Progressive paraparesis due to thoracic extramedullary hematopoiesis in myelofibrosis

Case report

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A patient with myelofibrosis who developed a progressive paraparesis caused by spinal cord compression due to thoracic extramedullary hematopoietic tissue is reported. He recovered well after local radiotherapy alone.

Key Words • extramedullary hematopoiesis • paraparesis • myelofibrosis

SPINAL cord compression due to extramedullary hematopoiesis is a well-known complication in thalassemia, but extremely rare in myelofibrosis. We report a patient with extramedullary hematopoiesis caused by myelofibrosis, in whom radiotherapy was sufficient to obtain good symptomatic benefit.

Case Report

This 61-year-old man with a diagnosis of myelofibrosis of almost 10 years duration developed progressive paraparesis over a period of 2 months.

Examination. Physical examination showed a pale and cachectic man. Cardiac auscultation revealed a systolic murmur. Lung auscultation was normal. The spleen was enlarged without hepatomegaly. Neurological examination showed normal mental functions. There was no abnormality of the cranial nerves. Strength in the upper limbs was normal, but there was a global hypotonic paresis of the lower extremities. Tendon and lower and upper abdominal reflexes were absent, while plantar responses were extensor. There was loss of sensation below the T-8 level.

Laboratory examination revealed the following abnormal findings: hemoglobin 5.9 gm/dl; red blood cells $2 \times 10^6$/cu mm; hematocrit 17.5%; white blood cells $5.6 \times 10^9$/liter, with 55% polymorphonuclear cells, 26% lymphocytes, and 9% myelocytes, metamyelocytes, erythroblasts, and blast cells. A peripheral blood smear showed anisocytosis and anisochromasia with macrocytes. Liver and renal function tests were normal.

A chest x-ray film revealed only degenerative changes of the dorsal column. Magnetic resonance imaging disclosed a posterior epidural mass extending from C-5 to T-11 with compression of the spinal cord, especially at the midthoracic level (Fig. 1). Transverse images revealed a large right-sided paravertebral mass at the T-8 level extending into the spinal canal through the intervertebral canal (Fig. 2).

Biopsy and Course. A needle biopsy of the thoracic paravertebral mass was performed with computerized tomography guidance. Histological investigation of the specimen revealed the presence of extramedullary hematopoietic tissue with micromegakaryocytes and precursors of the white and red blood cells (Fig. 3). The patient received a transfusion with 6 U of packed cells. Radiotherapy (300 Gy total dose in 10 fractions) was administered from Day 6 to Day 12 with concomitant administration of four doses of 4 mg dexamethasone. The patient's strength gradually recovered completely over the ensuing 2 weeks. He died 4 months later of a pulmonary infection.

Discussion

The most common localizations of extramedullary hematopoiesis are the liver and spleen. The kidneys, mediastinum, pericardium, adrenal glands, and sclera are less frequently involved. Intrathoracic extramedullary hematopoiesis is rather uncommon and most cases have been reported in connection with thalassemia. The hematopoietic tissue may reach the spinal
Fig. 1. Magnetic resonance T1-weighted image, sagittal projection, showing an intraspinal hyperintensive mass extending from C-5 to T-11. The spinal cord is compressed mostly at the T5-7 level.

Fig. 2. Magnetic resonance T1-weighted image, transverse projection at the T-8 level, showing a hyperintensive paravertebral intrathoracic mass, infiltrating the spinal canal through the intervertebral canal.

Fig. 3. Photomicrograph showing a paravertebral thoracic mass with an islet of erythropoiesis, small megakaryocytes, and precursors of the white blood cells. H & E × 125.

that radiotherapy alone can produce complete neurological recovery in 70% of cases.1,6,9,10 Myelofibrosis is a much less common cause of spinal cord compression due to extramedullary hematopoiesis. Decompressive laminectomy followed by radiotherapy or radiotherapy alone have been used to treat extramedullary hematopoiesis in association with myelofibrosis.8

Radiotherapy can produce tissue edema and worsening of the symptoms during the first days of treatment, but this can be prevented by the administration of steroids. It can also produce marked myelosuppression with a drop in the white blood cell count due to a decrease of the extramedullary hematopoietic tissue or irradiation of the remaining normal bone marrow in the spine. This side effect is often the determining factor in the choice for further treatment. Radiotherapy produced no decrease in the number of white blood cells in our patient, which suggests that the intraspinal mass of hematopoietic tissue was ineffectively producing blood cells. The course of this patient confirms the opinion expressed by others that in cases of extramedullary hematopoiesis caused by myelofibrosis a good outcome can be obtained by radiotherapy alone.

References

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