Pulmonary Extramedullary Hematopoiesis Presented as Massive Hemoptysis, A Rare Entity: A Case Report

Masif Hemoptizi ile Beliren Ektrameddüller Hematopoez, Nadir Bir Antite: Bir Olgu Sunumu

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ABSTRACT

Extramedullary hematopoiesis occurs as a complication of hematological diseases such as myelofibrosis, sickle cell anemia and thalassemia. Extramedullary tissue usually consists of the liver, spleen and lymph nodes. Although it has been described to occur in almost every organ / region in the body, it is rarely reported in the thorax. Thoracic extramedullary hematopoiesis has been reported in the literature as paravertebral masses usually seen on chest radiography. In this report, we present a rare case of pulmonary extramedullary hematopoiesis in a 66-year-old male patient who did not have hematological or rheumatological comorbid disease and presented with hemoptysis.

Keywords: Extramedullary, hematopoiesis, hemoptysis, lung

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ÖZET

Ekstramedüller hematopoez miyelofibroz, orak hücreli anemi ve talasemi gibi hematolojik hastalıkların bir komplikasyonu olarak ortaya çıkar. Ekstramedüller doku genellikle karaciğer, dalak ve lenf nodlarından oluşur. Vücutta hemen hemen her organda/bölgede ortaya çıktığı anlatılmakla birlikte toraksta nadiren bildirilmektedir. Torasik ekstramedüller hematopoez literatürde genellikle akciğer grafisinde görülen paravertebral kitleler olarak bildirilmiştir. Bu yazıda hematolojik veya romatolojik ek hastalığı olmayan ve hemoptizi ile başvuran 66 yaşındaki erkek hastada nadir görülen pulmoner ekstramedüller hematopoez olgusunu sunuyoruz.

Anahtar Sözcükler: Ekstramedüller, hematopoez, hemoptizi, akciğer

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INTRODUCTION

Extramedullary hematopoiesis (EMH) is a rare condition and is most common in patients with myeloid metaplasia, myelofibrosis and chronic myeloproliferative disorders. Other underlying causes include hemoglobinopathies, such as thalassemia syndromes, sickle cell anemia, and other chronic hemolytic anemias (1). The most common locations of EMH are spleen and liver (2). It is rarely reported in the thorax. Here we reported a case of pulmonary EMH presented with massive hemoptysis.

CASE REPORT

A 66-years-old, male patient admitted to us with hemoptysis complaint. There was no considerable abnormality on physical examination and his initial laboratory findings were normal. He had a history of 50 packet / year smoking and coronary artery disease and percutaneous transluminal coronary angioplasty (PTCA). The ground glass density (possible alveolar hemorrhage) was observed in the right middle lobe and lower lobe basal segments on thorax computed tomography. Fiberoptic bronchoscopy was performed to find the source of bleeding and evaluate the presence of endobronchial lesion and it was detected that; the middle lobe bronchus was clogged with the clot. The patient was interned and medical treatment was started also, his antiaggregant was stopped. Trans-thoracic echocardiography was performed to exclude pulmonary hypertension and mitral valve disease and no abnormality was detected. There were no detected signs related with inflammation, mass, nodule, pneumonic infiltration and vascular pathologies. The patient was investigated for underlying rheumatological diseases but there were no physical examination and laboratory findings of its. Bronchial artery embolization was planned for control the bleeding. However, since active bleeding site could not be seen on angiography, embolization could not be performed. Surgical approach was planned because continuing sub-massive hemoptysis despite the medical treatment and VATS right middle lobectomy due to its congestion and destroyed nature and bronchial artery ligation procedure was performed. The patient's hemoptysis stopped and did not occur again in the postoperative period. Histopathologic examination was reported as pulmonary EMH (Figure 1). In the 6-month follow-up of the patient, physical examination and laboratory findings were evaluated as normal.

