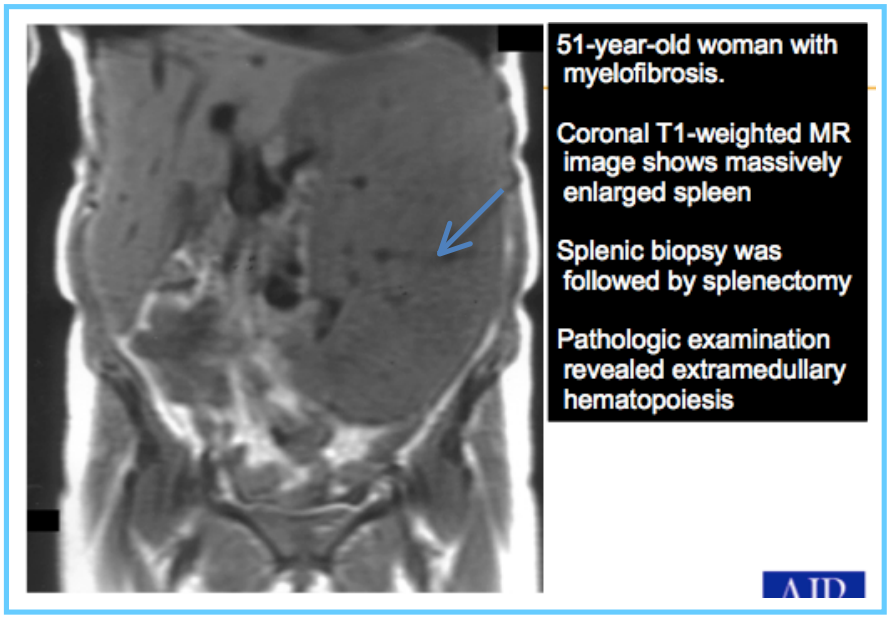
There is also extramedullary hematopoiesis.

- Reasons for extramedullary hematopoiesis:-

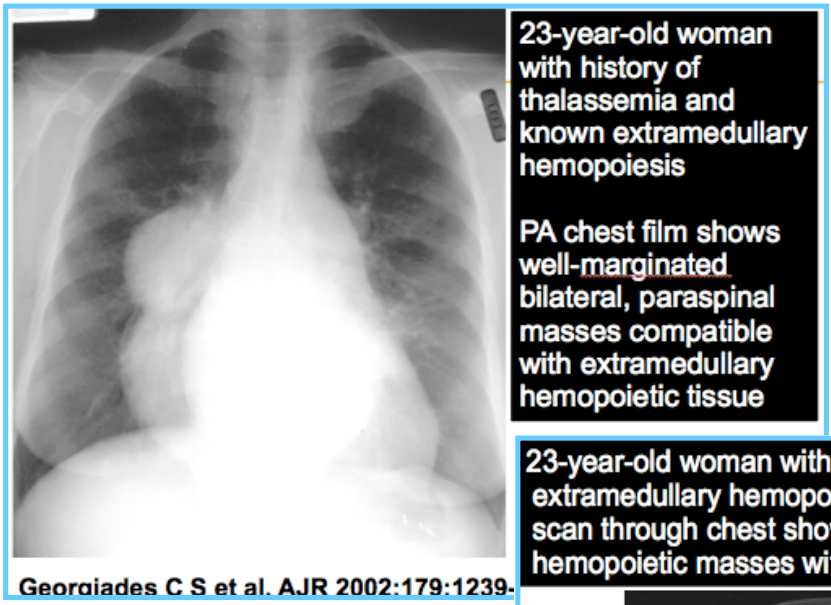
1. When the intramedullary is not enough.
2. When the intramedullary hematopoiesis is not occurring.

- How to differentiate between a hematopoietic area and a malignancy? Its based on the clinical scenario, if a patient has chronic anemia then most likely its extramedullary and with proper treatment, it will disappear within a few weeks. If it doesn't disappear then it's probably a different reason and a biopsy will be needed.

- There are relatively no complications to the extramedullary hematopoietic areas, except bleeding, due its high vascularity. Therefore biopsies are not recommended.

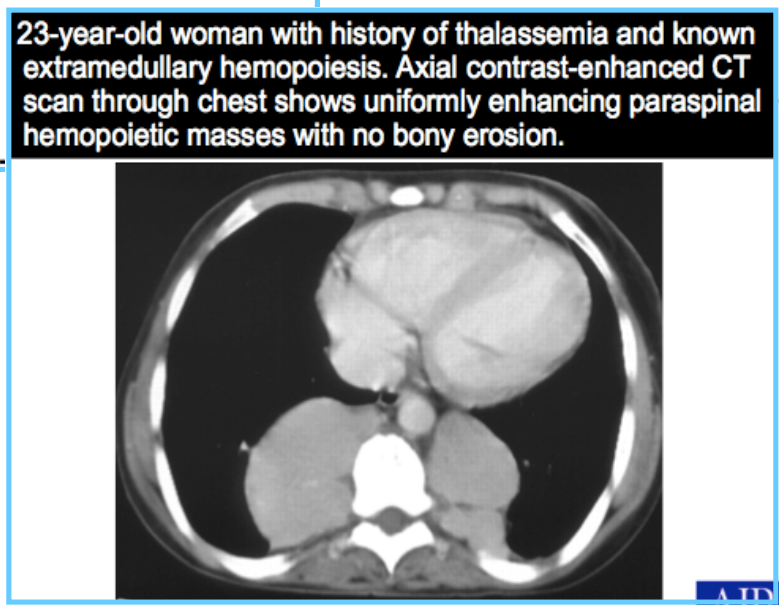


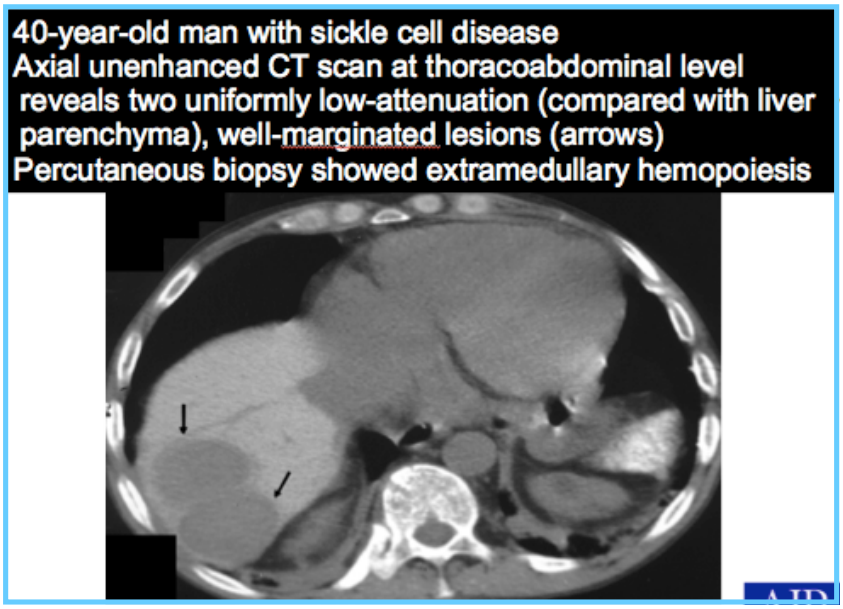
The patient has myelofibrosis → which is fibrosis in the marrow cavity → no RBC production in the marrow >> Spleen is enlarged and taking over, and is the new site for RBC production. (extramedullary hematopoietic areas.)



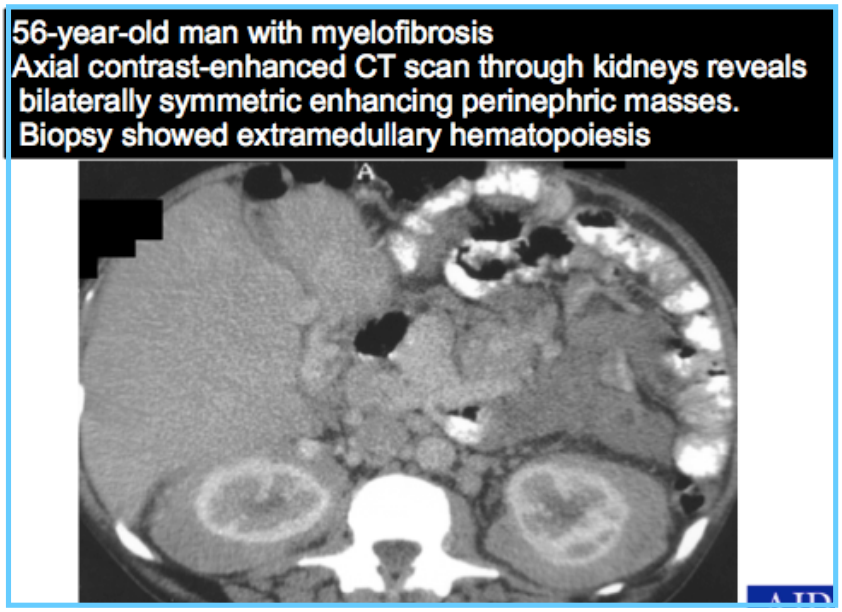
There are some signs of thickening and expansion of the ribs.

