CASE REPORT

Spinal epidural extramedullary haematopoiesis in β -thalassaemia intermedia

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SUMMARY

A 22-year-old man known to have β -thalassaemia intermedia since childhood presented with bilateral lower limb weakness after spinal anaesthesia for an elective minor operation of his left leg. MRI and CT scans were performed to rule out acute epidural haematoma; coincidental imaging features of marrow hyperplasia and spinal epidural extramedullary haematopoiesis were found. This article will present and discuss the imaging features, differential diagnosis, management and literature review of the rare occurrence of extramedullary haematopoiesis in the spinal epidural space.

BACKGROUND

The occurrence of extramedullary haematopoiesis (EMH) in the sacral epidural space in patients with β -thalassaemia intermedia is rarely reported. Knowledge of this disease entity and its imaging features enables correct clinical management of patients, thereby alleviating unnecessary biopsies or even spinal operations that have their own inherent risk of mortality and morbidity.

CASE PRESENTATION

A 22-year-old male patient known to have β -thalassaemia intermedia since childhood presented with bilateral lower limb weakness after spinal anaesthesia for elective debridement of a traumatic ulcer of his left leg. Relevant medical history included microcytic hypochromic anaemia that was not transfusion dependent, secondary haemochromatosis of the liver and silent lacunar infarcts in the brain. At around 4 h postoperatively, the patient had persistent bilateral lower limb weakness and subjective numbness at left L4 to S1 dermatome and the perianal region. On physical examination, lower limb power was grade 2/5. Anal tone was, however, preserved. Bladder ultrasound volume measurement showed residual urine of 456 mL. With the clinical suspicion of acute epidural haematoma complicating spinal anaesthesia causing cauda equina compression, urgent MRI and CT were performed.

INVESTIGATIONS

Apart from microcytic hypochromic anaemia, pre-operative blood tests were all normal. The patient had no bleeding tendency with normal platelet counts and clotting profiles.

Cross-sectional imaging with MRI and CT scan did not show any evidence of acute epidural haematoma. Instead, there were presence of multiple well-defined epidural masses demonstrating intermediate signal on T1-weighted (T1W) and

short tau inversion recovery (STIR) images, intermediate to low signal on T2-weighted (T2W) images, with mild-to-moderate degree of homogeneous enhancement after gadolinium contrast administration at S1 and S2 levels. These masses caused with mild extrinsic compression of the thecal sac and sacral nerve roots (figures 1 and 2). No erosive changes were seen at the vertebral endplates. The intervening intervertebral discs retained their hydration signal and height. Multiple epidural and paraspinal soft tissue masses of same signal intensity and enhancement were also seen along the thoracic spine (figure 3). With longstanding β -thalassaemia intermedia, the patient also had typical imaging features of marrow hyperplasia along the spinal column and rib cage with generalised osteopenia, trabecular coarsening and bony expansions at bilateral posterior ribs (figure 4).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of epidural masses includes metastasis, haematoma, sequestered disc material, abscess and spinal lymphoma.¹ The patient had no known history of primary malignancy. Epidural haematoma and sequestered disc materials do not enhance homogeneously with contrast. Epidural abscess seldom exists without adjacent vertebral endplate erosive changes or T2-hyperintense diskal intensity changes. Epidural lymphoma has more intense gadolinium enhancement and there may be involvement of the adjacent vertebrae. The diagnosis of EMH can therefore be made through the constellation of characteristic MRI findings, appropriate clinical history and associated radiological signs that highlight abnormal haematopoiesis in other parts of the body.¹

OUTCOME AND FOLLOW-UP

The patient's symptoms resolved spontaneously with a resumption of lower limb power the subsequent day. A follow-up MRI performed 6 months later showed no disease progression. No biopsy was performed. His lower limb power and sensation were normal in all clinical follow-up by the orthopaedic surgeons in the subsequent 12 months.

DISCUSSION

EMH was reported in patients with chronic haemolytic anaemias, such as thalassaemia and sickle cell disease², and myeloproliferative disorders, such as polycythaemia vera, myelofibrosis and myelodysplastic syndrome.³ Association with tumours, such as hepatocellular carcinoma, was also observed.³ The incidence of EMH in patients with thalassaemia

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Figure 1 Sagittal T1-weighted (T1W; A), postgadolinium T1W fat-suppressed (FS; B), T2-weighted (T2W; C) and short tau inversion recovery (STIR; D) MRI of lumbosacral spine showed two oval nodules at anterior epidural space at S1 and S2 levels. They show T1W-intermediate, T2W-low to intermediate and STIR-intermediate signal. Mild-to-moderate homogenous enhancement is noted after gadolinium administration.

