CASE REPORT

Extramedullary haematopoiesis causing spinal cord compression

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Background

Extramedullary haematopoiesis (EMH) is a rare cause of spinal cord compression. When a patient with a haematological disorder that causes chronic anaemia (particularly thalassaemia) presents with neurological deficits referable to the spine, EMH with paraspinal masses should be considered and imaging planned appropriately.

Case history

We present a case of thalassaemia intermedia with marked paraspinal EMH and spinal cord compression.

Clinical presentation

A 38-year-old man with thalassaemia intermedia presented with a 6-week history of progressive muscle weakness, back pain, paraesthesia and spasm in both legs. He did not have any bladder dysfunction. The patient had a cholecystectomy at age 34. One of his brothers had died from an unknown cause at age 7 years. However, he had also had thalassaemia. His other brother died from severe cardiac failure brought on by chronic anaemia at age 40 years.

On examination, the patient had normal vital signs and normal cardiac and respiratory examination. Hepatosplenomegaly was present. Neurological examination revealed muscle power of 3/5 in the lower limbs in all the muscle groups, brisk tendon reflexes and bilateral positive Babinski reflexes. The sensory level was at T3.

Biochemical evaluation showed microcytic hypochromic anaemia. Haemoglobin was 9.5 g/decilitre (normal=13.8 - 18.8 g/decilitre), mean cell volume was 73.2 fl (normal=79 - 100 fl), and mean cell haemoglobin was 25.5 picograms (normal=27 - 35 pg). He also had an elevated total bilirubin of 62 µmol/l (normal=2 - 26 µmol/l) and elevated conjugated bilirubin of 11 µmol/l (normal=1 - 7 µmol/l). The serum urate was also elevated at 0.45 mmol/l (normal=0.21 - 0.43 mmol/l). These changes may be secondary to haemolysis.

Imaging findings

A chest radiograph (Fig. 1) demonstrates lobulated soft-tissue posterior mediastinal masses, medullary expansion of the ribs and clavicles with thinning of the cortices. A biopsy of one of the masses showed hypercellular marrow with erythroid hyperplasia, moderate dyserythropoiesis, megaloblastic changes and overloaded iron stores which were in keeping with EMH.