But there are also bilateral paraspinal areas → extramedullary hematopoietic areas.

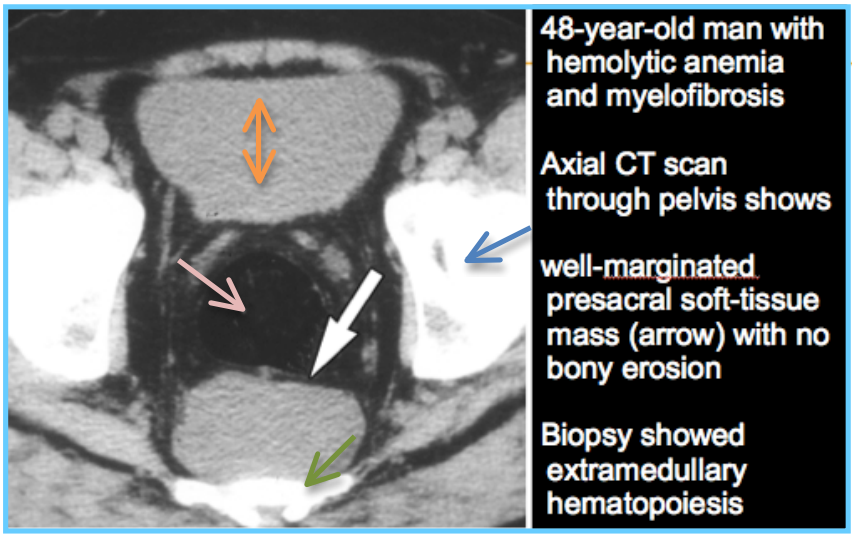




There are focal masses within the liver, which are also extramedullary hematopoiesis.



There is some soft tissue around the kidneys, this is perinepheric extramedullary hematopoiesis.



Urinary Bladder, Pelvic bones, Rectum and Sacrum.

There is a soft tissue density between the rectum and Sacrum [which is abnormal] → extramedullary hematopoiesis.

Sickle Cell Anemia:-

Sickle cell disease may be manifested as
- ANEMIA
  - growth failure, hyperkinetic heart failure
  - expansion of intramedullary hematopoiesis
  - Presence of extramedullary hematopoiesis
- VASO-OCCLUSION
  - Infarcts in spleen, bone marrow, kidney, bowel, brain, muscles
- SUPERIMPOSED INFECTION

Pneumonia (*Pneumococcus, H. influenzae, Staph. aureus, Chlamydia, and Salmonella*)

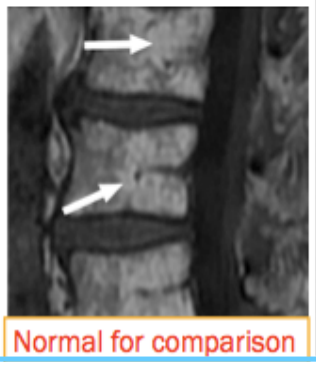
Osteomyelitis (*Salmonella*)

Red marrow in vertebral bodies in a 7-year-old girl with Sickle Cell Anemia.

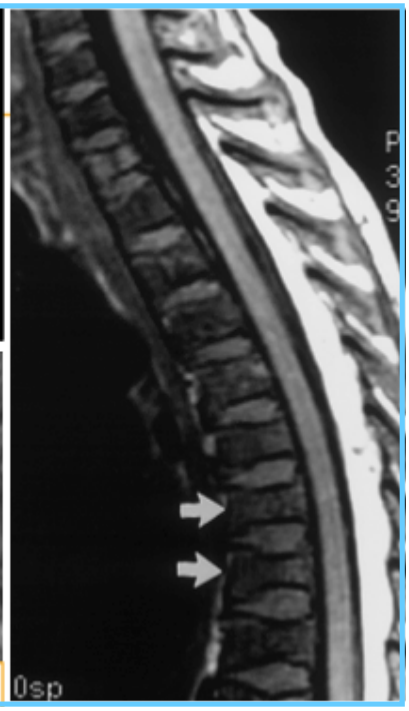
Sagittal T1-weighted MRI of spine shows

Low signal intensity in vertebral bodies compared to discs

H-shaped vertebrae (arrows in right image) due to osteonecrosis of vertebral endplates



Normal for comparison



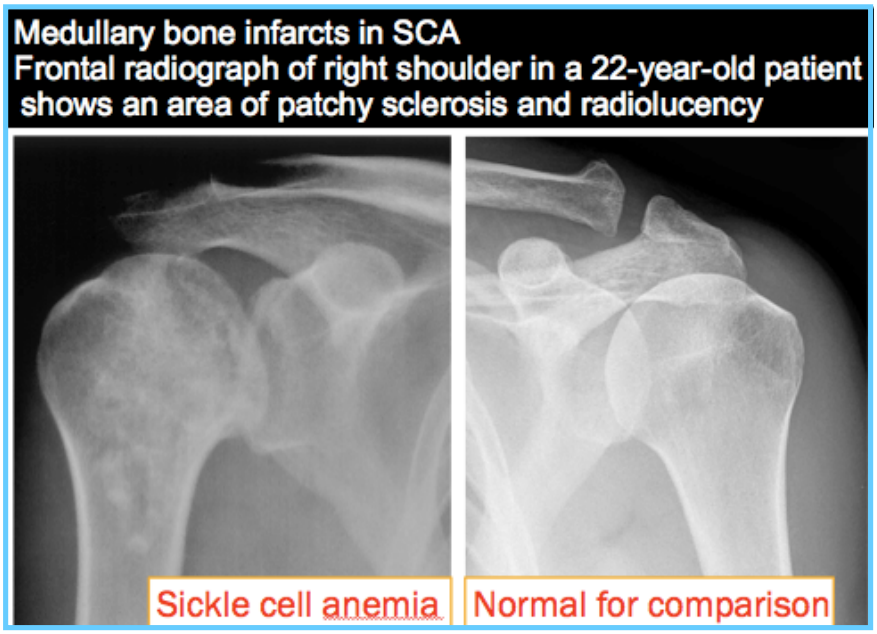
Normal:- the vertebral body is relatively bright compared to the disc.

Abnormal:- the vertebral body is darker compared to the disc (this indicates Fatty Marrow), which is a sign of most types of anemia. [non specific]

The vertebral body is also H-shaped which is central depression due to osteonecrosis → specific to sickle cell anemia.

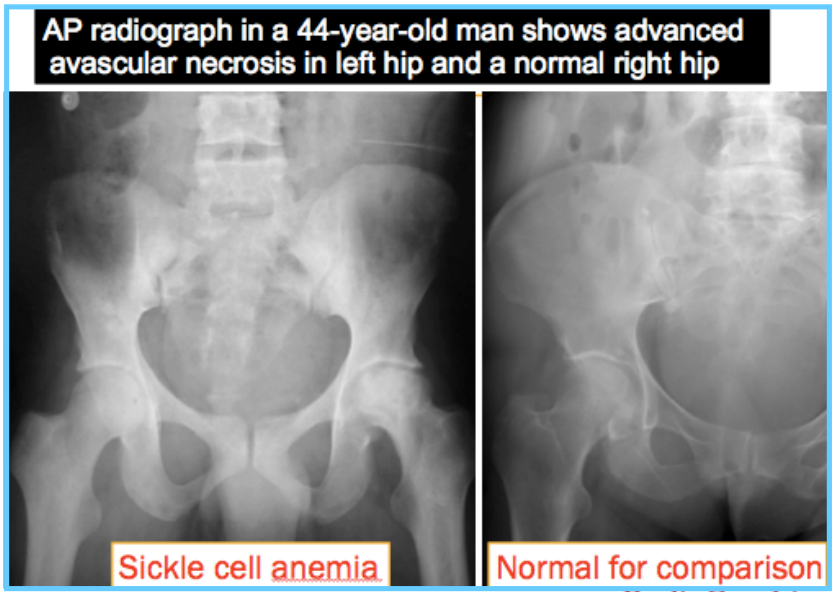
- Bone infarcts typically occur in the medullary cavities and epiphyses. (More vascular areas)

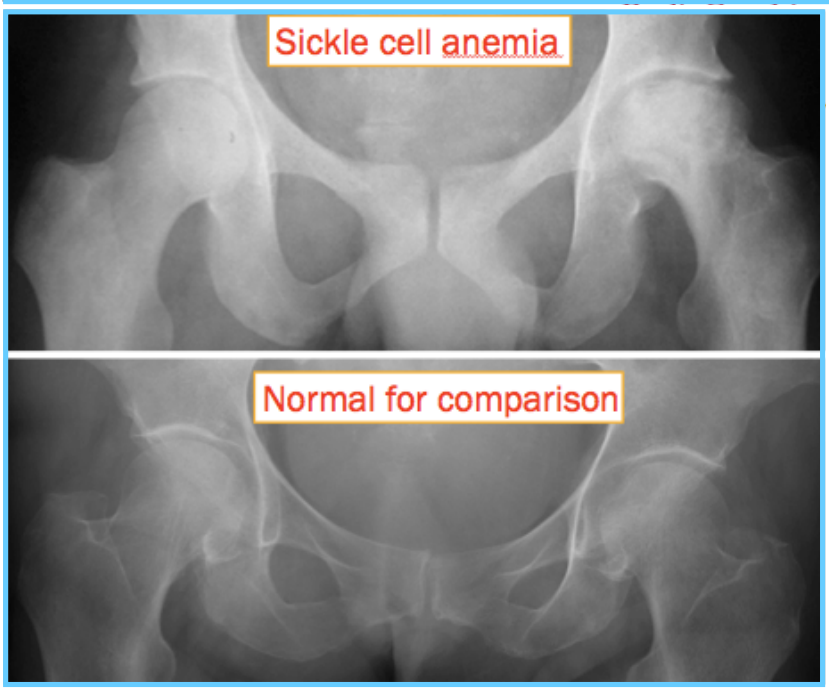
- Epiphyseal infarcts are frequently seen in the femoral and humeral heads, and more often bilateral than avascular necrosis due to other diseases.



