RADIOLOGY
TEAM 435

Radiology of Hematopoietic disorders

[Color index: Important | Notes | Extra | Editing file]

- **Objectives:**

- **Resources:**
  - 435 slides
  - 434 team

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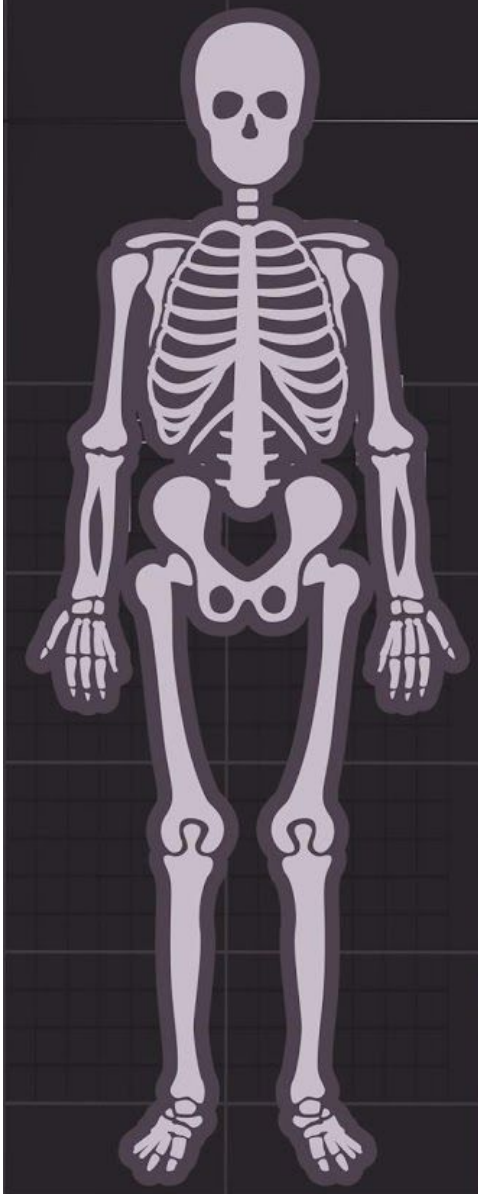
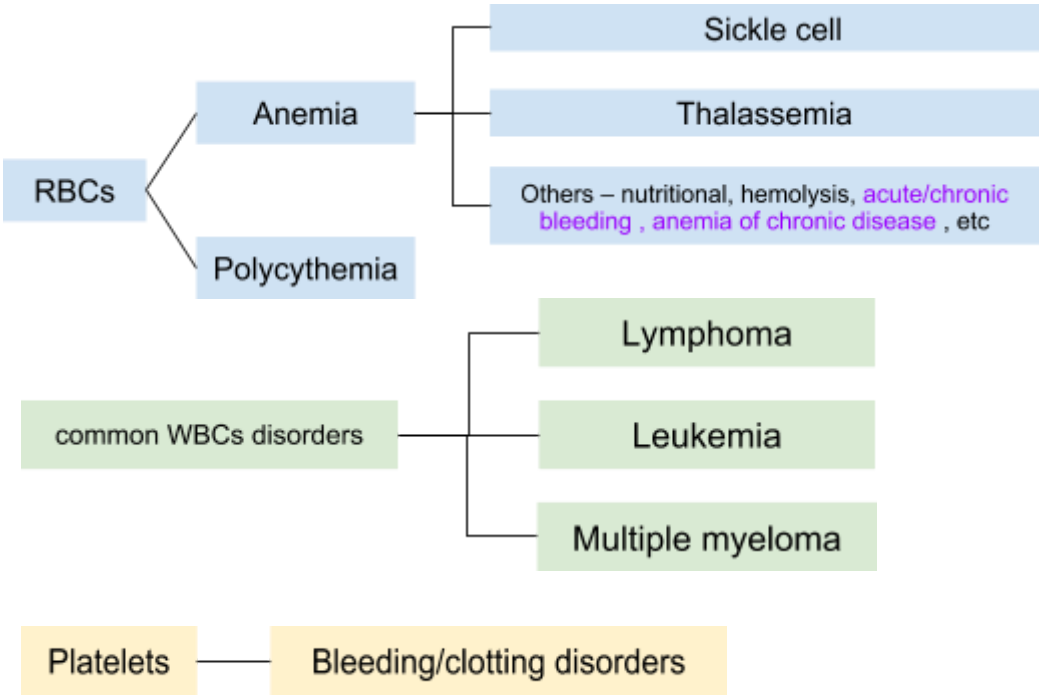


Introduction

blood contents :

1. Cells: RBCs, WBCs, platelets.
2. plasma



- long standing/genetic anemia such as thalassemia and SCA, which are chronic (stay with the patient), will produce some signs(vs the others which are temporary types (hemolytic and nutritional)
- No Radiologic signs in polycythemia.