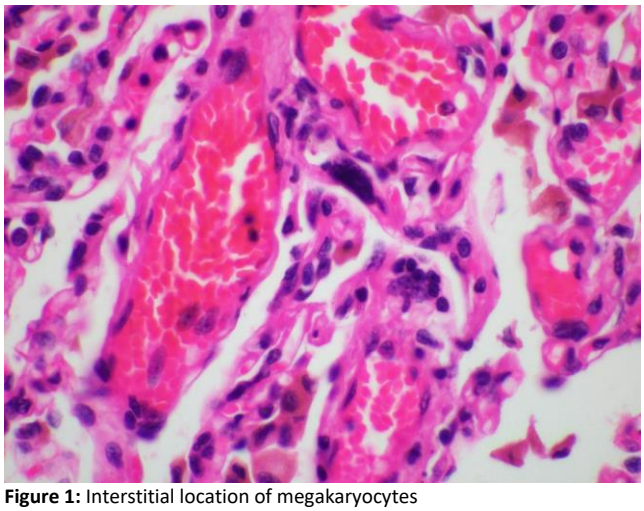

Figure 1: Interstitial location of megakaryocytes

DISCUSSION

Extramedullary hematopoiesis is usually a compensatory hematopoietic tissue expansion that includes the reticulo-endothelial system (1). Extramedullary hematopoiesis, which is normal in the fetal life, especially in the reticuloendothelial system, is considered abnormal after birth (2). EMH is most common in the liver and spleen, but has been documented in other organs such as mediastinum, central nervous system, lymph nodes, prostate and breast (2). Pulmonary EMH has been rarely reported.

Almost all of the reported cases occurred after myeloproliferative disorders, especially myelofibrosis. Other less common causes are thalassemia syndromes and other hemoglobinopathies. However Ghosh et al (3) reported a primary case of extramedullary hematopoiesis present with massive bilateral chylothorax without an underlying hematological disease. In addition, Sjögren's Syndrome and cases reported to be related to asthma have also been reported (4,5). Most pulmonary EMH masses are asymptomatic. However, sometimes patients may present with hemoptysis, acute or progressive dyspnea or chest pain. Rarely, life-threatening complications such as massive pleural effusion, hemothorax, chylothorax, or spinal compression (posterior mediastinal EMH) can occur (6). The most common radiographic finding in thoracic EMH is a well-confined, lobulated, paravertebral mass containing fat density without calcification. In the literature, cases presented in the form of bilateral pulmonary nodules, interstitial infiltrates, fibrosis or lung mass are presented. In general, the characteristics of EMH reported in ultrasound, CT and MR are quite variable and not sufficient even in the presence of clinical suspicion (5). It has also been reported that 18F-PET-CT and 99m Tc-SC scintigraphy are used as noninvasive methods for the diagnosis of EMH. Zade et al(7) used PET-CT and 99mTc-SC for diagnosis in a 55-year-old male patient with steroid sensitive hypoplastic anemia presenting with bilateral paravertebral mass. They detected increased FDG uptake in PET CT in the lesion area and diffuse FDG uptake in the bone marrow. Afterwards, they detected increased uptake in 99m Tc scintigraphy. Thus, they diagnosed with history, clinical findings and scintigraphic evaluation(7). The definitive diagnosis of EMH is made histopathologically. Usually, video assisted thoracoscopic surgery (VATS) or imaging guided fine needle aspiration provide tissue samples(8). Intrathoracic EMH treatments include regular follow-up, transfusion specifically for the treatment of concomitant hematological diseases and use of hydroxyurea, low-dose radiation therapy, surgical excision or combination of them (8). In our case, extramedullary hematopoiesis was not suspected preoperatively, because there was no evidence of hematological disorder in the patient. There was neither hepato-splenomegaly nor pleural, mediastinal mass on his computed tomography. The operation was planned for persistant hemoptysis. VATS right middle lobectomy and bronchial artery ligation procedure was performed. In the follow-ups made after postoperative diagnosise, no findings suggesting hematological disease were found. Moreover, no findings of hemotological disorders were detected in his laboratory studies performed after this histopathological diagnosis.

CONCLUSION

Pulmonary EMH should also be considered in the differential diagnosis in patients presenting with hemoptysis and the cause of bleeding.

Conflict of interest

No conflict of interest was declared by the authors.

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