Normal:- there's homogenous density and a very sharp outline of the humerus.

Abnormal:- its very patchy, with dark and bright areas which indicates osteonecrosis of the humeral head.





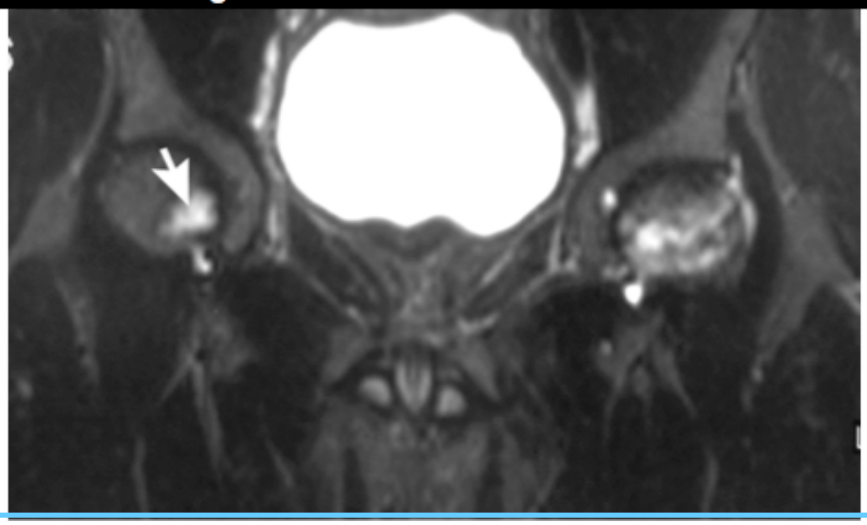
Abnormal:- the femoral head is more dense, patchy, and the outline is not smooth. Also there's some compression and it started to collapse → Osteonecrosis.

On X-Ray the right femoral head looks normal while the left looks abnormal.

However on MRI *next page* it shows minor changes on the right side.

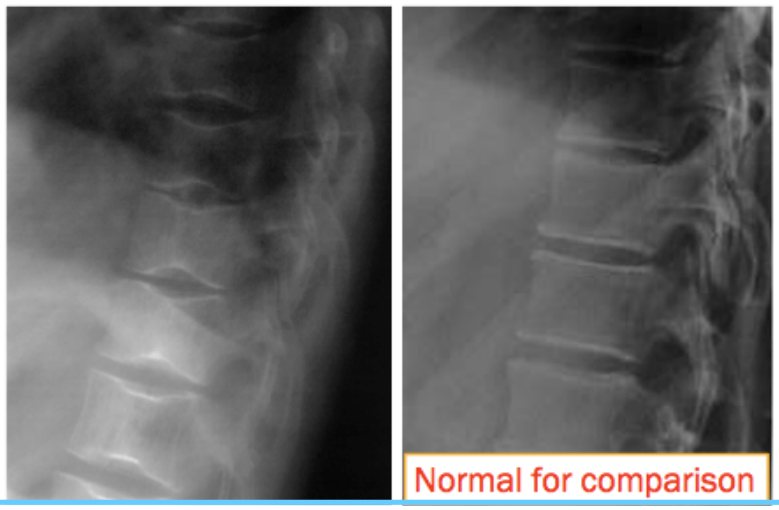
So the x-ray is good for screening but it will NOT show all the lesions, some can be missed. (X-rays should not be considered as a final investigation.)

Coronal STIR MR image in the same patient shows stage 1 avascular necrosis in right hip (arrow) as well, in addition to advanced changes of avascular necrosis of left femoral head



On MRI there was advanced changes on the left femoral head, but minor changes on the right femoral head.

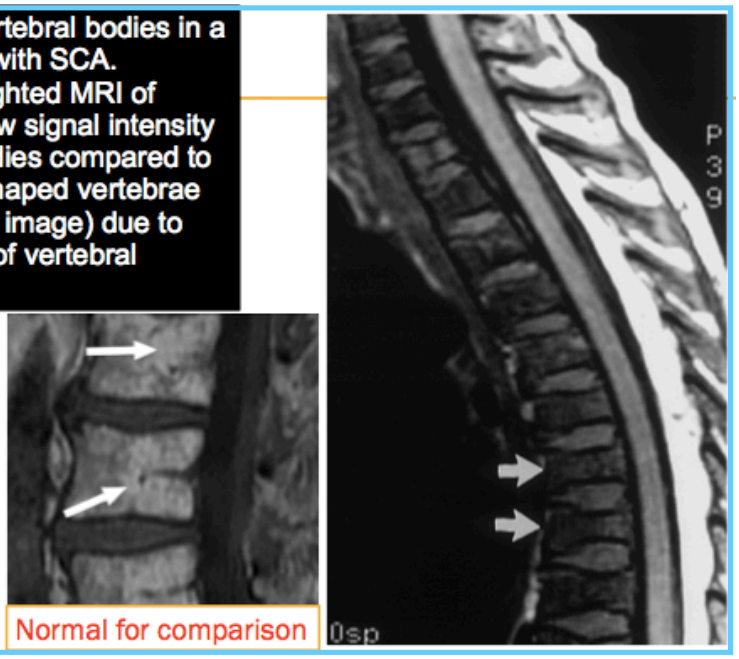
H-shaped vertebrae in a 15-year-old patient with SCA Lateral radiograph of spine shows classic boxlike endplate depressions in middle portion (see the lowest vertebra shown) due to osteonecrosis of the vertebral endplates

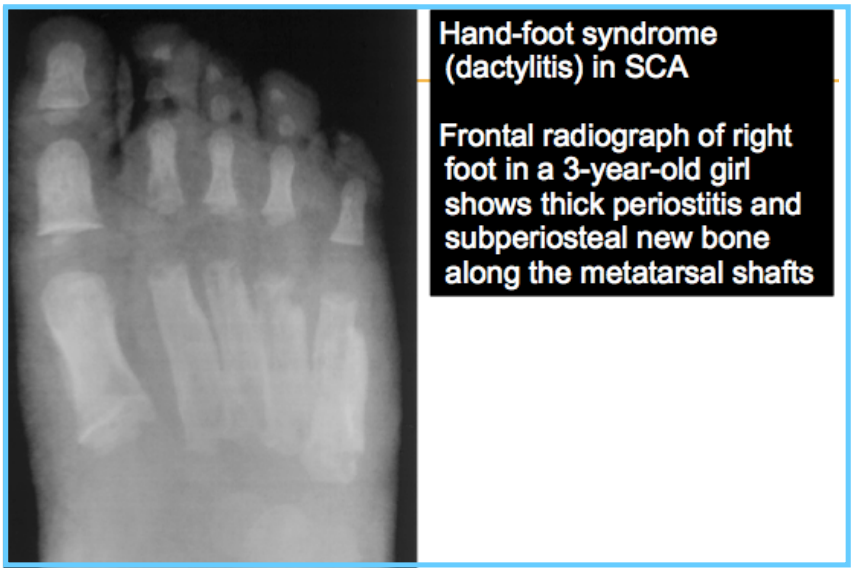


Central depression.

Nowadays, it is rare to see patients with H-shaped vertebrae due to early diagnosis and treatment; however if you don't treat the patient and he stays anemic for 5-10 more years, there will be very well defined H-shaped vertebrae.

