

Unusual cause of mediastinal widening

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DESCRIPTION

A 63-year-old woman presented to a local centre after an episode of transient chest pain. A chest X-ray revealed mediastinal widening with well-rounded margins (**figure 1**). A contrast-enhanced CT was done which showed multiple paravertebral masses (**figure 2**). She was subsequently seen in Pulmonology clinic where a guided fine-needle aspiration cytology revealed extramedullary haematopoiesis and she was referred to the Haematology clinic.

On reviewing the history, she gave history of chronic fatigue and had been transfused 2 units of packed red blood cells (PRBCs) during childbirth. She also complained of on-and-off jaundice which had never been evaluated. Examination revealed mild pallor, icterus and spleen was palpable 6 cm below the left costal margin. Complete hemogram revealed isolated anaemia (haemoglobin 7.5 g/dL) with microcytosis (mean corpuscular volume 61 fL). Reticulocyte count was 15% with a reticulocyte production index of 4. Peripheral blood smear was suggestive of moderate anisopoikilocytosis and showed multiple teardrop cells, elliptocytes and occasional polychromatophils. The presentation and basic investigations were consistent with a chronic haemolytic anaemia with microcytosis, thalassemia being the most obvious differential. High-performance liquid chromatography (HbA0 34.4%, HbA2 7.8% and HbF 49.4%) confirmed the diagnosis of non-transfusion-dependent thalassemia. She was started on hydroxyurea at 10 mg/kg/day and administered 2 PRBC transfusions. At 6 months of follow-up, her haemoglobin had risen to 8.6 g/dL with marked improvement in fatigue.



Figure 1 Chest X-ray showing presence of mediastinal widening.

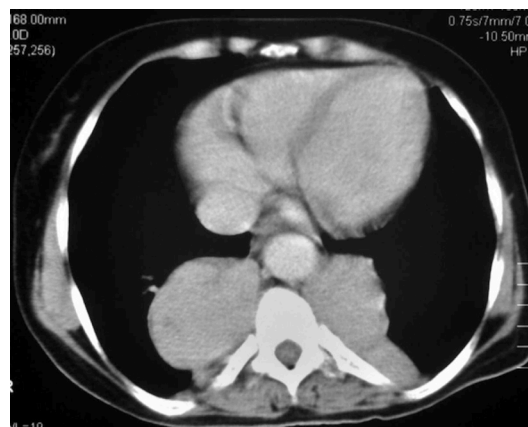


Figure 2 Contrast-enhanced CT chest showing rounded, well-defined paravertebral masses.

Mediastinal widening on chest X-ray is not an uncommon finding. In patients more than 60 years, bronchogenic carcinoma and lymphoma are the most important differentials. Paravertebral extramedullary haematopoiesis as a first manifestation of non-transfusion-dependent thalassemia presenting at this age is exceedingly rare.¹ Similar 'mediastinal tumours' have been described with other chronic haemolytic anaemias as well.² Well-rounded margins, lumpy nodular opacities and obliteration of cardiac shadow are subtle points that can help differentiate paravertebral extramedullary haematopoiesis from the more common superior mediastinal tumours.

Extramedullary haematopoiesis in patients with thalassemia intermedia occurs as a consequence of ineffective erythropoiesis. It may occur at various sites, including lung, spinal cord, heart and pleura, and may present with a pseudotumour or a mass. A higher rate of extramedullary haematopoiesis has been reported in older patients with thalassemia intermedia.³ Management includes regular transfusions and therapies directed at increasing the fetal haemoglobin levels. Local radiation may be considered, especially in those with paraspinous masses causing spinal cord compression.⁴

Learning points

- ▶ Paravertebral extramedullary haematopoiesis can be a presenting manifestation of non-transfusion-dependent thalassemia.
- ▶ A detailed clinical history and complete physical examination along with subtle radiological differences can help differentiate it from the more common malignant causes of mediastinal widening.

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