ANEMIA features on imaging :	
Reactive increase in red bone marrow	• first response: your body will increase the RBCs production in bone marrow, which is called intramedullary hematopoiesis. • Expanded bone marrow in bones including long bones , more obvious in hands, feet, limbs, skull when the anemia has been there for sufficiently long time and SEVERE • visible on X-ray • Decreased T1 MRI signal in vertebral body bone marrow than adjacent discs
New marrow areas in potential organs	- Extramedullary hematopoiesis: if the previous mechanism wasn't enough and the patient is still anemic, some other sites will try to synthesize RBCs. Or if the previous mechanism isn't working (aplastic anemia/myelofibrosis) - BUT in normal adult these sites don't do hematopoiesis - these sites : Liver, Spleen, Lymph nodes, Thymus, Paraspinal areas with possible extension into spinal canal outside the dura, Kidneys, Meninges, Skin - it's rare nowadays to see extramedullary hematopoiesis in unusual sites like kidney , meninges ,,etc because patients are diagnosed and treated early before they reach this severity
Transfusions Iron overload	• some patients will develop iron overload if they have been treated with blood transfusion multiple time, because our bodies can't get rid of the iron, so it will be deposed somewhere, usually in liver and spleen brain (basal ganglia), pancreas ,,etc • Increased CT density (brightness) • changes in MRI signal of liver & spleen

Sickle Cell Anemia :	
Infections	Pneumonias, Osteomyelitis
Infarcts	anywhere in the body : Spleen "commonest", Kidneys, Brain, Bones

- You can't specify which type of anemia the patient has depending on the radiologic features above, just say chronic/severe anemia.
- But if it's associated with infection/infarcts then the probability of SCA is high.
- If we correct the anemia , the extramedullary hematopoiesis will disappear

cases:

classic appearance of anemia

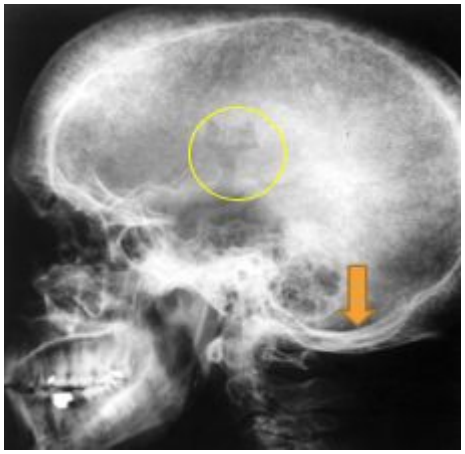
features :

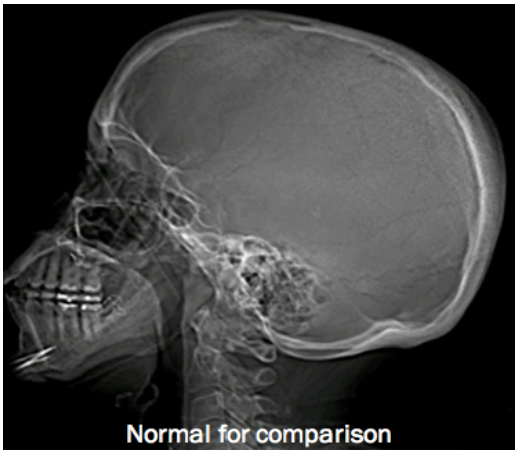
- **Decreased** bone density with coarse trabeculae.
- **Wide** medullary cavity with thin cortex.
- spongy bone.
- remember that x-ray doesn't give you the diagnosis of thalassemia

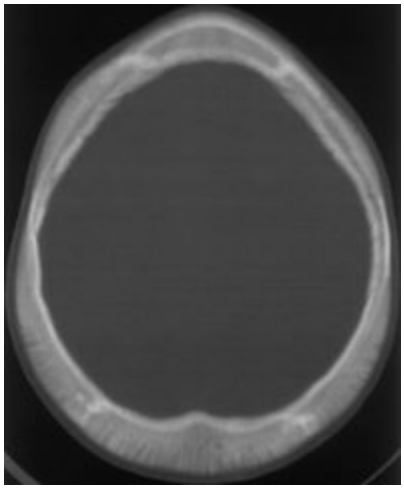
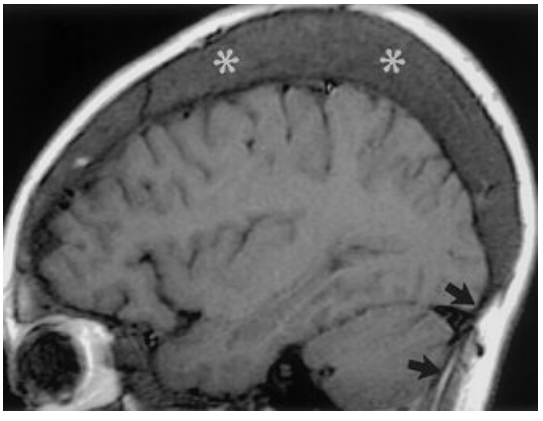
case: 25-year-old man with β -thalassemia.