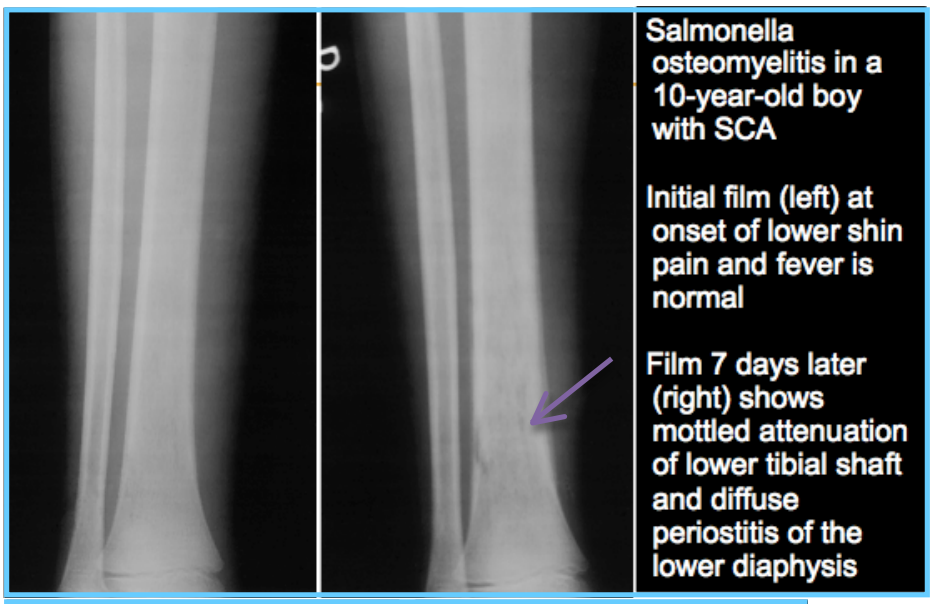
Red marrow vertebral bodies in a 7-year-old girl with SCA. Sagittal T1-weighted MRI of spine shows low signal intensity in vertebral bodies compared to discs, and H-shaped vertebrae (arrows in right image) due to osteonecrosis of vertebral endplates

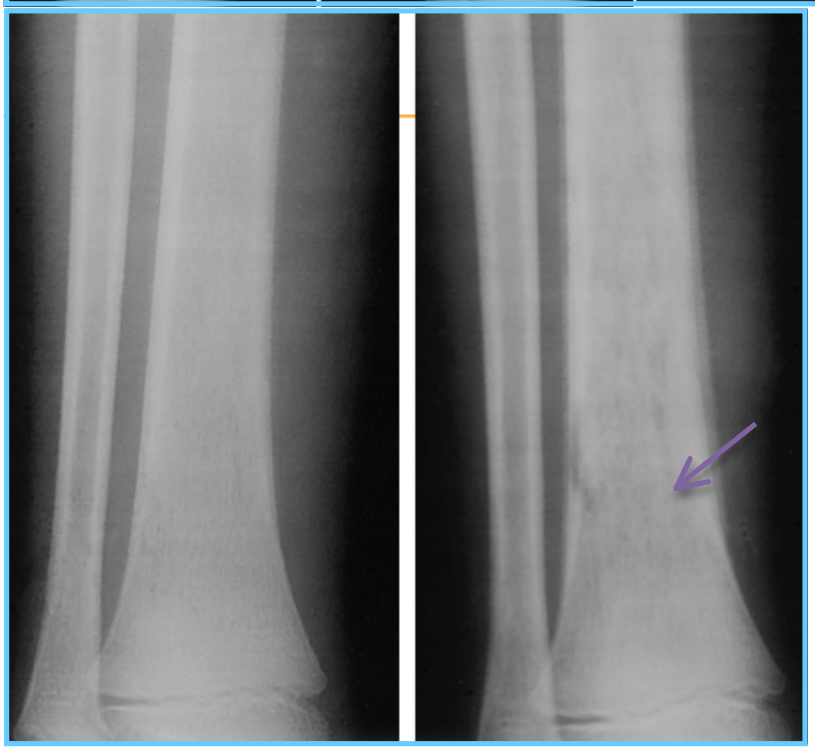




The doctor said you can skip it.

Dactylitis is a condition in which the fingers or toes develops inflammation. There's swelling, tenderness and fever. It's usually the first sign of sickle cell anemia in infants.





Is it an infection or infarction??

On x-rays its difficult to differentiate between infection and infarction.

There's some abnormality in the marrow cavity, you can see some black areas.

Bone infarcts and osteomyelitis are difficult to differentiate on history, clinical examination and plain x-ray images but are very important to avoid complications of osteomyelitis.

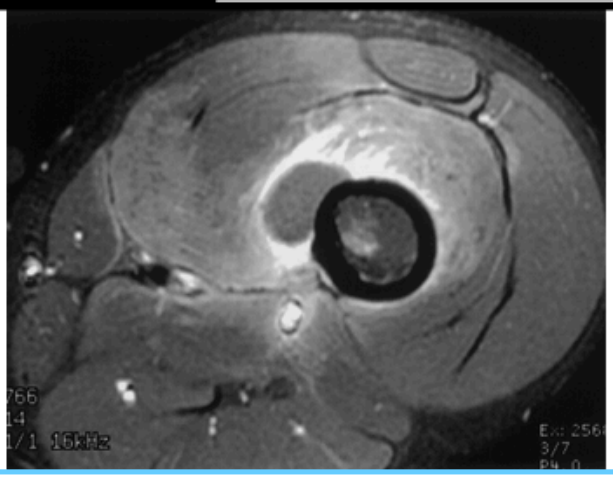
MRI findings of:-

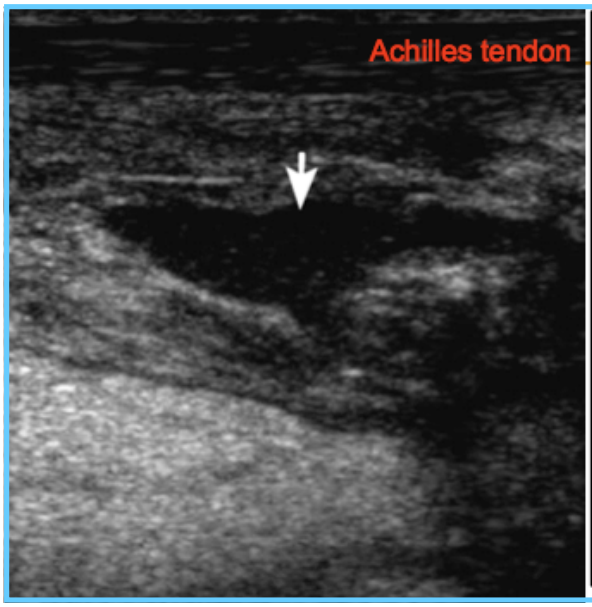
- Cortical defects in bone
- Adjacent fluid collections in soft tissue
- Bone marrow enhancement

→ Are highly suggestive of infection

Ultrasound guided aspiration of fluid collection around the involved bone can be confirmatory.

Osteomyelitis of femur in a 24-year-old patient with SCA
Axial T1-weighted MRI after contrast shows heterogeneous enhancement of marrow cavity, a rounded low-signal-intensity area adjacent to the shaft that is non-enhancing (fluid collection), and enhancement of the soft tissues around the shaft and of the adjacent musculature. Areas of enhancement are likely infected





Soft-tissue infection in a 52-year-old man with homozygous sickle cell disease.

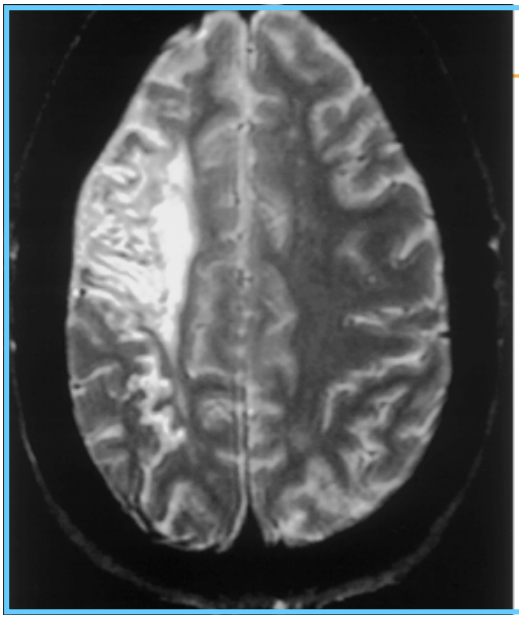
Longitudinal high-resolution ultrasound image of left ankle shows a hypoechoic (dark) fluid collection (arrow) deep to Achilles tendon. Thick pus was aspirated from this area under ultrasound guidance

MRI → any fluid collection in a patient with known sickle cell anemia indicates infection.

Even on ultrasound you can see the fluid accumulation.

If a patient with known sickle cell anemia came in with pain in his knee or ankle, and there's fluid accumulation on imaging, you can assume that it's an infection.

(The doctor didn't focus on these images. Just that to diagnose an infection you use MRI or ultrasound)



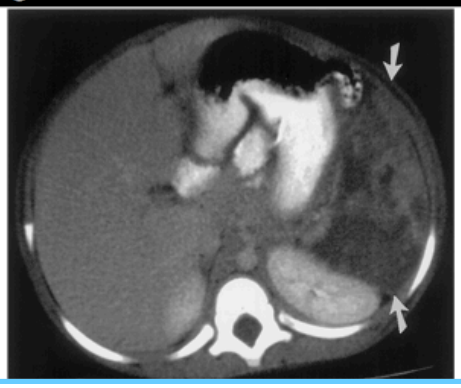
Chronic infarct in a 19-year-old patient with SCA and longstanding mild left sided weakness

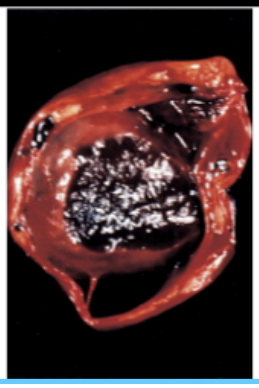
Axial T2-weighted MRI shows an area of high signal intensity and enlargement of overlying CSF spaces, compatible with chronic infarction and atrophy

Infarctions can occur anywhere.

Here it's in the Brain.

