EJMI

DOI: 10.14744/ejmi.2017.68552
EJMI 2017;1(1):16-18

Case Report

Co-existence of Unilateral Intrathoracic Extramedullary Hematopoiesis with COPD and Respiratory Failure: A Case Report

Nursen Yasayancan

Department of Pulmonary Medicine, Gaziosmanpasa University, Tokat, Turkey

Abstract

Extramedullary hematopoiesis (EMH) is a compensation mechanism in which blood cells are produced in regions other than the bone marrow in response to deficient bone marrow production. It commonly occurs in diseases that lead to chronic anemia such as sickle cell anemia, beta thalassemia and myelofibrosis. A 68-year-old male patient was referred by the hematology outpatient clinic for investigation of secondary polycythemia etiology. Contrast-enhanced thoracic computed tomography revealed a space-occupying formation with an approximate size of 26x31 mm in right lateral adjacency to vertebra IV. The density of the mass was measured as -13 HU and it was considered to be a mass with fatty content. Mediastinal magnetic resonance imaging (MRI) was performed to clearly reveal the relationship of the mass to the spinal tract. Based on the radiological findings detected on the MRI, it was determined to exhibit results resembling EMH.

Keywords: Chronic obstructive pulmonary disease, extramedullary hematopoiesis, thoracic imaging

Extramedullary hematopoiesis (EMH) is a compensation mechanism whereby blood cells are produced in regions other than the bone marrow in response to deficient bone marrow production. It occurs commonly in diseases which lead to chronic anemia such as sickle-cell anemia, beta thalassemia and myelofibrosis. While it often occurs in regions responsible for hematopoiesis in fetal life such as the spleen and the liver, it may rarely manifest as bilateral masses in the intrathoracic region of the paravertebral field.[1, 2]

In this case report, we aimed to present a case of lateral EMH in a male patient diagnosed with chronic obstructive pulmonary disease (COPD) and respiratory failure who was referred by the hematology outpatient clinic for investigation of secondary polycythemia, and also present the relevant literature.

Case Report

A 68-year-old male patient was referred by the hematology outpatient clinic for investigation of secondary polycythemia etiology. The hematologist did not detect any hematologic disorder like leukemia, lymphoma, sickle cell anemia, thalassemia, myelodysplasia as a result of physical examination and labaratory tests. He had been under follow-up for COPD for approximately 15 years and for respiratory failure for the last 2 years. He was receiving long-term oxygen therapy and using bilevel positive airway pressure devices at home. His physical examination revealed no pathologic findings other than bilateral rhonchi. He had no hepatosplenomagaly. Laboratory test results were as follows: Hb: 11.6 gr/dL (13-18), Hct: 49.2% (35-45), erythropoietin >750 mIU/mL (3.5-17.6). Arterial blood gas investigation detect-

Address for correspondence: Nursen Yasayancan, MD. Gaziosmanpasa Universitesi, Gogus Hastalıkları Anabilim Dalı, Tokat, Turkey
Phone: +90 356 212 95 00 **E-mail:** nursenkokturk@hotmail.com
Submitted Date: September 31, 2017 **Accepted Date:** October 31, 2017 **Available Online Date:** November 20, 2017

ed a pH of 7.33, PCO_2 of 58.4 mmHg, PO_2 of 47.8 mmHg and sO_2 of 76.6%. Lung radiography indicated extension in the right upper mediastinum (Fig. 1). Contrast-enhanced thoracic CT (Fig. 2) showed a space-occupying formation at an approximate size of 26x31 mm in right lateral adjacency to vertebra IV. The density of the mass was measured as -13 HU and the mass was considered to be a mass with fatty content. Mediastinal magnetic resonance imaging (MRI) was performed to clearly reveal the relation of the mass to the spinal tract (Fig. 3). As a result of the radiologic findings of the mass detected on MRI, it was determined to exhibit results in favor of EMH and it was deemed as not associated with neural phenomenon. Fine needle aspiration biopsy (FNAB) was recommended to the patient to confirm the definite diagnosis but he did not accept biopsy. PET CT was also recommended but he did not want to get whole body radiation. Follow-up was planned since the patient had no neurologic symptoms. Follow-up radiologic investigations performed at 6 months and one year later revealed no changes in EMH, therefore the patient remains under follow-up without treatment. He has been living without worsening in his clinic for 4 years and does not want to have any intervention during this time.

Written informed consent was obtained from the patient participated in this case.

Discussion

While EMH commonly occurs in patients with myeloproliferative disease, it may also be observed in those with COPD and respiratory failure due to chronic anemia and hypoxia. Thoracic area is a rare region of involvement for EMH. Intrathoracic EMH usually occurs as posterior mediastinal masses between the vertebras T6 and T12. While it is mostly bilateral, it may rarely be unilateral. In patients with severe anemia, benign production of blood cells outside the bone marrow may also be seen. They may manifest as lobulated masses of the bone marrow as a result of protrusion of the bone marrow from cortical defects.

