Hemoptysis as a presentational finding of intrathoracic paravertebral mass of extramedullary erythropoiesis in a patient with β-thalassemia intermedia

Beta talasemi intermedia tanılı bir hastada ekstramedüller eritropoeze ikincil intratorasik paravertabral kitlelerin başvuru bulgusu olarak hemoptizi

Turan Bayhan[1], Şule Ünal[1], Tuncay Hazırolan[2], Fatma Gümrük[1]
[1]Hacettepe Üniversitesi Çocuk Hematolojisi Bölümü
[2]Hacettepe Üniversitesi Radyoloji Bölümü

Dergiye Ulaşma Tarihi: 21.09.2016 Dergiye Kabul Tarihi: 15.12.2016 Doi: 10.5505/aot.2017.88597

ÖZET

İntratorasik ekstramedüller eritropoez beta talasemi intermedianın nadir ama iyi bilinen bir komplikasyonudur. İntratorasik ekstramedüller eritropoezi olan hastalar genellikle asemptomatiktir. Burada beta talasemi intermedia tanısıyla izlenirken hemoptizi ile başvuran ve sonrasında intratorasik ekstramedüller eritropoez kitleleri tespit edilen bir erişkin vaka sunulmaktadır. Torakal ekstramedüller eritropoez kitleleri olan beta talasemi hastalarında hemoptizi ile başvuru daha öncesinde bildirilmemiştir. Hastanın eritrosit transfüzyon programına alınması ve hidroksiüre tedavisinin başlanması ile hem hemoptizi şikâyeti kaybolmuş hem de kitleleri küçülmüştür. Beta talasemi intermedia tanılı hastalarda hemoptizi intratorasik ekstramedüller eritropoezin bir bulgusu olarak akılda bulundurulmalıdır.

Anahtar Kelimeler: Beta talasemi intermedia, ekstramedüller eritropoez, hemoptizi

ABSTRACT

Intrathoracic extramedullary erythropoiesis (EME) is a rare but well-known complication of β-thalassemia intermedia. Patients with intrathoracic EME are generally asymptomatic. Herein, we report an adult patient with β-thalassemia intermedia who was diagnosed to have intrathoracic EME masses and presented with hemoptysis. Upto our knowledge, hemoptysis has not been reported previously as a presentational complaint among patients with β-thalassemia who have thoracal EME masses. Initiation of chronic transfusion programme and hydroxurea treatment caused regression in the masses, in addition to disappearance of the complaint of hemoptysis. Hemoptysis should be kept in mind as a rare finding of of EME in β-thalassemia intermedia.

Keywords: Beta thalassemia intermedia, extramedullary erythropoiesis, hemopthysis

Introduction

Extramedullary erythropoesis (EME) generally occurs secondary to conditions with high turn-over in bone marrow like chronic hemolysis and ineffective erhytropoesis. Extramedullary erythropoiesis develops as a compensatory mechanism to supply the increased demand for erythrocyte production. Patients with β-thalassemia major are treated with regular erythrocyte transfusions due to low levels of hemoglobin and their erhytropoeisis is suppressed. However, patients with β-thalassemia intermedia are morerisky to develop EME because of their lower needs for transfusions and unsupressed erythropoietic activity (1). The masses of EME are usually asymptomatic (2). Herein, we report an adult patient with β-thalassemia intermedia and intrathoracic paravertebral masses who presented to our outpatient clinic with mild to moderate hemoptysis as the sole complaint.

Case

A 36-year-old man with β-thalasemia intermedia presented with hemoptysis. He was diagnosed to have β-thalassemia intermedia at four years of age and underwent splenectomy at five years of age. Anti-arrhythmic treatment was initiated because of atrioventricular dissociation at 17 years of age. He didn't require any transfusion after splenectomy. The course was uncomplicated until the admission

with mild to moderate hemoptysis of approximately 100 ml of hemoptysis. Hemoptysis occurred spontaneously without dyspnea or chest pain. On physical examination he had no dyspnea, chest sounds were normal and no rales were heard at auscultation. Thoracal magnetic resonance imaging (MRI) revealed multiple paravertabral masses like with the greatest diameter of 11.4 cm (Figure 1A). Echocardiography revealed no pulmonary hypertention that could explain the hemoptysis. He was put on regular erythrocyte transfusion programme and additionally hydroxyurea was initiated in order to cause regression in the paravertebral mass of EME. Hemoptysis did not recur during the follow-up of seven months and the control MRI revealed decrease in dimensions of EME masses to 7x7.5 cm (Figure 1B).