Sequestration syndrome with splenic infarction in SCA
Axial CT after contrast shows enlarged spleen that enhances heterogeneously and minimally with large non-enhancing areas (arrows)
Photograph of spleen in a different patient shows areas of congestion and central necrosis

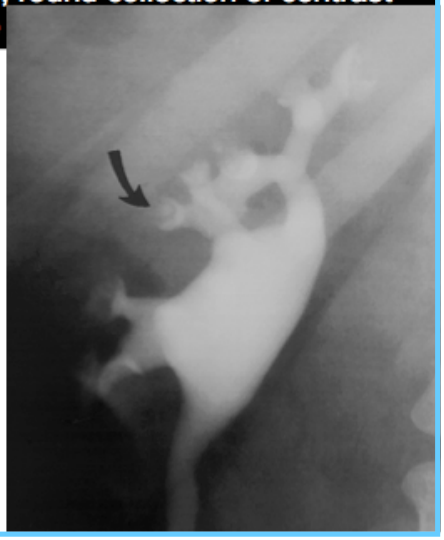




<< Spleen. The entire spleen was infarcted.

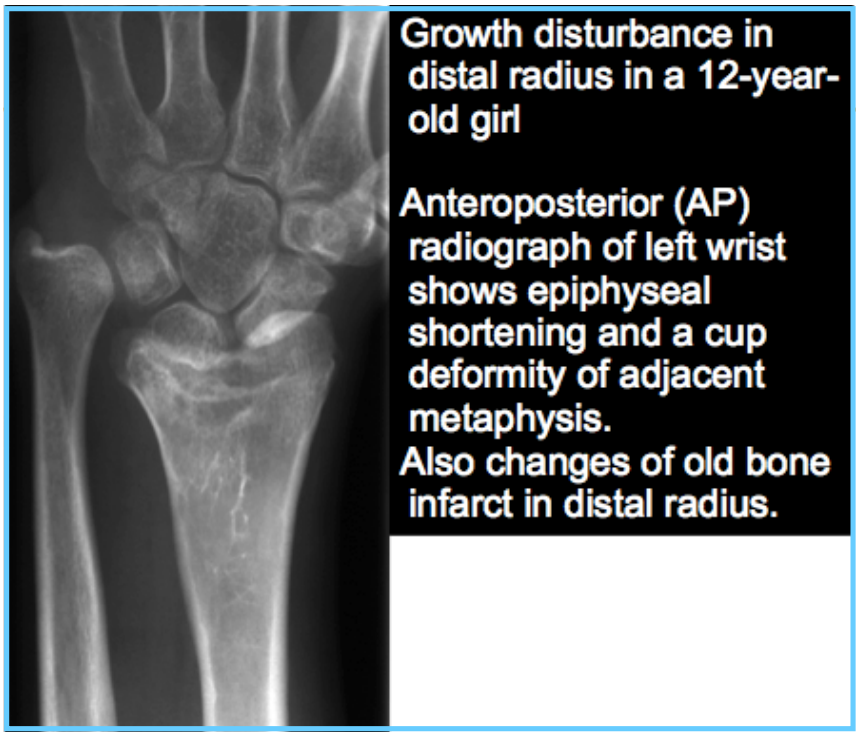
Papillary necrosis in SCA

Frontal view of kidney during excretory urography in a 32-year-old man with SCA shows a small, round collection of contrast material in a missing papillary tip



Photograph of a kidney from a different patient shows loss of papillary tips in some upper pole pyramids (arrows).

It can also occur in the renal papilla. There are some infarcted areas.



Infarction can also occur in the epiphysis. When the epiphysis becomes infarcted (its dead) it will stop growing, furthermore if the child is still growing deformities will occur.

<< Here the radial epiphysis is infarcted and it stopped growing, however the ulna is still growing so that it's overgrown, when usually the ulna and radius should grow parallel to each other.

When there are some focal high-density and some low-density areas that are mixed, it indicates infarction.

Lymphoma:-

Hodgkin's Disease
- Lymphocytic predominance
- Mixed cellularity
- Lymphocytic depletion
- Nodular sclerosis - the most common

Non Hodgkin's Lymphoma
- Burkitt lymphoma (jaw and abdomen)
- Burkitt-like lymphomas (abdomen and nodes)
- Large B-cell lymphomas (abdomen and nodes)
- Lymphoblastic lymphoma (Mediastinum, nodes, bone marrow)
- Anaplastic large cell lymphoma (Nodes, skin, soft tissue, bone)
- Other peripheral T-cell lymphomas
- MALT lymphoma

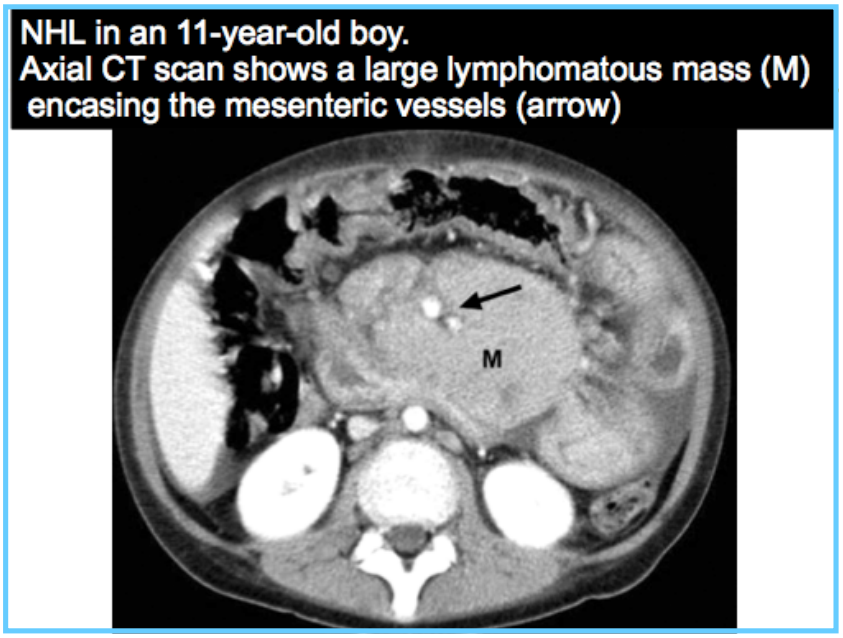
Lymphoma can present as mass anywhere in the body

Lymphoma is diagnosed on a hematological basis, not radiological.

The role of radiology in lymphoma:-

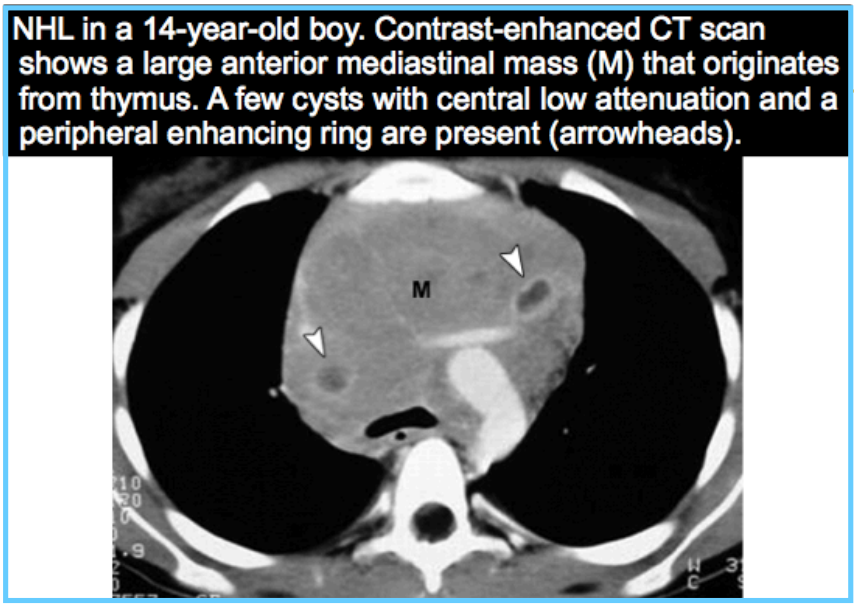
1. Stage the disease. (Stage the extent of the disease, if it's in one group of lymph nodes or multiple, if its in both sides of the diaphragm or one side, and whether it has visceral involvement or not, for example the liver, spleen, etc.)
2. CT-guided biopsy, if the mass is not easily accessible. (Patients initially present with a mass, and to diagnose they have to get a biopsy. if that mass is in the chest or abdomen, further imaging of the axial and inguinal area will be needed, and if a mass was found that is more easily accessible a surgical biopsy will be performed. However, if the mass was localized to chest or abdomen, a CT-guided biopsy will be done.)

Lymphoma can appear as mass-like lesion ANYWHERE in the body.



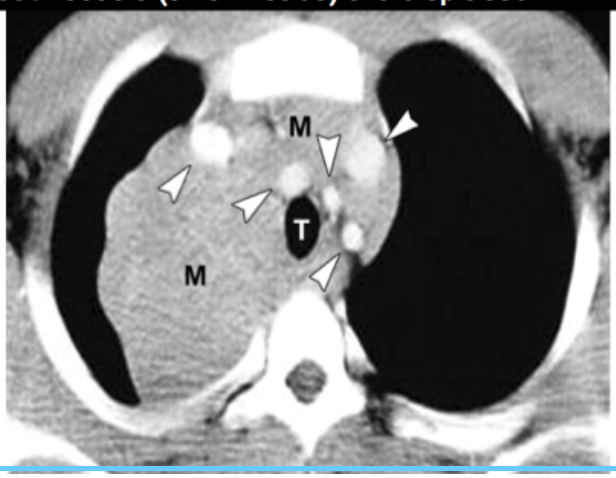
A mass in the abdomen.

