

## A Hemothorax and Hemo-Retroperitoneum Revealing Para-Vertebral Thoracic Extra-Medullary Hematopoiesis

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### Abstract

The authors report the case of a 55-year-old patient, admitted for management of dyspnea with deterioration of the general state. The thoracic computed tomography (CT) finds para-vertebral masses associated with a hemothorax and a hemoperitoneum. The diagnosis of extramedullary hematopoiesis (EMH) complicated by bleeding is evocated.

**Keywords:** Extra-Medullary Hematopoiesis; Hemothorax; Hemo-Retroperitoneum; CT Scan

### Introduction

Extra-medullary hematopoiesis (EMH) is also called "Medullary heterotopia" or even "hematopoietic lipoma", it represents a classic entity complicating the evolution of many chronic hematological conditions such as thalassemia, sickle cell anemia or neoplasia.

### Case Report

55-year-old patient, without notable history, consults in the emergency for management of progressive worsening dyspnea evolving in a context of deterioration of the general state. Biological assessment finds a predominantly lymphocytic hyperleukocytosis (40.000 cells/mm<sup>3</sup>). Chest radiograph shows mediastinal enlargement with bilateral paravertebral lobulated masses. A thoraco-abdomino-pelvic computed tomography is performed, which objective bilateral paravertebral, above and under diaphragmatic, tissue masses, circumscribed and lobulated, moderately enhanced after injection, without underlying bone lysis or endocanal extension (Figure 1 and 2). A hemothorax and a hemoretroperitoneal are associated (Figure 1 and 3). Hepatosplenomegaly, periportal infiltration and diffuse bone demineralization of the entire skeleton is noted (Figure 2 and 4). Due to key-information provided by imaging, a para-vertebral thoracic extramedullary hematopoiesis complicated by bleeding is diagnosed.

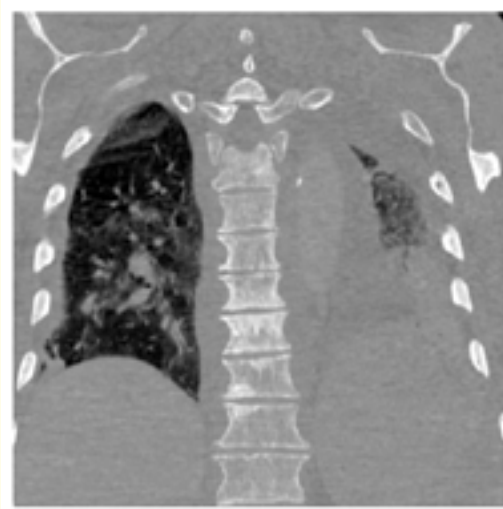
### Discussion

Extra-medullary hematopoiesis (EMH) is also called "Medullary heterotopia" or even "hematopoietic lipoma", it represents a classic entity complicating the evolution of many chronic hematological conditions such as thalassemia, sickle cell anemia or neoplasia. It reflects a compensatory phenomenon of a spinal dysfunction in other hematopoietic organs - this process is activated when a dysfunction of production in red blood cells occurs [1].



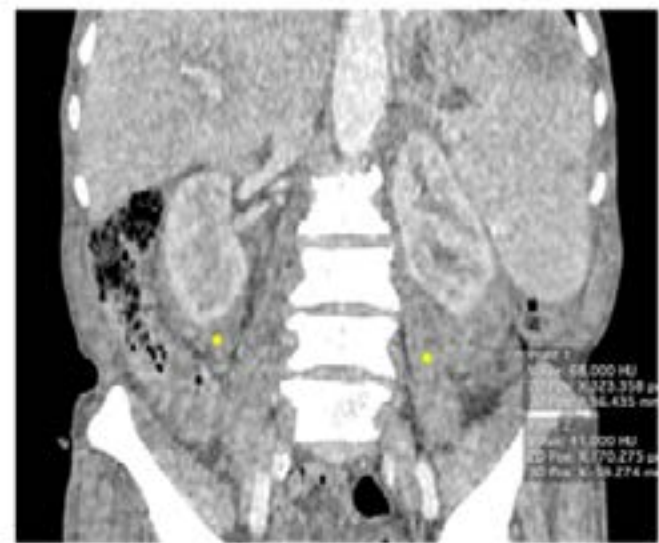
**Figure 1:** Thoracic CT scan with iodized contrast agent injection in axial section showing bilateral posterior mediastinal tissue masses opposite T10-T11, with regular, heterogeneous contours, without associated calcifications, increasing moderately after injection of product contrast (→). Note the bilateral hemothorax, more marked on the left (->).



