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Radiodiagnosis

An Unusual Cause of Nerve Root Compression in AML: Paraspinal and Epidural EMH

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Abstract **Case Report**

Extramedullary hematopoiesis is characterized by the presence of hematopoietic tissue outside the bone marrow. Extra-thoracic extramedullary hematopoiesis is a rare and usually asymptomatic condition. We report a case of a 37-year-old male with paraspinal and presacral extramedullary hematopoiesis with acute myeloid leukemia. Clinical and laboratory evaluation, along with radiological and histopathological findings, are described. The diagnosis of the disease was confirmed by a CT-guided biopsy. A review of the literature is presented.

Keywords: hematopoiesis tissue, bone marrow, CT-guided biopsy.

INTRODUCTION

Acute myeloid leukemia (AML) is a kind of blood cell cancer that causes fast proliferation of aberrant cells in the bone marrow and blood, interfering with normal blood cell synthesis. EMH is an uncommon condition in people with protracted anemias such as hemolytic anemias, myeloproliferative illnesses, and certain neoplasms as a compensatory mechanism required for adequate erythrogenesis [1]. It is uncommon in the thorax and significantly rarer in extra-thoracic areas such as the paraspinal (lumbar) and presacral regions [2, 3]. Extramedullary hematopoiesis (EMH) is a unique condition that causes spinal cord compression syndrome. In this report, we describe a case of paraspinal (lumbar) and presacral EMH in a man with acute myeloid leukemia.

CASE PRESENTATION

A 37-year-old guy reported to the Department of Radiodiagnosis at our institution with the inability to move both lower limbs for one month, as well as bowel bladder incontinence and grade 4 bed sore across the sacral area. The patient was being treated for pancytopenia.

Table 1: Laboratory data

Hemoglobin	6.9 g/dl,
RBC	2.24 x 10^6
WBC	0.54X10^3
Neutrophil	13%
Lymphocyte	87%
Monocyte	0%
Eosinophils	0%
basophils	0%
Platelet	30 x 10^3
HCT	21.2%
MCV	94.6%
MCH	30.8pg
MCHC	32.5g/dl
RDW	17.2%

Peripheral Smear –

- RBCs – Normocytic Normochromic.
- WBCs – Marked Leucopenia with 50 Cells Counts.
- Neutrophils – 6/50%, Lymphocyte -44/50%- No Blast Seen.
- Platelets: Reduced (Count = 30000/Ul).
- Reticulocyte count -4.48%.

The contrast-enhanced computed tomography (CECT) of the thorax, abdomen, and pelvis was done from the diaphragm to the coccyx. CECT indicated moderate to mild hepatomegaly and several paravertebral soft tissue density enhancing lesions ranging from the T2-L1 vertebral levels. Transmural extension into the epidural area is present, with an augmenting component within the spinal canal. There is

no evidence of bone erosion. Only a few lesions have central hypodensity. Post-contrast and non-contrast HU are comparable to splenic tissue. There is no erosion or expansion seen on the Bone window. Similar lesions can be detected along the right paravertebral area from L5 to S1.

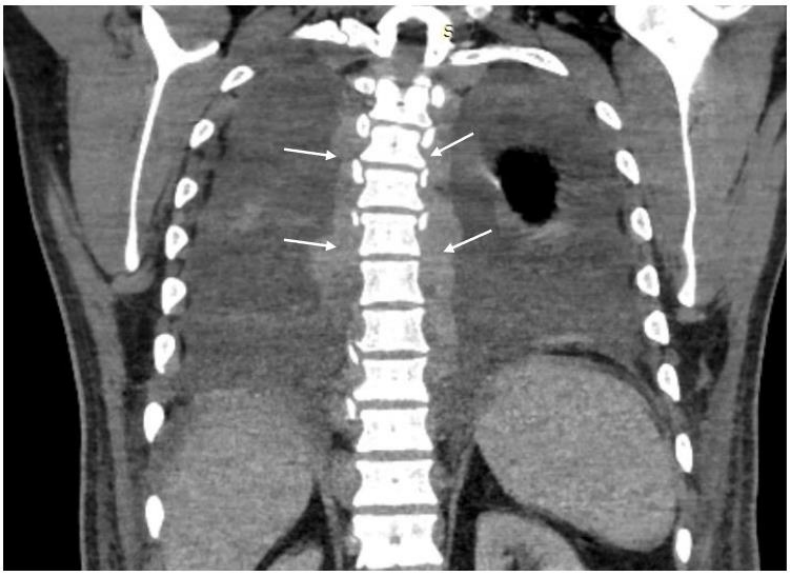

Fig. 1: Contrast enhanced CT coronal sections of thorax and upper abdomen shows multiple paravertebral soft tissue density enhancing lesions are seen extending from T2-L1 vertebral levels (arrows)

