

Posterior mediastinal masses in a patient with beta-thalassaemia intermedia

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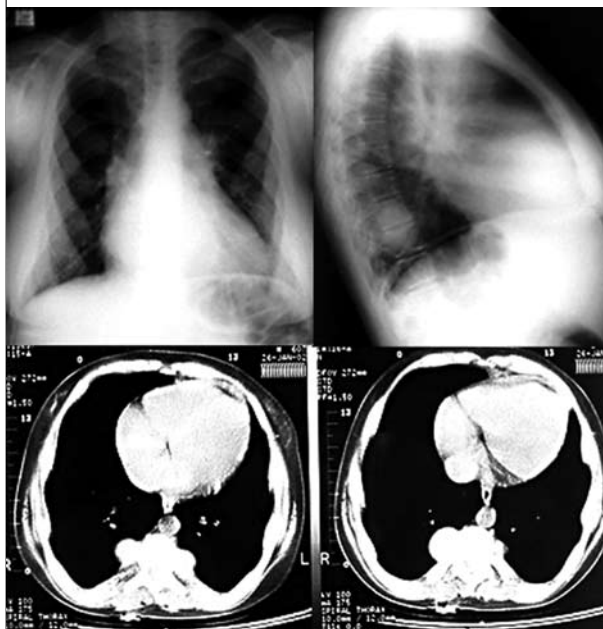
KEYWORDS

Beta-thalassaemia intermedia, extramedullary haematopoiesis, posterior mediastinal mass

CASE REPORT

A 23-year-old female patient was admitted for evaluation of a posterior mediastinal mass on her chest X-ray. She had been suffering from dyspnoea on effort and chest pain for three months. Beta-thalassaemia intermedia was diagnosed when she was four years old. Later a splenectomy was performed. She had been on treatment with folic acid and iron supplements for 13 years. Because of transfusion reactions she had only had four units of blood transfusion in her lifetime. On examination, the patient had a small stature, a typical thalassaemic face and spoon nails. Haemoglobin electrophoresis revealed levels of HbF of 99.7% and HbA₂ of 0.3%. The chest X-ray demonstrated a mass silhouette behind the cardiac shadow and widened ribs. The mass could be seen posteriorly on the lateral chest X-ray. The chest computed tomography disclosed bilateral paravertebral masses and expansion of especially the posterior ribs (*figure 1*).

Figure 1. A mass shadow and widening of the ribs were seen in the posterior-anterior chest X-ray of our patient



The lateral chest X-ray revealed the posterior location of the mass. Computed tomography of the chest revealed bilateral paravertebral masses.

WHAT IS YOUR DIAGNOSIS?

See page 273 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (ON PAGE 271)

POSTERIOR MEDIASTINAL MASSES IN A PATIENT WITH BETA-THALASSAEMIA INTERMEDIA

DIAGNOSIS

There were three paravertebral masses, the largest of which was 3 x 4 cm in diameter, located at T6 to T9. No distortion in the vertebral bodies and no extension to the spinal cord were seen on magnetic resonance imaging of the chest (figure 2). The nature of the masses was established by the bone marrow scan with ^{99m}Tc sulphur colloid, which revealed intense uptake of the radioisotope by the paravertebral masses, which was identical to that of bone marrow. Posterior mediastinal mass lesions of the patient were considered to be caused by extramedullary haematopoiesis (EMH).

EMH, a rare cause of posterior mediastinal mass, may occur in response to insufficient erythropoiesis. Thalassaemia major or intermedia, spherocytosis, sickle cell anaemia and congenital haemolytic anaemia are responsible in most cases of EMH.¹⁻⁴ Commonly involved organs are the liver and spleen where red cells are produced in the foetus during gestation. Intrathoracic involvement is rarely seen and the most common localisation is the lower paravertebral region.^{1,4,5} It is thought to arise from the remains of primitive blood-forming precursor cells present particularly in the posterior thoracic epidural space.⁶ EMH is usually asymptomatic since the organ involvement is most often microscopic; however, it may progress to an organomegaly or sometimes a mass-like lesion. It may even cause spinal cord compression or haemothorax.^{3,4,7}

The radiological diagnostic clues of thoracic EMH are widening of the ribs and lobulated paravertebral masses with no calcification. MRI may be useful in establishing a diagnosis especially by demonstrating the presence of adipose tissue within the mass and by confirming that the bony cortex

is intact.¹ On MRI, EMH masses appear as an isointense mass with high signal intensity on T1-weighted images and a mass of high signal intensity on T2-weighted images.⁶

It is important to recognise the possibility of intrathoracic EMH in the differential diagnosis of posterior mediastinal masses, especially in patients with chronic haemolytic anaemia, before attempting to undergo invasive procedures for the evaluation of the nature of a posterior mediastinal mass; otherwise, it may cause life-threatening bleeding into the pleural space.

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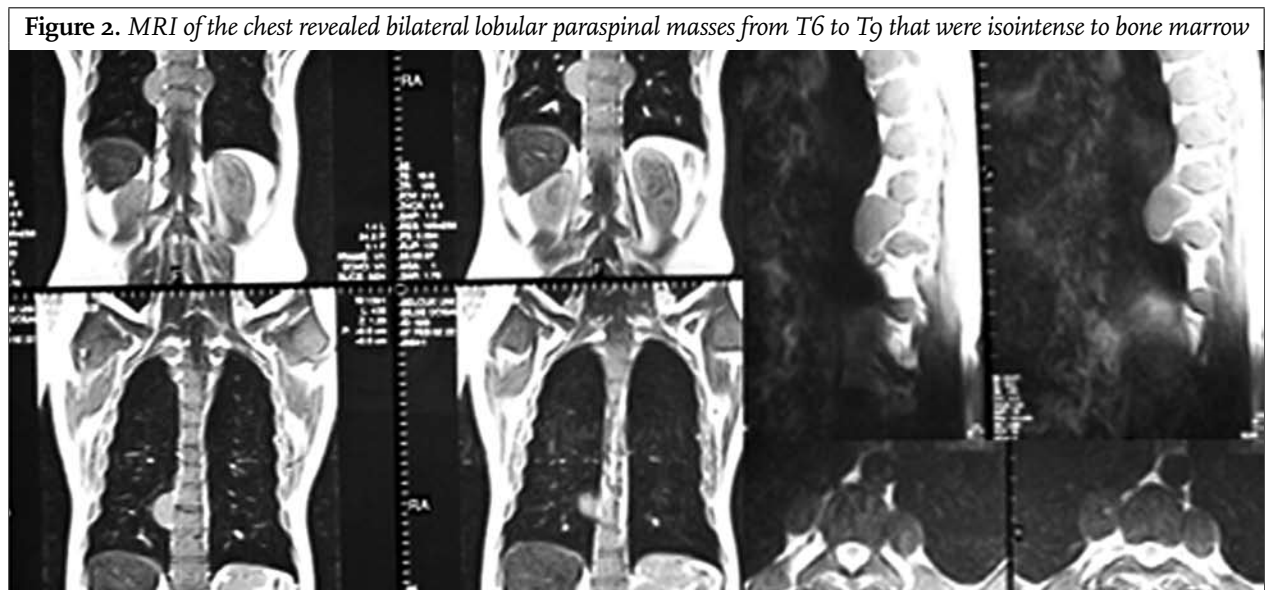


Figure 2. MRI of the chest revealed bilateral lobular paraspinal masses from T6 to T9 that were isointense to bone marrow