Letter to the Editor

Extramedullary haematopoiesis: An unusual etiology of backache

Dear editor, backache is a common complaint encountered by an orthopaedic surgeon, the differential diagnosis of which encompasses a wide array of conditions. A focused history, meticulous physical exam and a directed imaging are invaluable in dissecting out the culprit in a significant proportion of cases. Most common causes of backache are enlisted in Table 1.^{1,2} A 28-year-old man complained of dull aching diffuse upper back pain since 1 year. He denied any fever, night sweats, weight loss or morning stiffness. Past history was significant for easy fatigability since 10 years of age. There was no history of prior blood transfusion. Examination revealed frontal bossing and maxillary prominence along with pallor and a mild icterus. Liver (4 cm) and spleen (5 cm) were palpable on abdomen examination. Chest radiograph identified the presence of two paravertebral masses along a scoliotic spine (Fig. 1a). Complete blood counts were: haemoglobin-78 gm/l, white cell counts- $14 \times 10^9/l$, differential counts-75% neutrophils, 25% lymphocytes and platelets- $350 \times 10^9/l$. Red cell indices were-mean corpuscular volume-71 fl, mean corpuscular hemoglobin-18 pg, mean corpuscular hemoglobin concentration-24 gm/dl, red cell differential width-18% and corrected reticulocyte-4%. Peripheral blood film suggested marked aniso-poikilocytosis, microcytic hypochromic red cells and the presence of target cells (Fig. 1b). Iron profile revealed serum iron: 171 $\mu\text{g/dl}$, total iron binding capacity: 320 $\mu\text{g/dl}$, transferrin saturation: 57% and serum ferritin: 1050 ng/ml. High performance liquid chromatography (HPLC) findings were suggestive of β -thalassemia intermedia (HbA-15%, HbF-80%, and HbA2-5%). Magnetic resonance imaging (MRI) of the thoracic spine was consistent with features of extramedullary hematopoiesis (EMH). Patient was treated with a regular transfusion regimen (350 ml leucodepleted packed red cells every 3–4 weeks) maintaining a pre-transfusion hemoglobin of 100 g/l along with oral iron chelation therapy (Deferasirox at 20 mg/kg/day single morning dose) and oral hydroxyurea (20 mg/kg/day in two divided doses).

Blood counts, liver and kidney function tests were monitored monthly. Patient experienced a significant improvement in backache within 3 months of treatment. EMH is a common (20%) complication of thalassemia intermedia (TI) arising as a consequence of ineffective erythropoiesis.³ TI patients are particularly prone to these sequelae due to an infrequent blood transfusion requirement resulting in a delayed clinical presentation.⁴ Paraspinal mass accounts for 11%–15% of all EMH cases and may culminate in spinal cord compression. Although biopsy is the gold standard, characteristic imaging findings may clinch the diagnosis without the need for an invasive procedure. Regular red cell transfusions and hydroxyurea are the initial therapeutic options aimed at suppressing the EMH. Low-dose radiotherapy and surgical decompression may be used in refractory cases or for an urgent symptomatic relief.³ A physician must maintain a high index of suspicion for this entity as an underlying etiology of backache. This is particularly important considering that thalassemia is quiet prevalent in India with an estimated 30 million cases of β -thalassemia trait, 65,000–67,000 cases of β -thalassemia intermedia and 10,000 cases of β -thalassemia major.⁵ A history suggestive of long standing easy fatigability, presence of microcytic hypochromic anaemia with target cells on a peripheral blood film and a normal/high serum ferritin value provide an initial clue which should be confirmed by HPLC. EMH should be considered in the differential diagnosis of backache in an appropriate clinical context and a prompt referral of such cases by an orthopedic surgeon to an expert hematologist may avoid the catastrophic cord compression.

Consent for publication

A written and informed consent was obtained from the patient prior to the publication.

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None.

Table 1
classification of various causes of acute and chronic backache.

Acute backache ¹	Chronic backache ²
Intrinsic spine: compression fracture, herniated nucleus pulposus, lumbar strain/sprain, spinal stenosis, Spondylolisthesis, spondylolysis, spondylosis, Systemic: connective tissue disease, inflammatory spondyloarthropathy, malignancy, vertebral diskitis/osteomyelitis Referred: abdominal aortic aneurysm, gastrointestinal conditions, herpes zoster, pelvic conditions, retroperitoneal conditions.	Non-specific/idiopathic (70%): lumbar strain/sprain Mechanical (27%): degenerative processes, herniated disc, osteoporotic fracture, spinal stenosis, traumatic fracture, spondylosis, discogenic pain, presumed instability Referred pain (2%): aortic aneurysm, pelvic conditions, gastrointestinal disorders, renal diseases Non-mechanical (1%): neoplasia, inflammatory arthritis, infection, Scheuermann disease, Paget disease of bone

Fig. 1. Chest radiograph of the patient showing two well defined paraspinal masses (black arrows) along a scoliotic spine (1a). Peripheral blood film of the patient showing marked anisocytosis, poikilocytosis, microcytic, hypochromic red cells along with large number of target cells (black arrow), spherocytes (red arrow) likely post-transfusion and tear drop cells (green arrow) [Leishman stain, $\times 1000$] (1b).

Ethical clearance

obtained from the institute as per Helsinki's declaration.

Patient consent disclosure

We state that a written and informed consent was obtained from the patient prior to submission of the article in the journal for publication.

Conflicts of interests

The authors declare no conflicts of interests.

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