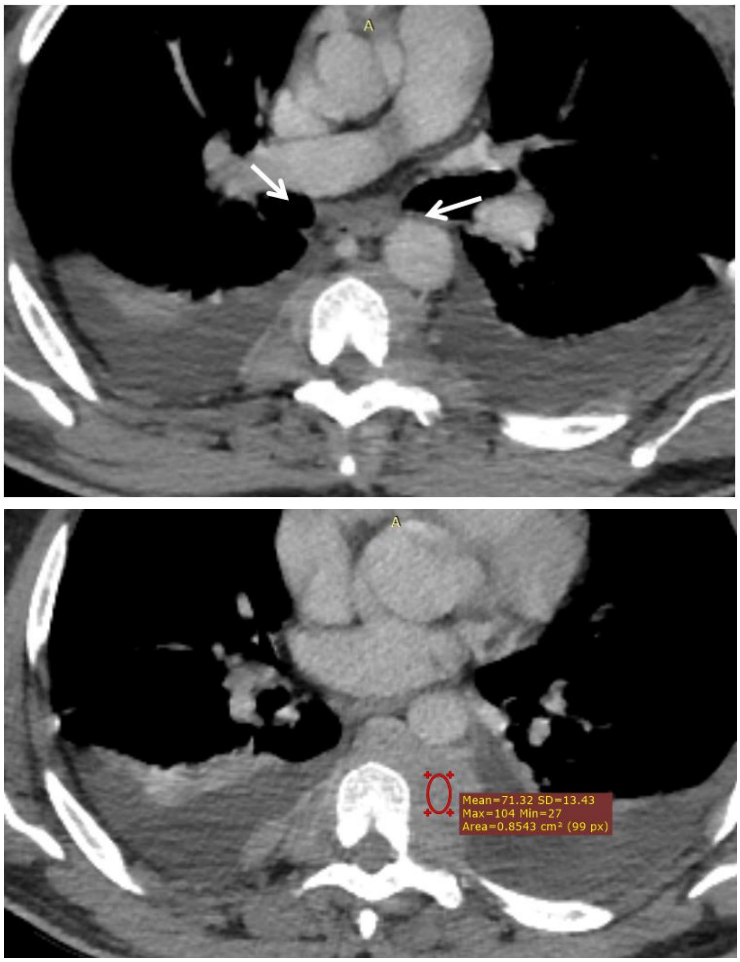

Fig. 2(a): contrast enhanced CT axial sections shows there is transmural extension into epidural space with enhancing component within spinal canal. 2(b): Post contrast HU and non-contrast HU is similar to splenic tissue

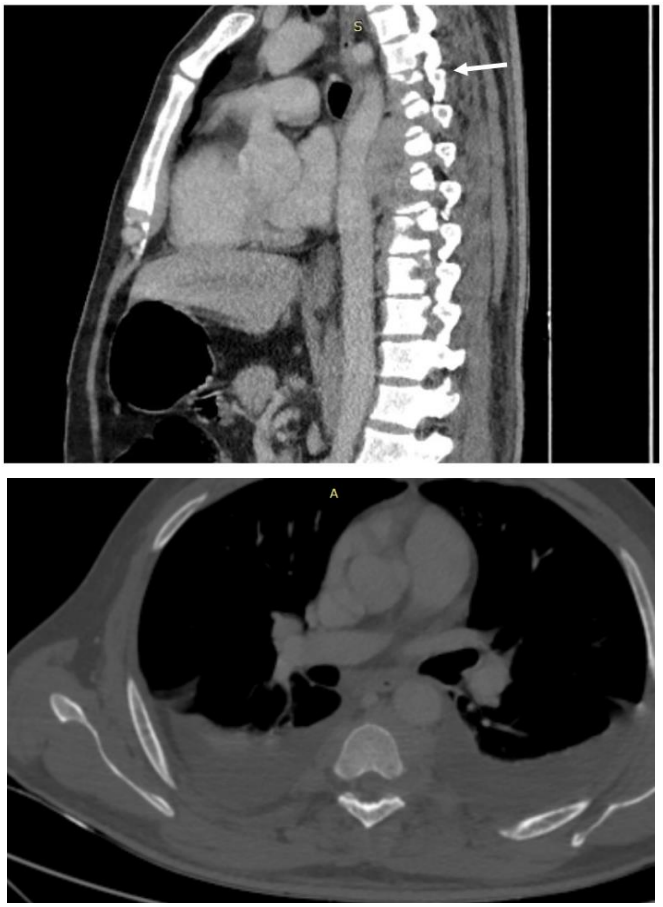

Fig. 3: (a) contrast enhanced CT sagittal sections shows transmural extension into epidural space with enhancing component within spinal canal. (b): On bone window, no bony erosion is seen

CEMRI at 1.5 T found that -CEMRI exhibits widespread T1 and T2 hypointense marrow signals throughout the spine. Posterior elements and dorsal vertebral bodies show heterogeneous post-contrast enhancement.