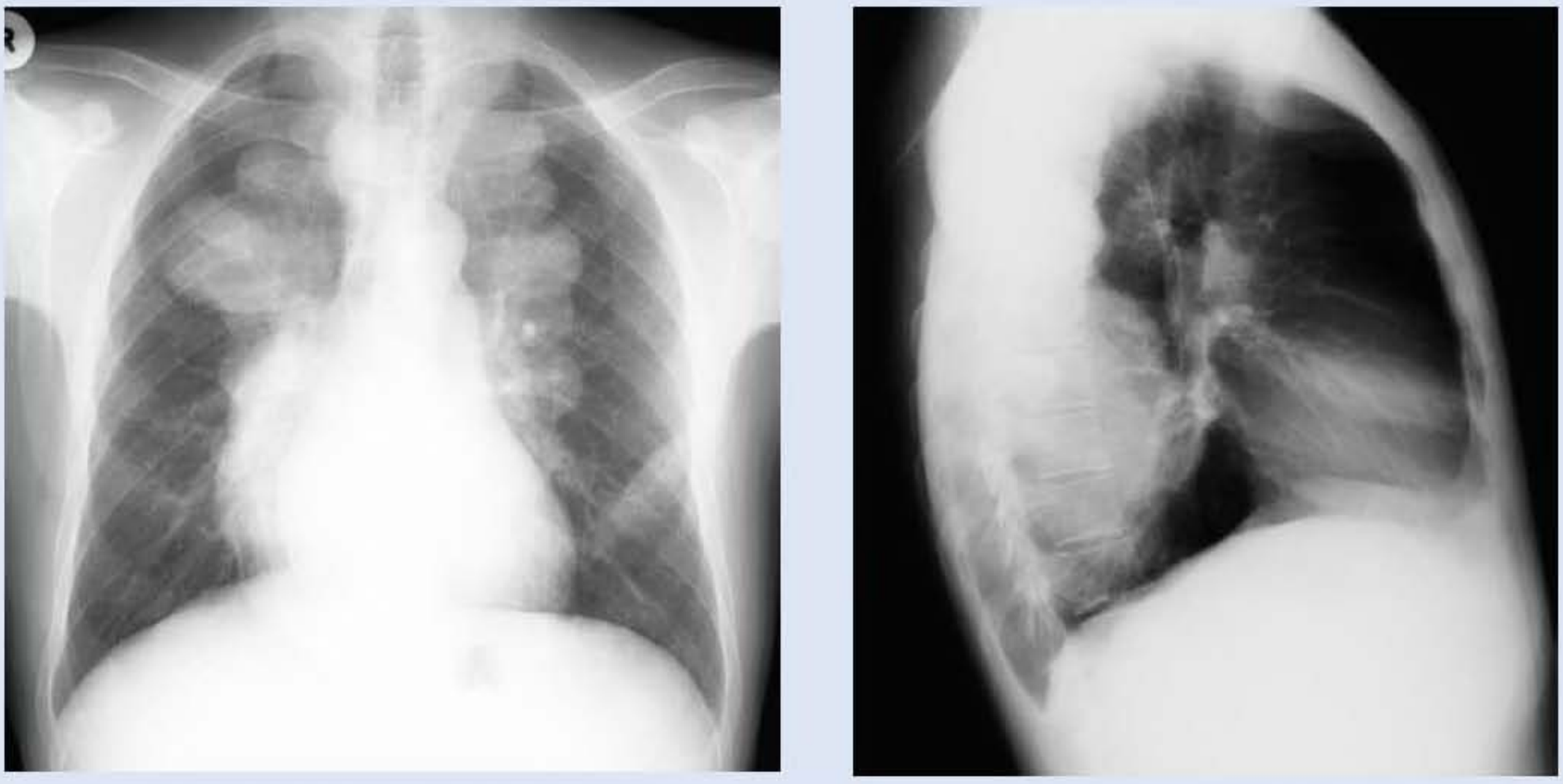

Fig. 1. Frontal and lateral radiographs of the chest show lobulated soft-tissue posterior mediastinal masses and medullary expansion of the ribs and clavicles with thinning of the cortices.

