MR of Thoracic Cord Compression Caused by Epidural Extramedullary Hematopoiesis in Myelodysplastic Syndrome

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Summary: Spinal cord compression caused by extramedullary hematopoiesis is a rare complication of chronic anemic states, most frequently occurring in patients with \(\beta\)-thalassemia. We report the MR appearance of extramedullary hematopoiesis resulting in cord compression in a patient with a myelodysplastic syndrome, which was isointense with the spinal cord on T1-weighted images and markedly hypointense on fast spin-echo T2-weighted images, and that demonstrated enhancement.

Index terms: Spinal cord, abnormalities and anomalies; Spinal cord, compression

Rarely, extramedullary hematopoiesis may complicate chronic anemic states. Most frequently, this occurs in patients with \(\beta\)-thalassemia, although it has also been described in patients with sickle-cell anemia, myelofibrosis, and polycythemia vera (1). Abnormal hematopoietic tissue usually develops in sites involved in hematopoiesis during fetal development, such as the spleen, liver, and kidneys (2, 3); however, other locations, such as the paraspinal tissue, especially in the posterior mediastinum, may be involved (4). Rarely, the epidural space around the spinal cord may be a site of extramedullary hematopoiesis. The resulting mass may cause neurologic impairment due to spinal cord compression (1). We describe the magnetic resonance (MR) findings of thoracic cord compression caused by extramedullary hematopoietic tissue in a patient with myelodysplastic syndrome.

Case Report

A 79-year-old man with a history of chronic anemia caused by myelodysplastic syndrome had cellulitis of the right leg. His medical history was notable for chronic inflammatory demyelinating polyneuropathy with symptoms of gait disturbance and a distal sensory neuropathy in the lower extremities. Physical examination revealed a fever to 101.5°F as well as a markedly swollen, erythematous right leg. His neurologic examination showed diminished perception to light touch bilaterally below the knees, thought to be related to his chronic inflammatory demyelinating polyneuropathy. His motor examination was nonfocal. Deep tendon reflexes were intact and symmetric throughout with downgoing toes. Laboratory studies were notable for a leukocytosis with bandemia (15 100 white blood cell count with 26% bands), normochromic/normocytic anemia (hemoglobin, 10.3 mg/dL), and thrombocytopenia (95 000 platelets).

The patient was treated with intravenous antibiotics as well as surgical incision and drainage. Ten days after admission, he noted onset of numbness in the buttocks and leg weakness. Neurologic examination revealed a T-6 sensory level, mild proximal lower extremity weakness, diminished achilles and patellar tendon reflexes, and bilateral Babinski responses. MR imaging showed multiple well-circumscribed, enhancing lesions in the posterior epidural space extending from the T-5 through the T-8 levels and resulting in thoracic cord compression (Fig 1A–D). Focal T2 hyperintensity in the compressed cord was also noted.

The patient was given high-dose intravenous steroids and 2 days later underwent a T-5 through T-8 laminectomy. Discrete flesh-colored extradural masses were embedded in thickened, partially degenerated epidural fat. The dura mater was hemosiderin-stained along the extent of exposure, but was intact. Histopathologic analysis of the epidural masses revealed extramedullary hematopoiesis with numerous ringed sideroblasts. This was consistent with the previous diagnosis of refractory anemia with ringed sideroblasts, a form of myelodysplastic syndrome. The morphologic appearance of the epidural hematopoietic tissue was similar to the bone marrow biopsy specimen, including the degree of cellularity and the presence of ringed sideroblasts (Fig 1E and F). The patient’s postoperative course was complicated only by a brief period of
Fig 1. Extramedullary hematopoiesis resulting in thoracic cord compression in a 79-year-old man with myelodysplastic syndrome.

A, Sagittal spin-echo T1-weighted (550/17 [repetition time/echo time]) MR image of the mid-thoracic spine shows three well-demarcated masses embedded within the posterior epidural fat that are isointense with the spinal cord. Cord compression is present. Also note the extensive decreased signal intensity throughout the vertebral bodies, consistent with diffuse replacement of the marrow. Multiple vertebral body wedge deformities are noted in the mid-thoracic spine.

B, On a sagittal fast spin-echo T2-weighted (2700/85) MR image the epidural masses are markedly hypointense, probably related to excessive iron deposition within the cellular elements as well as ringed sideroblasts constituting a large portion of the extramedullary hematopoietic tissue. The dura is nicely illustrated (arrow). High signal intensity in the cord is seen at levels of compression.

C, Axial spin-echo T1-weighted (600/11) MR image shows a mass (M) within the epidural fat. The compressed cord is anteriorly displaced within the spinal canal (arrow). Also note extramedullary hematopoiesis in the right paravertebral region (arrowheads).

D, Contrast-enhanced, fat-suppressed sagittal spin-echo T1-weighted (600/11) MR image shows enhancement of the epidural masses (M).