It encases the appropriate neurovascular bundles along the posterior ribs posteriorly. It enters the posterior epidural space through bilateral neural foramina.

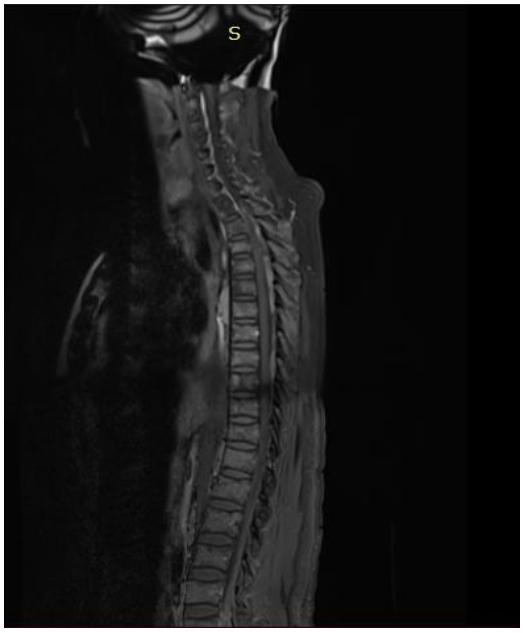

Fig. 4: Post gadolinium MRI (T1+C) shows diffuse altered marrow signal is seen throughout spine, suggestive of diffuse infiltration

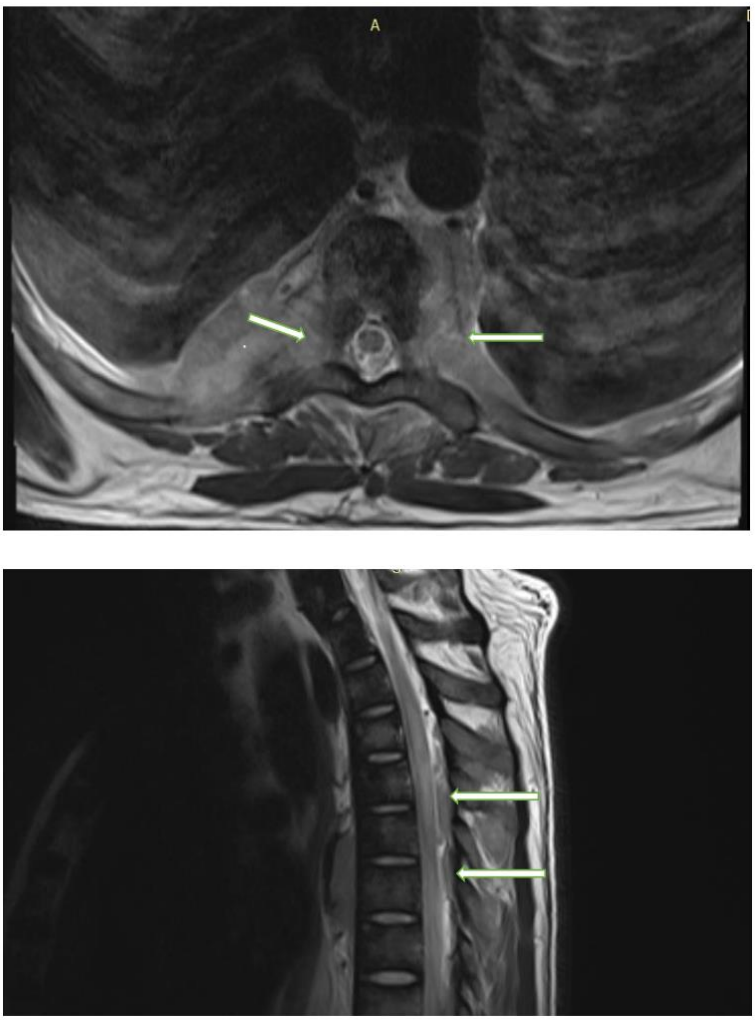

Fig. 5 (a): post contrast axial sections shows the mass is extending into posterior epidural space through bilateral neural foramina. (b): Sagittal sections show well defined mass in posterior epidural space