Lateral skull radiograph shows :

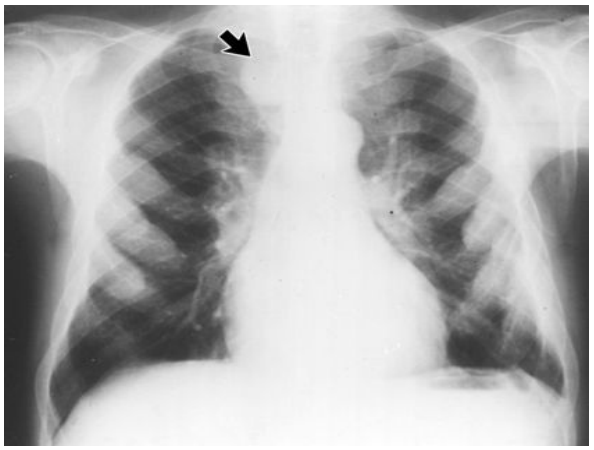
- outward expansion of diploic space with hair-on-end appearance
- widened groove for middle meningeal artery (circle)
- Spared occipital bone (arrow)





Axial CT image of upper skull	Sagittal MRI of brain
	
- **diploic space widening** - **trabecular prominence**	- **diploic space widening representing red marrow (*).** - **arrows → spared occipital bone which has no marrow elements**

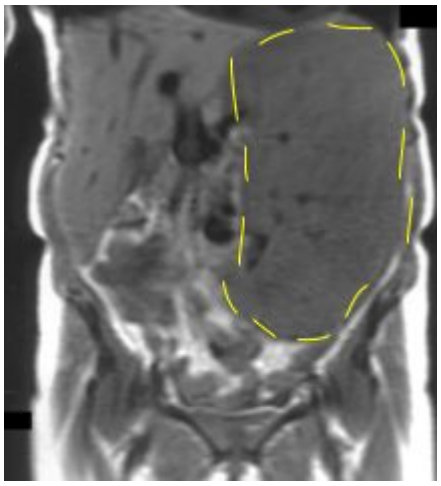
case : 25-year-old man with β -thalassemia.
PA radiograph of chest (left)



- **diffuse expansion of ribs**
- **arrow → right upper paraspinal thoracic mass compatible with extramedullary hematopoiesis.**

normal for comparison

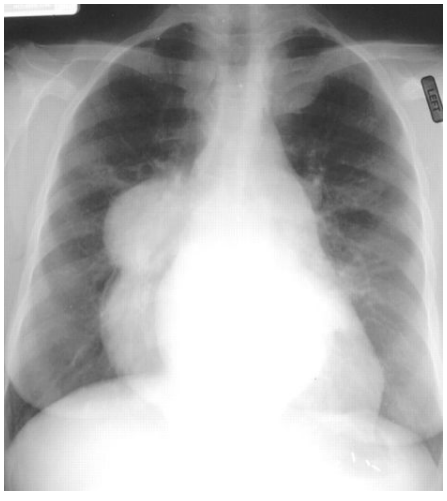
case : 51-year-old woman with myelofibrosis
Coronal T1-weighted MR image :



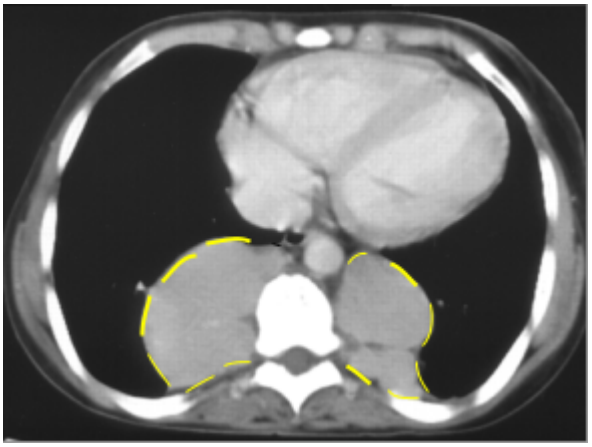
- massively enlarged spleen
- Splenic biopsy was followed by splenectomy
- Pathologic examination revealed extramedullary hematopoiesis
- also common in non-functional bone marrow.

case: 23-year-old woman with history of thalassemia and known extramedullary hemopoiesis

PA chest film (left) & Axial contrast-enhanced CT (right)



well-marginated bilateral, paraspinal masses compatible with extramedullary hematopoietic tissue