The doctor skipped this slide.



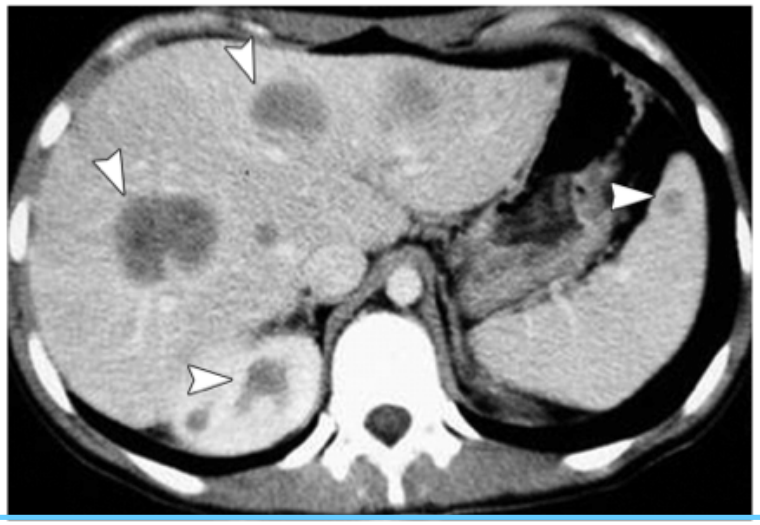
This patient has multiple chest lesions, which are matted together and are compressing the airway posteriorly. If this patient has no other mass they'll perform a CT-guided biopsy from here.

HD in a 17-year-old boy. Contrast-enhanced CT scan shows a large mediastinal mass (M). Trachea (T) is compressed, and great vessels (arrowheads) are displaced

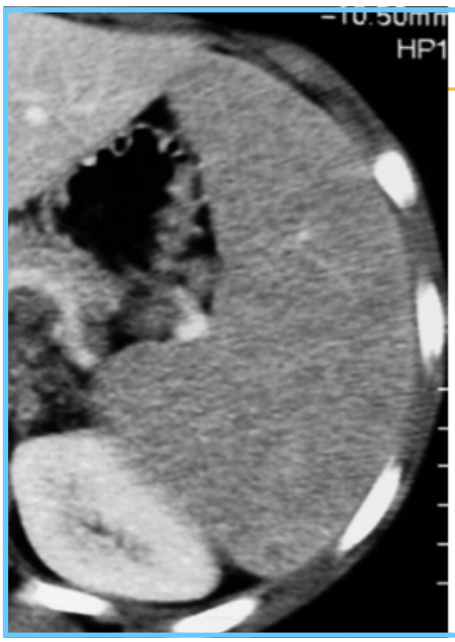


Multiple mediastinal masses.

NHL in a 16-year-old girl. Contrast-enhanced CT scan shows low-density lesions (arrowheads) in both hepatic lobes, with small nodules in spleen and right kidney.



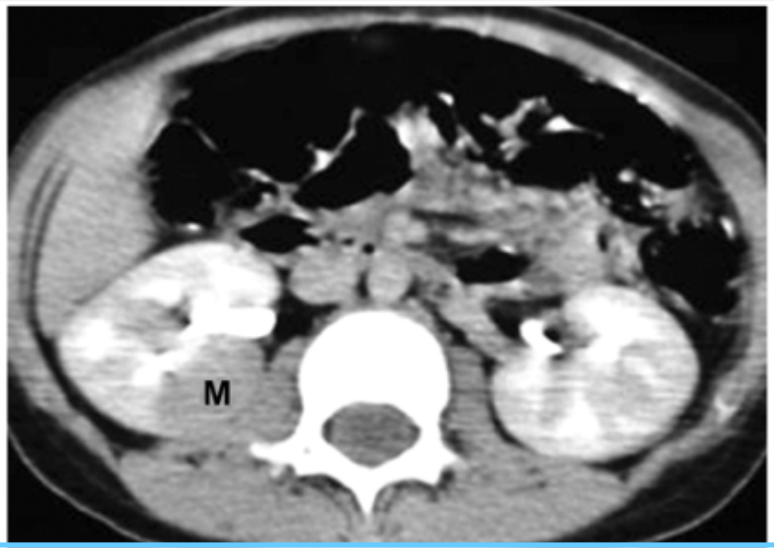
This patient has multiple liver lesions as well as spleen lesions.



HD in a 12-year-old girl Contrast-enhanced CT scan shows an enlarged spleen with a diffusely inhomogeneous appearance.

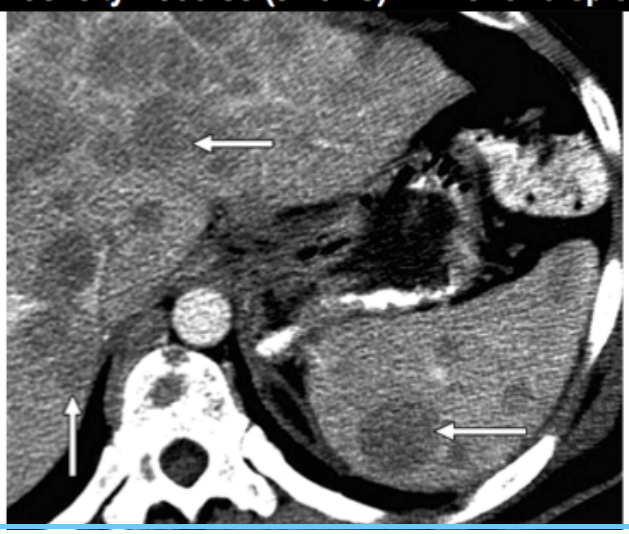
This patient has splenomegaly with small multiple lesions.

NHL in a 14-year-old boy. Contrast-enhanced CT scan shows single well-defined, hypoattenuating mass (M) in right kidney

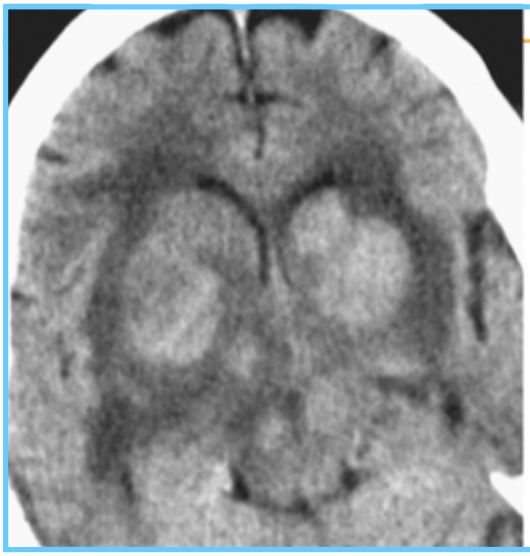


A mass in the kidney.

Diffuse hepatosplenic involvement in lymphoma
Axial CT scan shows multiple round, homogeneous, low density nodules (arrows) in liver and spleen



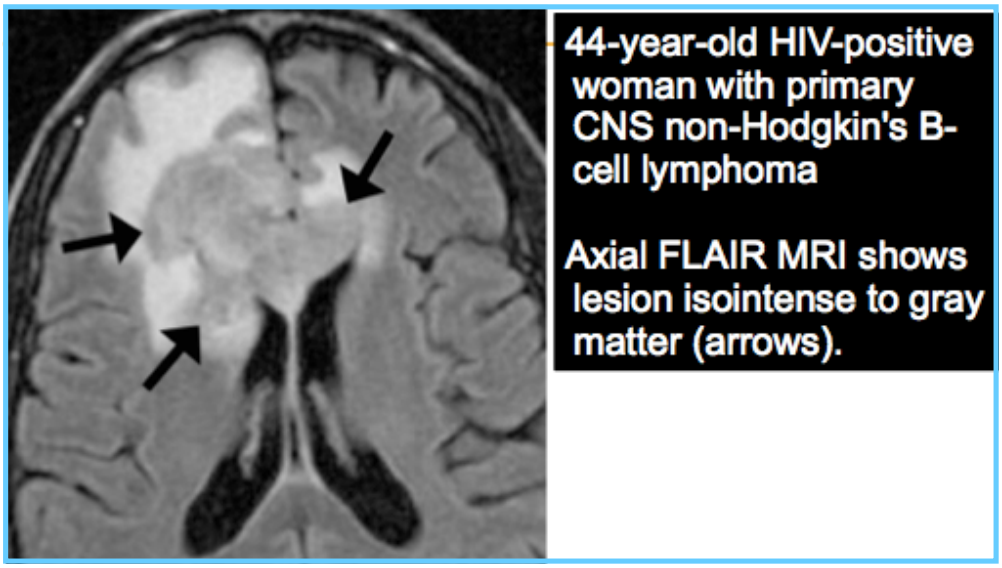
Liver and splenic masses.



