Extramedullary Haematopoiesis Presenting with Backache and Paraparesis - A Case Report

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PRESENTATION OF CASE

A 49-year-old female patient was admitted in the medicine department of Assam Medical College with complaints of paraparesis of 15 days duration and backache of 3 months' duration. She had history of dizziness and low blood pressure. On general examination, she had tachycardia (heart rate of 126 beats / min), tachypnoea (respiratory rate 24 / min), severe pallor and bilateral non-pitting pedal oedema. The patient had low haemoglobin (6.6 gm / dl) with red blood cell (RBC) count of 2.5 million / mm^3.

She was sent to the radio-diagnosis department for non-contrast computed tomography (CT) scan of thorax and contrast enhanced magnetic resonance (CEMR) of dorsolumbar region. On CT scan, soft tissue density lesions were noted in paraspinal region extending from lower thoracic, lumbar region without destruction of vertebral bodies. Additional findings include hepato-splenomegaly and left sided pleural effusion. On CEMR, there was T1 hypo and T2 / T2FSiso-intense soft tissue lesion in paraspinal region extending from lower thoracic, lumbar and sacral vertebrae. The lesions were showing diffusion restriction, diffusion weighted imaging (DWI) with low apparent diffusion coefficient (ADC) value and mild enhancement on post contrast scan. The soft tissue component was noted to extend into the epidural space in multiple vertebral levels in thoracic, lumbar and sacral regions causing spinal canal stenosis, maximum (7.0 mm) at L4 vertebral level. Based on the imaging findings, a diagnosis of extramedullary haematopoiesis in paraspinal region with extension of the soft tissue in the epidural space leading to compression of spinal cord at multiple levels was made.

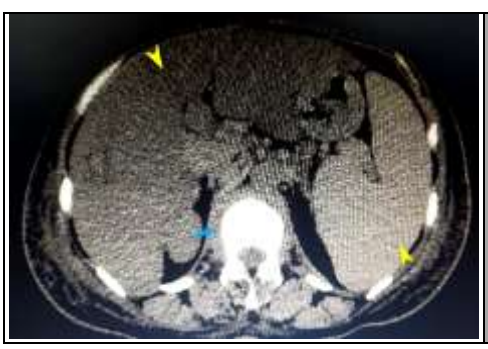

Figure 1.
Axial NECT Image Shows Paraspinal Soft Tissue Thickening (Blue Arrow) with Hepatosplenomegaly (Arrowhead)

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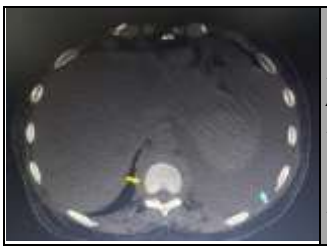

Figure 2.
Axial NECT Image Showing Paraspinal Soft Tissue Thickening (Yellow Arrow) with Left Sided Pleural Effusion (Blue Arrow)

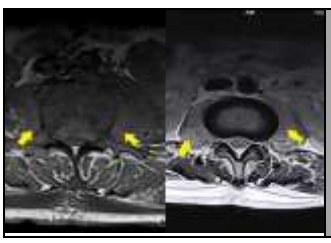

Figure 3.
T1W MR Image Showing Iso to Hypointense and T2WI Showing Iso to Mildly Hyperintense Soft Tissue Lesion in Paraspinal Region (Yellow Arrows)

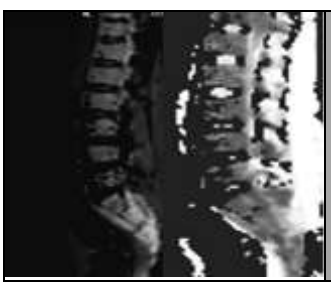

Figure 4.
DW Image Shows Diffusion Restriction in Paraspinal Region with Low ADC

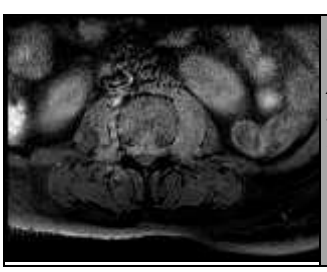

Figure 5.
On Gadolinium Contrast Administration, the Paraspinal Soft Tissue Shows Near Homogenous Enhancement with Extension of the Soft Tissue in the Epidural Space Causing Spinal Cord Compression

