

(1): RADIOLOGY of HEMATOPOIETIC DISORDERS

* Many thanks to 431 team for their helpful notes *

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 Color GUIDE:
 • Females' Notes
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Objectives

Not given "(

Classification:

Hematopoietic disorders are classified into disorders of:

RBC	anemia	Sickle cell – thalassemia	Only these
	Polycythemia		going to
WBC	Lymphoma		discusse
	Myeloma		this lectu
Plasma	Platelet	Bleeding- clotting	

e are be d in ure

ANEMIA :

There will be some changes that can be seen on radiological images:

(when sever and for long time)

- **1.** Reactive increase in red bone marrow : (1st response)
 - Expanded bone marrow in bones including long bones of hands, feet, limbs, and skull. 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*.)
 - Decreased T1 MwRI signal in vertebral body bone marrow than adjacent discs.
- 1. 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, Skin. This is called EXTRA-MEDULLARY HEMATOPOIESIS
- 2. Transfusions 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 for any type of anemia (we cannot determine the type of anemia based only on radiology)

Sickle Cell Anemia: has some additional features

- 1. Increased risk of Infections (Pneumonias, Osteomyelitis)
- 2. Increased risk of <u>Infarction</u> (Spleen, Kidneys, Brain, Bones. due to sickling of the RBCs inside the vessels, so the vessel will be occluded and ischemic.

1. B-thalassemia





<u>Normal</u>: For comparison

Normally bones are broad at the edges and narrow in the middle





Abnormal:

- Severe chronic anemia , Decreased bone density with coarse trabeculae, Wide medullary cavity with thin cortex
- Abnormal: expansion in the middle . (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.)



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)



Abnormal:

The overall skull is very thickened, expanded with cortical thinning. There are also some are as that are more Lucent and areas that are darker \rightarrow 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 is 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/superior meningeal artery is enlarged.

Not specific to thalassemia. Both cases are intramedullary hematopoiesis.



Normal: (For comparison) The bone marrow is between the inner and outer cortex.



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

(When we do MRI we can actually see the bone marrow with the skull and the CT has basically the same appearance)



25-year-old man with β -thalassemia. PA radiograph of chest (left) shows diffuse expansion of ribs and right upper paraspinal thoracic mass (arrow) compatible with extramedullary hemopoiesis.





Abnormal:

The whole rib is expanded and especially the anterior end \rightarrow intramedullary hematopoiesis. So, basically the same sign of anemia; expansion of the bones. There is also extramedullary hematopoiesis

Note(s):

- 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.

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



Axial contrast-enhanced CT scan through chest shows uniformly enhancing paraspinal hemopoietic masses with no bony erosion. There are some signs of thickening and expansion of the ribs but there are also bilateral parastinal areas \rightarrow there is extramedullary hematopoietic areas.



PA chest film shows well-marginated bilateral, paraspinal masses compatible with extramedullary hemopoietic tissue

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Lecture Title

51-year-old woman with myelofibrosis. Coronal T1-weighted MR image shows massively enlarged spleen Splenic biopsy was followed by splenectomy Pathologic examination revealed extramedullary hematopoiesis

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.)

56-year-old man with myelofibrosis Axial contrast-enhanced CT scan through kidneys reveals bilaterally symmetric enhancing perinephric masses. Biopsy showed extramedullary hematopoiesis

There ar some soft tissue around kidneys, this Is perinepheric extramedullary hematopoiesis.

48-year-old man with hemolytic anemia and myelofibrosis. Axial CT scan through pelvis shows well-marginated presacral soft-tissue mass (arrow) with no bony erosion. Biopsy showed 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.

40-year-old man with sickle cell disease. Axial unenhanced CT scan at thoracoabdominal level reveals two uniformly low-attenuation (compared with liver parenchyma), well-marginated lesions (arrows) Percutaneous biopsy showed extramedullary hemopoiesis

Focal masses within the liver \rightarrow extramedullary hematopoiesis. (liver is bright due to iron overload)









2. <u>Sickle cell disease</u>

May be manifested as:

1. ANEMIA

- growth failure, hyperkinetic heart failure
- expansion of intramedullary hematopoiesis
- Presence of extramedullary hematopoiesis

2. VASO-OCCLUSION

Infarcts in spleen, bone marrow, kidney, bowel, brain, muscles

3. SUPERIMPOSED INFECTION

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

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

Medullary bone infarcts in SCA. Frontal radiograph of right shoulder in a 22-year-old patient shows an area of patchy sclerosis and radiolucency



<u>Normal</u>: there's homogenous density and a very sharp outline of the humerus.