In 1912, Guizetti detected intrathoracic EMH during autopsy. In 1945, Ask-Upmark for the first time succeeded in diagnosing EMH patients by means of percutaneous needle biopsy.[3]

Clinically, these cases are generally asymptomatic. Patients may have anemia and a high erythropoietin level may be detected. They may grow slowly without clinical findings. In some cases, back pain secondary to spinal cord compression and neurologic findings may be observed. Mostly, they are detected incidentally with imaging methods.

The largest case series (510 cases) related to EMH was published by Koch et al.[4] In 94.7% of these patients, the hematopoietic focus was reported to be the liver or the spleen. In the remaining 5.3%, EMH foci included the paraspinal region, lymph nodes, pulmonary parenchyma and pleura. In this study, the authors reported that they found chronic anemia in most of the patients with EMH and that their mean hemoglobin level was 9.5 g/dL with normal hemoglobin values in a portion of patients. Our patient also had a low hemoglobin value.

Although most patients describe symptoms associated with the site of lesion, they are asymptomatic at the time of diagnosis. In case of intrathoracic EMH, dyspnea is the most common complaint of presentation.[5] In the case presented herein, the patient had also dyspnea and this condition was mainly associated with the patient's COPD and respiratory failure.

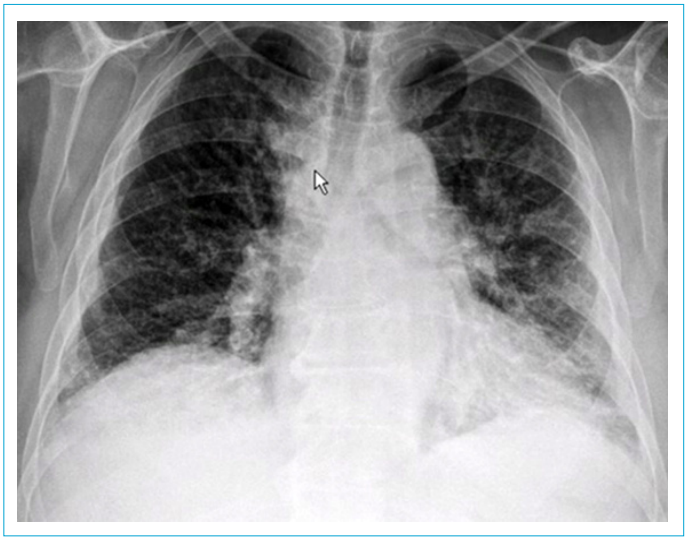

Figure 1. Extension in the right upper mediastinum as seen on posterior-anterior (PA) lung radiography (white arrow).

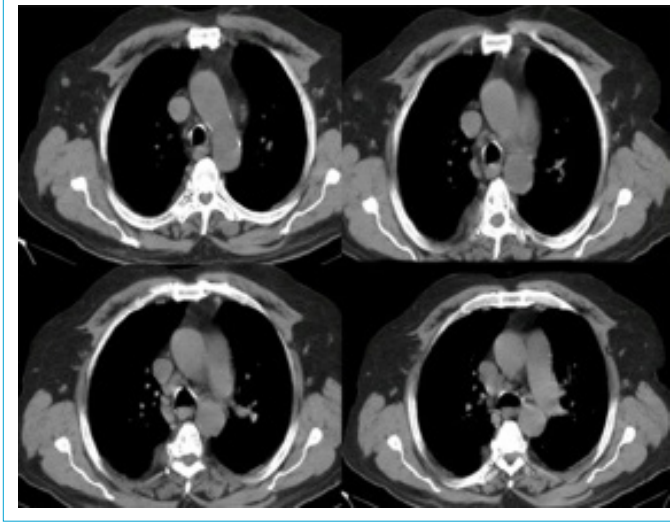

Figure 2. The formation of 26x31 mm in size with heterogeneous density in right lateral adjacency to the 4th vertebra on thoracic CT (white arrow).