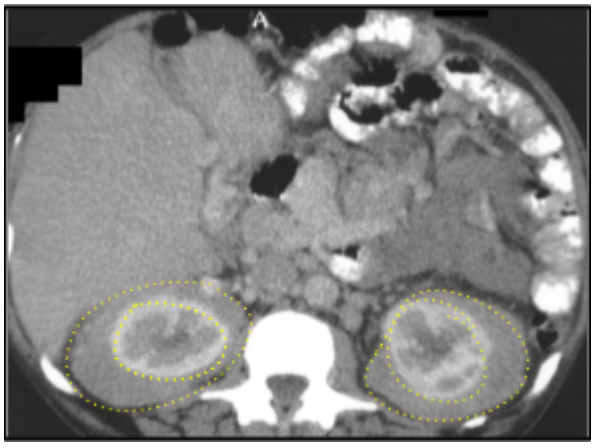


uniformly enhancing bilateral paraspinal hemopoietic masses with no bony erosion

case : 40-year-old man with sickle cell disease
Axial unenhanced CT scan at thoracoabdominal level

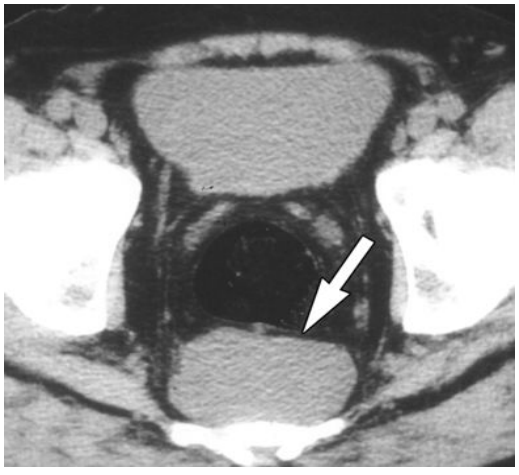
- arrows → two uniformly low-attenuation (compared with liver parenchyma), well circumscribed lesions.
- Percutaneous biopsy showed extramedullary hemopoiesis

case : 56-year-old man with myelofibrosis
Axial contrast-enhanced CT scan through kidneys:



- bilaterally symmetric enhancing perinephric masses. Biopsy showed extramedullary hematopoiesis
- if we don't have a history and we won't be sure; it will be doubtful and may be mistaken by tumor and further investigation is needed

case :48-year-old man with hemolytic anemia and myelofibrosis.
Axial CT scan through pelvis shows:

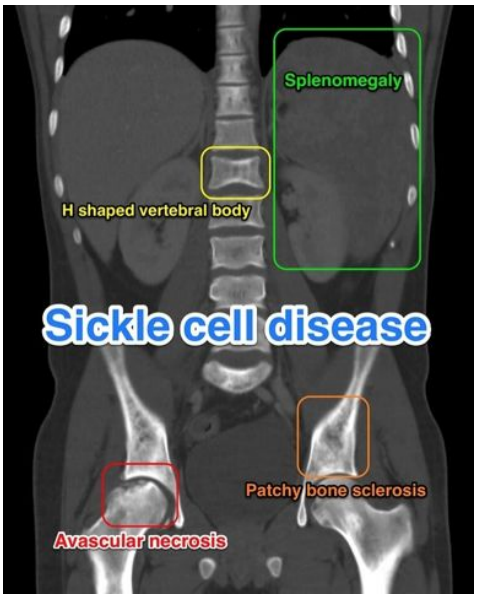


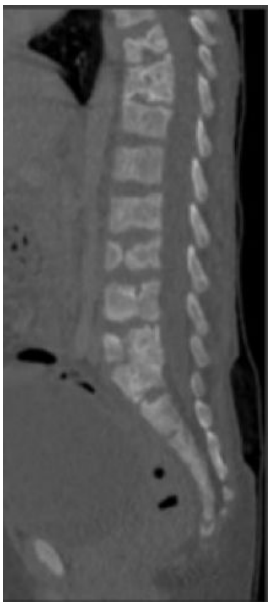
- arrow→ well-marginated presacral soft-tissue mass
- No bony erosion
- Biopsy (not often needed) showed extramedullary hematopoiesis

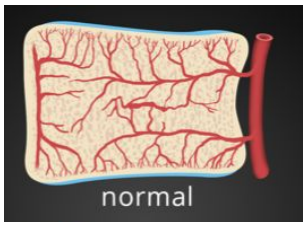
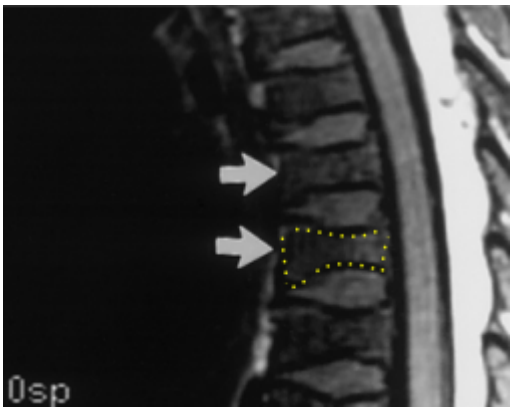
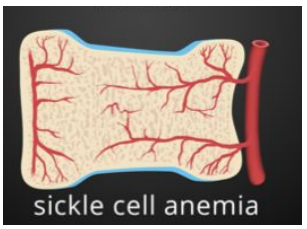
Sickle cell disease may be manifested as :

- **Anemia:**
  - Growth failure
  - Hyperkinetic heart failure.
  - Expanded intramedullary hematopoiesis.
  - Presence of extramedullary hematopoiesis.

