



Image Report

Lumbosacral extradural extramedullary hematopoiesis in thalassemia major causing spinal canal stenosis

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ABSTRACT

Background: Extramedullary hematopoiesis (EH) is common in patients with ineffective erythropoiesis like thalassemia major (TM). EH commonly involves intra-abdominal organs (e.g., spleen and liver), but rarely involves vertebral bodies. Here, we reviewed images of EH contributing to spinal canal stenosis.

Case Description: A 19-year-old male with beta-thalassemia major (TM) presented with pain and bilateral lower extremity neurogenic claudication. Bilaterally, on examination, he had positive straight leg raising to 30°, a loss of the Achilles responses, and decreased pain appreciation in the L5/S1 distributions. The lumbar MR showed anterior epidural lobulated mass lesions at L5 and S1, contributing to marked canal stenosis. Following an L5/S1 laminectomy for decompression, the biopsy revealed extramedullary hematopoietic tissue.

Conclusion: Patients presenting with the lower extremity symptoms/signs, ranging from low back pain to neurogenic claudication and even paraplegia, may have EH secondary to TM. Treatment options include hypertransfusion, local radiation therapy, and/or surgical decompression.

Keywords: Beta-thalassemia, Extramedullary hematopoiesis, Radiculopathy, Spinal stenosis

INTRODUCTION

Extramedullary hematopoiesis (EH) is observed in many hemoglobinopathies, including beta-thalassemia major (TM). Here, we report a patient with TM who, while receiving chronic transfusions, developed epidural EH resulting in spinal canal stenosis at the L5/S1 level requiring operative decompression.

CASE DESCRIPTION

A 19-year-old male with beta-TM complicated by chronic iron overload, presented with bilateral lower extremity pain, numbness, and neurogenic claudication. Bilaterally, on examination, he had positive straight leg raising to 30°, a loss of the Achilles responses, and decreased sensation in the L5-S1 distributions. When the lumbar MR demonstrated L5/S1 EH responsible for spinal canal stenosis [Figure 1], the patient underwent an L5/S1 laminectomy; postoperatively, all preoperative symptoms and signs resolved. The histopathological evaluation of the EH mass revealed hyperplastic hematopoietic tissue.

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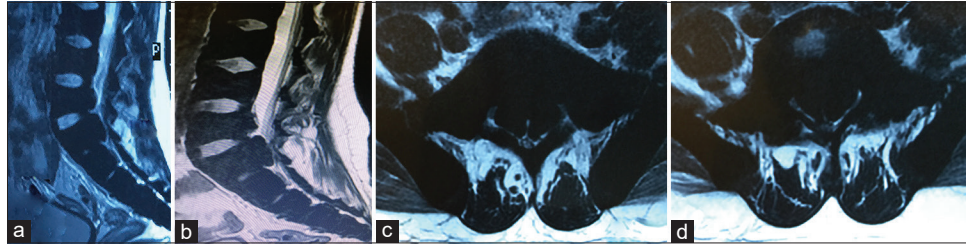


Figure 1: (a and b) Sagittal T2-weighted images showing L5, S1 level anterior epidural hypointense lobulated mass lesion attached to the posterior surface of the vertebral body with severe canal stenosis. (c and d) Axial T2-weighted images showing bi-lobulated lesion protruding from the vertebral body with near total canal stenosis.

DISCUSSION

Location and frequency of occurrence of spinal EH with TM

EH has a prevalence in transfusion-dependent beta-thalassemia of <1%.^[4,6] EH typically occurs systemically (e.g., in many abdominal and/or thoracic organs). In the central nervous system (CNS), it commonly involves the choroid plexus, cranial dural folds, and peripheral nerves and can even be found within some brain tumors.^[1] In the spinal canal, EH often involves contiguous vertebral bodies and can extend into the spinal canal where it may present as a pseudotumor.^[2]

Differential diagnosis and MR findings of EH with TM

The differential diagnoses for EH spinal lesions include an epidural infection, vertebral fracture, or epidural hematoma.^[3] MRI, the study of choice, frequently demonstrates a lobulated hypointense mass dorsal to a vertebral body that does not enhance with contrast.

Treatment options for EH with TM

Treatment of EH with TM include; hypertransfusion, local radiation therapy, and/or surgical decompression. Blood transfusions can prevent further progression of EH pseudotumor.^[5] Radiotherapy reduces the volume of pseudotumor, thus relieving local compressive symptoms/signs. Surgery, such as decompressive laminectomy, offers immediate relief of cord compression, and/or canal obstruction.^[1,3]

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Amirjamshidi A, Abbassioun K, Ketabchi S. Spinal extradural hematopoiesis in adolescents with thalassemia. *Childs Nerv Syst* 1991;7:223-5.
2. Chehal A, Aoun E, Koussa S, Skoury H, Koussa S, Taher A. Hypertransfusion: A successful method of treatment in thalassemia intermedia patients with spinal cord compression secondary to extramedullary hematopoiesis. *Spine (Phila Pa 1976)* 2003;28:E245-9.
3. Munn RK, Kramer CA, Arnold SM. Spinal cord compression due to extramedullary hematopoiesis in beta-thalassemia intermedia. *Int J Radiat Oncol Biol Phys* 1998;42:607-9.
4. Taher A, Isma'el H, Cappellini MD. Thalassemia intermedia: Revisited. *Blood Cells Mol Dis* 2006;37:12-20.
5. Taher A, Vichinsky EP, Musallam K, Cappellini MD, Viprakasit V, Thalassaemia International F. Guidelines for the Management of Non Transfusion Dependent Thalassaemia (NTDT). Cyprus: Thalassaemia International Federation; 2013.
6. Taher AT, Musallam KM, Karimi M, El-Beshlawy A, Belhoul K, Daar S, *et al.* Overview on practices in thalassemia intermedia management aiming for lowering complication rates across a region of endemicity: The optimal care study. *Blood* 2010;115:1886-92.

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