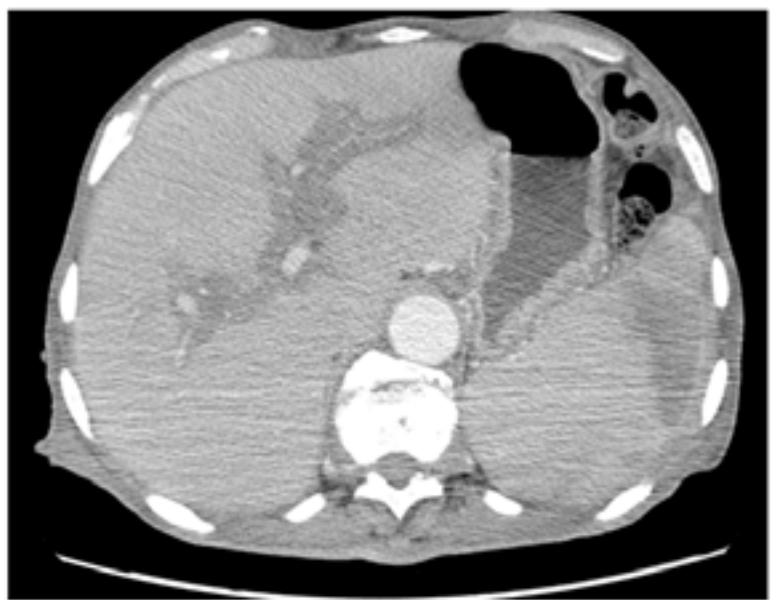


**Figure 2:** Chest CT in coronal sections, showing bilateral para-vertebral enlargement. Note the bone demineralization extended to the entire spine.





**Figure 3:** Abdominal CT scan with injection of iodinated contrast agent in axial and coronal sections, showing a bilateral hemo-retroperitoneum, filling the infra-renal spaces (→).



**Figure 4:** Abdominal CT scan, in axial section, showing hepatosplenomegaly with periportal infiltration. The spleen is increased in size and seat of peripheral infarction zones.

The pathophysiological mechanism is still poorly understood, however two hypotheses try to explain the EMH: An extension of a hyperplastic marrow through the ribs and vertebral bodies covered with the periosteum or a transformation of the remaining osteogenic tissue into hematopoietic tissue [2].

The intra-thoracic location is almost exceptional and generate a diagnostic problem. Indeed, its posterior para-vertebral mediastinal location, should suggest a good number of diagnoses: nerve or esophageal tumor, tuberculous abscess, pleural fibrosis, lymphoma, etc.

In its typical form, the EMH is asymptomatic, with incidental discovery. If the hematopoiesis is massive and bulky, it manifests as chest pain, cough, dyspnea or spinal cord compression. In the event of a hemorrhagic complication, it is responsible for a hemothorax and a hemo-retroperitoneum.

The chest x-ray shows a mediastinal enlargement, or more rarely a retro-lobulated mass of the heart, a widening of the anterior arches of the ribs or an exaggeration of the trabeculation of the shoulder blades and ribs.

Computed tomography finds heterogeneous bilateral vertebral masses, well limited, of variable size, fatty density without calcification, moderately enhanced after injection, without locoregional lymphadenopathies and underlying vertebral lesion [3]. The bleeding in EMH results in a united or bilateral pleural effusion, increased unenhanced attenuation density in spontaneous contrast. When heavy, the bleeding may extend to the retro-peritoneal spaces. This was the case in our patient, where the effusion was so abundant, that we found a total and bilateral filling of the peri-renal spaces without associated renal parenchymal disease.

Diagnostic imaging of EMH foci is essential, because it makes it possible to make the differential diagnosis with malignant tumor lesions and thus avoids additional invasive investigations [4].

Abstention from therapy is the rule, especially when faced with asymptomatic HEM and incidental discovery [5].

The evolution is generally favorable with regression of the volume of the paravertebral masses with the correction of hematopoietic disorders.

### Conclusion

HEM is an extremely rare entity. Its symptomatic nature should raise fears of a complication. The evolution is favorable after treatment of hematopoietic disorders. It is therefore fundamental to know how to evoke the diagnosis in the face of hematopoietic dysfunction and characteristic imaging.

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