- **Vaso-occlusive** due to sickling **:**
  - Infarcts in spleen, bone marrow, kidney, bowel, brain, muscles etc.
- **superimposed infection** due to splenic malfunction **:**
  - Pneumonia (*Pneumococcus, H. influenzae, Staph. aureus, Chlamydia, and Salmonella).*
  - Osteomyelitis *(Salmonella).*
  - resistance against malaria.





Red marrow in vertebral bodies in a 7-year-old girl with Sickle Cell Anemia.	
Sagittal T1-weighted MRI of spine	
 	 
Normal for comparison	- Low signal intensity in vertebral bodies compared to discs H-shaped vertebrae (arrows in right image) due to osteonecrosis of vertebral endplates. - depressed center due to avascular necrosis. - if you see infarction with signs of anemia then it's highly suggestive of SCD - Other causes of infarction include idiopathic avascular necrosis , steroids ..etc

Frontal radiograph of right shoulder in a 22-year-old patient

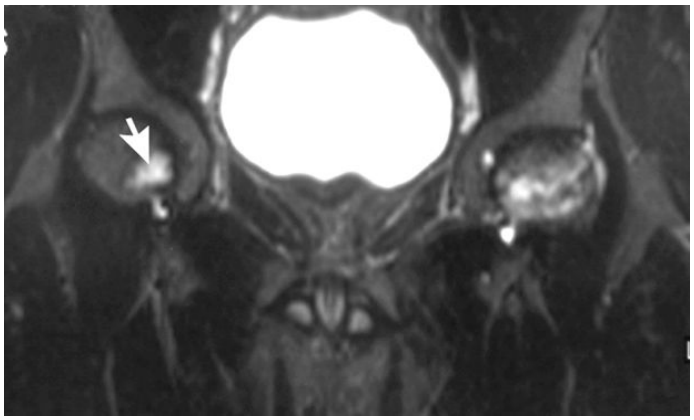
- Medullary bone infarcts in SCA
- area of patchy sclerosis and radiolucency
- infarcts usually seen in humours head / femoral head (big long bone)

- Bone infarcts typically occur in the medullary cavities and epiphyses
- Epiphyseal infarcts are frequently seen in the femoral and humeral heads and more often bilateral than avascular necrosis due to other diseases

AP radiograph in a 44-year-old man

- left hip → advanced avascular necrosis
- right hip → normal

Coronal STIR MRI image in the same patient



-Right hip → stage 1 avascular necrosis
-left hip → advanced changes of avascular necrosis
-It's important to keep in mind that X-ray modality is not sensitive to early stages of bone necrosis !! In this 44-year old case, x-ray only was able to detect the necrosis in its late stages, while MRI detected it even the early changes in the right hip!

Lateral radiograph of spine shows : H-shaped vertebrae in a 15-year-old patient.

-classic boxlike endplate depressions in middle portion (see the lowest vertebra shown) due to osteonecrosis of the vertebral endplates .

- **Hand-foot syndrome (dactylitis) in SCA**
- Frontal radiograph of right foot in a 3-year-old girl

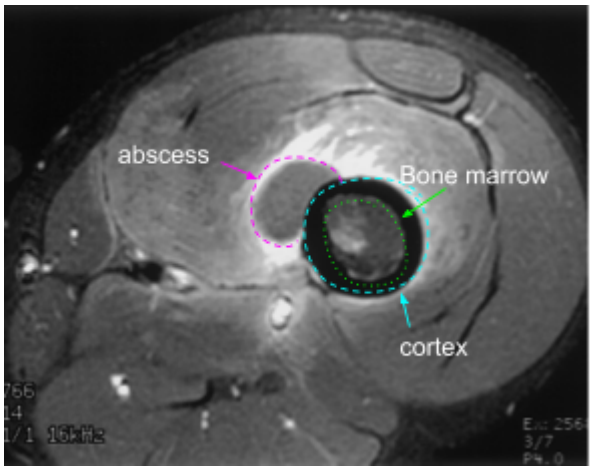
- thick periostitis and subperiosteal new bone along the metatarsal shafts