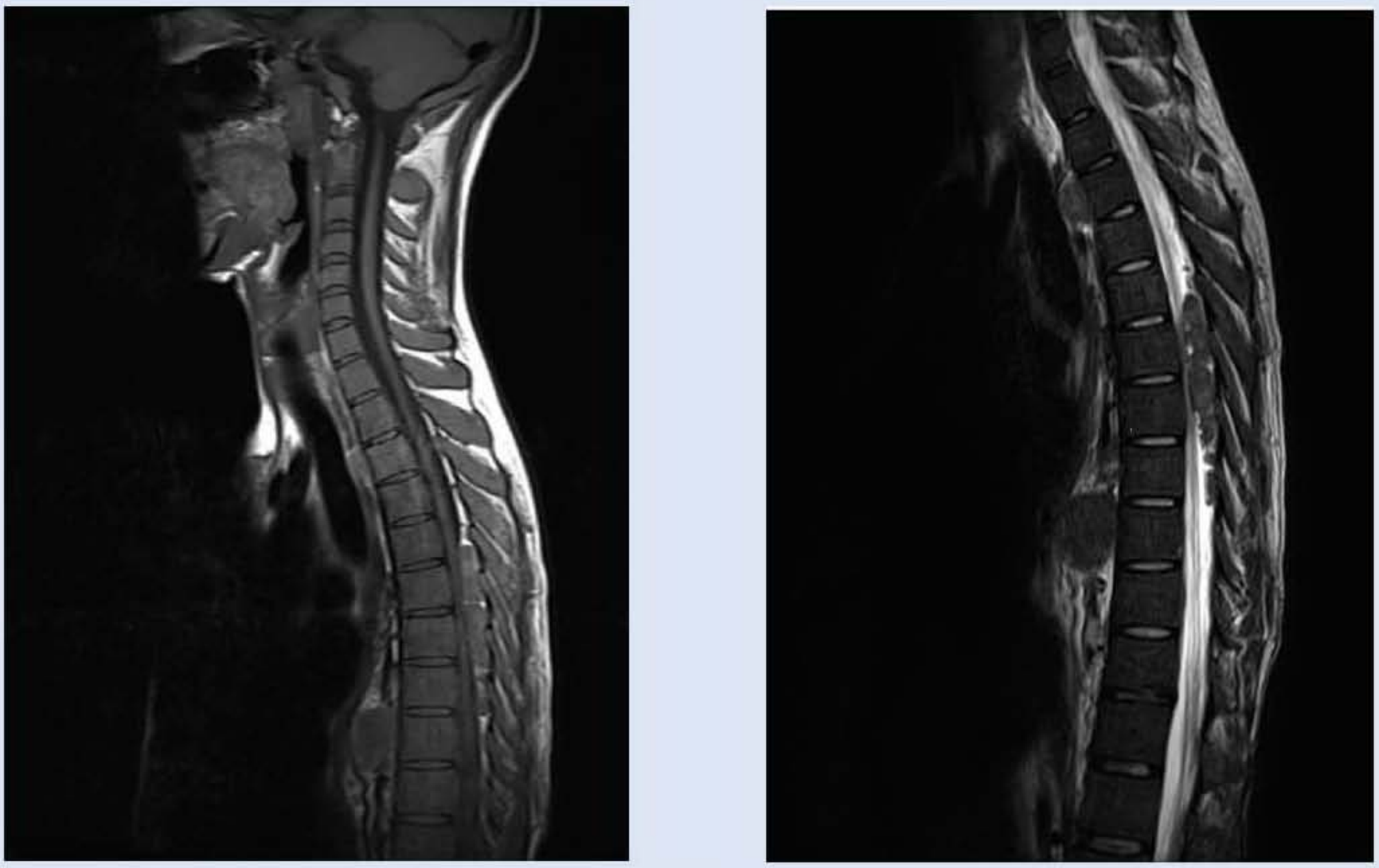

Fig. 2. Sagittal T1- and T2-weighted sequences demonstrate extradural lobulated soft-tissue masses which cause compression of the spinal cord at levels T5 - T8. These masses are iso-intense to the marrow within the adjacent vertebral bodies. There is enlargement of the central canal distal to the region of compression. There is also a mass anterior to the T9 vertebral body.

The patient also underwent magnetic resonance imaging (MRI) which showed extradural lobulated soft-tissue masses causing compression of the spinal cord at vertebral body levels T5 to T8 (Fig. 2). The mass lesions are iso-intense to the marrow within the adjacent vertebral bodies on T1- and T2-weighted sequences. The masses extend from the vertebral bodies laterally and anteriorly into the posterior mediastinum (Figs 3 and 4).

The patient underwent surgical decompression of his thoracic spine (posterior laminectomies) and multiple blood transfusions.

Discussion

Thalassaemia (the Greek etymon literally means 'anaemia of the sea') occurs in a broad geographical band from the Mediterranean ('Mediterranean anaemia') through Asia. The cause is an inherited defect in the synthesis of one of the alpha or beta globin chains. Homozygous (major) or heterozygous (minor) forms of the condition exist.[1]

Defective globin chains lead to excessive haemolysis and compensatory haematopoiesis within bone marrow, which in turn leads to enlargement of the medullary cavities and thinning of cortices. Extrusion of bone marrow through a defect of a severely thinned cortex may be a mechanism in the development of EMH.[2] Another possible theory for the development of EMH is transformation of haematopoietic precursors of mesodermal origin, located in the thoracic epidural space, into marrow.

The liver, spleen, kidney, posterior mediastinum and lymph nodes are the most common sites of extramedullary haematopoiesis.[3] It may also occur in uncommon locations such as the skin, central nervous system, adrenal glands, middle ear and paratracheal region.[3]

In the spine, EMH is commonly located in the mid- or lower thoracic regions; the differential diagnosis for these lesions includes metastases, abscesses, haematomas and lymphoma.[3]

EMH is radiosensitive, responding to low doses of radiotherapy, and therefore a favourable prognosis depends on early diagnosis.[3] Other treatment options include blood transfusions and surgery. Our patient underwent both treatment options.