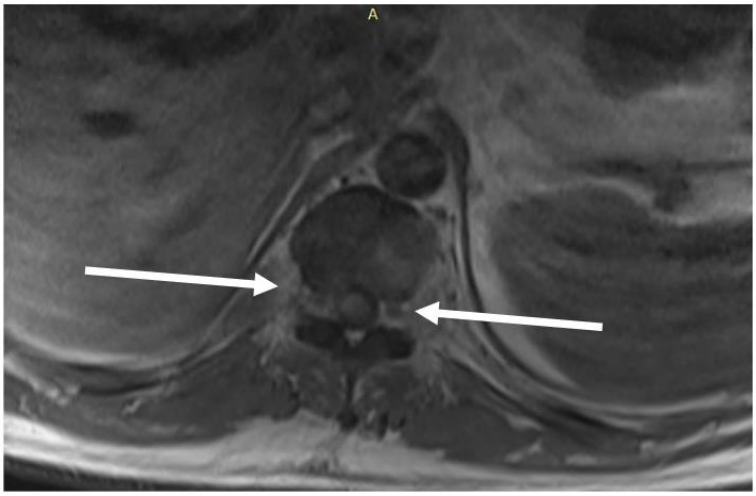

Fig. 6: Axial sections of thoracic spine showing the lesion is posteriorly encasing the corresponding neurovascular bundles (arrows) along the posterior ribs

A CT-guided biopsy of the right paraspinal (lumbar) and presacral masses revealed hematopoietic tissue with M: E - 5.4:1 ratio and inhibited myelopoiesis, erythropoiesis, and megakaryopoiesis.

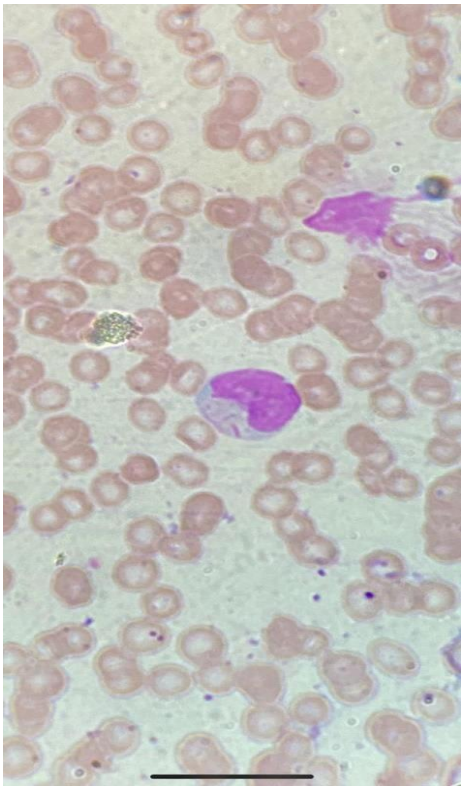

Fig. 7: Shows large blast showing folded nuclei, 3-4 prominent nucleoli and auer rod in the peripheral smear

Atypical cells/anything else: Large-sized blasts with modest amounts of cytoplasm, bent nuclei, and 1-3 nucleoli replace bone marrow. There are a few blasts with monocytic shapes. Auer rods have been seen.

Table 2: Bone marrow biopsy

CELLS	Observed %	NORMAL %
Blast	65%	0-5%
Promyelocytes	0%	3-12%
Myelocytes	3%	2-16%
Meta-myelocytes	3%	2-16%
Neutrophils	4%	22-66%
Lymphocytes	10%	5-20%
Mono-	1%	1-3%
Eosinophils+precursors.	0%	1-4%
Erythroid precursors	14%	10-35%
Basophils	0%	0-0.5%
Plasma cells	0%	0-3%

Flow Cytometry Report

Peripheral blood was used as the sample, and the cell preparation method was stain-lyse-wash. SSC vs. CD45 gating technique was used. Events with modest side scatter and dim to moderate CD45 expression account for 15% of all events.

Marker Expression Study:

- The cells of interest are POSITIVE for:
  - CD38, CD34, CD117, HLA-DR, CMPO, CD13, CD33, and DIM positive for CD45, CD14, CD56.

- The cells of interest are NEGATIVE for:
  - nTdt, CD 10,CD19,CD 20,CD79a, Surface CD3, CCD3, CD4, CD 5,CD7

Impression

Acute myeloid leukemia characteristics correlate with bone marrow imprint morphology. The definitive diagnosis of acute myeloid leukemia (AML) with extramedullary hematopoiesis manifesting as paraspinal (lumbar) and presacral masses was reached after correlating the clinical, laboratory, radiographic, and histopathological data.