DISCUSSION

Extramedullary haematopoiesis (EMH) is a compensatory mechanism that occurs in the human body in response to low blood RBC count which is associated with many hematologic disorders and bone marrow dysfunction. EMH primarily occurs in liver, spleen, pleura, lymph nodes, adrenal glands, breast, thymus, kidneys, gastrointestinal tract, intracranial structures and paraspinal regions. Although it is asymptomatic, involvement of various organs in EMH may mimic as symptomatic mass within the organ.[1,2] EMH may occur as a result of several clinical causes which include haemolytic anaemia, primary and secondary myelofibrosis, leukemia, lymphoma and bone metastasis, and myelodysplastic syndromes, characterised by abnormal morphology and inadequate production of blood cells.[3]

Two main categories are seen in EMH. The first category shows para-osseous foci that may be due to herniation of medullary tissue from the underlying bone and is generally seen in haemolytic disorders such as thalassemia and sickle cell anaemia, where the marrow has good cellularity. The second category shows extra osseous soft tissue foci arising from multipotential stem cells when the bone marrow activity is ineffective, seen in idiopathic myelofibrosis or, rarely, with toxic or tumoral marrow destruction.[4] A paraspinal location in hematopoietic tissue occurs in 11 – 15 % of cases with EMH.[5] Involvement of spinal cord may occur by hematopoietic tissue, most frequently in the lumbar region and to a lesser extent in thoracic region.[6]

Paraspinal EMH has a typical presentation of soft tissue thickening or pseudo-tumour and may sometimes appear as symptomatic tumour like mass causing variety of neurological symptoms due to secondary spinal cord compression. More than 80 % of cases don't have the signs and symptoms related directly to the disorder and the lesions are typically seen incidentally during radiological investigations. Clinical presentation of the spinal cord involvement includes back pain, lower extremity pain, paraesthesia, abnormal proprioception, exaggerated or brisk deep tendon reflexes, Babinski response, Lasegue's sign, paraparesis, paraplegia, ankle clonus, spastic gate, urgency of urination and bowel incontinence. The severity, acuteness and multiplicity of signs and symptoms depends on the size and location of the soft tissue and extent of involvement of spinal cord.[1] The development of neurologic symptoms is directly proportional to the chronicity of the disease, however, third and fourth decades of life has highest predilection of developing neurologic symptoms.[7]

History and physical examination are important to narrow the differential diagnosis. Radiographic imaging like CT-scan and MRI (magnetic resonance imaging) gives confirmation of the existence of hematopoietic tissue. MRI is the diagnostic investigation of choice. Paraspinal EMH appears as unique, multiple masses usually lobular, well-circumscribed of intermediate signal intensity on T1-weighted images and iso to low signal intensity on T2-weighted images with homogeneous mild enhancement following contrast administration.[1] The CT appearance is characterised by the homogenous soft tissue density mass. The diagnosis is confirmed following surgical removal of the mass followed by biopsy.[8]

There is no standard treatment protocol or evidence-based guidelines for the treatment of EMH. The therapy typically depends on the severity of symptoms, size of the mass, patient's clinical condition and previous treatment. Transfusion therapy, laminectomy, radiotherapy and the use of fetal haemoglobin, inducing agents are the treatment options which decrease the need of EMH. Although biopsy remains the gold standard, in radiological point of view cross-sectional imaging like CT and MRI are the primary investigation to diagnose the EMH and its extent of spinal cord involvement.

To conclude, EMH is a rare entity involving various organs and paraspinal regions that causes variety of signs and symptoms which should be essentially differentiated from the neoplastic masses or secondaries. In our study, we presented a case of EMH which was diagnosed on the basis of findings in CT scan and CEMRI. On blood investigation, she was confirmed to have anaemia. This case has indicated that EMH should be considered as a differential diagnosis when paraspinal masses with characteristic radiologic appearance are identified in patients with various blood dyscrasias.[9]

FINAL DIAGNOSIS

Extramedullary haematopoiesis

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Disclosure forms provided by the authors are available with the full text of this article at jebmh.com.

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