Salmonella osteomyelitis in a 10-year-old boy with SCA

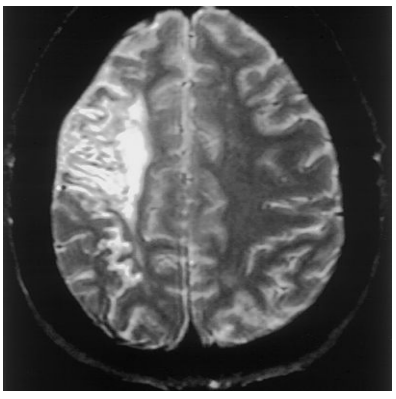
-Left ⟶ Initial film at onset of lower shin pain and fever is normal
-Right ⟶ Film 7 days later shows mottled lower tibial shaft and diffuse periostitis of the lower diaphysis

- 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.
- the treatment will be different in both, so we need an accurate Diagnose, how ? additional imaging (best by MRI , can be done by US) because in X-ray they are similar , if you see fluid/abscess collection ⟶ infection , NO ⟶ infarction

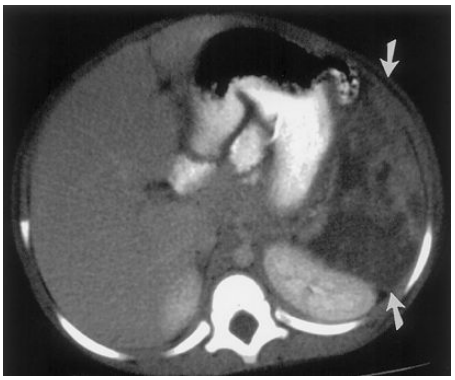
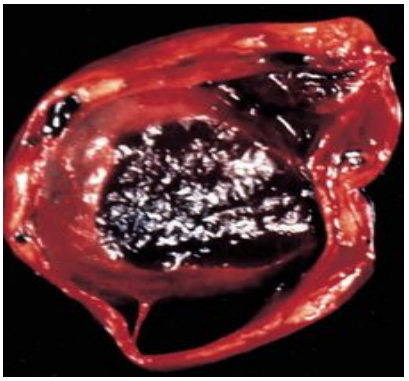
- infarction → conservative and observation while infection → antibiotic
- **MRI** findings that highly suggest **infection** :
  - Cortical defects in bone
  - Adjacent fluid collections (abscess) in soft tissue
  - Bone marrow enhancement
- Ultrasound guided aspiration of fluid collection around the 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 - rounded low-signal-intensity area adjacent to the shaft that is non-enhancing (fluid collection) - enhancement of the soft tissues around the shaft and of the adjacent musculature - Areas of enhancement are likely infected

case : Soft-tissue infection in a 52-year-old man with homozygous sickle cell disease.	
 arrow → Achilles tendon	**Longitudinal high-resolution ultrasound image of left ankle shows :** - arrow → hypoechoic fluid collection deep to Achilles tendon - Thick pus was aspirated from this area under ultrasound guidance 

case: Chronic infarct in a 19-year-old patient with SCA and longstanding mild left sided weakness	
	**Axial T2-weighted MRI shows:** area of high signal intensity and enlargement of overlying CSF spaces, compatible with chronic infarction and atrophy

Sequestration syndrome with splenic infarction in SCA

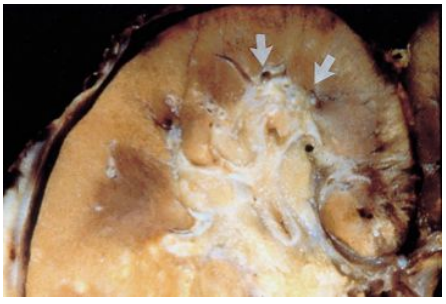



Axial CT after contrast shows :
enlarged spleen that enhances heterogeneously and minimally with large non-enhancing areas (arrows)
Photograph of spleen in a different patient shows areas of congestion and central necrosis
In Acute setting, splenectomy has to be done to prevent perforation & internal bleeding! because of high chance of rupture .

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 (arrow)

papillary necrosis has a lot of causes , like pain killers (commonest) , alcohol and SCA



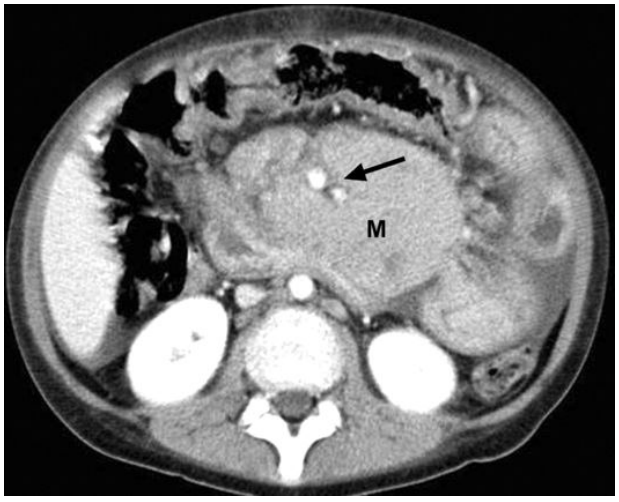
Photograph of a kidney from a different patient shows loss of papillary tips in some upper pole pyramids (arrows).