3. DISCUSSION

EMH is a physiological compensating mechanism in which normal blood cells are formed

outside of the bone marrow because the bone marrow is unable to satisfy everyday circulatory demands [4]. EMH can be found in a variety of haematological illnesses, including myelofibrosis, polycythemia vera, lymphoma, leukaemia, and post-bone marrow irradiation. The spleen, liver, thymus, breast, lung pleura, heart, kidneys, prostate, suprarenal glands, ovaries, gut, sclera, lymph nodes, retroperitoneal soft tissues, skin, peripheral and cranial nerves, and the spinal canal are all implicated in EMH. Active hematopoiesis occurs in these areas during the gestation period, which ends after delivery. However, if unsuccessful erythropoiesis persists, the extramedullary hematopoietic organs retain their potential to make red cells. Extramedullary hematopoietic tissue, according to some writers, can be extruded through the trabecular bone of the vertebral body or via the thinning trabeculae at the proximal ends of the ribs [5]. Immature and mature cells (predominantly erythroid and myeloid series) and dilated sinusoids containing precursors of red cells are found at the site of paraspinal EMH during the early stages of its evolution, and the lesions later become inactive, revealing some fatty tissue, fibrosis, or massive iron deposits.

EMH is related to chronic anemic states, most frequent in thalassemia; however, it is less common in other anemic and myeloproliferative illnesses such as acute myeloid leukemia, myelofibrosis, and polycythemia vera. It can occasionally result in cord compression, pleural effusion, severe hemothorax, and respiratory failure. EMH is an uncommon cause of paraspinal mass that should be distinguished from more frequent causes such as neurogenic tumors, lymphoma, metastasis, paravertebral abscess, and lateral meningocele. In 11-15% of EMH instances, the hematopoietic tissue is located paraspinal. Approximately 80% of these instances are asymptomatic and are frequently discovered by chance during imaging. Paraspinal EMH is often observed as a solitary mediastinal mass or in conjunction with abdominal paraspinal tumors. Because these masses are highly vascular, contrast-enhanced CT scans reveal significant homogenous enhancement [6]. MacCallum *et al.*, investigated a proliferative polycythemia vera in a 52-year-old man who developed EMH in the cervical and lumbar paravertebral areas and died as a result of cervical cord compression. However, in our investigation, the masses are in the thoracic and lumbar paravertebral areas, with cord compression. Masses in the thoracic and lumbar paravertebral regions of the EMH are uncommon.

The strength of the EMH signal on MRI is determined by the activity of the hematopoietic cells. Lesions in active hematopoiesis have intermediate signal intensity on T1-weighted images, and high signal intensity on T2-weighted images, indicating either immature or adult erythroid or myeloid series. Lesions in chronic inactive hematopoiesis are hypointense on T1 and T2 weighted images due to iron accumulation or display high signal intensity when fatty tissues predominate. The active hematopoietic lesions show strong enhancement after gadolinium delivery, but the dormant hematopoietic lesions show no enhancement. MRI is currently the gold standard for demonstrating spinal EMH; it produces superior soft tissue delineation and is highly sensitive [7, 8].

In acute myeloid leukemia. Most patients' treatment is typically divided into two chemotherapy (chemo) phases. If there is compressive myelopathy, laminectomy is done. In our case, the patient was later diagnosed with acute myeloid leukemia; the presence of paraspinal and presacral masses in the thoracic and lumbar regions was supposed to be EMH.

MRI is now the gold standard for showing spinal EMH; it gives more excellent soft tissue delineation and is very sensitive [7, 8].

In cases of acute myeloid leukemia. Most patients' treatment is usually divided into two chemotherapy (chemo) periods. Laminectomy is performed if there is compressive myelopathy. The patient in our case was eventually diagnosed with acute myeloid leukemia; the existence of paraspinal and presacral masses in the thoracic and lumbar areas was thought to represent EMH.

CONCLUSION

Our case illustrates a distinctive form of AML that causes nerve root compression. Extramedullary hematopoiesis should be considered in the differential diagnosis of paraspinal (lumbar) and presacral masses in acute myeloid leukemia based on distinctive clinical, biochemical, radiological, and histopathological features.

Abbreviations

AML: Acute myeloid leukemia; EMH: Extramedullary hematopoiesis; CECT: Contrast-enhanced computed tomography; CE-MRI: Contrast enhanced magnetic resonance imaging.

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Availability of Data and Materials: Not applicable.

Ethics Approval and Consent to Participate

Ethical approval was obtained from the institutional Ethics Committee.

Consent for Publication

The written and informed consent to publish the personal and clinical details of the participant was obtained from the patient.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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