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MR of Extramedullary Hematopoiesis Causing Cord Compression in Beta-Thalassemia

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Summary: The authors detail the MR findings in a 31-year-old woman with beta-thalassemia intermedia (a posterior epidural mass hyperintense to contiguous hyperplastic marrow, cord compression from T4-T11, expansion of L5 and the sacrum, and widening of the diploic space), and emphasize the need for early diagnosis.

Index terms: Spinal cord, magnetic resonance; Spinal cord compression

While spinal cord compression by an epidural extramedullary hematopoietic mass has been described, there is only one description of the MR findings in beta-thalassemia intermedia (1). In addition to recurrent spinal cord compression by extramedullary hematopoiesis, this patient also demonstrates the MR appearance of involvement in the clivus, skull and orbits.

Case Report

A 30-year-old woman presented with back pain, weakness and numbness in both lower extremities, and difficulty initiating voiding. The patient was diagnosed at age 3 with beta-thalassemia intermedia.

Physical examination revealed frontal bossing, a prominent mandible, T4-level tenderness, decreased strength, light touch, pin prick and temperature in both lower extremities, abdomen and back with a T7-T9 level, and decreased vibratory and position sense in both toes. Bilirubin was 2.3/0.3 mg% with a serum iron of 241. Emergency magnetic resonance (MR) was performed of the entire spine (Figs. 1 and 2). This was followed by MR of the brain (Fig. 3). Intravenous dexamethasone and radiation therapy was initiated emergently. The patient's lower extremity strength improved and sensation was regained. One year later, she returned with similar complaints and MR demonstrated a recurrent posterior epidural mass compressing the spinal cord from T6-T9.

Discussion

Extramedullary hematopoiesis rarely occurs in the spinal epidural space. Although well described, spinal cord compression is considered an unusual complication of thalassemia with an unexplained predilection for the thoracic region. The diagnosis can be made confidently except in the patient with a primary malignancy, where differentiation from epidural metastases can be difficult. If the lesion is isolated, metastatic disease or an epidural abscess should be considered. With more diffuse involvement, lymphoma also needs to be excluded. The role of contrast enhancement in narrowing the differential diagnosis needs to be further investigated.

Several theories have been proposed for the etiology of extramedullary hematopoietic tissue within the spinal epidural space: 1) extrusion of proliferating, hypertrophied marrow into a subperiosteal location (2-6); and 2) transformation of hematopoietic precursors of mesodermal origin, located in the thoracic epidural space, into marrow (2, 4–7).

Patients with thalassemia intermedia seem to be more likely to develop extramedullary hematopoiesis (3). With a milder clinical form and an ability to maintain hemoglobin levels near normal without transfusions, they have a longer life expectancy. However, this is at the expense of severe hematopoietic stress (8).

Marrow erythroid hyperplasia and hypertrophy result in enlargement and decreased T1 signal of the vertebral bodies. Thinning of the outer table and widening of the diploic space cause a marked increase in skull thickness, especially in the frontal region. The occipital bone is spared secondary to lack of hematopoietic marrow. Clival and periorbital marrow expansion may result in neu-
Fig. 1. Sagittal T1-weighted image 600/20/2 (TR/TE/excitations) shows a posterior epidural mass (arrows), slightly hyperintense to the adjacent hyperplastic marrow (perhaps due to infiltration into epidural fat), compressing the spinal cord from T4-11.

Fig. 2. Expansion of L5 and the sacrum secondary to marrow hyperplasia with adjacent extramedullary hematopoiesis (arrows) causing encroachment of the spinal canal.

Fig. 3. Sagittal (A) and coronal (B) T1-weighted images (600/20) demonstrate widening of the diploic space, greatest in the frontal bone (arrows). Note the normal appearance of the occipital bone. The clivus and sphenoid bone are expanded (small arrows), resulting in cavernous sinus compromise.

References
2. Jackson DV Jr, Randall ME, Richards F II, et al. Spinal cord compromise due to extramedullary hematopoiesis in thalassemia: long-term rologic compromise due to brain stem, basilar artery, or optic nerve impingement. Epidural extramedullary hematopoietic masses are radiosensitive with a rapid response to low doses. Therefore, a favorable prognosis depends on an early diagnosis.