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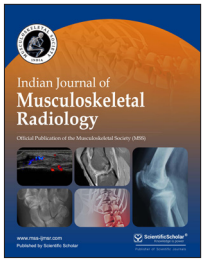

Case Series

Imaging features in extramedullary hematopoiesis - A case series

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ABSTRACT

Extramedullary hematopoiesis (EMH) is a rare compensatory process associated with many hematologic disorders and bone marrow dysfunction. Through this article, we aim to present various imaging features of EMH and its complications including spinal cord compression. Three cases of EMH in hemoglobinopathies (thalassemia and sickle cell anemia) have been described. Cases 1 and 2 of our series include paraspinal masses causing cord compression. Case 1 of thalassemia intermedia presented with various skeletal deformities in long bones, skull, and facial bones (Chipmunk facies). Case 2 was diagnosed with extrapleural lung mass with other paraspinal lesions. Case 3 presented with multiple enlarged retroperitoneal lymph nodes due to EMH. Common and uncommon imaging features of EMH have been described. Lymph node involvement was difficult to diagnose as it mimicked tubercular and neoplastic etiologies; hence, histopathological diagnosis was mandatory.

Keywords: Extramedullary hematopoiesis, Thalassemia, Spinal cord compression, Lymph nodes, Paraspinal masses

INTRODUCTION

In contrast to the fetus, where the yolk sac, spleen and liver are the primary sites of hematopoiesis, adult hematopoiesis occurs in the marrow of long bones, ribs and vertebrae.[1,2] Extramedullary hematopoiesis (EMH) mainly results from insufficient blood element production happens when the bone marrow is replaced, which is most often caused by myelofibrosis, widespread metastatic illness, leukemia, and conditions that can reduce the amount of functional red blood cells (RBCs) such as thalassemia, sickle cell anemia (SCA), acquired hemolytic anemia, and Vitamin B12 or folate deficiency.[3] It is critical to recognize imaging abnormalities that are consistent with extramedullary hematopoiesis since they may resemble a neoplasm, requiring biopsy to rule out a neoplasm and altering treatment and prognosis. We describe patients with a range of imaging symptoms of EMH in this paper.

CASE REPORTS

Case 1 - EMH in case of thalassemia with spinal cord compression (SCC)

A 21-year-old girl with thalassemia intermedia came with a 2 years history of bilateral lower extremity weakness and numbness. Her development was stunted and she had a short stature, according to the assessment. A typical thalassemia chipmunk face was noticed [Figure 1a]. She was pale and jaundiced to a degree. We discovered widespread enlargement of the diploic space

of the calvarial bone on skull head X-ray and computed tomography (CT) of the head [Figure 1b-d]. Multiple lobulated paraspinal masses and rib enlargement were seen on the chest X-ray [Figure 2]. We discovered hepatosplenomegaly on ultrasonography (USG). A 6.4 g/dl hemoglobin and an increased reticulocyte count were found in the lab tests. A plain X-ray of the dorsal spine revealed significant osteopenia with an exaggerated trabecular pattern. There was also medullary enlargement of the ribs, scapula, clavicles, and bones of the hands and other long bones [Figure 2]. CT thorax was done which showed medullary expansion with a prominence of trabeculae in all the visualized bones. Posterior ends of all the ribs appear expanded with surrounding overlying soft-tissue components [Figure 3a and b]. Lobulated soft tissue density was seen in the extradural location of the spinal canal from D3 to D8 vertebral levels causing compression of the spinal cord for which MRI spine was done.

MRI spine examination showed T1 and T2 isointense (to the cord) homogeneously enhancing expanded posterior ends of visualized ribs [Figure 3a, c, and d]. Similar lobulated intraspinal extradural lesions were seen arising from posterior elements of D3 to D8 vertebrae causing spinal canal narrowing and compression of the spinal cord at D4 to D7 vertebral levels [Figure 3e and f]. All the visualized vertebrae appeared hypointense on T2 which was likely due to active red bone marrow [Figure 3e and f].

The patient was initially infused with four units of packed RBCs. Numbness and weakness were lessened as a sign of improvement. To remove epidural and para-spinal masses and relieve cord compression, a D4-D9 laminectomy was performed. She was then discharged home and treated as an outpatient. After 10 weeks, the patient was able to walk properly again.

