CASE REPORT

Thoracic extramedullary hematopoiesis: A diagnosis to not forget in a patient with posterior mediastinal mass with anemia

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Summary
The most common masses occurring in the posterior mediastinum are tumors of nerve-cell origin. Other several etiologies can be evoked such as tuberculous abscess, bronchogenic malignancies, esophageal tumors, lymphomas and ectopic hematopoiesis.

We report a case of extramedullary hematopoiesis presenting as posterior mediastinal masses in a 63-year-old woman with congenital dyserythropoietic anemia. The diagnosis was confirmed by a CT-guided fine needle biopsy.

Thoracic extramedullary hematopoiesis represents a rare phenomenon complicating several chronic hemopathies. This diagnosis should be born in mind in front of posterior mediastinal masses in a patient with chronic hemolytic anemia in order to avoid dangerous surgical interventions.

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Introduction

The most common masses occurring in the posterior mediastinum are tumors of nerve-cell origin. Other several etiologies can be evoked such as tuberculous abscess, bronchogenic malignancies, esophageal tumors, lymphomas and ectopic hematopoiesis. Their exact frequency is unknown. We report a case of extramedullary hematopoiesis (EMH) presenting as posterior mediastinal masses that were symptomatic and confirmed by fine needle biopsy.

Case report

A 63-year-old woman was referred to the Pulmonary Department of Sfax Hospital (Tunisia) because of a 2-week history of increasing breathlessness, dry cough, pleuritic
left chest pain and fatigue. Past history disclosed anemia diagnosed at the age of 10 years requiring blood transfusions and since the patient was lost sight of.

Initial physical examination revealed a blood pressure of 120/70 mm Hg, pulse rate of 80 beats/min, respiratory rate of 24 breaths/min. Chest examination showed decreased breath sounds at the right lower lung. Splenomegaly was noted on abdominal palpation without hepatomegaly.

Laboratory data revealed a normocytic, normochromic anemia (hemoglobin: 6.5 g/dL, mean corpuscular volume: 89 fL, mean corpuscular hemoglobin concentration: 30.6 g/dL), nonregenerative (83,000 reticulocytes/mm³). The peripheral blood smear examination disclosed marked anisocytosis, poikilocytosis, microcytosis and polychromatophilia. The liver function test showed total bilirubin of 29 µmol/L (normal: 4–18), direct bilirubin of 6 µmol/L (normal: 0–4). Serum ferritin level was 352 ng/mL (normal: 20–200).

Chest radiography (Fig. 1) revealed widening of the upper mediastinum, large right paravertebral mass and paracardiac lobulated mass located at the posterior mediastinum. Computed tomographic scanning of the chest (Fig. 2) demonstrated two large, well circumscribed, paravertebral thoracic masses, measuring 6 cm at the right and 8 cm at the left, containing fat and associated with left pleural effusion. CT scan showed no adenopathy, or erosion of the vertebral bodies and the ribs. A CT-guided needle biopsy of the left mass was performed. Histological findings (Fig. 3) revealed normal hematopoietic tissue comprising erythroblasts, megakaryocytes and myeloid precursors without neoplastic infiltration.

As part of exploration of the anemia, a bone marrow aspirate was performed. It showed wealthy marrow with marked signs of dyserythropoiesis and nuclear fragility (bi and multinucleated cells) occurring in mature erythroid cells. Serum B12, folate and hemoglobin electrophoresis were normal. All these findings suggested the diagnosis of congenital dyserythropoietic anemia type II.

The final diagnosis of EMH presenting as posterior mediastinal masses in a patient with congenital dyserythropoietic anemia was established. Regular blood transfusions were performed.

**Comments**

EMH is a physiologic response to compensate for bone marrow dysfunction. It can occur in patients with either neoplastic process or chronic hemolytic anemia especially thalassemia, hereditary spherocytosis and sickle cell anemia. It is rarely associated with congenital dyserythropoietic anemia; few cases are reported in the literature. Any site of the body can be involved by this phenomenon but the most common sites are the liver, spleen and lymph nodes. Less frequently locations include the kidneys, adrenal glands, intrathoracic cavity, presacral.

**Figure. 1** Chest radiography: a wide upper mediastinum with large right paravertebral mass and paracardiac lobulated mass.

**Figure. 2** Computed tomographic scanning of the chest: two large, well circumscribed, paravertebral thoracic masses, containing fat and associated with left pleural effusion.

**Figure. 3** A normal hematopoietic tissue including erythroblasts, megakaryocytes and myeloid precursors without dysplasia (HE × 200).
region, peritoneum, skin, breast, central nervous system and paravertebral areas.\textsuperscript{2,3,7}

The pathogenesis of this outside-bone marrow hematopoiesis is unclear. It may originate from extension of hyperplastic marrow through the thin cortex of ribs and vertebral bodies; the capsule of the mass is formed by the periosteum. Another explanation is that EMH results from transforming of embryonal rests of osteogenic tissue into hematopoietic one under stress conditions in order to maintain sufficient red cell production.\textsuperscript{2,8,9}

Intrathoracic EMH is most often silent, discovered by chance. In rare cases, it can compress neighboring organs and leads to clinical signs such as symptoms of spinal cord compression, dyspnea, cough or pleuritic chest pain.\textsuperscript{4,7,10} in our case, symptoms are due to the large size of the masses. Radiographic exams are useful for recognizing the diagnosis. The chest X-ray shows smooth lobulated masses located at the posterior mediastinum without bony erosion.\textsuperscript{10,11} On computed tomography scanning, intrathoracic EMH appears as unilateral or bilateral well circumscribed, paravertebral masses, lying between vertebra T6 and T12 and having soft tissue density with homogeneous contrast product’s enhancement. These masses contain sometimes adipose tissue as it was seen in our case; calcification is rare. Bony structures can be widened but not eroded in contrast to neurogenic tumors which are associated with osseous destruction in about 50 percent of cases.\textsuperscript{7,9,12}

Technetium 99 m-labeled sulfur colloid scan is also a non-invasive technique of detecting areas of EMH; this radioactive agent is taken up by reticulo-endothelial cells.\textsuperscript{3,10}

The diagnosis of EMH can be established on the basis of radiographic features especially when they occur in a patient with a long history of anemia. Although, if the diagnosis is not certain or if complications require surgical intervention, biopsy is mandatory.\textsuperscript{3,10,12} Histological proof can be obtained using transthoracic biopsy, video-assisted thoracoscopy or thoracotomy. All theses procedures should be carried out carefully because of the high risk of bleeding complications.\textsuperscript{8,13}

Treatment of EMH remains controversial and it is generally required only in the presence of complications such as spinal cord compression, massive hemotherax or recurrent pleural effusion. Low-dose radiation, repeated blood transfusions, surgery, corticosteroids and recently hydroxyurea therapy are the main therapeutic tools allowing the inhibition of hematopoiesis and the decrease of recurrence.\textsuperscript{3,8,10}

\textbf{Conclusion}

Thoracic EMH represents a rare phenomenon complicating several chronic hemopathies. This diagnosis should be born in mind in front of posterior mediastinal masses in a patient with chronic hemolytic anemia in order to avoid dangerous surgical interventions.

\textbf{Conflict of interest statement}

We confirm that none of authors have a conflict of interest to declare in relation to this work.

\textbf{References}