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Presacral, Renal and Intrathoracic Extramedullary Hematopoiesis in Myelofibrosis: A Case Report

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Abstract: Extramedullary hematopoiesis (EMH) refers to the location of hematopoietic elements in locations other than the bone marrow andperipheral blood. It may be seen as a compensatory condition in many hematological conditions like myeloproliferative disorders included myelofibrosis, haemoglobinopathies. Intrathoracic EMH commonly develops in the posteroinferior mediastinum. Presacral EMH is an extremely rare condition and there are a limited number of case reports published in the literature. We are reporting a case of EMH that involves the posterior, middle mediastinum and presacral region on computed tomography in a patient of myelofibrosis.
Keywords: adult, ciliary body tumor; medulloepithelioma

INTRODUCTION

Extramedullary hematopoiesis (EMH) refers to the hematopoiesis that occurs in organs other than bone marrow [1]. EMH occurs under conditions of myelofibrosis and ineffective erythropoiesis, including that in thalassemia, hereditary spherocytosis and sickle cell disease [2-4]. While EMH may occur anywhere in the body, it most commonly presents as diffuse lesions in the liver, spleen and/or lymph nodes. In rare cases, EMH presents as a solitary mass [5], appearing as a tumor-simulating lesion in an atypical location, and is frequently misdiagnosed.

The most typical localization can be detected in the posteroinferior mediastinum of the paravertebral region. On the other hand, presacral location is very uncommon, and only a limited number of cases have been described in the literature [6-9]. Myelofibrosis is a haematological disorder where there is the replacement of bone marrow with collagenous connective tissue and progressive fibrosis. It is also classified as a myeloproliferative disorder. It is characterised by extramedullary hematopoiesis (EMH), progressive splenomegaly, anaemia and variable change in the number of granulocytes and platelets including thrombocytopenia.

CASE REPORT

A 35 year-old male with severe aneamia (Hb 6.6 gm/dl) presented to casuality with chronic lower back pain with constipation not relieved by medication. Physical examination revealed a pale conjunctiva, an icteric sclera, and marked hepatosplenomegaly. The peripheral blood findings demonstrate moderate anemia of the normocytic, normochromic type. The red cells were characterized by poikilocytosis with numerous teardrops, elongated in comma forms with Anisocytosis. Giant platelet fragments of megakaryocytes were also seen. In view of complaints patient was advised to undergo CECT chest and abdomen to rule out any pathology. It revealed evidence of large (12*10cms) presacral mass with soft tissue and fat component and bilateral paraspinal posterior mediastinum masses with extension into middle mediastinum on right side. Heart was seen to be displaced anteriorly. There was evidence of two fat containing soft tissue lesions in relation to right kidney. There was absence of calcification within masses along with presence of adipose tissue. The masses did not erode the adjacent bony structures. There was associated hepatospleenomegaly. Bone window of patient revealed diffuse osteosclerosis with obliteration of marrow cavity of long bones. MRI wasn't done as the patient was claustrophobic. Biopsy was taken to confirm the diagnosis. Biopsy revealed fibrocartilagenous tissue with myeloid and erythroid cells along with megakaryocytosis.

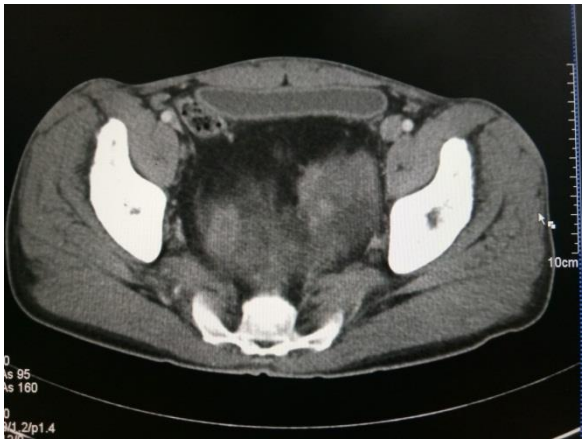

Fig-1: Axial CECT section revealing presacral fat containing soft tissue mass

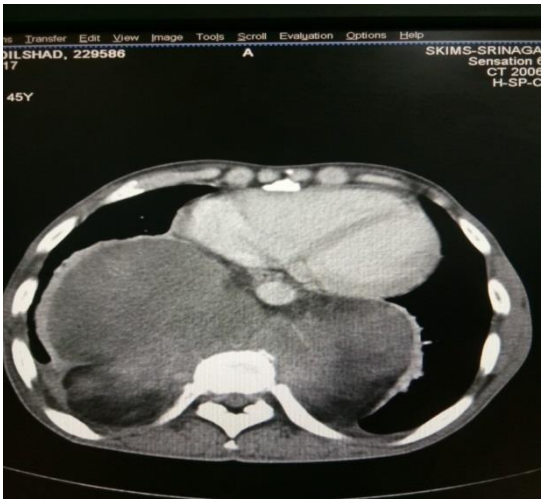

Fig-2: Axial CECT section revealing bilateral well defined lobulated posterior mediastinum masses with anterior displacement of heart.