On plain chest radiography, thoracic extramedullary haematopoiesis lesions appear as smooth or lobulated posterior mediastinal masses. These paraspinal masses can be unilateral or bilateral, isolated or extensive.[2] They have a predilection for the lower thoracic spine region, and rarely appear at anterior rib ends. Calcification and vertebral anomalies are rare, therefore other causes of posterior mediastinal masses, such as neurenteric cyst and neuroblastoma, can be excluded. In patients with chronic anaemia not requiring hyper-transfusion, the typical bony thorax appearance demonstrates osteopenia, medullary expansion, coarse trabecular pattern and cortical thinning. Rib changes include bulbous widening of the posterior ribs, localised lucencies, cortical erosions and the 'rib within rib' picture.[4]

On non-contrast computed tomography (CT) scanning, extramedullary haematopoiesis lesions appear as soft tissue masses that are denser than fat. If these lesions are adjacent to the posterior

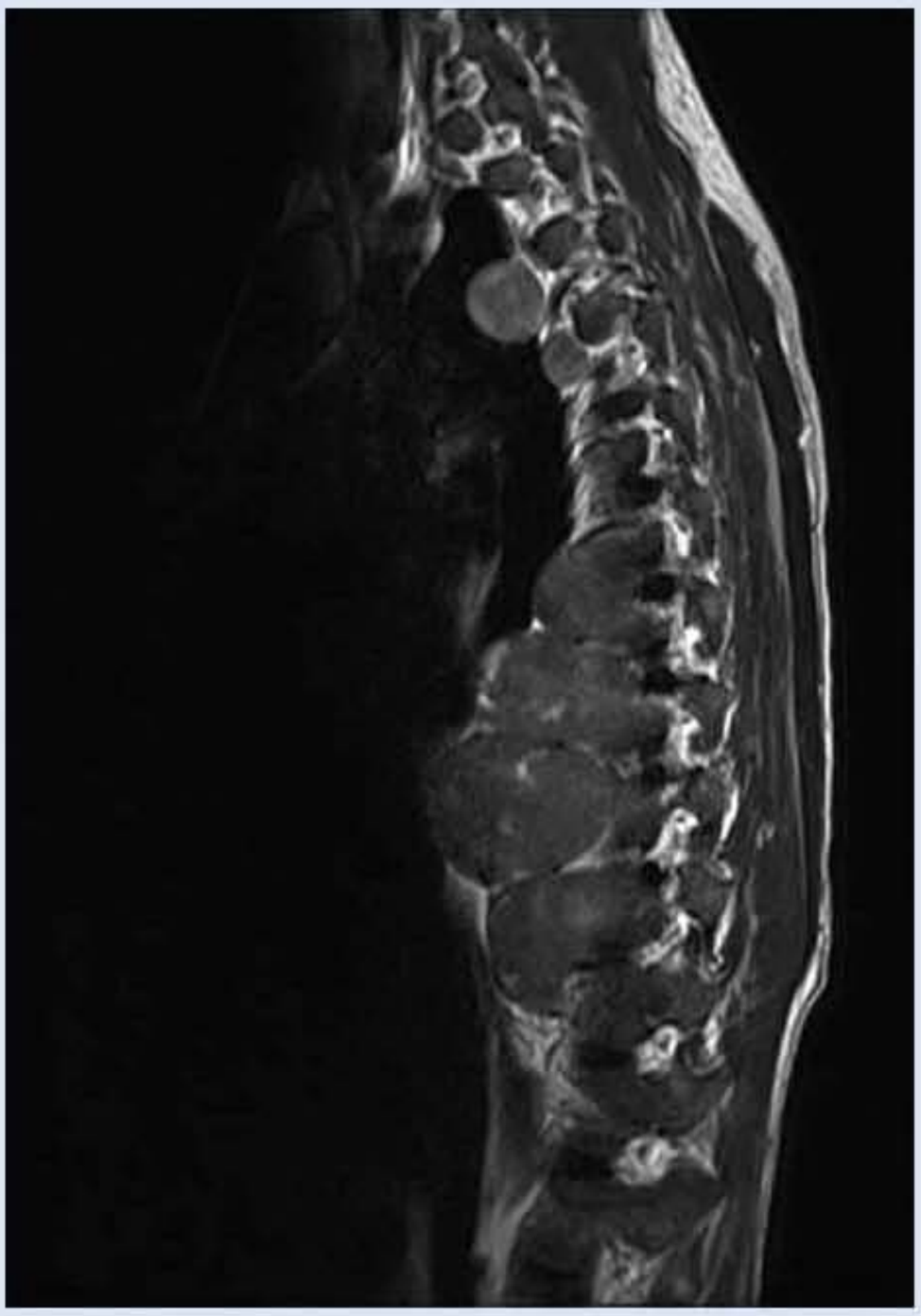

Fig. 3. Para-sagittal T1-weighted sequence demonstrates multiple lobulated masses extending from the thoracic vertebrae which are iso-intense to the vertebral bodies. The masses extend laterally and anteriorly into the posterior mediastinum.