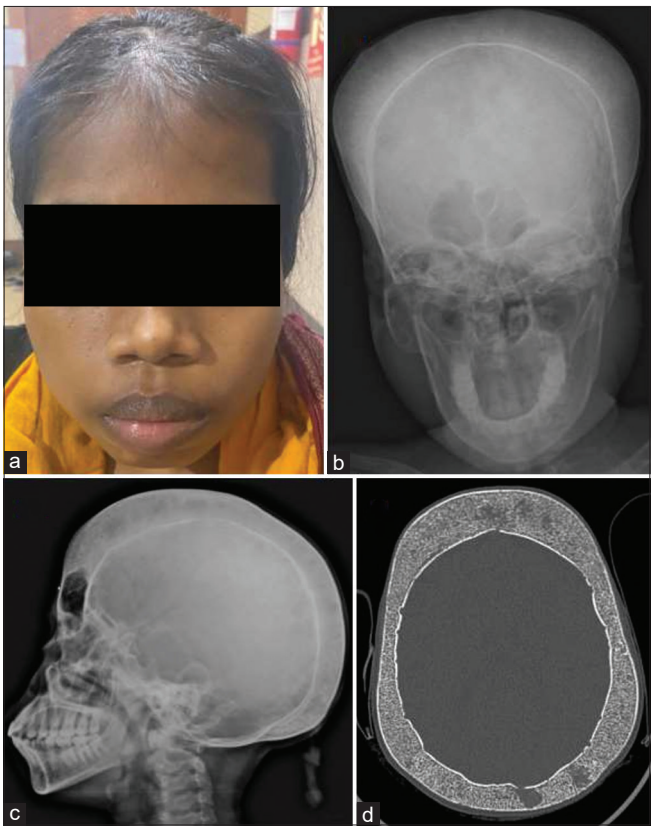

Figure 1: (a) Chipmunk/rodent facies due to over-expansion of maxilla and mandible. (b and c) AP and lateral skull radiograph, elicits the protrusion of the maxilla and mandible. (b, c, and d) X-ray AP and lateral view and CT axial view showing widening of the diploic space and thinning of the inner and outer table with prominent secondary trabeculae.

Case 2 - EMH in SCA with lung mass and spinal cord compression (SCC)

A 2-year-old male baby, with known SCA, presented with difficulty in walking, excessive crying, and difficulty in breathing. Hemoglobin was 5.6% and platelet count was 3.2 lakhs/cu.mm, according to routine tests.

On CT thorax, an extrapleural soft tissue mass of size approx. 5.2 × 3.7 × 2.1 cm along the left lateral chest wall, adjacent to

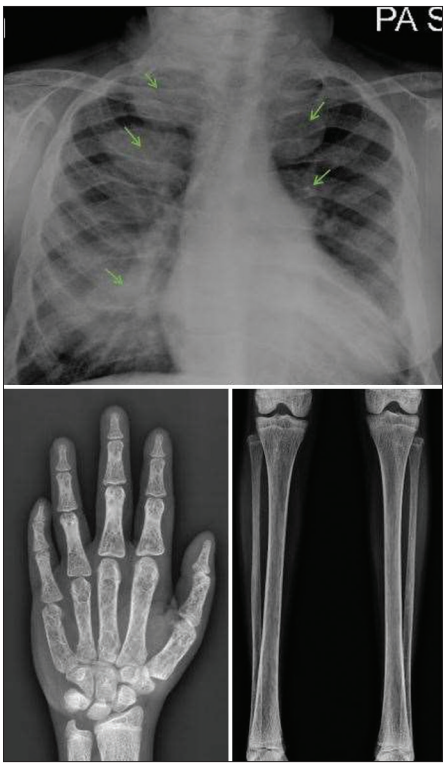

Figure 2: Chest X-ray PA views showing multiple mass-like lesions (green arrows) in the paraspinal in a patient with thalassemia are consistent with extramedullary hematopoiesis. Furthermore, note the expanded visualized bones including ribs and clavicles. AP view X-ray (hand and legs) showing marrow proliferation leading to expansion of all medullary spaces creating a lace-like trabecular pattern due to severe chronic anemia.