<u>Abnormal</u>: it is very patchy, with dark and bright areas which indicates <u>osteonecrosis</u> Of the humeral head. (flattened with some bone infarcts)



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Hand-foot syndrome (dactylitis) in SCA Frontal radiograph of right foot in a 3-yearold girl shows thick periostitis and subperiosteal new bone along the metatarsal shafts

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



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Salmonella osteomyelitis in a 10year-old boy with SCA. Initial film (left) at onset of lower shin pain and fever is normal. Film 7 days later (right) shows mottled attenuation of lower tibial shaft and diffuse periostitis of the lower diaphysis

Is it an infection or infarction ? On x-rays : It is difficult to defrenciate b/w infection and infarction. There's some abnormality In the marrow cavity you can see some black areas. Also there is a sign of extra line



AP radiograph in a 44-year-old man shows advanced avascular necrosis in left hip and a normal right hip



<u>Abnormal</u>: The femoral head is more dense, patchy, and the outline is not smooth. Also there is some compression and it started to collapse (not round) \rightarrow Osteonecrosis. On X-Ray : The right femoral ead looks normal while the left looks abnormal. However on MRI : 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 5

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: 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 the most types of anemia. [non specific]
- The vertebral body is also H-shaped which is central depression due to osteonecrosis \rightarrow specific to sickle cell anemia.

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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



Normal : for comparison

Central depression. Nowadays, It is rare to see patients With H-shaped Vertebrae due to early Dx and Rx ; 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



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: (highly suggestive for infection)

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

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

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Osteomyelitis of femur in a 24-yearold patient with SCA. Axial T1weighted MRI after contrast shows heterogeneous enhancement of marrow cavity, a rounded low-signalintensity 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. <u>Areas of</u> <u>enhancement are likely infected</u>

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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 \rightarrow any fluid collection In a patient with a known sickle cell anemia indicates infection. Even in US you can see the fluid accumulation.

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





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Lecture Title

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Chronic infarct in a 19-year-old patient with SCA and longstanding mild left sided weakness. Axial T2weighted MRI shows an 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



The entire spleen is infarcted

11 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). Analgesic medications can cause it.

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



12 Growth disturbance in distal radius in a 12year-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.

- Infarction can also occure in the epiphysis. when the epiphysis becomes infarcted (it is dead) → it Will stop growing.
- Furthermore, if the child is still growing deformities will occur.<< Here the radial epiphysis Is infarcted and 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 density and some low-density
- areas that mixed it indicates infarction



1. Hodgkin's Disease

- Lymphocytic predominance
- Mixed cellularity
- Lymphocytic depletion
- Nodular sclerosis the most common

2. 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



NHL in an 11-year-old boy. Axial CT scan shows a large lymphomatous mass (M) encasing the mesenteric vessels (arrow)

A mass in the abdomen

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

This patient has multiple chest lesions which are mattered together and are compressing the airway posteriorly.

If the patient has no other mass, they will perform a CT \rightarrow Gide biopsy from here.





Note(s):

- Lymphoma is diagnosed on a hematological basis, not radiological.
- The role f radiology in lymphoma :
- 1. Stage the disease.

(stage the extent of the disease ,

If it is in one group of lymph nodes or multiple,

If it is in both sides of the diaphragm or one side and whether it has visceral involvement or not; for example (liver, spleen, etc)

2. CT-guided biopsy if the mass is not easily accessible. \rightarrow for confirmation

(Patients initially present with a mass and to diagnose they have to get a biopsy. If that mass is in the chest or the 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.

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HD in a 17-year-old boy. Contrastenhanced 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. Contrastenhanced CT scan shows lowdensity lesions (arrowheads) in both hepatic lobes, with small nodules in spleen and right kidney.

Multiple lesions in liver, kidney, and spleen

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

Liver and splenic masses

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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

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

Splenomegaly with multiple lesions

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.

A mass in the brain





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

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Lateral skull radiograph showing multiple very sharply outlined (punched out) lytic lesions of multiple myeloma

Small numerous lesions in the skull \rightarrow multiple myeloma. The radiological role is staging the disease.



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



SUMMARY

1. <u>Anemia :</u>

- we cannot determine the type of anemia based only on radiology
- > Signs (not specific for any type of anemia):
 - Reactive increase in red bone marrow
 - New marrow areas in potential organs
 - Transfusions Iron overload
- Sickle Cell Anemia: has some additional features :
 - Increased risk of Infections (Pneumonias, Osteomyelitis)
 - Increased risk of Infarction (Spleen, Kidneys, Brain, Bones.
- MRI findings: (highly suggestive for infection)
 - Cortical defects in bone
 - Adjacent fluid collections in soft tissue
 - Bone marrow enhancement

2. lymphoma:

CT is used to Stage the disease, or to guide the biopsy for confirmation

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Lecture Title

Questions

1) What is your diagnosis?

- a. any severe chronic anemia
- b. thalassemia
- c. Sickle cell anemia
- d. Lymphoma

2) What is your diagnosis ?

- a. any severe chronic anemia
- b. thalassemia
- c. Sickle cell anemia
- d. Lymphoma





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Answers:

1st Questions: a

2nd Questions: a