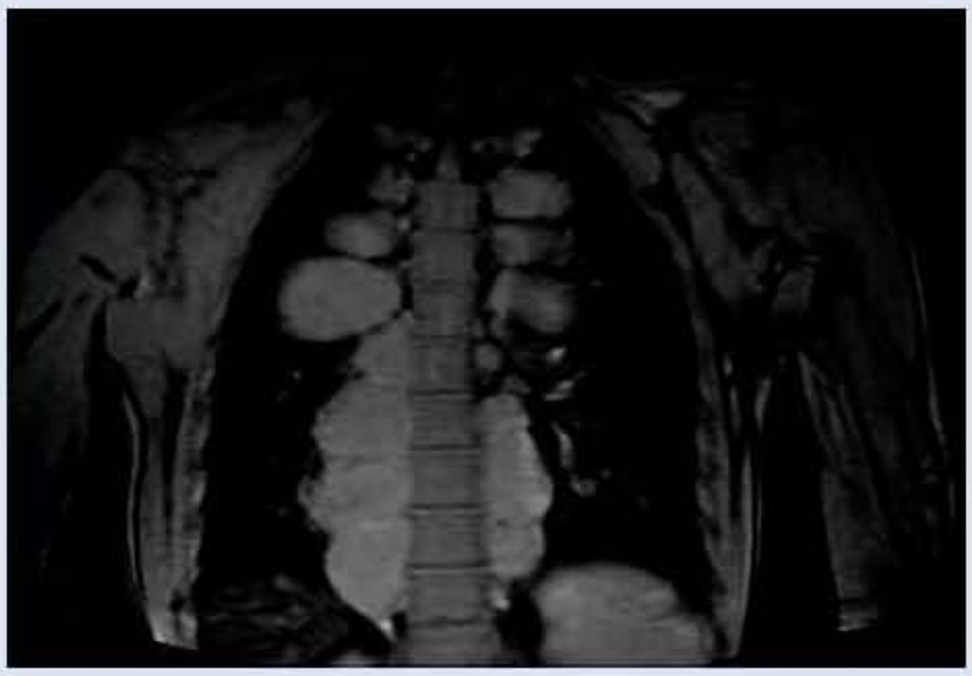

Fig. 4. Coronal T3-weighted sequence shows lobulated masses extending laterally into the posterior mediastinum, with similar signal to the adjacent vertebral bodies.

ribs, the underlying rib will be widened with a thin cortex.[4] On CT scan with contrast, extramedullary haematopoiesis lesions enhance inhomogeneously owing to the iron deposition and fat infiltration. As lymphadenopathy and neurogenic tumours enhance homogeneously and neurogenic tumours may have calcifications, these masses are differentiated from EMH masses.[2]

MRI is the most effective imaging modality for demonstrating extramedullary haematopoiesis.[3] The lesions are iso-intense with the bone marrow and may show different degrees of hyperintensity on T1- and T2-weighted images.[3] However, mixture of blood products of different age within the extramedullary haematopoiesis foci results in increased or decreased T1 or T2 signal. For example, ferrous or ferric iron shortens T1 and T2 signal, methaemoglobin causes an increased T1 signal and variable T2 signal whereas haemosiderin decreases T1 and T2 signal.[5] Some masses show a peripheral hyperintense rim which is attributed to fat. This distinguishes extramedullary haematopoiesis lesions from other lesions such as metastasis. In patients with a known primary malignancy, it may be difficult to differentiate EMH from an epidural metastasis.[3]

With positron emission tomography – computed tomography (PET-CT), extramedullary haematopoiesis lesions demonstrate mild to moderate metabolic activity when compared with malignant lesions which demonstrate high metabolic activity.[5]

Conclusion

EMH is a rare cause of posterior mediastinal masses[6] but in patients with thalassaemia should be considered as part of the differential diagnosis. In patients with thalassaemia who present with spinal cord symptoms, MRI is the study of choice for evaluation of the spine.

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