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PARASPINAL EXTRAMEDULLARY HAEMATOPOIESIS IN A PATIENT WITH SICKLE CELL DISEASE: A RARE CLINICAL PRESENTATION

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ABSTRACT

Background: Extramedullary haematopoiesis refers to the formation of blood cells outside the bone marrow, typically as a compensatory response to chronic anaemia or bone marrow failure. It is a well-documented phenomenon in hemoglobinopathies with ineffective erythropoiesis, such as thalassemia major and intermedia, where it occurs frequently due to persistent marrow stress. However, in sickle cell disease, extramedullary haematopoiesis is considered a rare phenomenon. This rarity is attributed to the fact that, in sickle cell disease, anaemia is primarily due to haemolysis rather than ineffective erythropoiesis. When extramedullary haematopoiesis does occur in sickle cell disease, it often involves unusual sites such as the paraspinal regions, potentially causing compressive symptoms. Recognition of this rare complication is crucial for timely diagnosis and management.

Case presentation: We report a case of a 43-year-old male with homozygous sickle cell disease who presented with progressive mid back pain and bilateral lower limb weakness. Magnetic resonance imaging of the thoracic spine revealed a well-circumscribed paraspinal mass causing spinal cord compression.

Discussion: Paraspinal extramedullary haematopoiesis should be considered in the differential diagnosis of spinal masses in patients with chronic haemolytic anaemias. While often asymptomatic, mass effect can lead to neurological deficits requiring urgent intervention. Diagnosis is based on imaging and requires histological confirmation. Treatment options include blood transfusion to suppress erythropoiesis, radiotherapy, and surgical decompression in severe cases.

Conclusion: This case underscores the importance of recognizing paraspinal extramedullary haematopoiesis as a potential cause of neurological symptoms in sickle cell disease patients. Timely diagnosis and multidisciplinary management are essential to prevent irreversible complications.

KEYWORDS

Extra-medullary haematopoiesis, sickle cell disease, paraspinal mass, backache

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LEARNING POINTS

- Consider extramedullary haematopoiesis in sickle cell disease patients with paraspinal masses to avoid unnecessary biopsy.
- Use non-invasive imaging for diagnosis and follow-up in stable cases.
- Address modifiable crisis triggers such as diet and exertion.
- Employ multidisciplinary care to optimize outcomes and prevent complications.

INTRODUCTION

Sickle cell disease is an autosomal recessive hemoglobinopathy caused by point mutation in the beta-globin gene, resulting in formation of haemoglobin S (HbS). It leads to chronic haemolytic anaemia, vaso-occlusive crises, and progressive multisystem organ damage. In response to persistent anaemia and bone marrow stress, Extramedullary haematopoiesis may occur as a compensatory mechanism, characterized by hemopoietic proliferation outside the marrow cavity, most commonly in the liver, spleen, or lymph nodes.

While extramedullary haematopoiesis is well recognized in thalassemia and myeloproliferative disorders, it has historically been considered rare in sickle cell disease. Recent evidence, however, suggests that extramedullary haematopoiesis in sickle cell disease may be more prevalent than previously thought, with paraspinal and visceral sites increasingly reported in the literature[1,2].

Although extramedullary haematopoiesis is observed in up to 7% of hospitalized adults with homozygous sickle cell disease, symptomatic paraspinal involvement with potential neurological complications remains rare, occurring in fewer than 5% of cases, and is typically managed conservatively unless cord compression develops[3-5]. We report a case of incidental paraspinal extramedullary haematopoiesis in a 43-year-old male with sickle cell disease, discovered during evaluation for a vaso-occlusive crisis, emphasizing the diagnostic and therapeutic considerations in this uncommon entity.

CASE DESCRIPTION

A 43-year-old male with a history of sickle cell disease diagnosed at 16 years of age presented with back pain in the setting of a vaso-occlusive crisis. His disease course had been characterized by approximately two pain crises per year, though he reported increasing frequency of episodes over the past two years, requiring multiple hospital admissions.

On this admission, he complained of severe back pain, consistent with his prior crises. He denied fever, chest pain, dyspnoea, abdominal pain, or constitutional symptoms. Physical examination revealed diffuse musculoskeletal tenderness, particularly over the spine, but no neurological deficits or cardiopulmonary abnormalities. He reported dissatisfaction with prior pain control and noted difficulty with outpatient follow-up due to transportation barriers. He also mentioned recent lifestyle changes, including transition to a vegan diet and increased strenuous exercise, which he believed contributed to symptom exacerbation.

A computed tomography (CT) scan of the chest and spine was obtained and compared with prior imaging (19/02/2024). Findings demonstrated no pulmonary embolism or right heart strain. The spleen was shrunken and calcified, consistent with autosplenectomy secondary to sickle cell disease. There was diffuse sclerosis of osseous structures compatible with chronic bone infarcts. Importantly, a lobulated paraspinal soft-tissue density lesion adjacent to the T9–T10 vertebral bodies, measuring up to 2.8 × 2.7 cm, was identified *(Fig. 1)*. This lesion was stable compared with previous imaging and suggesting extramedullary haematopoiesis though a peripheral nerve sheath tumour remained a differential consideration.