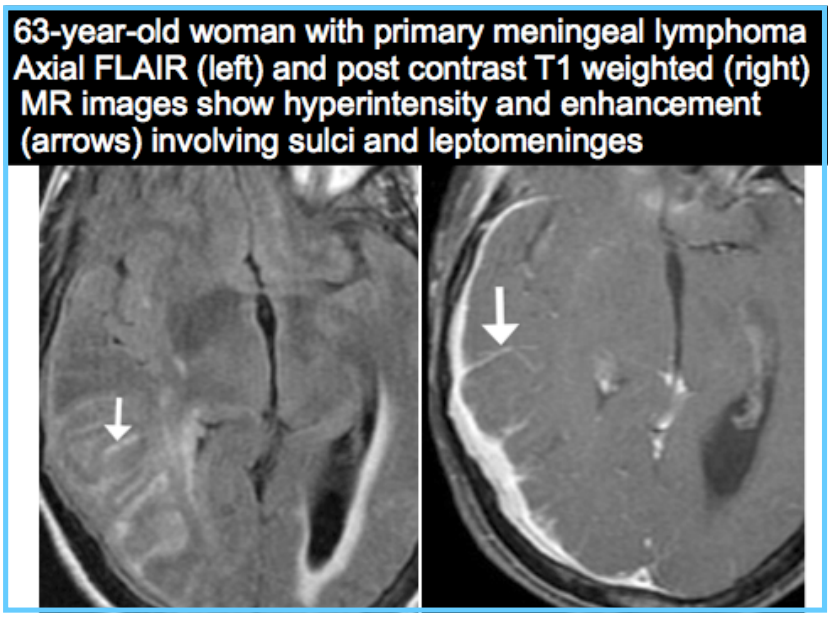
72-year-old immunocompetent woman with primary CNS non-Hodgkin's B-cell lymphoma

Unenhanced CT image shows classic hyperdense masses involving deep white and gray matter.

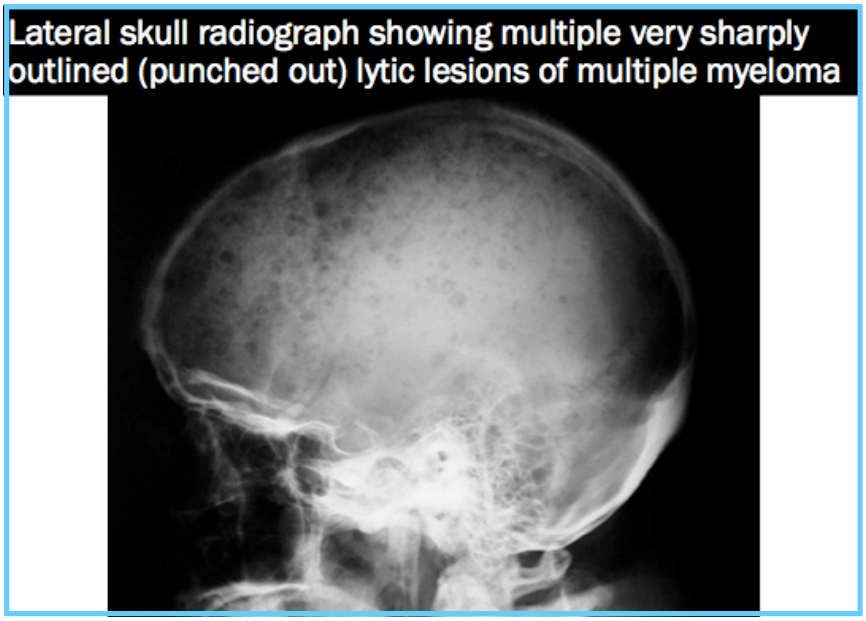
Mass in the brain.



A mass in the brain.



Meningeal lymphoma.



Small numerous lesions in the skull >> multiple myeloma. The radiological role is to stage the disease.

The Doctor's Summary From The Lecture

- INTRA medullary hyperplasia can be seen in Thalassemia, Sickle cell anemia, Iron deficiency anemia, Any severe chronic anemia except bone marrow failure.

- Signs of INTRA medullary hyperplasia include:
    - Expanded bone marrow in bones including long bones of hands, feet, limbs, and skull.
    - Decreased T1 MRI bone marrow signal than adjacent discs.

- EXTRA medullary hematopoiesis can be seen in ALL SEVERE CHRONIC ANEMIAS.

- Sites of EXTRA medullary hematopoiesis include Liver, Spleen, Paraspinal areas with possible extension into spinal canal outside the dura, Kidneys, Meninges, Skin, Lymph nodes, Thymus.

- EXTRA medullary hematopoiesis appears as homogeneous soft tissue masses on imaging.

- INFARCTS and INFECTIONS are additional findings in Sickle cell anemia.

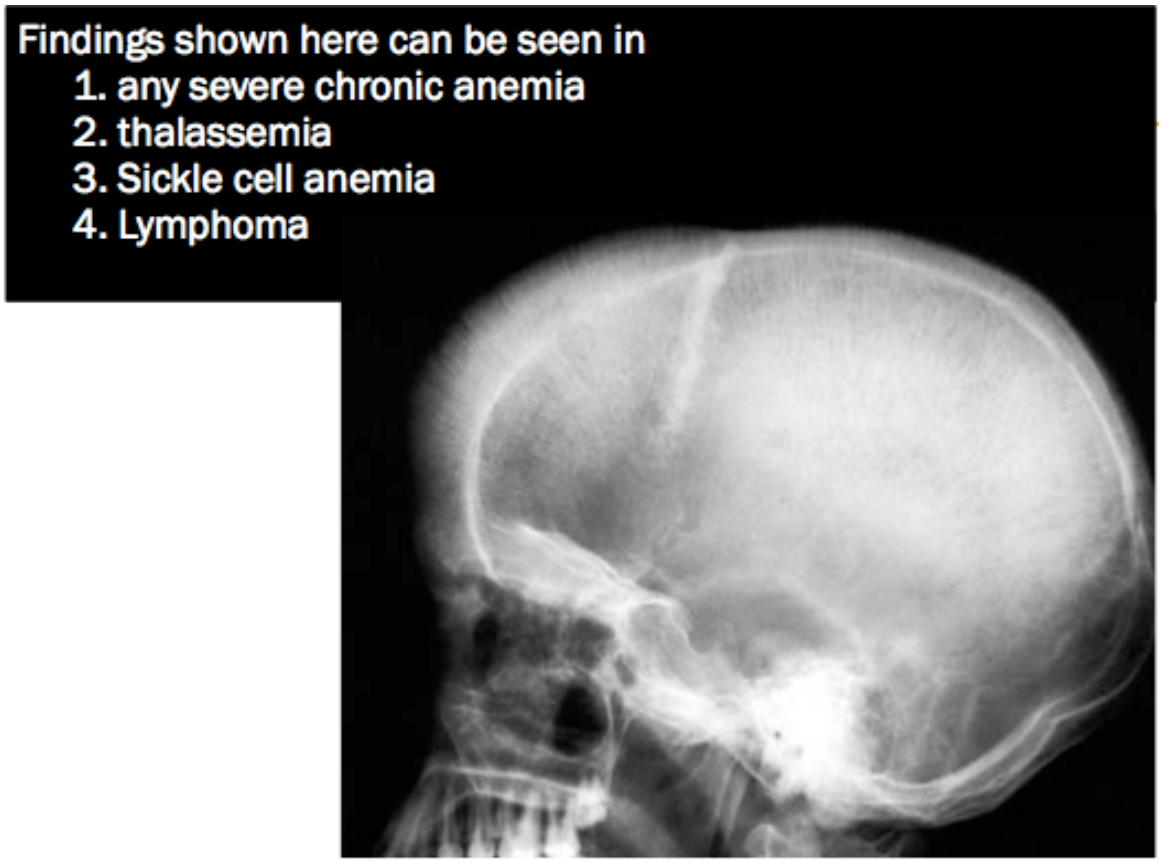
- Bone infarct vs infection is important to diagnose early so that antibiotics can be started early to prevent complications. MRI with contrast and ultrasound /CT guided aspiration of fluid collections are very helpful if imaging is unable to differentiate these two.

- Multiple myeloma produces punched out lytic lesions in bones with background bone appearing normal. Opposite to bony metastases, myeloma more often involves intervertebral discs and mandible, and less often involves pedicles.

