Case Report

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Spinal cord compression secondary to intraspinal extramedullary hematopoiesis

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Abstract:

Extramedullary hematopoiesis (EMH) is a rare cause of spinal cord compression (SCC) occurring in various hematological illnesses such as myeloproliferative disorders, thalassemias, and various types of anemia. EMH represents the growth of blood cells outside the bone marrow. Common EMH locations include the spleen, liver, lymph nodes, and paravertebral regions. When this occurs in the spinal cord, the mass effect can compress the spinal cord and cause different neurological symptoms depending on the area of the spinal cord affected. This report describes a 27-year-old female with a known case of beta-thalassemia major, who presented with mid-thoracic back pain, weakness, and paresthesia at the T10 level. In addition, this report illustrates the importance of considering EMH in the differential diagnosis of SCC in patients with thalassemia.

Keywords:

Beta-thalassemia, extramedullary hematopoiesis, spinal cord compression

Introduction

Extramedullary hematopoiesis (EMH) is known as proliferation of noncancerous hematopoietic tissue outside the bone marrow. Occasionally, this condition results when the functioning blood cells to meet the body oxygen demand are inadequate. EMH particularly occurs when the patient has thalassemia, acquired hemolytic anemia, sickle cell anemia, folate, or Vitamin B deficiency. It also occurs in myelophthisic diseases in which the bone marrow is disrupted, such as leukemia, lymphoma, or myelofibrosis.[1] EMH rarely occurs in the spinal canal but commonly presents in the liver, spleen, and lymph nodes.[2,3] In 1954, Gatto *et al*.,[4] discovered the first case of spinal cord compression (SCC) secondary to EMH. Intraspinal EMH in thalassemic patients accounts for 11%–15% of overall cases.[5,6] In the spinal region, the most common site affected is the thoracic region owing to its narrowed space, followed by the lumbar region.[3]

Case Report

A 27-year-old Saudi female, a known case of beta-thalassemia major, presented to the emergency department with mid-thoracic back pain, weakness, and paresthesia at T10 level. The patient was admitted to the medical ward because of symptoms of severe anemia, palpitation, generalized fatigue, and exertional dyspnea in association with back pain. The patient had had splenectomy 6 years previously but had no history of a sphincter abnormality.

On physical examination, the vital signs were within normal range, but the patient looked ill, was severely pale, and slightly jaundiced. The abdomen was soft, lax

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without hepatomegaly. The neurological examination found the patient alert, conscious, and cooperative, with a Glasgow Coma Scale of 15/15. Motor examination showed normal power and tone with hyperreflexia bilaterally at knee reflex and ankle reflex, with bilateral extensor planter response. Finally, the sensory examination showed sensory level at T10 level with no coordination or gait abnormality otherwise.

The complete blood count results were as follows: white blood cell 34.6; Hb 3.5; platelets 272; mean corpuscular volume 83.2; mean corpuscular hemoglobin (MCH) 22.6; MCH concentration 27.1; urea 3.5; creatinine 31; Na 134; K 4.7; total bilirubin 52.9; direct bilirubin 9.7; and lactate dehydrogenase 377. Renal function tests and liver function tests were within normal range.

Magnetic resonance imaging (MRI) findings showed sagittal postcontrast T1 with fat sat-weighted image showing low bone marrow signal intensity, enhancing posterior epidural masses in the thoracic region, which are isointense signal to bone marrow compressing the thoracic cord. There were similar masses in the ventral epidural space at the lumbosacral spine [Figure 1a]. Axial T1-weighted image showed paraspinal masses isointense signal to bone marrow [Figure 1b].

The patient was diagnosed with multi-level intraspinal EMH at level of D3, D6, L5, S1, and S2 in addition to direct and indirect autoimmune hemolysis. The management regimen was rituximab 375 mg IV in 500 mL N.S. once weekly, calcium PO 600 mg BID, folic acid PO 5 mg PO OD, hydroxyurea PO 500 mg BID, Nexium PO 20 mg OD, and prednisolone PO 50 mg OD.

The patient was transferred to a tertiary hospital and started on radiotherapy, and the SCC improved. Follow-up MRI findings after radiation therapy showed sagittal T1 [Figure 2a] and postcontrast T1 with fat sat [Figure 2b]-weighted images showed interval regression in size of enhancing posterior epidural masses in the thoracic region with interval improvement of compression on the thoracic cord which showed subtle high T2 signal alteration.