- Growth disturbance in distal radius in a 12-year-old girl
- Anteroposterior (AP) radiograph of left wrist shows epiphyseal shortening and a cup deformity of adjacent metaphysis.
- Also changes of old bone infarct in distal radius

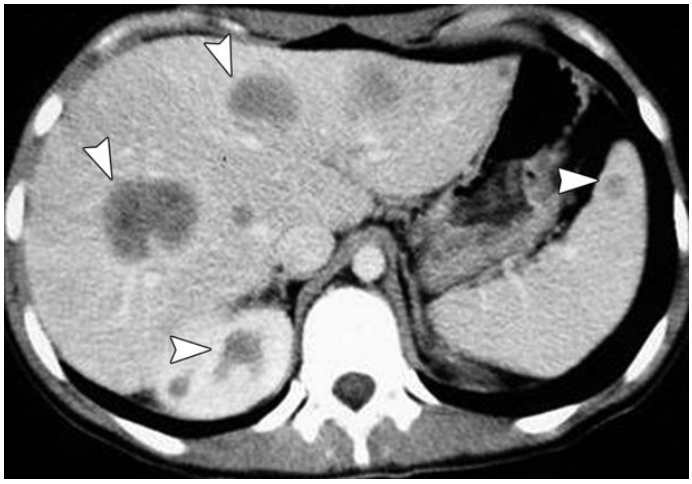
Lymphoma

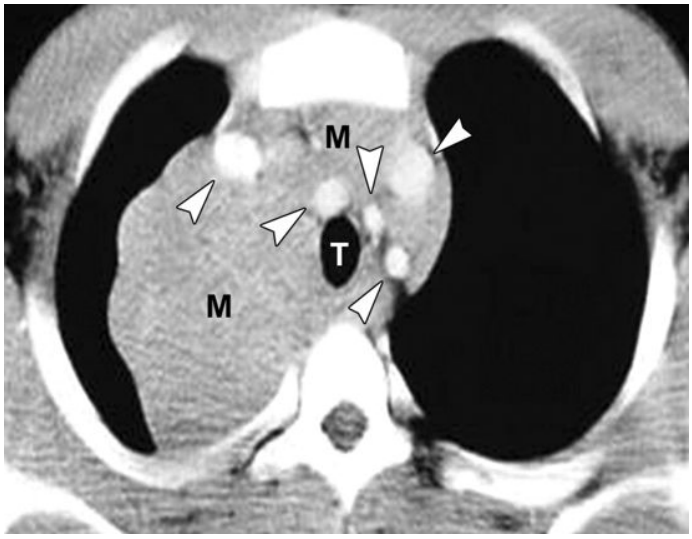
- it's neoplastic proliferation of the lymphocytes
- again , radiology has no role in diagnosing the subtypes of lymphoma, but we can identify the disease **extent** because sometimes it's hard to do physical examination of these masses especially if it is in a deep location ex: near the aorta.
- most of the time we do CT ,,, MRI takes long time.
- imaging has two role : staging and guiding the biopsy if no other accessible lymph node is available

Hodgkin's Disease	Non Hodgkin's Lymphoma
- Lymphocytic predominance - Mixed cellularity - Lymphocytic depletion - Nodular sclerosis - the most common	- Burkitt lymphoma (jaw and abdomen) - Burkitt-like lymphoma (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	

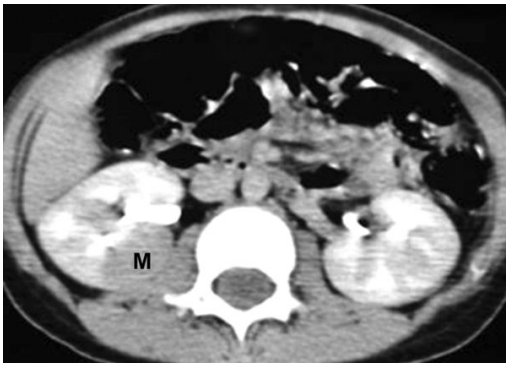
Non Hodgkin's Lymphoma (NHL) in an 11-year-old boy.	
	Axial CT scan with contrast shows: - large lymphomatous mass (M) encasing the mesenteric vessels (arrow)

NHL in a 14-year-old boy.	
	Contrast-enhanced CT scan shows: - large anterior mediastinal mass (M) that originates from thymus. A few cysts with central low attenuation and a peripheral enhancing ring are present (arrowheads).

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.

