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Paravertebral Masses Due To Extramedullary Hematopoıesıs (EMH) in A Subject With Beta Thalassemia

Beta Talasemili Olguda Ekstramedüller Hematopoeze Bağlı Paravertebral Kitleler
Hematoloji

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Özet

Talasemi inefektif eritropoeze bağlı olarak ekstramedüller hematopoez(EMH)oluşumu ile seyreden bir gurup kalıtsal kan hastalığıdır. EMH'e bağlı oluşan paravertebral kitleler ağrı , paresteziden total paraliziye kadar uzanan geniş bir klinik spektrumla karşımıza çıkabilmektedir. Tedavide kan transfüzyonu, radyoterapi ve cerrahi tedavi (laminektomi, hemilaminektomi) ve çeşitli medikal ajanlar kullanlmaktadır. Sırt ağrısı ile başvuran olgumuzda paravertebral kitlelerin klinik yansımasını, kitlelerin radyolojik görünümlerini ,hastalığın patofizyolojik gelişimi ile birlikte sunmayı amaçladık.

Anahtar kelimeler: *Talasemi, Paravertebral kitleler Ekstramedüller hematopez MR görüntüleme*

Abstract

Thalassemias are a group of inherited anemias in which extramedullary hematopoiesis associated with ineffective erythropoiesis can be found as a complication of the disease paravertebral masses may be confronted within a broad spectrum extending from paresthesia to total paralysis. Blood transfusion, radiotherapy and surgical therapy (laminectomy, hemilaminectomy), and various medical agents can be used in its treatment. In our case presented with back pain, we aimed to present radiological images of masses formed by the focuses of paravertebral extramedullary hematopoiesis together with the pathophysiologic development of the disease.

Keywords: *Thalassemia, Paravertebral masses Extramedullary haematopoeiesis MRI imaging*

Introduction

Thalassemias are a heterogeneous group of inherited anemias characterized by defects in the synthesis of one or more of the globulin chain subunits of the adult hemoglobin tetramer hemoglobin A. In contrast to its major forms, its intermedia forms manifest themselves in later stages of life and rarely require transfusion. Over the following years, it appears as storage of iron in parenchymal tissues due to transfusion and as focuses of extramedullary hematopoiesis associated with ineffective erythropoiesis. Here, we will discuss a twenty-seven year old thalassemia patient with multiple paraspinal extramedullary hematopoiesis.

Case Report

A twenty-seven year old male patient was admitted with back pain being aggravated by movement. In addition, he had pallor and subicterus. His medical history revealed multiple blood transfusions from age two to age fourteen considering his diagnosis of heterozygous beta thalassemia. During physical examination, the palpation along the thoracolumbar vertebra trace revealed increasing sharp pain radiating to the sides bilateraly on anteflexion. His abdominal palpation revealed hepatomegaly and mild splenomegaly with patient claimed to have vague abdominal discomfort. The rest of his medical examination findings was normal.

Laboratory results revealed hemoglobin levels of 7 gr/dl, hematocrite levels of 24%, mean corpuscular volume

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(MCV) of 64 fL, platelet levels of 370,000 and indirect bilirubin levels of 4 mg/dl. A chest x-ray was perfomed, showing opacities, one on the right side measuring about 10 cm in diameter and extending towards the mediastinum over pulmonary hilus, the other one was on the left side being superposed with the pulmonary hilus on medial border , indistinguishable from the contour of the vertebral column (Figure 1).

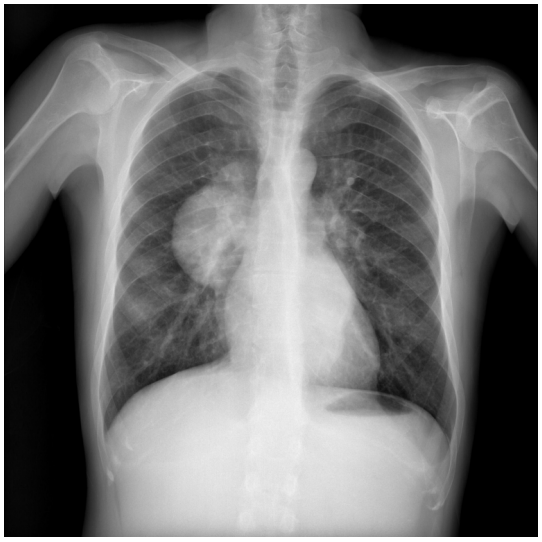

Figure 1
Chest x-ray shows homogenous round opasity superimposed on pulmonary right hilum . Note that the hilar pulmonary vasculature is visualized through the mass. On left side mediastinal mass with lobulated borders is apparent also which overlaps with the cardiac shadow.

In order to diagnose these masses an MRI was performed . MRI revealed showing apparent homogenous contrasting. Paravertebral masses along the thoracic vertebra were iso-, hyperintense on T1-W and hypointense on T2-W images in comparison with the muscle signal. There were apparent homogenous contrast enhancement of masses (Figure 4a, 4b).

