Spinal Cord Compression Due to Extramedullary Hematopoiesis Associated With Polycythemia Vera

—Case Report—

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Abstract

A 69-year-old woman with a 14-year history of polycythemia vera suffered progressive paraparesis due to epidural involvement of hematopoietic tissue. Magnetic resonance (MR) imaging demonstrated extensive epidural masses. Decompressive surgery and radiotherapy were performed and she made an almost complete clinical recovery. Serial MR imaging showed no regrowth of the other epidural masses.

Extramedullary hematopoiesis occurs in patients with various hematologic disorders involving a chronic increase in the production of red blood cells, and is often associated with thalassemia, but is less common with polycythemia vera. The most frequent sites are the spleen, liver, and kidney. Extramedullary hematopoietic tissue occurring within the spinal canal and causing cord compression is very rare. Total surgical excision is not usually feasible because of the diffuse nature of extramedullary hematopoietic tissue and the possibility of recurrence, but acute neurological deterioration does require emergency surgery. Extramedullary hematopoiesis is radiosensitive and displays a rapid response to low dosages, so radiation therapy is recommended for residual tumors. Considering the possibility of central nervous system extramedullary hematopoiesis in patients with polycythemia vera, an early diagnosis is necessary for a favorable prognosis.

Key words: polycythemia vera, extramedullary hematopoiesis, spinal cord compression, paraparesis

Introduction

Extramedullary hematopoiesis is a well-recognized pathological process occurring in various hematologic diseases. Extramedullary hematopoietic tissue usually develops in sites related to hematopoiesis during fetal development. However, spinal cord compression caused by epidural extramedullary hematopoiesis is a rare complication of polycythemia vera. We treated one patient with polycythemia vera who suffered from progressive paraplegia due to spinal cord compression by hematopoietic tumors.

Case Report

A 69-year-old woman had a 3-month history of progressive weakness in the lower limbs, urinary disturbance, and severe constipation. She was admitted to our hospital in March 2000. Polycythemia vera had been identified in 1986. Neurological examination revealed increased muscle tone with hyperreflexia, bilateral Babinski signs, gait disturbance, and sensory disturbance below the T-5 level. Laboratory data included a white blood cell count of 25,000/µl, a red blood cell count of 7.8 × 10⁶/µl, and a platelet count of 44.8 × 10⁴/µl. Hemoglobin level was 17.6 mg/dl. Magnetic resonance (MR) imaging of the spine revealed multiple extradural masses extending from T-4 through T-9. The spinal cord was compressed severely at T-4 through T-8. Cord compression was not observed at the other levels. The epidural tumors appeared as isointensity on T₁ and mixed intensity on T₂-weighted MR images compared with the spinal cord (Fig. 1A, B). T₁-weighted MR imaging showed heterogeneous enhancement after gadolinium administration (Fig. 1C, D).

Laminectomy was performed from T-4 to T-8 because of progressive neurological deterioration.
Well-circumscribed, flesh-colored tumors were embedded in thickened epidural fat (Fig. 2). The dura mater was intact. Histological examination showed that the epidural masses were composed of hematopoietic tissue, with pronounced megakaryocytic, erythroid, and myeloid cell lines (Fig. 3).

Her neurological condition dramatically improved. Radiotherapy was delivered to the thoracic spine with a total dose of 20 Gy over 14 days to treat the residual tumors. The target for irradiation was based on the MR imaging findings. MR imaging revealed no regrowth of the tumors after irradiation.

**Discussion**

Polycythemia vera is a myeloproliferative disorder of clonal origin appearing at the level of the pluripotential hematopoietic stem cell and resulting in neoplastic proliferation of erythroid, myeloid, and megakaryocytic elements in the bone marrow. Laboratory examinations reveal elevated hemoglobin and hematocrit values as well as thrombocytosis.

Extramedullary hematopoiesis is a complex disease associated with chronic anemic states, frequently accompanied by thalassemia and less commonly by other anemic and myeloproliferative disorders such as myelofibrosis and polycythemia vera. The most common sites of extramedullary hematopoiesis include the spleen, liver, and kidney, but also the adrenals, thymus, mediastinum, and lymph nodes. Spinal epidural hematopoiesis is rare. The source of the epidural hematopoietic tissue in extramedullary hematopoiesis is contentious. The dura mater retains hematopoietic capacity in the fetus and extramedullary hematopoiesis may therefore develop from primitive rests. Most cases of spinal cord compression due to extramedul-
Extradural tumors have been associated with thalassemia and myelofibrosis.\(^\text{1,6,8,16,17,19,21}\) The lesions are frequently localized in the mid or lower thoracic regions.\(^\text{1,4,5,10,21}\)

In the present case, the appearance of the tumors on MR imaging and the known presence of an underlying hematological condition suggested the diagnosis of extramedullary hematopoiesis. The differential diagnosis of an extensive epidural mass includes epidural hematoma, infection (abscess), extramedullary hematopoiesis, and neoplasms such as metastasis. The diffuse form of lymphoma or leu-

kemic mass (granulocytic sarcoma) is the major differential consideration.\(^\text{6,10,20}\)

Histological examination can mistake granulocytic sarcoma for extramedullary hematopoiesis, hemorrhage, and other malignant tumors.\(^\text{18}\) Granulocytic sarcoma is uncommon in polycythemia vera but has been found within a tumor of extramedullary hematopoiesis in polycythemia vera.\(^\text{13}\) Granulocytic sarcoma is ominous in terms of the prognosis because transformation to acute myelocytic leukemia of blastic crisis frequently occurs.\(^\text{7,14}\)

It is critical that the initial diagnosis is correct.

Spinal cord compression caused by extramedullary hematopoiesis has been treated with intravenous steroid administration on an emergency basis, blood transfusion, chemotherapy, radiotherapy, and decompression laminectomy.\(^\text{8,10,12,20,21}\) Blood transfusion may temporarily reduce the size of the hematopoietic tumor.\(^\text{20,21}\) Hydroxyurea acts as a gyrostatic agent to increase production of hemoglobin F, so administration may be beneficial in patients for whom radiotherapy is not possible.\(^\text{12,20}\) Epidural extramedullary hematopoiesis is radiosensitive with a rapid response to low dosages.\(^\text{8,10,20}\) Radiation therapy has been used by itself as the primary therapy.\(^\text{8}\) The dose of radiation ranged from 9 Gy to 35 Gy for different fraction sizes.\(^\text{8,20}\) MR imaging allows the radiation field to be accurately delineated. Total surgical excision is not always possible because of the diffuse nature of the hematopoietic tumor and the possibility of recurrence.\(^\text{8,10}\) Emergency surgery may be necessary in patients suffering acute neurological deterioration.\(^\text{9}\) Extramedullary hematopoiesis should be considered for in the differential diagnosis of patients with hematologic disorders and spinal cord symptoms, but a favorable prognosis depends upon early diagnosis.

References


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