

# A rare case of reversible paraplegia due to extramedullary hematopoiesis in a patient with thalassemia intermedia

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**Received** 20 July 2013

**Accepted** 10 January 2014

**The Egyptian Society of Internal Medicine**  
2014, 26:27–29

30 yrs old man is known to have thalassemia intermedia presented with bilateral lower limb weakness and sphincteric troubles with loss of deep sensations, the motor power of the muscles and precipitancy of urine improved after blood transfusion, laboratory investigations and MRI dorsolumbar spine confirm the diagnosis of extramedullary haematopoiesis.

## Keywords:

extramedullary hematopoiesis, paraplegia, thalassemia

Egypt J Intern Med 26:27–29

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1110-7782

## Introduction

Extramedullary hematopoiesis (EMH) is a rare condition encountered in chronic hemolytic disorders. To compensate for long-lasting anemia in thalassemia intermedia, EMH in the liver, spleen and lymph nodes, and in rare circumstances in the paravertebral and retroperitoneal regions, spinal cord, scalp, pelvis, suprarenal glands, pleura, thymus, breast, prostate, and kidneys can be seen. Spinal cord compression due to EMH is a very rare complication [1,2].

## Case report

A 30-year-old man diagnosed as thalassemia intermedia since the age of 13 and receiving blood transfusion on demand was admitted to Kasr ELAini hospital because of back pain and weakness in both lower limbs that had progressed over the month before the admission with urinary precipitancy. He experienced similar condition 10 months before the current one with marked improvement after blood transfusion. On examination, he had characteristic mongoloid facies, pallor, and jaundice. His blood pressure was 120/80 mmHg, pulse rate was 100/min regular big pulse volume, and temperature was 37.2°C. Abdominal examination revealed evidence of hepatosplenomegaly, with resonant flanks and normal intestinal sounds. Neurologic examination revealed symmetrical hypertonia of both lower limbs, and muscle strength was 4/5 on the proximal and 3/5 on the distal muscles on both sides with flexors weaker than extensors. Deep tendon reflexes were hyperactive and the plantar responses were extensor on both sides. Lost vibration sense up to anterior superior iliac spine and lost joint sense (both sense of position and movement).

Laboratory investigations were: hemoglobin 6.1 g/dl, white blood cell count  $21.8 \times 10^9/l$ , platelet count  $582 \times 10^9/l$ , and mean corpuscular volume 79.9 fl.

Reticulocyte count was 3.9% and ESR was 5 mm/h 1st hr.

Biochemical investigations were: serum bilirubin 2.7 mg/dl (unconjugated bilirubin –2.5 mg/dl), blood urea 32 mg/dl, serum creatinine 0.6 mg/dl, serum calcium 8.8 mg/dl, serum phosphorous 4.6 mg/dl, alanine aminotransferase 14.4 IU/dl, aspartate aminotransferase 47 IU/dl, and serum ferritin 400 ng/ml. Tests for viral hepatitis markers (HBsAg, HCV antibody) were negative. Chest radiography showed widened mediastinum with evidence of bilateral paravertebral opacities (Fig. 1), and chest computed tomography and dorsal spine showed bilateral symmetrical enhancing soft tissue paravertebral masses extending from T5 to T8 into the spinal canal and compressing the spinal cord at D10 and D11(Fig. 2).

MRI lumbosacral spine showed paravertebral and intraspinal extraosseous soft tissue masses at level of L5 (Fig. 3). The appearances of the masses along with the known underlying hematological condition were strongly suggestive of EMH.

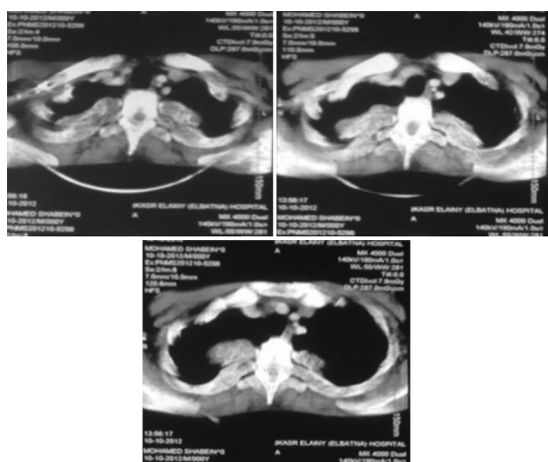
The patient received packed red blood cell transfusion to keep his hemoglobin level above 10 g/dl. A neurosurgical consultation was sought, but the patient was considered to be a poor candidate for surgical decompression in view of large extent of lesion, possibility of hemorrhage, and hemodynamic instability due to anemia. After packed red cell transfusion, the patient condition markedly improved and he could walk without support.