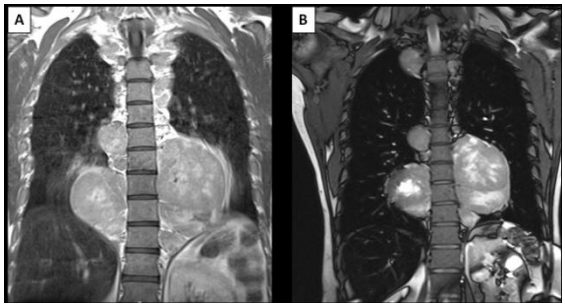

Figure1A-B

Discussion

Extramedullary erythropoiesis leads to development of erthropoietic masses commonly in liver, spleen and lymph nodes (3)**.** These masses can be detected in thorax rarely. Intrathoracic EME masses usually present in the lower paravertebral area with multiple and bilateral distribution. They have distinctive radiologic appearance and characteristic topography that make differential diagnosis easy (4). Besides, high vascularity of EME masses may end-up with hemorrhage subsequent to biopsy which encourages noninvasive diagnostic procedures, instead of interventional diagnostic procedures (2).

Patients with intrathoracic EME masses are usually asymptomatic. In some patients these masses can be detected incidentally (2, 5). However, pleural effusions, hemothorax, and neurologic deficiencies secondary to spinal cord compression have been reported (2, 3). In the review of the English literature, we found nine reports of intrathoracic EME masses presented with hemothorax development and of these reports six of the patients had thalassemia as the underlying cause of EME (6, 7). However, upto our knowledge, hemoptysis has not been reported as a presentational finding of thoracal EME mass.

In our patient the intrathoracic masses had no anatomical relationship with the pulmonary vasculature. EME develops from stem cells that extruded through the thin cortex of vertebral bodies and ribs (6), thus EME occurs out of lung parenchyma and vasculature. Due to high vascularity of EME masses they might cause hemothorax outside the lung parenchyma (6, 7). Interestingly, our patient presented with hemoptysis without hemothorax. We can explain pathophysiology of hemopthysis with compression of paranchymal small vessels by EME masses. Mild clinical presentation of hemopthysis also may be secondary to damage of paranchymal small vessels because directly hemorrhage from EME masses might have provoked more severe hemoptysis.

Treatment modalities for EME are surgery, regular transfusions, hydroxyurea, radiotherapy, or combination of them. Regular erythrocyte transfusion programme is the first line treatment of EME (5). Radiotherapy can be chosen in conditions when rapid response is needed like spinal cord compressions with considering its toxic effects. Surgical removal of masses also should be reserved for acute and progressive situations because of risk for hemorrhage (6). Hydroxyurea is a well- known myelosuppressive agent that has been used in patients with sickle cell anemia and β-thalassemia intermedia/major. Effectiveness of hydroxyurea has been shown in regression of masses of EME in β-thalassemia intermedia (5).

In our patient there was an uncomplicated course and regression of the masses was achieved with the introduction of the combined use of regular transfusions and hydroxyurea.

Conclusion

Hemoptysis may be a rare presentational finding of thoracal EME masses in patients with β-thalassemia intermedia and hydroxyurae plus regular transfusions may be used successfully in order to regress the masses.

Disclosure

There is no any interest to disclose.

References

1. Smith PR, Manjoney DL, Teitcher JB, Choi KN, Braverman AS. Massive hemothorax due to intrathoracic extramedullary hematopoiesis in a patient with thalassemia intermedia. Chest 1988; 94: 658-660.
2. Castelli R, Graziadei G, Karimi M, Cappellini MD. Intrathoracic masses due to extramedullary hematopoiesis. Am J Med Sci 2004; 328: 299-303.
3. Cappellini MD, Musallam KM, Taher AT. Insight onto the pathophysiology and clinical complications of thalassemia intermedia. Hemoglobin 2009; 33 Suppl 1: S145-159.
4. Tsitouridis J, Stamos S, Hassapopoulou E, Tsitouridis K, Nikolopoulos P. Extramedullary paraspinal hematopoiesis in thalassemia: CT and MRI evaluation. Eur J Radiol 1999; 30: 33-38.
5. Meo A, Cassinerio E, Castelli R, Bignamini D, Perego L, Cappellini MD. Effect of hydroxyurea on extramedullary haematopoiesis in thalassaemia intermedia: case reports and literature review. Int J Lab Hematol 2008; 30: 425-431.
6. Pornsuriyasak P, Suwatanapongched T, Wangsuppasawad N, Ngodngamthaweesuk M, Angchaisuksiri P. Massive hemothorax in a beta-thalassemic patient due to spontaneous rupture of extramedullary hematopoietic masses: diagnosis and successful treatment. Respir Care 2006; 51: 272-276.
7. Tantraworasin A, Saeteng S. Massive hemothorax due to intrathoracic extramedullary hematopoiesis in a patient with beta thalassemia hemoglobin E disease. J Med Assoc Thai 2013; 96: 866-869.

Adress for correspondence: Uzm. Dr. Turan Bayhan, Hacettepe Üniversitesi İhsan Doğramacı Çocuk Hastanesi Çocuk Hematolojisi Bölümü Ankara - Türkiye
e-mail: **turanbayhan@yahoo.com**
Available at www.actaoncologicaturcica.com