Case 11002

Extra medullary haematopoiesis
(ECR 2013 Case of the Day)
Published on 21.05.2013

DOI: 10.1594/EURORAD/CASE.11002
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Thorax Mediastinum Musculoskeletal bone
Procedure: Diagnostic procedure
Technique: Conventional radiography
Technique: CT-Angiography
Technique: CT
Special Focus: Haematologic diseases Case Type: Clinical Cases
Authors: Revel M-P, Fedida B
Patient: 45 years, female

Clinical History:
A 45-year-old woman of Italian descent presented with dyspnoea. The physical examination revealed 26 respirations per minute, a heart rate of 103 beats per minute, clear lungs. Laboratory analyses showed a haemoglobin level of 8g per decilitre. She had chronic anaemia and had undergone splenectomy several years before.

Imaging Findings:
Posteroanterior chest-X ray (Fig. 1)
Comment: Posteroanterior chest radiograph demonstrates smooth soft-tissue masses projecting along the lower dorsal spine behind the cardiac silhouette.
CT angiography mediastinal window (Fig. 2 and 3)
Comment: Bilateral paravertebral soft-tissue masses are demonstrated and are associated with soft tissue masses along the inner part of the anterior part of right rib, with focal cortical erosion.
Coronal reformation, bone window (Fig. 4)
Comment: The vertebral cortexes are thinned due to medullary expansion.

Discussion:
A. Background
Extramedullary haematopoiesis (EMH) is regarded as a physiologic compensatory mechanism that occurs when the bone marrow is unable to maintain sufficient red cell production to supply body demand, particularly in congenital haemolytic anaemia such as thalassaemia and hereditary spherocytosis. The CT appearance is characterised by the presence of homogeneous round or lobulated soft tissue density masses usually in the posterior mediastinum. It has also been reported in various extra thoracic locations, most commonly in the liver, spleen, and thorax [1, 2]. Though symptoms related to intrathoracic EMH are uncommon, there are reported cases of spinal cord compression [3].
B. Clinical Perspective
The thalassaemia syndromes are inherited disorders of haemoglobin synthesis, typically seen in individuals of Mediterranean descent. Most patients with thalassaemia have the homozygous form (thalassaemia major). The total
annual incidence of symptomatic individuals is estimated at 1 in 100,000 throughout the world [4].

C. Imaging Perspective
Thoracic EMH characteristically appears on the plain chest radiograph as posterior mediastinal masses that are lobulated and smooth. They may be unilateral or bilateral and exhibit a predilection for the lower thoracic spine region. On computed tomography scans, EMH appears as an area of soft-tissue attenuation adjacent to the posterior ribs [5]. Calcification and associated vertebral anomalies are rare, which helps distinguish EMH from many other causes of posterior mediastinal masses. EMH may also manifest as masses at expanded anterior rib ends as in the present case, though this site is less common.

D. Outcome
Complications include those due to iron overload, post splenectomy infectious complications and spinal cord compression. Pulmonary arterial hypertension is another complication in patients with thalassaemia major and is due to chronic anaemia, haemolysis, and to an increased tendency for microscopic thrombi to form within the pulmonary vasculature.

E. Take Home Message, Teaching Points
Posterior mediastinal masses of soft-tissue attenuation, in the lower part of the thorax in patients with chronic haemolytic anaemia are highly suggestive of EMH and must be distinguished from other causes of posterior mediastinal masses.

**Differential Diagnosis List:** Extramedullary haematopoiesis (EMH), here due to thalassaemia major, Lymphoma, Pleural plaques, Sarcoidosis, Pleural metastatic disease

**Final Diagnosis:** Extramedullary haematopoiesis (EMH), here due to thalassaemia major

**References:**

Description: Posteroanterior chest radiograph demonstrates smooth soft-tissue masses projecting along the lower dorsal spine behind the cardiac silhouette Origin: Revel mp, Department of Radiology, Pompidou hospital, Paris, France
Description: There is a soft tissue mass along the inner part of the anterior part of right rib, with focal cortical erosion. Origin: Revel mp, Department of radiology, Pompidou hospital, Paris France
Description: Bilateral paravertebral soft-tissue masses are demonstrated. Origin: Revel mp, department of Radiology, Pompidou hospital, Paris, France
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