Figure 1



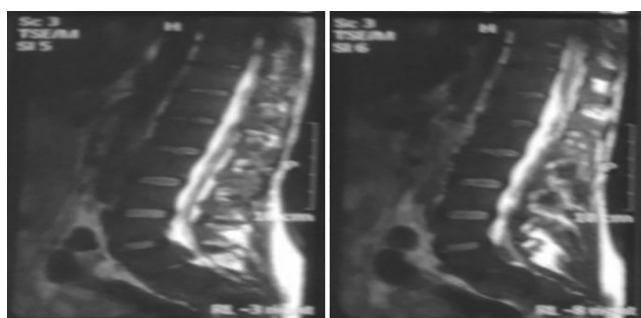
Chest radiograph showing bilateral paravertebral opacities.

Figure 2



Chest computed tomography showing paravertebral masses.

Figure 3



MRI lumbosacral spine showing paravertebral and intraspinal masses.

## Discussion

The occurrence of EMH is a well-known complication in primary and secondary myelofibrosis. However, this phenomenon is rare in association with other hematological disorders such as hemolytic anemia and chronic myeloproliferative disorders without evolving into myelofibrosis [3].

Unlike thalassemia major patients who receive regular blood transfusion from an early age, which suppresses excessive activity of bone marrow, patients with thalassemia intermedia do not receive regular blood transfusion exactly like our patients. Chronic and inefficient erythropoiesis result in EMH [4]. EMH as a compensatory phenomenon usually develops in the sites involved in hematopoiesis during fetal life, such as liver, spleen, and lymph nodes. Involvement of other sites has been reported less frequently. Among the various body regions reported, paraspinal involvement deserves special attention because of the debilitating clinical consequences and challenges in management.

The origin of the spinal epidural hematopoietic tissue is controversial. It has been hypothesized that this tissue could be extruded through the trabecular bone of the vertebral body with a circumferential involvement of the vertebra, or it may have extended through the thinned trabeculae at the proximal rib ends [5,6]. Others have proposed some embryological hematopoietic cell remnants within the epidural space, which would be stimulated along the course of chronic hemolytic anemia. Development of hematopoietic tissue from branches of the intercostal veins has also been suggested [7], whereas others still attribute the masses to embolic phenomena [8,9]. Early in its evolution, the paraspinal extramedullary site of hematopoiesis reveals immature and mature cells mainly of the erythroid and myeloid series and dilated sinusoids containing precursors of red cells. The lesions eventually become inactive and reveal some fatty tissue and fibrosis or massive iron deposits.

A paraspinal location for the hematopoietic tissue occurs in 11–15% of patients with EMH. Since the first case described by Gatto *et al.* [10] in 1954, a large number of cases has been reported in the literature [11].

Paraspinal EMH mainly presents as pseudotumors. However, it is believed that more than 80% of cases may remain asymptomatic and the lesions are usually discovered incidentally by the radiologic techniques [12]. Neurological complication resulting from cord or neural foramen compression is exceedingly rare.

MRI is considered the method of choice for the diagnosis and follow-up evaluation of spinal cord compression cases resulting from EMH [13]. MRI can clearly show anatomical details with high quality including both site and extent of the masses within the spinal canal, while producing soft tissue delineation with high sensitivity.

Because of its rarity, no evidence-based guidelines for the treatment of paraspinal pseudotumors caused

by EMH exist. Management options include blood transfusion, radiotherapy, surgical decompression, hydroxyurea, or a combination of these modalities. Therapy usually depends on the severity of symptoms, size of the mass, patient's clinical condition, and previous treatment [14,15].

## Acknowledgements

### Conflicts of interest

There are no conflicts of interest.

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