

## CASE REPORT

## Myelofibrosis presenting as spinal cord compression

E Horwood, H Dowson, R Gupta, R Kaczmariski, M Williamson

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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.

Extramedullary 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,<sup>1</sup> adrenals, skin, lung, pleura,<sup>2</sup> ovary, gastrointestinal tract, thyroid,<sup>3</sup> 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 parasthesia 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,  $14.5 \times 10^9$ /litre (90% neutrophils); and platelets,  $354 \times 10^9$ /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.

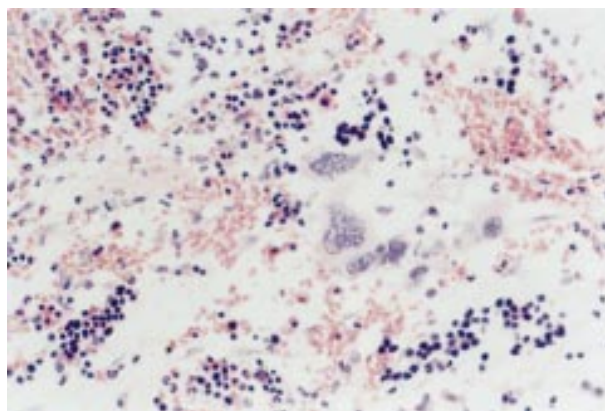
An abdominal ultrasound scan and computed tomography confirmed mild hepatomegaly of 13 cm and 15 cm splenomegaly. An 8.5 cm mass arising from the pelvis was also shown. A chest radiograph showed extensive lobulated paraspinal soft tissue extending from the thoracic inlet to the diaphragm,



**Figure 1** Sagittal magnetic resonance imaging of spine. Extensive space occupying disease within the spinal canal involving almost the entire length of the lumbar canal, and the dural sac is pinched and narrowed. There is also extensive retroperitoneal disease infiltrating the psoas and disease in the pelvis with posterior mediastinal and retrocrural adenopathy. There is an absence of marrow fat throughout the scanned spine.

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



**Figure 2** Open biopsy of retroperitoneal mass. Sclerotic areas are present with elongated megakaryocytes in a myxoid and hyalinised background. Most of the tumour has a classic sclerosing haemopoietic appearance, with readily recognisable trilineage haemopoiesis and a little collagen deposition only in the fat.

haemopoiesis in a background of fatty replacement by collagen (fig 2). A histological diagnosis of myelofibrosis with extensive sclerosing extramedullary haemopoiesis was made.

He was started on hydroxyurea 2 g/day. At follow up three weeks later there was a pronounced improvement in his neurological symptoms and signs and the previously large palpable abdominal mass and organomegaly was much reduced in size. He developed pancytopenia and the hydroxyurea was stopped, but was subsequently restarted when this resolved.

Ferokinetic studies showed considerable medullary activity in the vertebrae and pronounced activity over the liver, but little activity in the spleen.

He remained well for nine months after diagnosis, but died suddenly after a presumed myocardial infarction. We were unable to perform a necropsy.

## DISCUSSION

Extramedullary haemopoiesis is seen in many haematological disorders associated with bone marrow dysfunction. Spinal cord compression as a result of extramedullary haemopoiesis has been reported on several occasions,<sup>4-7</sup> more commonly secondary to thalassaemia than to myelofibrosis. The cord compression in our case was in the lumbar spinal canal accounting for the lower motor neurone pattern of weakness. This probably also accounts for the good recovery seen once treatment was started with hydroxyurea.

Previously reported unusual sites of extramedullary haemopoiesis and spinal cord compression in myelofibrosis have usually occurred between two and 20 years after diagnosis.<sup>5-7</sup> Myelofibrosis presenting as spinal cord compression resulting from extramedullary haemopoietic tissue is very unusual.

A striking feature of our case was the extent of the retroperitoneal mass contiguous with the tissue surrounding the spinal cord. This is not a feature of the other reported cases of myelofibrosis with myelopathy secondary to extramedullary haemopoiesis. Our patient had extramedullary haemopoietic tissue extending the whole length of the spine, from the thoracic to the presacral region. This soft tissue was seen to infiltrate through the lateral spine recesses and was in continuity with the large retroperitoneal mass surrounding the aorta and inferior vena cava, which was easily palpable on examination.

Another atypical feature was the relatively minor degree of splenomegaly as demonstrated by clinical examination and radiological investigations. In a case of myelofibrosis with such extensive extramedullary haemopoiesis a far greater

## Take home messages

- We describe a very unusual case of myelofibrosis, which presented with a lower abdominal mass and abnormal lower limb neurology
- It was also unusual in that the extramedullary haemopoietic tissue extended the whole length of the spine, from the thoracic to the presacral region, and passed through the lateral spine recesses to the large retroperitoneal mass surrounding the aorta and inferior vena cava
- Despite this extensive extramedullary haemopoiesis only a minor degree of splenomegaly was seen
- Hydroxyurea produced an extremely rapid improvement, although severe pancytopenia required blood and platelet transfusions and temporary cessation of treatment, demonstrating the balance that needs to be achieved between the side effects and the therapeutic effect

degree of splenomegaly would normally be expected. Abdominal ultrasound and computed tomography scan demonstrated the moderate enlargement and this was reflected by the surprisingly small amount of medullary activity seen in the spleen in ferrokinetic studies.

*“Myelofibrosis presenting as spinal cord compression resulting from extramedullary haemopoietic tissue is very unusual”*

Therapeutic options for this condition include surgical decompression, radiotherapy,<sup>3-7</sup> and myelosuppression with agents such as hydroxyurea.<sup>8</sup> 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.<sup>9</sup>

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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