The patient was managed with intravenous fluids, multimodal pain control (nonsteroidal anti-inflammatory drugs and opioids), bowel regimen, and incentive spirometry. Hydroxyurea was held during hospitalization and planned for resumption upon discharge. Blood transfusion was deferred as there were no indications. His pain improved with supportive measures, and he was discharged with instructions for outpatient follow-up and surveillance of the paraspinal lesion.

DISCUSSION

Extramedullary haematopoiesis is a compensatory process in which hematopoietic tissue proliferates outside the

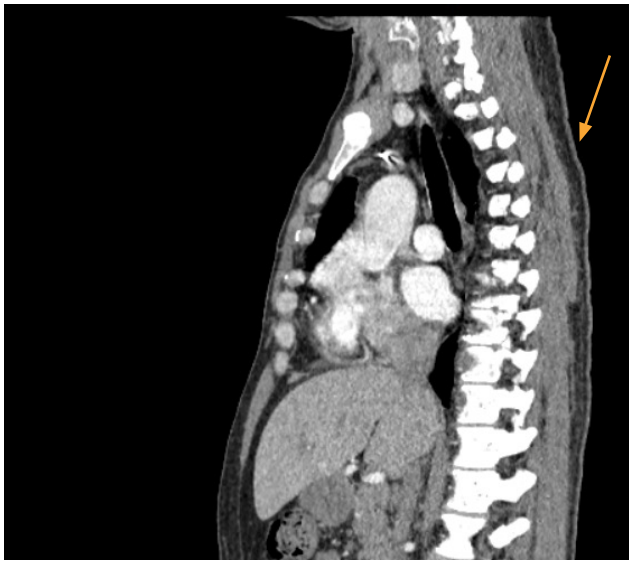


Figure 1. Axial computed tomography scan of the chest showing a lobulated paraspinal soft-tissue mass at T9–T10 (arrow), consistent with extramedullary haematopoiesis.

marrow, typically in response to chronic anaemia or ineffective erythropoiesis. While common in thalassemia and myeloproliferative disorders, extramedullary haematopoiesis occurs in ~7–15% of sickle cell disease patients, most often in the liver, spleen, or lymph nodes[2,4,6-8]. Paraspinal involvement, particularly in asymptomatic forms, remains rare.

In sickle cell disease, chronic haemolysis and vaso-occlusion drive marrow hyperplasia and eventual exhaustion. Persistent hypoxia and elevated erythropoietin promote extramedullary proliferation, often extending through cortical defects or reactivating foetal hematopoietic sites. In the thoracic spine (T8–T12), this results in paraosseous lobulated masses containing trilineage hematopoietic elements[3,5].

Clinically, paraspinal extramedullary haematopoiesis is frequently asymptomatic, discovered incidentally during imaging for other complications, as in our 43-year-old man with sickle cell disease presenting with recurrent vaso-occlusive crises. Symptoms, when present, include back pain, dyspnoea, or neurological deficits from cord compression. CT scan and magnetic resonance imaging (MRI) typically reveal smooth, lobulated paravertebral masses with fatty components (30–65 HU on CT scan; iso- to hyperintense on T1/T2 MRI)[5]. In our patient, chest CT scan—performed to exclude pulmonary embolism—showed stable right paraspinal lesions at T9–T10 (2.8 × 2.7 cm) alongside classic features of sickle cell anaemia, including cardiomegaly, splenic calcification, and sclerotic infarcts.

The differential for paraspinal masses includes lymphoma, myeloid sarcoma, schwannoma, abscess, or infarcts. Extramedullary haematopoiesis is distinguished by stability on imaging, absence of destructive bone changes, and characteristic fatty signals; biopsy is reserved for cases with high suspicion for malignancy.

Management depends on symptom burden. Asymptomatic lesions warrant observation with serial imaging, as in our case[1,2]. For the concurrent vaso-occlusive crisis, supportive care with intravenous hydration, multimodal analgesia, incentive spirometry, and ambulation was effective, and hydroxyurea was resumed on discharge. Symptomatic extramedullary haematopoiesis may require hypertransfusion, hydroxyurea, low-dose radiotherapy (10–20 Gy), or surgical decompression, with symptom resolution reported in up to 80% of cases[2,5].

A limitation of our report is that histological confirmation was not obtained. However, biopsy was considered technically challenging and potentially hazardous due to the delicate paraspinal location of the lesion. In addition, the characteristic radiological appearance and stability of the mass across serial imaging studies were strongly consistent with extramedullary haematopoiesis, making biopsy unnecessary in this case.

The novelty of this case lies in the incidental discovery of stable, asymptomatic paraspinal extramedullary haematopoiesis during crisis evaluation in a sickle cell disease patient with recent lifestyle triggers (strenuous exercise and vegan diet). This highlights how routine imaging for acute complications may reveal rare chronic manifestations. Unlike symptomatic thoracic masses described in thalassemia and sickle cell disease, this case underscores the underrecognized prevalence of extramedullary haematopoiesis and its benign course when asymptomatic.

CONCLUSION

Paraspinal extramedullary haematopoiesis, though rare, should be considered in sickle cell disease patients presenting with back pain or paravertebral masses. Non-invasive imaging combined with clinical context often provides a sufficient diagnosis. Conservative management is appropriate for stable, asymptomatic lesions, with intervention reserved for compressive symptoms. Increased awareness of extramedullary haematopoiesis in sickle cell disease will reduce misdiagnosis and avoid unnecessary procedures.

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