Figure 2 Axial T1-weighted (T1W; A), postgadolinium T1W (B) and T2-weighted (T2W; C) MRI showed a bilobed mass at anterior epidural space at S1 level. They showed intermediate signal on T1W and low to intermediate signal on T2W images with mild-to-moderate degree of homogenous gadolinium enhancement.

Figure 3 (A) Sagittal T2-weighted (T2W) MRI showed masses at posterior epidural space from T2 to T8 levels with low-to-intermediate T2 signal. (B) Coronal T2W MRI showed masses at the bilateral paraspinal region at T4–T10 levels showing intermediate T2 signal.

Figure 4 (A) Axial CT scan at T4 level with soft tissue window showed soft tissue masses at the left paraspinal region and posterior epidural space. (B) Axial CT scan at T6 level with bone window showed osteopenia with trabeculated pattern. Focal expansion of posterior rib is seen. (C) Axial CT scan at S1 level with soft tissue window showed soft tissue mass at the anterior epidural space.

intermedia was reported up to 20%, in comparison with transfusion-dependent thalassaemia major patients where the incidence is less than 1%.⁴ The peak prevalence occurs in the fourth decade of life, with a male to female ratio of up to 5 : 1.¹

Epidural EMH tissue is usually situated posteriorly in the thoracic region. This is thought to be due to direct extension of hyperplastic marrow through ribs or adjacent vertebral bone marrow, or due to haematopoietic rest cells which are most prevalent in the thoracic canal.^{1 3 5} Sacral-based and anterior-based EMH such as the present case is rarely reported.¹ Sacral EMH has been found in patients with underlying thalassaemia,⁶ sickle cell disease¹ and in patients with no identifiable haematological disorder.⁷

If severe, sacral and cervicothoracic EMH can have neurological complications including cauda equina syndrome and spinal cord compression, respectively.^{1 7–10}

MRI is the method of choice for diagnosis of spinal EMH.² Spinal EMH are well-defined, epidural soft tissue lesions that may be single or multiple, discrete or elongated extending along a length of vertebral column.¹ Multifocal and bilateral involvement in paraspinal and epidural regions are characteristic features of EMH.⁴

T1W and T2W signal intensities of EMH are highly variable² and non-specific.¹ It may appear heterogeneous or homogenous,³ intermediate to high signal intensity on T1W and T2W images.^{1 3–5 8 10} One article reported the T1W signal intensity as hypointense.⁹ Appearance on STIR is described as moderately hyperintense.¹ Gadolinium enhancement is also variable, from absent,^{1 4} minimal,⁴ slight⁹ and moderate to strong.¹ The variable signal characteristic is related to the activity and vascularity of the lesion. Active lesions show intermediate signal intensity in T1W and T2W imaging, while older inactive lesions may show high signal or low signal due to fatty infiltration or iron deposition.⁴

Previous reports suggested that absent or minimal enhancement of some EMH can be used to differentiate them from other epidural masses such as abscess or metastasis.^{4 9} Some spinal EMH have a peripheral hyperintense fat rim which may help distinguish EMH from other lesions such as spinal metastasis.^{2 8}

CT can be performed for a patient in whom MRI is contraindicated or unavailable.⁹ Paraspinal EMH is shown, in CT scan, as soft tissue mass with well-defined margin and homogenous density. It displays homogenous enhancement after intravenous contrast administration.¹¹ Tissue diagnosis is not advocated due to risk of catastrophic haemorrhage.^{9 10} Tc99m bone scan or Tc99m-labelled sulfur colloid has been used to diagnose

paraspinal EMH, but diagnosis within epidural space may be difficult due to its limited spatial resolution and proximity to bone marrow.^{1 9}

Management options of spinal EMH include blood transfusion, radiotherapy, surgical decompression and oral hydroxyurea.^{2 4 12} As there is no formal study to compare their effectiveness, treatment would be multidisciplinary and tailor-made to the patient.¹⁰

Learning points

- Extramedullary haematopoiesis represents abnormal compensatory haematopoietic tissue that usually occurs in the liver, spleen and paraspinal regions. It can occur in the spinal epidural space presenting as acute spinal cord compression.
- In patients with a longstanding history of β -thalassaemia intermedia and imaging features of marrow hyperplasia, extramedullary haematopoiesis present in the spinal epidural space has distinguishing imaging features.
- Knowledge of this disease entity can alleviate patients from unnecessary biopsy or surgery.

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