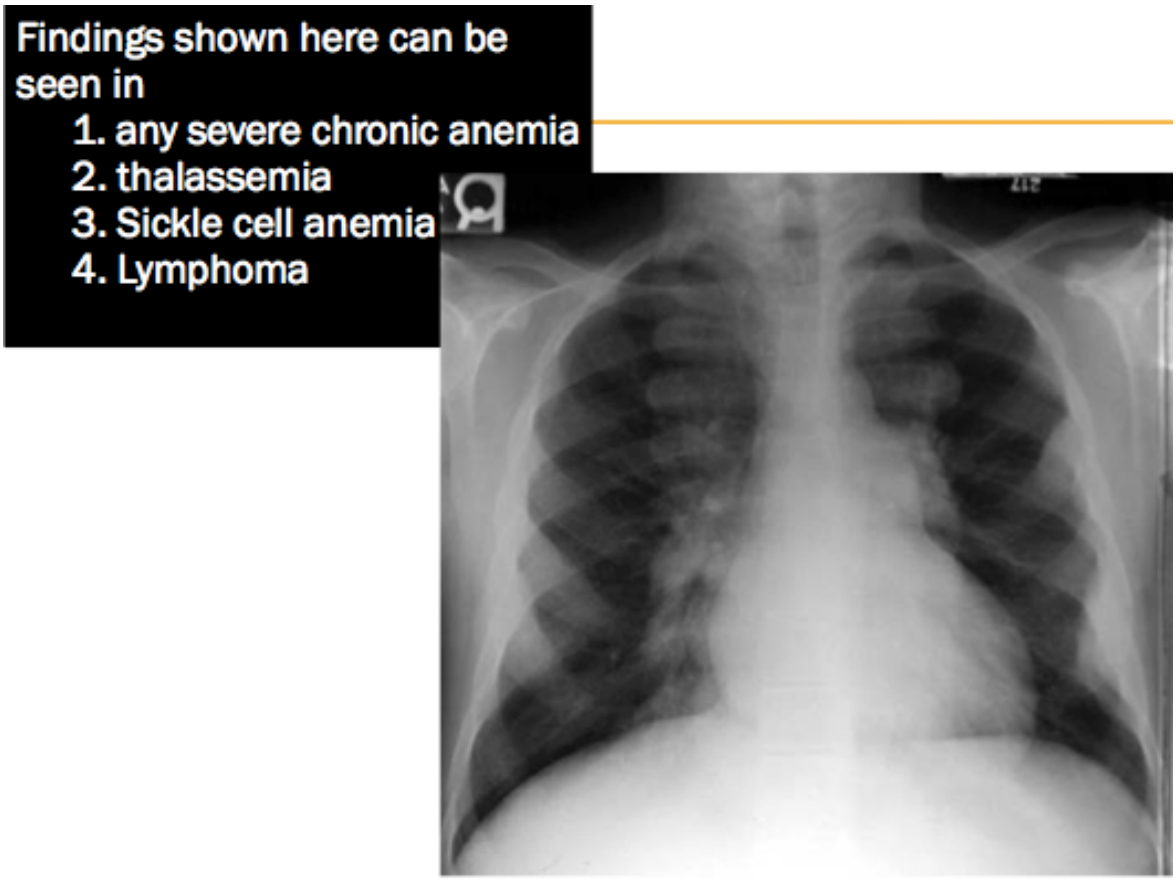
- Lymphoma can produce a mass anywhere in the body. CT is often used to scan whole body to evaluate the disease extent (staging), and to do CT-guided biopsy to make tissue diagnosis if not already diagnosed. Rest is all by laboratory and clinical based.

The Doctor's Questions

1)

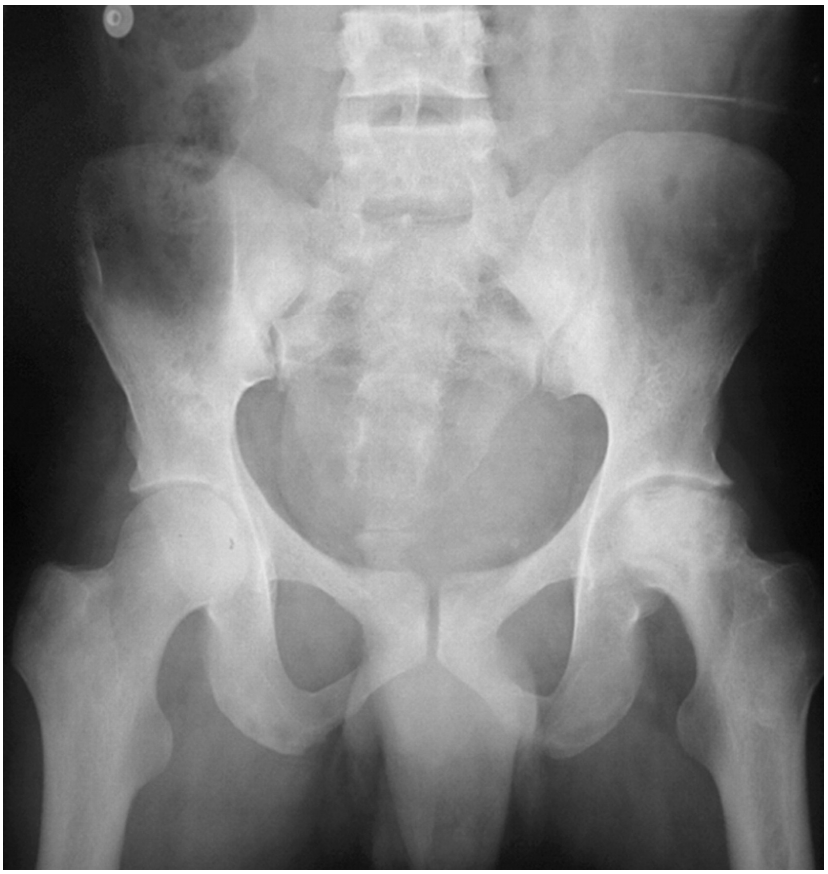


2)



The Team's Questions

3) In the following plain anteroposterior (AP) view of the pelvis. Which one is correct?

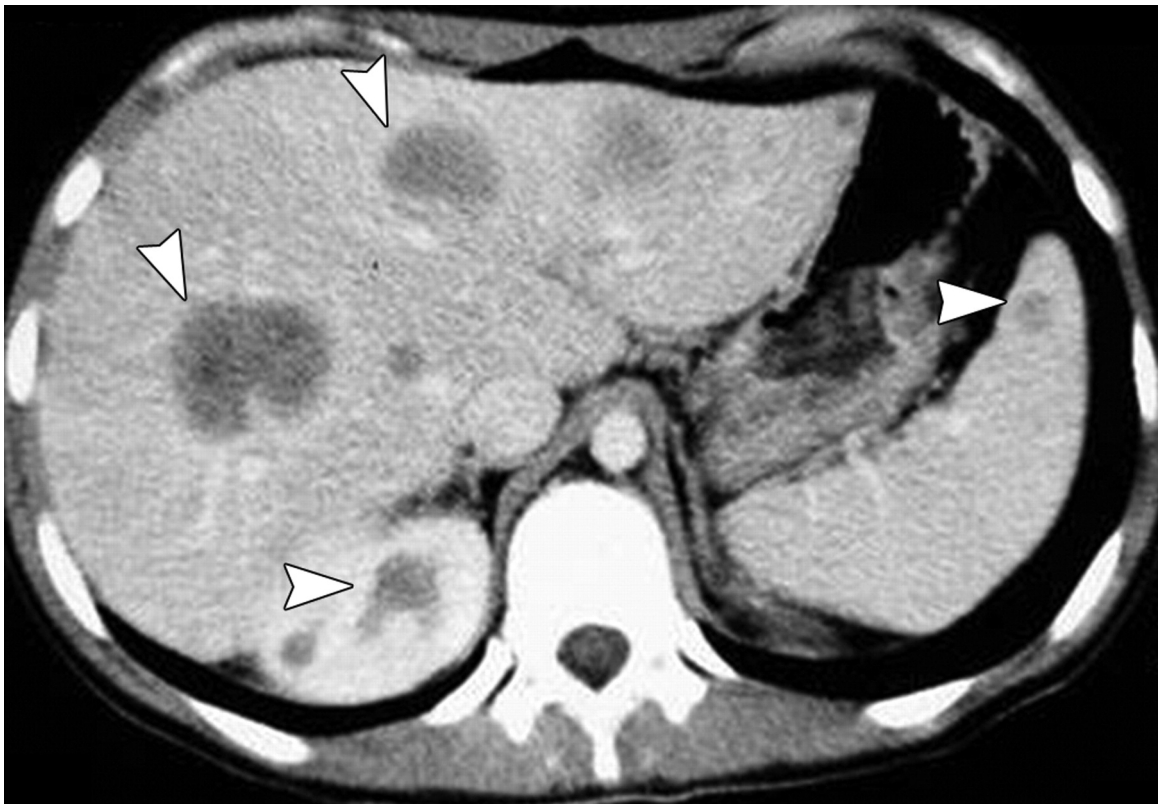


a) Right femoral head is larger than left due to focal bone marrow hyperplasia and favors diagnosis of Lymphoma.
b) Left femoral head is partially collapsed and is very characteristic of Osteomyelitis.
c) Left femoral head is partially collapsed due to bone infarct and favors diagnosis of Sickle Cell Anemia.
d) Right femoral head is normal and excludes possibility of early infection or infarction.

4) Which ONE of the following is true in a patient with chronic thalassemia?

a) Paraspinal bilateral soft tissue nodules in lower thoracic region are non-specific and needs biopsy for confirmation.
b) Hair-on-end appearance of skull due to bone marrow hyperplasia classically affects the occipital bone.
c) Hematopoietic tissue can be seen in the spinal canal sometimes causing compression of the spinal cord.
d) Spleen is often shrunken due to repeated infarcts.

5) Which one of the following is true regarding the multiple masses seen in liver, spleen, and kidney in the following image?



a) Highly specific for Lymphoma.
b) Very likely of Multiple Myeloma.
c) Non-specific and needs biopsy for confirmation.
d) Normal if patient is anemic.

Answers:

1) 1.
2) 1.
3) C.
4) C.
5) C.