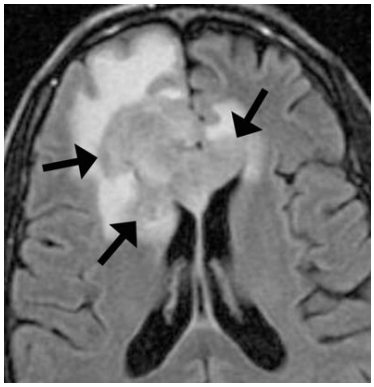
HD in a 17-year-old boy	
	Contrast-enhanced CT scan shows: - large mediastinal mass (M). Trachea (T) is compressed, and great vessels (arrowheads) are displaced

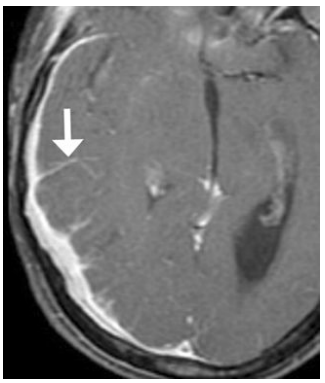
HD in a 12-year-old girl	
	Contrast-enhanced CT scan shows: - enlarged spleen with a diffusely inhomogeneous appearance.

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

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

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.

44-year-old HIV-positive woman with primary CNS non-Hodgkin's B-cell lymphoma	
	Axial FLAIR MRI shows : - lesion isointense to gray matter (arrows).

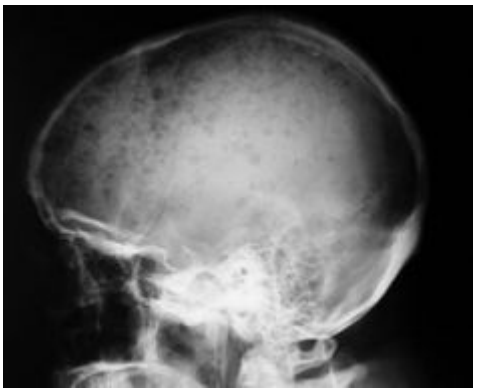
63-year-old woman with primary meningeal lymphoma	
	
Axial FLAIR MR images	post contrast T1 weighted MR images
hyperintensity and enhancement (arrows) involving sulci and leptomeninges	

Diagnosis of lymphoma:

- If accessible area, then open-surgery-Biopsy is often performed (e.g. Axilla and groin)
- If not accessible, CT guided biopsy is done to reach final diagnosis
- MRI is not commonly used unless you suspect a lymphomatous lesion in a specific area (e.g.brain)!!

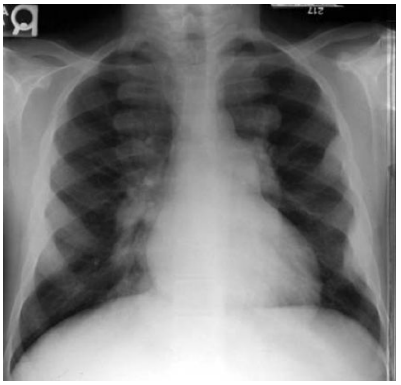
(forget about MRI, it's NOT used to assess lymphoma (pt. can't wait for 3-4 hours for you to visualize his body, while you have an excellent modality (CT scan) that takes only 2- 3 mins!!!)

Lateral skull radiograph shows:
multiple very sharply outlined (punched out) lytic lesions of multiple myeloma
very characteristic for multiple myeloma
looks like raindrops



- Findings shown can be seen in:

1. any severe chronic anemia
2. thalassemia
3. Sickle cell anemia
4. Lymphoma
5. Lymphoma



SUMMARY

INTRA medullary hyperplasia :	EXTRA medullary hematopoiesis :
- seen in : 1. Thalassemia 2. Sickle cell anemia 3. Iron deficiency anemia 4. Any severe chronic anemia - except bone marrow failure 'aplastic anemia'.	Appears as homogeneous soft tissue masses on imaging seen in : - ALL severe chronic anemias
- **Signs** of INTRA medullary hyperplasia: - Expanded bone marrow in bones including long bones of hands, feet, limbs, skull. - Decreased T1 MRI bone marrow signal than adjacent discs	Sites of EXTRA medullary hematopoiesis: 1. Liver 2. Spleen 3. Paraspinal areas with possible extension into spinal canal outside the dura 4. Kidneys 5. Meninges 6. Skin 7. Lymph nodes 8. Thymus

INFARCTS and INFECTIONS are additional findings in Sickle cell anemia
Bone infarct vs infection. It is important to diagnose these two as early as possible so that management can be started early to prevent complications - infarction → conservative and observation while infection → antibiotic MRI with contrast and ultrasound/CT guided aspiration of fluid collections are very helpful if imaging is unable to differentiate between 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 the patient is not already diagnosed. Rest is all by laboratory and clinical based.