CASE REPORT

Myelofibrosis presenting as spinal cord compression

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This report describes a case of myelofibrosis presenting as spinal cord compression on account of extramedullary haemopoietic tissue encroaching upon the spinal cord from a large pelvic mass.

Extra medullary haemopoiesis is a recognised accompaniment of many haematological diseases. The sites most commonly affected are liver, spleen, and lymph nodes but other sites have been described including the kidney, adrenal glands, skin, lung, pleura, ovary, gastrointestinal tract, thyroid, and the dura mater. Spinal cord compression as a result of extramedullary haemopoiesis has been reported, but extension of this abnormal tissue into the spinal canal from the retroperitoneum is rare.

CASE REPORT

A 75 year old Indian man presented with a two month history of weight loss, bilateral leg weakness, and paraesthesia below the right knee. There was no bowel or bladder dysfunction. Previously he had been diagnosed with ischaemic heart disease but this was currently asymptomatic. On examination he was afebrile and in sinus rhythm. He was noted to be cachectic but there was no lymphadenopathy. The cardiovascular and respiratory systems were normal, but abdominal examination revealed hepatosplenomegaly and a large, firm mass arising from the pelvis. Examination of the cranial nerves and upper limbs was entirely normal. There was considerable muscle wasting of the right quadriceps and power was 2/5 in all muscle groups of the leg. The right knee reflex was absent; other tendon reflexes were normal. Power was 4/5 in all muscle groups of the left leg but examination of that limb was otherwise normal. Both plantars were flexor and there was no objective sensory deficit.

Initial investigations revealed: haemoglobin, 10.7 g/litre; white blood cell count, 14.5 × 10^9/litre (90% neutrophils); and platelets, 354 × 10^9/litre. The erythrocyte sedimentation rate and lactate dehydrogenase value were marginally raised but blood tests were otherwise unremarkable. Serum ferritin, vitamin B12, red cell folate, and haemoglobin electrophoresis were normal. The blood film showed polychromasia and nucleated red blood cells, pronounced anisopoikilocytosis with occasional teardrop cells, dysplastic neutrophils with occasional pelger forms, and 5% basophilia.

Attempted bone marrow aspiration resulted in a dry tap. A trephine biopsy showed grade 4 reticulin fibrosis with foci of haemopoietic tissue showing trilineage hyperplasia, consistent with a myeloproliferative or myelofibrotic disorder. Bone marrow cytogenetic studies could not be performed but reverse transcription polymerase chain reaction analysis of peripheral blood for the BCR–ABL fusion transcript was negative.

An abdominal ultrasound scan and computed tomography confirmed mild hepatomegaly of 13 cm and 15 cm splenomegaly. An 8.5 cm mass arising from the pelvis was also shown. A chest radiograph showed extensive lobulated paraspinal soft tissue extending from the thoracic inlet to the diaphragm, which was also seen on computed tomography. In addition, a T2 weighted magnetic resonance imaging scan demonstrated confluent space occupying disease throughout the lumbar spinal canal. This tissue extended through exit foramina at all levels and was contiguous with retroperitoneal disease, which infiltrated the psoas and paraspinal muscles and surrounded the aorta and inferior vena cava (fig 1).

An open biopsy of the abdominal mass was performed. Histological examination showed a classic sclerosing haemopoietic appearance, with readily recognisable trilineage...
Myelofibrosis presenting as spinal cord compression resulting from extramedullary haemopoietic tissue is very unusual.

Therapeutic options for this condition include surgical decompression, radiotherapy, and myelosuppression with agents such as hydroxyurea. In our patient, local treatment was not possible because of the extent of the disease. Hydroxyurea did result in an extremely rapid improvement. This result was achieved with the side effect of severe pancytopenia at follow up, which required blood and platelet transfusions and temporary cessation of treatment. This demonstrates the balance that needs to be achieved between the side effects and the therapeutic effect.

In view of the predominantly sclerotic background with atypical megakaryocytes, the mass could be mistaken for a sarcoma, carcinoma, or other neoplasm because these can be morphologically similar. It is the identification of maturing haemopoietic precursors of all three myeloid cell lines that enables the pathologist to distinguish between sclerosing extramedullary haemopoiesis and other tumours.

Our case, with the confusing clinical picture of a lower abdominal mass and abnormal lower limb neurology, represents a very unusual presentation of myelofibrosis, and demonstrates how prompt diagnosis and treatment can result in rapid neurological recovery.

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REFERENCES