2nd and 3rd intercostal space was seen [Figure 4a, c, and d]. Erosion and remodeling of the 3rd rib were noted on the left side [Figure 4b]. Soft tissue causing expansion of posterior elements of the D5 vertebra was noted, predominantly on the left side [Figure 4b].

MRI spine showed heterogeneously hyperintense extrapleural soft tissue mass on T2 and STIR along the left lateral chest wall [Figure 5b]. Extension of soft tissue was noted into the spinal canal (Extradural from D4 to D6 vertebral level) causing moderate to severe spinal canal narrowing and SCC at D5 vertebral level [Figure 5a-e]. Enhancement was seen in the post contrast study. Soft tissue extension was noted, causing moderate to severe left neural foraminal narrowing at D4-D5 and D5-D6 disc levels. Spine decompression surgery was done to relieve the cord. At present, the patient is on regular follow-up.

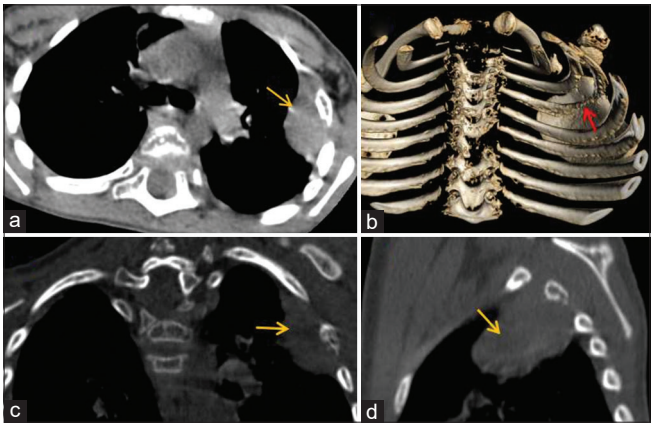

Figure 4: (a-d) Showing an extrapleural soft tissue mass lesion (yellow arrows in a, c and d) along the left lateral chest wall, adjadent to 2nd and 3rd intercostal space with the remodeling of 3rd left rib (red arrow in b).

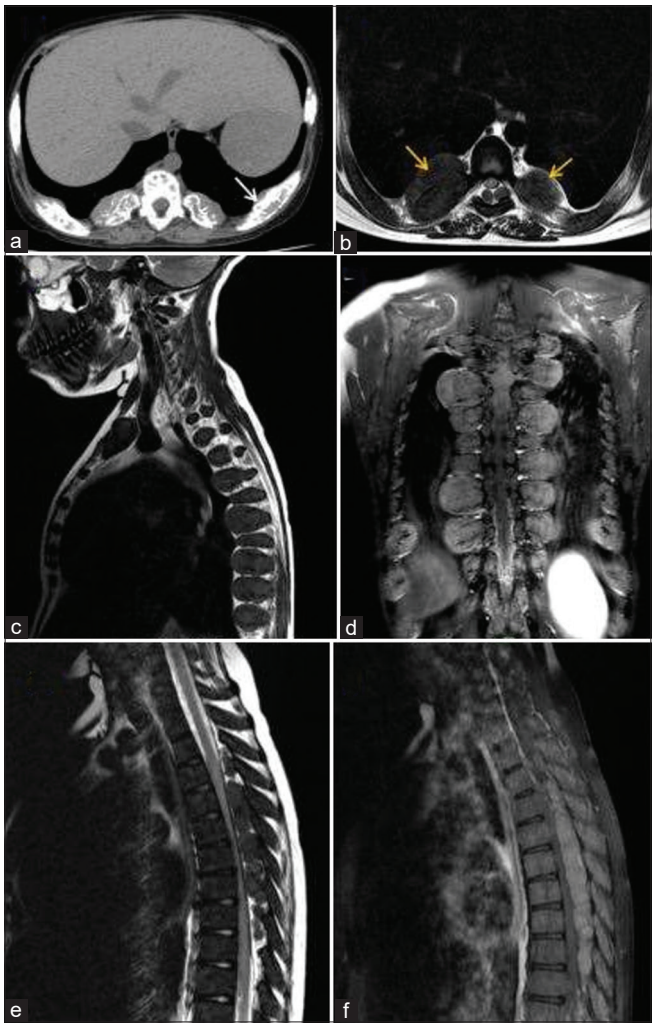

Figure 3: (a and b) CT and MRI T2 axial images showing soft tissue paraspinal masses with the expansion of ribs. Also, note hepatomegaly in (a). (c and d) MRI T2 sagittal and coronal T1 contrast images showing widened posterior ribs and paraspinal masses, respectively. Masses enhance on contrast study (d). (e and f) MRI sagittal T2, T1 contrast images respectively showing cord compression by the masses at D4 to D7 levels.

