Extramedullary hematopoietic tumor mimicking a thoracic nerve root schwannoma

Case report

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Extramedullary hematopoiesis secondary to chronic anemia is well reported throughout the literature. A rare presentation of this condition is in the central nervous tissue reported most frequently as an epidural mass causing spinal cord compression. The authors report the case of a 51-year-old man with β-thalassemia and chronic anemia who was found to have a 4-cm paravertebral mass suggestive of a schwannoma. The patient underwent transthoracic resection of the mass. Histological examination confirmed an extramedullary hematopoietic tumor. In this article, the authors propose a method to distinguish extramedullary hematopoietic tumors from schwannomas. To the authors' knowledge, this is the first reported case in the neurosurgical literature of this phenomenon.

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KEY WORDS • schwannoma • extramedullary hematopoiesis • β-thalassemia

Abbreviation used in this paper: EMHT = extramedullary hematopoietic tumor.

Case Report

History and Presentation. This 51-year-old Asian man presented with a several-year history of right-hand numbness and paresthesias. Pertinent medical history included β-thalassemia associated with chronic anemia. He underwent a splenectomy at the age of 30 and received several blood transfusions during his lifetime for his chronic anemia. A review of systems was significant for a persistent dry cough.

Physical Examination. On examination, the patient was found to have hyperreflexia in his lower extremities, but not in his upper extremities. Muscle bulk, tone, and strength were normal. Sensory examination revealed hypoesthesia in his right flank in the distribution of the T-10 dermatome.

Laboratory Data. The results of a complete blood count performed on admission were consistent with chronic anemia secondary to β-thalassemia: hemoglobin

This article contains some figures that are displayed in color online but in black and white in the print edition.
level 9.4 g/L, hematocrit 30.0%, mean corpuscular volume 71.4 fl. Manual differentiation showed 2+ target cells, 1+ hypochromic red blood cells, 1+ schistocytes, and occasional sickle cells, consistent with β-thalassemia.

**Neuroradiological Imaging.** Magnetic resonance imaging of the cervical and thoracic spine demonstrated mild cervical spondylosis, mild foraminal stenosis at the C5–6 level, and a 4-cm ovoid paravertebral mass in the right intervertebral foramen at T-10. There was no clear violation of the parietal pleura. After gadolinium administration, the mass exhibited intense, homogeneous enhancement (Fig. 1). The images were reviewed independently by 2 neuroradiologists and interpreted as showing thoracic schwannoma.

**Surgical Intervention and Histological Findings.** The patient underwent gross-total resection of the tumor through a right T9–10 transthoracic approach. Macroscopically, the lesion was a soft, purple, encapsulated, and nodular mass. It was grossly removed without violation of its capsule. The tumor abutted the neural foramen but did not enter the spinal canal. The intercostal nerve was visualized after resection and judged to be intact. Microscopically, the tumor consisted of a series of vascular channels and sinuses with a connective tissue capsule and reticulin-staining matrix. There was an abundance of red pulp with scattered lymphoid nodules and extensive trilineage hematopoiesis characterized by cell lines that demonstrated complete maturation (Fig. 2). These findings were consistent with the diagnosis of extramedullary hematopoiesis (EMHT).

**Postoperative Course.** The patient’s postoperative course was unremarkable. He was discharged on postoperative Day 6, and surgical pain was controlled well with oral analgesics. He remained at his neurological baseline. There were no postoperative complications. At 1-year follow-up, he reported no pain and had no symptoms, and neurological examination demonstrated intact function; he was not taking any pain medications. Repeat imaging at this time showed no recurrence of the mass (Fig. 3).
Discussion

Beta-thalassemia is the most common cause of EMHT occurring near the spinal cord. The incidence is greatest in the thoracic region.\textsuperscript{7} It is important, therefore, to consider the diagnosis of EMHT when evaluating para-vertebral masses in patients with a history of this hematological disorder.

