Intrathoracic extramedullary haematopoiesis and skeletal involvement in a case of thalassaemia intermedia

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Section: Chest imaging
Area of Interest: Haematologic
Procedure: Diagnostic procedure
Procedure: Contrast agent-intravenous
Imaging Technique: Conventional radiography
Imaging Technique: CT
Special Focus: Haematologic diseases
Case Type: Clinical Cases
Authors: Eleonora Tricarico1, Raffaella Peschechera2, Francesco Tricarico3
Patient: 25 years, male

Clinical History:

A 25-year-old boy affected by thalassemia intermedia underwent regular follow-up. The patient was asymptomatic, regularly received transfusions and was positive for HCV. A routine chest radiograph was performed.

Imaging Findings:

Chest radiograph revealed a peripheral right basal opacity abutting the costal arches and bilateral well-defined opacities overlying the spine (Fig. 1). A chest CT (computed tomography) was ordered for further characterization, which showed multiple bilateral well-marginated paravertebral and pericostal soft tissue masses demonstrating mild enhancement after intravenous injection of contrast medium (Fig. 2). A moderate amount of soft tissue diffusely surrounded the sternum (Fig. 2b, 2c, 4b). A diffuse structural remodelling of ribs, vertebral bodies, sternum and scapulae was also seen, consisting of rib widening, destruction of medullary trabeculae with trabecular thinning and multiple cortical interruptions, especially in the costal arches adjacent to the soft tissue masses (Fig. 3, 4). The diagnosis of extramedullary haematopoiesis and skeletal abnormalities related to bone marrow expansion was made.

Discussion:

Thalassemia is a chronic, inherited, microcytic anaemia characterized by a defective haemoglobin synthesis associated with ineffective erythropoiesis, leading to bone marrow hyperplasia and extramedullary haematopoiesis, which represents the proliferation of hematopoietic cells outside of the bone marrow [1]. Extramedullary haematopoiesis (EMH) usually involves liver, spleen and lymph nodes, but other possible locations are thorax and kidneys. Uncommon locations are skin, central nervous system and adrenal gland [3]. Bone marrow hyperplasia leads to skeletal changes characterized by expansion of the medulla, cortical thinning and resorption of trabecular bone, with loss of bone density that causes a greater risk of fracture [1, 2]; these bone anomalies manifest with a typical widening of the ribs. Secondary cortical breaks may occur because of the expanded bone marrow and could also be attributed to the presence of adjacent foci of erythropoietic tissue. In fact,
the presence of masses of EMH adjacent to bone and cortical bone disruption are strongly related [4]. Our patient suffered from thalassaemia intermedia and presented diffuse skeletal anomalies with multiple cortical interruptions that were associated with the adjacent masses of intrathoracic extramedullary haematopoiesis. Intrathoracic EMH is characterized by paraosseous masses adjacent to the vertebrae (frequently bilaterally), ribs (commonly multiple), or both, and may also manifest with interstitial lung changes and haemothorax, either alone or in combination. The presence of paraosseous mass is more frequent in thalassaemia than in other causes of intrathoracic EMH [5]. Chest radiograph reveals smooth or lobulated paraspinal or rib masses. On CT images, paraspinal EMH may manifest with isolated or multiple mediastinal masses. Occasionally these can be continuous with abdominal paraspinal masses. The lesions may present mild enhancement, which may be heterogeneous in long-standing lesions representing iron deposition and fat infiltration [3]. Magnetic resonance imaging is an accurate technique in characterization of intrathoracic EMH, showing with great detail both the topography and extent of the masses; in T1 and T2 weighted images, active lesions show intermediate signal intensity, while inactive lesions may reveal high signal intensity in both sequences because of fatty infiltration or low signal intensity secondary to iron deposition [6]. EMH may be asymptomatic or present with symptoms related to the mass-effect of the lesions [5]. The diagnosis of EMH is more straightforward when the lesions are multifocal or bilateral, or present iron deposition or fatty replacement, while in EMH manifesting as a single mass, differentiation from neurogenic or mesenchymal tumour may be challenging and may require biopsy [6].

**Differential Diagnosis List:** Intrathoracic extramedullary haematopoiesis associated with skeletal involvement in thalassaemia intermedia, Neurogenic tumour (for EMH), Mesenchymal tumour (for EMH), Lymphadenopathy (for EMH), Metabolic bone diseases (for skeletal abnormalities), Multiple myeloma (for skeletal abnormalities)

**Final Diagnosis:** Intrathoracic extramedullary haematopoiesis associated with skeletal involvement in thalassaemia intermedia

**References:**

**Description:** Chest radiograph shows bilateral well-defined opacities overlying the spine (yellow arrows) and right basal peripheral opacity abutting the costal arches (green arrow). **Origin:** Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Description: Chest axial image shows bilateral paravertebral well-margined soft tissue masses, demonstrating mild enhancement after intravenous injection of contrast medium; diffuse decreased density of bones with structural remodelling is also seen. Origin: Tricarico C, Radiological Center “Raggi X”, Modugno, Italy
Description: Chest axial image shows bilateral paravertebral well-marginated soft tissue masses and soft tissue in parasternal location (arrow); diffuse decreased density of bones with structural remodelling is also seen. Origin: Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Description: Chest axial image shows bilateral paravertebral well-marginated soft tissue masses and soft tissue in parasternal location (arrow); diffuse decreased density of bones with structural remodelling is also seen. Origin: Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Description: Chest axial image shows bilateral paravertebral and right pericostal well-marginated masses, demonstrating mild enhancement after intravenous injection of contrast medium; diffuse decreased density of bones with structural remodelling is also seen. Origin: Tricarico C, Radiological Center “Raggi X”, Modugno, Italy
Description: Chest axial image shows bilateral paravertebral and left pericostal well-margined masses, demonstrating mild enhancement after intravenous injection of contrast medium; diffuse decreased density of bones with structural remodelling is also seen. Origin: Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Figure 3

Description: Chest axial image shows diffuse structural remodelling of bones with destruction of trabeculae and multiple cortical interruptions; ribs appear widened (arrow). Origin: Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Description: Chest axial image shows diffuse structural remodelling of vertebral bodies with trabecular thinning. Origin: Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Description: Chest sagittal image shows diffuse structural remodelling of vertebral bodies and sternum, that reveals trabecular thinning and multiple cortical interruptions; a moderate amount of soft tissue is seen in parasternal location. **Origin:** Tricarico C, Radiological Center "Raggi X", Modugno, Italy

Description: Chest sagittal image shows diffuse structural remodelling of scapula with trabecular thinning. **Origin:** Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Description: Chest CT axial image shows the rib's cortical interruption (arrow) next to the soft tissue mass. Origin: Tricarico C, Radiological Center "Raggi X", Modugno, Italy
Description: Chest CT sagittal image shows multiple cortical interruptions of the sternum (arrows), which appears surrounded by soft tissue. Origin: Tricarico C, Radiological Center "Raggi X", Modugno, Italy