Discussion

EMH is a compensatory mechanism of the body characterized by the presence of hematopoiesis in sites other than the bone marrow.[7] EMH is known to be caused by hematological disorders such as myelofibrosis, sickle cell anemia, polycythemia vera, and thalassemia among others. The most common sites of EMH are organs involved in the process of fetal hematopoiesis, such as the liver, spleen, kidneys, and lymph nodes.[1,7] However, the occurrence of EMH can involve any body site including the heart, breasts, thymus, adrenal glands, retroperitoneal tissue, pleura, skin, intracranial structures, and paraspinal regions.[8,9] EMH in the paraspinal region is a rare cause of SCC.

The clinical presentation of EMH can range from being asymptomatic to being found incidentally to back pain, paraplegia, paresthesia, bowel incontinence, or other neurological symptoms as a result of significant SCC. The most severe cases could lead to permanent neurological injury.[1,8,9] History and physical examination play a significant role in narrowing the differentials. Radiological investigations, preferably MRI, remain essential in establishing the diagnosis.[8] MRI can show multiple distinctive isointense to hyperintense masses on T1–T2-weighted imaging. On administration of a contrast, it exhibits homogeneous enhancement.[1,8] Occasionally, computed tomography-guided needle biopsy could be utilized to confirm the diagnosis.[1]

The review of the literature revealed several management options in patients with SCC due to EMH. However,

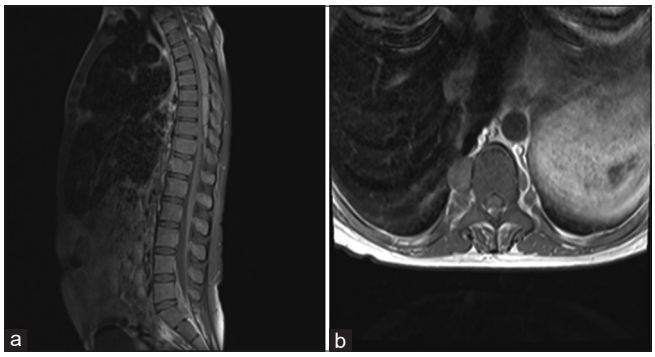

Figure 1: (a) Sagittal postcontrast T1 with fat sat-weighted image showing low bone marrow signal intensity, enhancing posterior epidural masses in the thoracic region which are isointense signal to bone marrow compressing the thoracic cord. Similar masses in the ventral epidural space at the lumbosacral spine. (b) Axial T1-weighted image showing paraspinal masses isointense signal to bone marrow

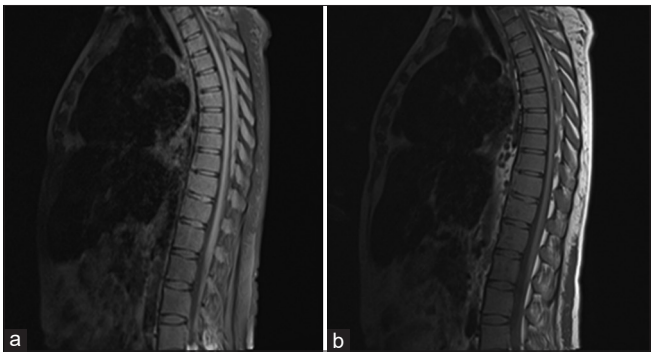

Figure 2: Follow-up magnetic resonance imaging after 2 years of radiation therapy (a) sagittal T1 and (b) postcontrast T1 with fat sat-weighted images showing interval regression in size of enhancing posterior epidural masses in the thoracic region with interval improvement of compression on the thoracic cord which shows subtle high T2 signal alteration

since it is considered a rare entity, the most appropriate therapeutic management is still under debate.[8] The treatment options include surgical decompression, radiotherapy, blood transfusions, hydroxyurea, or any combination of those therapies. Some studies have reported significant neurological improvement following either single or combined surgical decompression with radiotherapy.[1] Other studies have reported remarkable neurological recovery upon the administration of hypertransfusion and/or hydroxyurea.[9]

The presented case is SCC due to EMH in a patient known to have beta-thalassemia major. The patient presented with mid-thoracic back pain, weakness, and paresthesia at T10 level. After establishing the diagnosis with MRI, the patient underwent radiotherapy resulting in a dramatic improvement of her neurological symptoms.

Conclusion

Although rare, EMH should be suspected in a patient, known to have chronic hematological disorders, presenting with signs of SCC. Therefore, appropriate radiological investigations are essential in establishing the diagnosis. There are several treatment options, but the standard therapeutic management remains controversial. Early diagnosis and proper management in patients with SCC owing to EMH is crucial in the prevention of permanent neurological damage.

Declaration of patient consent

The author certifies that all appropriate patient consent forms to publish the case report were obtained. In the forms, the patient gave her consent for her images and other clinical information to be reported in the journal. She understood that her name and initials will not be published, and every effort would be made to conceal her identity, though anonymity could not be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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