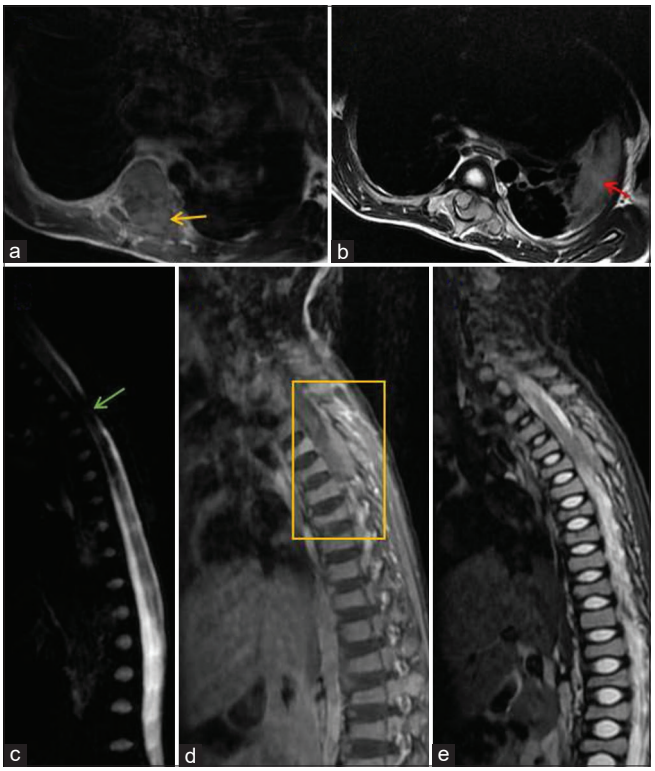

Figure 5: (a and b) MRI T1 contrast and T2 axial images showing soft tissue masses causing spinal cord compression and moderate to severe left neural foraminal narrowing (yellow arrow in a). Furthermore, note the heterogenously hyperintense extra pleural soft tissue mass along the left lateral chest wall on axial T2 MRI image (arrow in b). (c-e) MRI sagittal myelogram, T1 contrast, and T2 images showing spinal cord compression (green arrow in c).

Case 3 - EMH in case of SCA with retroperitoneal lymph nodal involvement

A 13-year-old male, known case of SCA, presented with chief complaints of weakness and loss of appetite for 1 month. Laboratory investigations showed TLC count: 2.6 × 10^3/ul, hemoglobin: 7.5 g/dl, RBC count: 3.44 × 10^6/ul, platelet count: 74 × 10^3/ul, and ESR: 5 mm. Peripheral smear revealed microcytic hypochromic anemia. Moderate anisopoikilocytosis, elliptocytes, teardrop cells, schistocytes, macrocytes, and polychromatic cells were also seen.

USG abdomen revealed mild hepatosplenomegaly. CT whole abdomen (contrast) revealed multiple enlarged necrotic paraaortic, aortocaval, left external iliac, retro crural, retro-aortic and retrocaval nodes [Figure 6a and b] which were found to be rich in hematopoitic cells on histopathology. Extramedullary hematopoiesis was noted in the left iliac bone and multiple ribs on the right side [Figure 6c and d]. Bone marrow aspiration showed erythroid hyperplasia with megaloblastoid changes.

DISCUSSION

In the thorax, the most common imaging manifestations are paraspinal masses and rib expansion, and these findings are more frequent in β-thalassemia than in other causes of extramedullary hematopoiesis. The paraspinal active hemopoietic masses are well marginated and show mild homogenous enhancement on contrast-enhanced CT,

