

# A rare case of thoracic extramedullary hematopoiesis associated with myelofibrosis: a diagnostic dilemma

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## Abstract

Extramedullary hematopoiesis (EMH) is the production of hematopoietic precursors outside the bone marrow cavity which may cause mass effects according to its localization. A 50-year-old male presented with non-specific symptomatology and a mass in both paravertebral regions and posterior mediastinum on chest X-ray and in the chest computed tomography (CT) scan. A CT guided percutaneous needle aspiration from the mass showed hematopoietic cells with fat spaces. A bone marrow biopsy revealed a hypercellular marrow with an increased fibrosis. The final diagnosis was myelofibrosis leading to extramedullary hematopoiesis in paravertebral region.

**Keywords:** Extramedullary hematopoiesis, paravertebral region mass, myelofibrosis.

**Abbreviations:** EMH - Extramedullary hematopoiesis, HSC - Hematopoietic stem cell, CT- Computed tomography

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open biopsy. We present a case of EMH presenting as a symptomatic mass in both paravertebral regions and posterior mediastinum diagnosed by CT guided fine needle biopsy, subsequently discovered to harbor myelofibrosis. A brief literature review relating to the pathogenesis, site, causes and incidence is also done.

## CASE REPORT

A 50-year-old man, non-smoker, presented with constitutional symptoms like fever, night sweats, weight loss, dry cough and worsening dyspnoea for last 6- 8 months. Physical examination showed pallor with mild hepato- splenomegaly. Bilateral crepitations were present on chest auscultation. Laboratory investigations revealed hemoglobin 8.9 gram percent, platelet count 80,000/mm<sup>3</sup>, and the WBC count 4,000/mm<sup>3</sup>, with a normal differential count. Abnormal RBCs, including teardrop cells and occasional nucleated RBC were seen on the peripheral smear. No other laboratory abnormality was present. Routine work-up included the postero-anterior chest radiograph showing a smoothly outlined mass posterior to the right border of the heart. The axial section of the CT of thorax revealed two solid oval soft tissue

## INTRODUCTION

Extramedullary hematopoiesis (EMH), production of blood cells outside the bone marrow, is a very rare cause of a posterior mediastinal mass and may be clinically confused with other benign or malignant mediastinal tumours. The commonest tumour of this region are neurogenic tumours, and is a close differential diagnosis.<sup>1</sup> Causes of EMH including various hematological conditions like chronic hemolytic anemia and myelofibrosis. CT guided FNAC is a simple and cost effective procedure to arrive at the diagnosis compared to

masses located symmetrically in the paravertebral space bilaterally encroaching the posterior mediastinum, without lymphadenopathy or bony erosion. Mild pleural reaction of the left side was present while there was no pleural effusion. (Figure 1a and 1b). A CT guided FNAC of the mass was performed and stained by MGG and PAP. The stained smears showed cells of myeloid series, nRBCs and megakaryocytes with admixed fat. (Fig. 2a and 2b) A diagnosis of EMH was made. The follow up of the procedure was uneventful. Because of the clinical symptoms and to explain EMH, a bone marrow trephine biopsy was performed and showed hypercellular bone marrow with grade 4 reticulin fibrosis. (Fig. 3) The final diagnosis of EMH presenting as posterior mediastinal mass in a patient with myelofibrosis was established.

### DISCUSSION

EMH is the formation and development of blood cells outside the medullary spaces of the bone marrow and is usually associated with various hematological conditions like myelofibrosis, myelophthisis, myelodysplasia, necrosis, infection, and metastatic malignancy. There are multiple hypothesis prevalent regarding the pathogenesis of EMH of which most prominent is the transformation of embryonic rests into hematopoietic tissue under circumstances of bone marrow stress.

The concept of a stem cell “niche” was first proposed in 1978 by Schofield, who hypothesized that a HSC in bone marrow acts as a fixed tissue cell whose further maturation is prevented by contact with the surrounding cellular environment.<sup>2</sup> The environmental conditions and cells (including resident populations of relevant stromal cells and possibly HSCs) in hemic tissues such as spleen and liver remain favorable for the initiation or reactivation of a stem cell niche. However, EMH—and, therefore, presumably the requisite stem cell microenvironment—can occur in any tissue.<sup>3</sup> It appears that subpopulations of cells within specific tissues may remain unrestricted and possess pluripotent potential that is reminiscent of embryonic stem cells.<sup>4</sup> Bone marrow disease or ablation causing damaged, disrupted, or inadequate medullary spaces and altered architecture, signaling, and microenvironment results in failure of

normal hematopoiesis and the mobilization and/or activation of HSCs and HPCs to extramedullary sites, resulting in marked EMH in multiple organs as in the causes mentioned above.<sup>3</sup> Primary myelofibrosis is the prototypical example of bone marrow failure associated with EMH. The disease arises from the transformation and clonal proliferation (hyperproliferation) of a single HSC in the marrow, with mobilization of stem cells to new or reactivated niches in various tissues.<sup>5</sup> The liver, spleen, kidney, lymph nodes, and posterior mediastinum are the most common sites of EMH, with EMH often occurring in more than one site.<sup>6</sup> Intrathoracic EMH may manifest as a paraosseous mass, interstitial pulmonary abnormality, pleural mass, or hemothorax, either alone or in combination. Paraosseous EMH is the most common manifestation of intrathoracic EMH. It appears as a small or large mass adjacent to the vertebrae (frequently bilaterally as in our case) ribs (commonly multiple ribs), or both. The absence of bone erosion in CT scan as in the present case, allows differentiation from neurogenic tumors.<sup>7</sup> In a case series of nonhepatosplenic extramedullary hematopoiesis done at Mayo clinic, the most common site involved was in or surrounding the vertebral column in the thoracic region.<sup>8</sup> These masses are usually asymptomatic and may be discovered incidentally<sup>9</sup>. 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.<sup>10</sup> In the present case, symptoms were due to the large size of the masses. Inhomogeneously enhancing bilateral paravertebral masses in patients with myelofibrosis or hemolytic anemia are highly indicative of EMH. These masses have to be differentiated from lymphadenopathy and neurogenic tumors.<sup>3</sup>

### CONCLUSIONS

In conclusion, paravertebral with mediastinal EMH represents a rare phenomenon associated with several hematological disorders. This diagnosis should be kept in mind in cases of posterior mediastinal masses in a patient with unexplained anemia and respiratory symptoms in order to avoid surgical interventions.



Figure 1a



Figure 1b

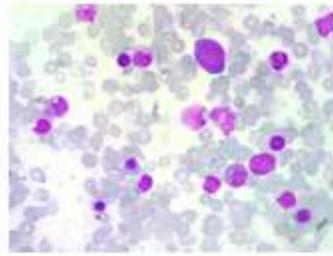


Figure 2a

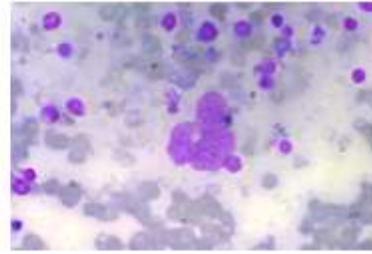


Figure 2b

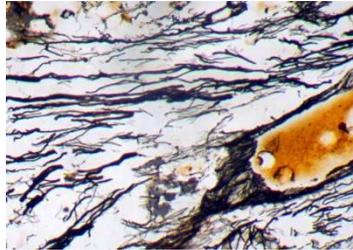


Figure 3

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