Beta-thalassemia typically occurs in people of Mediterranean, Asian, or African descent. It is a defect in the synthesis of $\beta$-globin, thought to be an evolutionary adaptation in areas prevalent for malaria. Beta-thalassemia major is a result of homozygous mutations in both $\beta$-globin genes and is much more severe than $\beta$-thalassemia minor, which results from a heterozygous mutation. Anemia develops because of defective erythropoiesis resulting from the insolubility of unbound $\alpha$-globin chains and their subsequent precipitation in red blood cell precursors.\textsuperscript{5}

As a result of the chronic anemia, extramedullary hematopoietic tissue develops. The site at which this occurs is often related to the embryological origin of the tissue: the spleen, liver, and kidney.\textsuperscript{2,3,12} Although rare, extramedullary hematopoietic tissue can also occur near the spinal cord. It is hypothesized that the dura mater may retain some ability to form hematopoietic tissue from primitive nests.\textsuperscript{3,6,12} Other possibilities are that hematopoietic tissue is extruded from weakened trabeculae in the vertebral bone marrow or that it arises from deposits of circulating immature myeloproliferative cells.\textsuperscript{3,3}

Schwannomas are often found in patients with neurofibromatosis Type 1 and Type 2. However, they are most commonly seen sporadically in men and women in the 5th to 7th decades of life.\textsuperscript{13} The most common presenting symptom is local vertebral pain that may be accompanied by radicular pain and paresthesias, since they can arise from dorsal sensory roots.\textsuperscript{1}

While schwannomas can occur extradurally, they occur most commonly intradurally.\textsuperscript{13} In contrast, EMHTs are exclusively extradural, a feature that cannot always be distinguished with imaging. Both schwannomas and EMHTs enhance with contrast.\textsuperscript{8} On T1-weighted images EMHTs appear isointense with the spinal cord, and on T2-weighted images they appear hypointense,\textsuperscript{3} mixed,\textsuperscript{10,12} or hyperintense,\textsuperscript{6} depending on the inciting condition. One distinctive difference that may help distinguish between the 2 types of lesions is radiographic evidence of bony erosions, which commonly accompany schwannomas. Therefore, osseous erosion is suggestive of an extradural schwanna.\textsuperscript{1} In addition, schwannomas typically have a “dumb-bell” shape, though this is not pathognomonic.\textsuperscript{8} Technetium sulfur colloid nuclear scans can be used to confirm the presence of hematopoietic tissue.\textsuperscript{2,3}

Definitive differentiation can be best achieved via fine needle aspiration or biopsy and a comparison of cytomorphicological characteristics. A smear from an EMHT will consist of red marrow elements, especially hyperplastic myeloid tissue.\textsuperscript{10} On the other hand, a preparation of a schwannoma will exhibit its classically compact cellularity with positive staining for S100 protein.

It is important to be able to distinguish EMHT from schwannomas because the treatment options differ. The treatment of choice for spinal schwannomas causing neurological deficit is resection or radiosurgery. Following gross-total resection, these lesions generally do not recur, and the patient usually remains disease free after remov-
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al.9 Histological examination of a schwannoma reveals palisading nuclei forming Verocay bodies and Antoni A and B regions rich in cytoplasm and spindle cells. Immunohistochemical staining of schwannomas is positive for S100 protein.1 On the other hand, treatment of EMHTs begins with medical therapies to improve the chronic anemia, such as hydroxyurea, blood transfusions, and intravenous steroids.3,12 The EMHTs are radiosensitive and respond well to low-dose radiation therapy.2,3,6,11,12 If the lesion is within the spinal canal and causing significant neural compression, surgical resection may be necessary despite the increased risk of hemorrhage. There have been no reported cases of recurrence following resection of EMHT causing spinal cord compression.

Conclusions

Rarely, an EMHT can be found in the paravertebral region and resemble a schwannoma on MR imaging studies. The distinction between schwannomas and EMHTs in this region can be difficult. We suggest that a careful evaluation of the patient’s medical history as well as laboratory studies. If there is a history of β-thalassemia or chronic anemia is discovered, EMHT should be considered in the differential diagnosis. Radiographic evidence of bone erosion is suggestive of a schwannoma. Technetium sulfur colloid nuclear scan may confirm the presence of extramedullary hematopoietic tissue. Medical management, surgical resection, and radiation therapy should be considered treatment options for EMHTs.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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