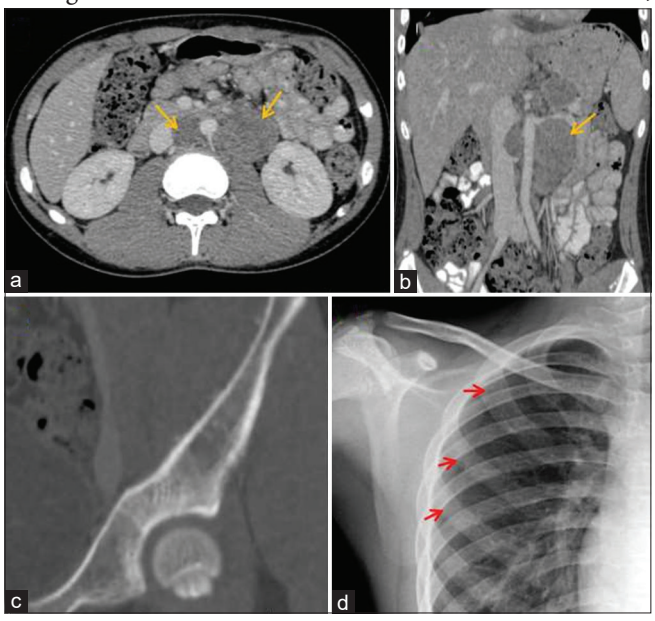

Figure 6: (a and b) Axial and coronal CT images of abdomen showing multiple enlarged retroperitoneal lymph nodes (yellow arrows in a and b). (c) Coronal CT abdomen (Zoom in view) showing expanded left iliac bone representing EMH. (d) PA view of chest X-ray (Cut section of right hemithorax) showing expansion of few ribs on right side (red arrows in d).

whereas old, burnt-out lesions may show iron deposition or fatty degeneration. Paraspinal hemopoietic tissues can extend into the central canal, especially in the thorax, and cause neurologic symptoms because of SCC. On MRI, the T1 sequence shows intermediate signal intensity but may be of low signal intensity due to massive iron deposition in repeated transfusions. T1 post-contrast shows variable enhancement of the masses but no enhancement in case of massive iron deposition. T2 intermediate to high signal intensity (relative to skeletal muscle) and low signal intensity due to massive iron deposition in repeated transfusions is seen within the lesions.

The first and second cases in this series had thalassemia and SCA, with paraspinal involvement and SCC, respectively. SCC induced by EMH was initially reported in 1954, and it was treated with surgical decompression and resection excision, followed by local radiation.[4] EMH etiologies associated with SCC include thalassemia, sickle cell anemia, myelofibrosis, polycythemia vera, acute myeloid leukemia, and myelodysplasia.[5] The most prevalent cause of these issues is thalassemia.[6,7]

When EMH is accompanied by neurological abnormalities, surgical intervention is necessary to decompress the neural components and get a tissue diagnosis. Decompression surgery was performed on cases 1 and 2 in our instance. The borderless character of the hematopoietic process, however, prevented large total removal of EMH masses. Surgical treatment of EMH is widely recognized to be reserved for acute, severe, or recurrent cases, despite differing viewpoints.[6,8] Surgical therapy was both appropriate and needed since cases 1 and 2 were having worsening symptoms linked to cord compression. Another promising therapeutic option for EMH-induced SCC is RBCs hypertransfusion.[9] It's said to be the least invasive method. Our case 1 benefited from blood transfusions as well, with indications of improvement such as reduced numbness and weakness within a few weeks following treatment.

While hematopoiesis is most normally observed in the bone marrow, EMH can also occur in lymph nodes. The third case in our study was a reported example of sickle cell anemia with lymphnodal involvement. On a CT scan of the abdomen, several enlarged retroperitoneal lymph nodes were visible. The left iliac bone and a few right-sided ribs had EMH. The presence of enlarged lymphnodes in this patient made it difficult to differentiate between tuberculosis and neoplasms. In these nodes, histopathology indicated an abundance of hematopoietic cells, suggesting EMH.

All of the patients had hepatosplenomegaly. Hepatomegaly might be caused by several hematological illnesses, such as EMH, hepatitis from chronic transfusion-related infections, and iron overload from excessive blood transfusion treatment. EMH and/or extravascular hemolysis can cause splenomegaly.

CONCLUSION

The imaging manifestations of EMH, as well as its relationship to hemoglobinopathies, have been shown. Understanding imaging characteristics aid in improved diagnosis and therapy response monitoring. Because EMH in lymphnodes might be mistaken for other inflammatory and neoplastic causes, a histopathological diagnosis is required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There is no conflict of interest.

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