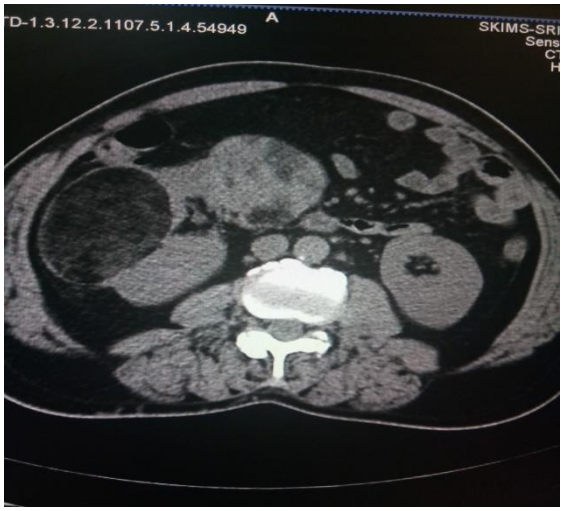

Fig-3: CECT Axial section revealing two lesions in relation to right kidney with one showing predominant fat component another showing predominant soft tissue component

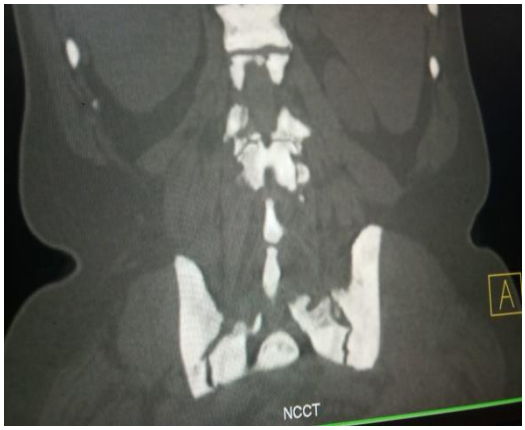

Fig-4: Coronal non contrast CT revealing evidence of diffusely increased bone density

DISCUSSION

Extramedullary hematopoiesis arises from the extrusion of a proliferating marrow through the cortex into a subperiosteal location. This feature explains its presence in a paravertebral or presacral location. Embryonic rests or totipotential cells are thought to be responsible for EMH in visceral sites [10]. EMH usually forms a soft, red mass resembling a hematoma on its cut surface. Histologically, EMH contains all hematopoietic elements including megacaryocytes and myeloid and erythroid cells at various stages of maturation [9].Myelofibrosis is characterized by fibrocartilagenous tissue infiltrated with erythroid and myeloid morphology of cells along with megakaryocytes. There may be multiple areas of hemorrhage with hemosiderin laden macrophages infiltrating adjoining fat.

Hepatosplenomegaly is the most common abdominal manifestation of extramedullany hematopoiesis. The spleen becomes massive; infarction is common. Portal hypertension, which develops in 1 0% of the cases of myeloid metaplasia, has the associated findings of ascites and vanices [1, 6].

Intrathoracic EMH commonly develops in the posteroinferior mediastinum. It is usually a multiple, bilateral, andasymptomatic condition. Destruction of adjacent ribs and vertebrae is rarely seen [11]. In our case, at the paravertebral area of the posteroinferior mediastinum, bilateral, regularly contoured EMH foci were determined that did not cause costal and vertebral destruction.In atypical cases with symptoms of compression or bone erosion, transthoracic and open biopsies are recommended. However, it should be noted that both procedures can cause a life-threatening hemorrhage. Because of the complication of hemorrhage, noninvasive methods including CT scans and MRI are preferred in the diagnosis of EMH [12, 13]. Renal extramedullary hematopoiesis needs to be differentiated from angiomyolipoma which also shows fat and soft tissue component.

Presacral EMH is an extremely rare condition, and there is a limited number of case reports published in the literature. More than 14 cases of EMH in the presacral region were reported. Among these cases, six patients were diagnosed as having thalassemia [14]. However, most of the patients were asymptomatic. Only two patients with a previous presacral EMH mass diagnosed as thalassemia were reported as symptomatic due to constipation and painful defecation [14].our case had constipation with chronic lower back pain with features of myelofibrosis.

On CT scan, presacral EMH is typically seen as a heterogeneous and lobulated mass with a smooth border of margin, adjacent and anterior to the sacrum, but formed without erosion. There may be fatty elements inside the mass, but calcifications are absent [7, 8, 14]. MRI is the technique of choice in evaluating this condition through its multiplanar capabilities and soft tissue resolution. On MRI, EMH masses may have intermediate signal intensity on both T1- and T2-weighted images. High signal intensity is seen around these masses due to the surrounding fat or inside the mass due to fat infiltration. Diffuse enhancement is seen after IV gadolinium chelate administration [7, 8, 15]. To support diagnosis, noninvasive methods including CT and/or MRI can be conducted instead of biopsy. In our case biopsy was taken as patient was extremely claustrophobic and alternate diagnosis in form of liposarcoma had to be ruled out. Liposarcomas are malignant tumours of fatty tissue and are the malignant counterpart to a benign lipoma. They are usually seen in the extremities (75%), most commonly the thigh, and are less commonly seen in the retroperitoneum, groin or elsewhere [17]. Differential diagnosis of Skeletal findings of myelofibrosis includes sclerosing dysplasias,fluorosis, renal osteodystrophy, lymphoma, osteoblastic metastasis etc.

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