E and F, Histologic features of resected epidural hematopoiesis, which are markedly similar to those of the bone marrow biopsy (not shown). In E, hematopoietic tissue consists of megakaryocytes (arrows), erythroid precursors (white arrowheads), and granulocytic precursors (black arrowheads) within scant remaining epidural fat (hematoxylin-eosin, original magnification ×40). In F, iron stain highlights numerous ringed sideroblasts (nucleated erythroid precursors with abnormal perinuclear iron stores) (arrows) and macrophages containing diffuse cytoplasmic iron (arrowheads) (Gomori stain, original magnification ×100).
Discussion

Extramedullary hematopoiesis is rare, occurring most commonly in chronic anemic states. The most frequent sites of extramedullary hematopoiesis include the spleen, liver, or kidney; less commonly, the adrenal gland, heart, lymph nodes, or thymus are involved (5). Spinal cord compression due to epidural hematopoietic tissue is extremely rare. According to reports in the literature, symptomatic cord compression most frequently involves the thoracic cord (1, 6–13). Epidural masses cause neurologic signs and symptoms by direct pressure on nerve roots and the spinal cord. Early in the clinical course, symptoms may include focal back pain and paresthesias, progressing to sensory impairment, spastic weakness, bladder and bowel dysfunction, and abnormal reflexes. Paraplegia or quadriplegia may ensue (14).

The source of epidural hematopoietic tissue in extramedullary hematopoiesis is controversial. In patients with chronic anemia, blood-forming elements in the vertebral marrow may be extruded through weakened trabecular bone into the epidural compartment, where they may proliferate. This hypothesis appears to be supported by reports of radiologic contiguity between these epidural masses and intramedullary marrow (8). Alternatively, extramedullary hematopoiesis occurring in the epidural space may arise from rests of primitive hematopoietic stem cells, which later expand under the extreme hematologic stress present in patients with severe chronic anemia (13).

The rare complication of cord compression due to extramedullary hematopoiesis has most often been reported in patients with β-thalassemia (6–10, 13, 15). Cord compression caused by extramedullary hematopoiesis has also been reported in cases of myelofibrosis (11), sickle-cell anemia (10), and polycythemia vera (12). In our case, thoracic cord compression was due to epidural extramedullary hematopoiesis caused by myelodysplastic syndrome.

Myelodysplastic syndrome is a preleukemic state characterized by a chronic, refractory normocytic anemia, often accompanied by neutropenia and thrombocytopenia. There are a variety of subtypes of this syndrome, which are based on the cellular and histologic characteristics of the peripheral blood and bone marrow aspirate smear (16). Patients with myelodysplastic syndrome have clonal abnormalities of hematopoietic stem cells, often containing various cytogenetic chromosomal abnormalities; certain subtypes may undergo evolution to frank acute leukemia (17). In our case, the patient had refractory anemia with ringed sideroblasts, which are notable for the presence of large amounts of mitochondrial iron in erythroid precursors (17).

Other entities need to be considered in the differential diagnosis of spinal cord compression from epidural masses. In this particular patient, other possibilities included lymphoma, metastases, and epidural abscess. In addition, extramedullary acute myeloid leukemia should be considered in patients with myelodysplasia. Nuclear scans (technetium sulfur colloid) may show ectopic foci of hematopoietic tissue (1). Before the advent of MR imaging, computed tomography (CT) (10, 15) and/or CT myelography (8–10) were used to help diagnose cord compression. Presently, MR imaging is the technique of choice in evaluating this condition. The multiplanar capabilities and excellent soft-tissue resolution afforded by MR imaging allow precise delineation of the location, size, and extent of lesions causing impingement of the spinal cord (7–10). Earlier case reports of MR findings in cases of cord compression due to extramedullary hematopoiesis noted epidural masses that were hyperintense relative to cord on T2-weighted images as well as the abnormally decreased marrow signal from erythroid hyperplasia in anemia (7–12).

In the present case, diffuse T1 hypointensity of the bone marrow throughout the vertebral column was noted, as is common in patients with chronic anemia. However, unlike in previously reported cases, the epidural masses representing extramedullary hematopoiesis were markedly hypointense on T2-weighted images. Since this patient’s subtype of myelodysplastic syndrome characterized by the presence of large amounts of mitochondrial iron (ringed sideroblasts) in erythroid tissue (17) is different from other anemias, iron deposition most likely accounts for the dramatic T2 hypointensity, as has been described in the deep gray matter (18). β-thalassemia, sickle-cell anemia, myelofibrosis, and polycythemia vera lack this unusual iron deposition in hematopoietic tissue,
accounting for their different MR imaging characteristics (7–12). T2 hyperintensity was present in the spinal cord of our patient. Others have also reported increased T2 signal intensity within the cord and have suggested that this is related to myelomalacia and/or gliosis (8). Other possibilities would include compressive or venous edema.

Treatment of cord compression due to extramedullary hematopoiesis is controversial. Intravenous steroids may be used on an emergency basis as a temporizing measure until definitive therapy can be arranged (7, 12). Decompressive laminectomy has been used often, as in this case (9). Surgery also allows for tissue biopsy and histopathologic examination. As hematopoietic tissue is composed of rapidly proliferating cells, radiotherapy has also been used either as an adjunct to surgical decompression (1) or, in some cases, alone as the primary therapy (7, 8, 10, 11). Advantages of treatment with radiation therapy include the avoidance of a surgical procedure and its risks. However, radiation may induce bone marrow suppression in an already anemic patient. In certain clinical situations, in which both surgery and radiation present special risks (such as in pregnancy), transfusion therapy has been successfully used to cause regression of hematopoietic tissue by relieving the anemic stress (6). A recent report has also suggested the novel use of hydroxyurea in this condition, which may act as a cytostatic agent and/or increase production of hemoglobin-F (15).

References