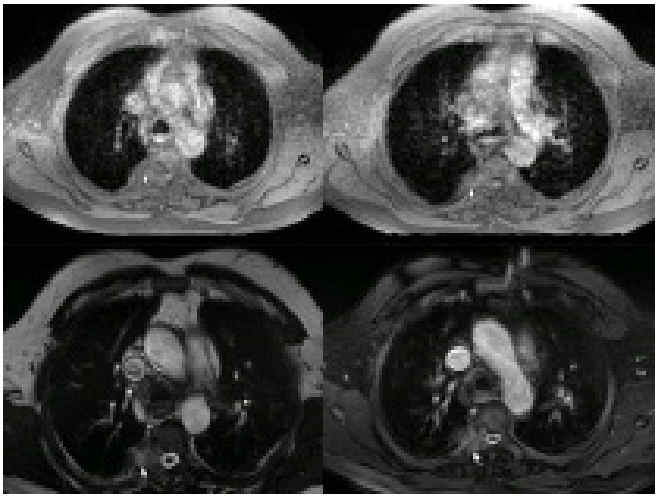

Figure 3. Space occupying, poorly demarcated lesion of approximately 28x26 mm size in the right lateral adjacency to vertebra IV extending to the neural foramen and spinal tract extradural space and causing extension in the neural foramen; mildly hyperintense on T2 axial series, intermediate intensity on T1 axial series; no suppression in fat-suppressed series and marked intense contrast uptake in the post-contrast series on mediastinum MRI.

During radiologic investigations, intrathoracic EMH commonly appears as well-demarcated, lobulated, non-calcified paravertebral masses of fatty density.[6] Thoracic CT is a beneficial method to reveal the localization and character of the masses seen in intrathoracic EMH and it is important for determining the presence or absence of an intraspinal extension of the lesion and potential bone destruction. The lesion exhibits fatty content and no calcification. Contrast uptake is observed. The size may vary from 5 mm to 5 cm, and these masses grow very slowly during follow-up.[7]

The diseases that should be considered in the differential diagnosis of EMH are neurogenic tumors, abscess arising from the spine, primary and metastatic malign tumors, Castleman's diseases. Neurogenic tumors are the most common cause of a posterior mediastinal mass, at least 50 per cent show either erosion or sclerosis of the adjacent bone. If it is abscess arising from the spine, involvement of the bone structure can be demonstrated, no lobulation is present. Primary and metastatic malign tumors don't show a lobulated contour and involvement of bone is common.[8]

In addition to co-assessment of clinical and radiologic findings, EMH diagnosis is also established through tissue biopsy or FNAB. However, if there is clinical or radiological suspicion of metastasis in patients with underlying malignancy, histopathologic diagnosis is deemed necessary. While histopathological investigation is the gold standard, it is not always required. In the presence of clinical anemia or polycythemia vera with characteristic findings detected on CT or MRI, intrathoracic EMH may be diagnosed without the need for invasive procedures. In our patient who does not accept further examination we think the diagnosis EMH because of a lobulated or rounded soft tissue density in the posterior medistinum with-out rib erosion, a history of chronic anemia and stabil radiological findings for one year.

In conclusion, review of the literature shows that EMH occurs mostly in thalassemia patients and clinically, it is usually asymptomatic while it may be bilateral or multiple in terms of radiological findings. In our case, in contrast to the literature, the patient had COPD and respiratory failure with only polycythemia detected without any underlying myeloproliferative disease. Radiologically, in contrast to the literature, it was lateral and not bilateral. Since the diagnosis was supported with CT and MRI findings following clinical suspicion.

Disclosures

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

References

1. Fucharoen S, Winichagoon P. Clinical and hematologic aspects of hemoglobin E beta-thalassemia. Curr Opin Hematol 2000;7:106–12.
2. Chehal A, Aoun E, Koussa S, Skoury H, Koussa S, Taher A. Hypertransfusion: a successful method of treatment in thalassemia intermedia patients with spinal cord compression secondary to extramedullary hematopoiesis. Spine (Phila Pa 1976) 2003;28:E245–9.
3. Catinella FP, Boyd AD, Spencer FC. Intrathoracic extramedullary hematopoiesis simulating anterior mediastinal tumor. J Thorac Cardiovasc Surg 1985;89:580–4.
4. Koch CA, Li CY, Mesa RA, Tefferi A. Nonhepatosplenic extramedullary hematopoiesis: associated diseases, pathology, clinical course, and treatment. Mayo Clin Proc 2003;78:1223–33.
5. Rumi E, Passamonti F, Boveri E, De Amici M, Astori C, Braschi M, et al. Dyspnea secondary to pulmonary hematopoiesis as presenting symptom of myelofibrosis with myeloid metaplasia. Am J Hematol 2006;81:124–7.
6. Fielding JR, Owens M, Naimark A. Intrathoracic extramedullary hematopoiesis secondary to B12 and folate deficiency: CT appearance. J Comput Assist Tomogr 1991;15:308–10.
7. Krag D, Reich SB. Heterotopic bone marrow (myelolipoma) of the mediastinum. Chest 1972;61:514–5.
8. Georgiades CS, Neyman EG, Francis IR, Sneider MB, Fishman EK. Typical and atypical presentations of extramedullary hemopoiesis. AJR Am J Roentgenol 2002;179:1239–43.