Spinal cord compression due to extramedullary haemopoiesis in myelofibrosis

G Cook, R A Sharp

Abstract
Extramedullary haemopoiesis resulting in spinal cord compression is rare. This report of extramedullary myeloid metaplasia in a patient with myelofibrosis serves to illustrate the value of magnetic resonance imaging (MRI) in the diagnosis and management of good neurological recovery.


Extramedullary haemopoiesis (EMH) or myeloid metaplasia is associated with many chronic haematological conditions, including thalassaemia, leukaemia, lymphoma, and myelofibrosis.1 It most commonly affects the liver, spleen, kidneys, and adrenal glands. Involvement of the dura mater of the spinal cord, resulting in spinal cord compression, has occasionally been described. We report a case of spinal cord compression due to EMH in association with myelofibrosis.

Discussion
Extramedullary haemopoiesis is a well described complication of many chronic haematological conditions. This haemopoietic tissue is rarely seen in sites other than those that normally occur in the foetus (table). Rarely, EMH has been found localised in the dura mater of the spinal cord, and can result in spinal cord compression. This was first reported in 1956 in a patient with myelofibrosis.2 Since then there have been more than 50 such reported cases, most of which occurred in association with thalassaemia.3 The dura mater has probably been involved more than this implies as this region is rarely, if ever, examined at necropsy. The exact origin of EMH in the dura mater is unknown, but as indicated the dura mater has haemopoietic capacity in the foetus and EMH may develop from primitive rests. An alternative explanation is the embolisation of haemopoietic stem cells to the dura mater.4 It is unlikely, however, that EMH arises from extrusion of vertebral bone marrow in the absence of bony erosions or fractures.

In spinal cord compression secondary to EMH, the lesions are commonly localised to the mid-lower thoracic region. This may be because of the narrow diameter of the

Figure 1 Balanced sagittal MRI: image illustrates extramedullary mass (A) at maximal depth T6-T7 level, displacing the spinal cord (C) anteriorly.
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spinal cord in this region. In earlier reports, diagnosis was made by myelography followed by decompression laminectomy and histological analysis of material gained. This served as both a diagnostic and therapeutic procedure. Although surgery is effective in most cases, haemopoietic tissue is radiosensitive and radiotherapy was introduced as an adjunct to surgery either because of incomplete excision or persisting neurological deficit. This dual approach was documented in several of the earlier cases, with satisfactory responses in most patients.

More recently this combined approach has been challenged with the advent of more complex radiological imaging techniques and by the use of radiotherapy alone. One group, the first to report the use of MRI in the diagnosis of spinal cord compression secondary to EMH, reported five patients with thalassaemia who responded to radiotherapy alone. This approach has the advantage of avoiding a major surgical procedure in patients whose risk of haemorrhage is compounded by thrombocytopenia, as in our patient, particularly when the mass of EMH is diffuse and complete resection is impossible.

The dose of radiation used has varied from 900 cGy to 3500 cGy with different fraction sizes. With the availability of MRI the radiation ports can be delineated more accurately to limit the extent of spinal cord at risk and thus low dose radiotherapy can be used to achieve a satisfactory clinical response. Recovery was prompt in most cases reported, beginning within a few days.

This case shows how MRI imaging can facilitate the diagnosis and assessment of the extent of extramedullary myeloid metaplasia in a patient with primary myelofibrosis leading to spinal cord compression.

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