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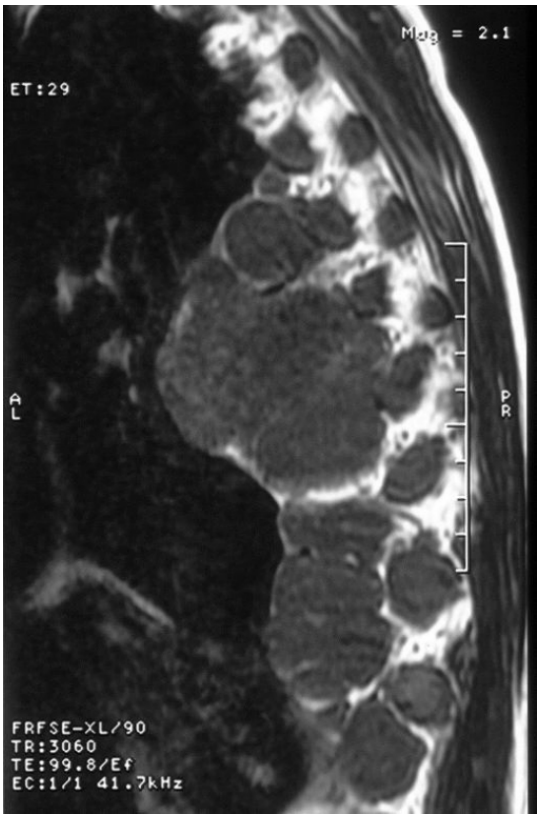

Figure 4A

4a,b,c,. In pre- and postcontrast T1W, T2W sagittal images and coronal T2W image nodular homogenous masses arelocated close to intercostal space and costovertebral junction.

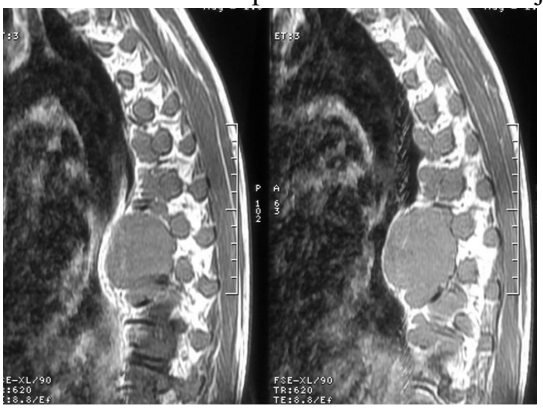

Figure 4B

4a,b,c,. In pre- and postcontrast T1W, T2W sagittal images and coronal T2W image nodular homogenous masses arelocated close to intercostal space and costovertebral junction.

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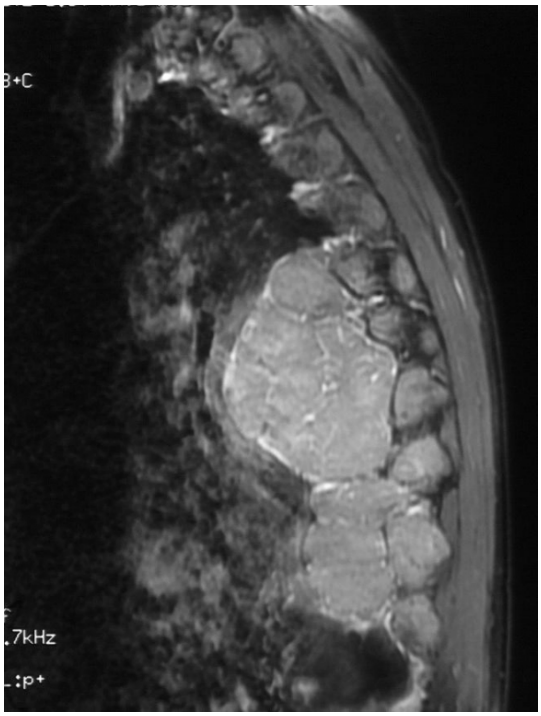

Figure 4C

4a,b,c,. In pre- and postcontrast T1W, T2W sagittal images and coronal T2W image nodular homogenous masses arelocated close to intercostal space and costovertebral junction.

The masses were shown to lean on the verterbra, protruding towards neural foramina without reaching into them. Hence, the masses did not reveal any sign of neural compression nor any vertebral or costal invasion (Fig. 5). In the light of these characteristic MRI findings and the result of the histopathological biopsy from patient's thoracovertebral region performed one year ago, the masses were confirmed to be *extramedullary hematopoiesis foci.*

After the diagnosis, several blood transfusions were administered to the patient and there was a decrease in patient's complaints. Even though the patient's presenting symptoms were kept under control, after four weeks of treatment, there wasn't any remission of the extramedullary hematopoiesis foci.

Discussion

Thalassemias are a heterogeneous group of inherited anemia characterized by defects in the synthesis of one or more of the globulin chain subunits of hemoglobin tetramer. Clinical syndromes in thalassemia manifest along with inadequate hemoglobin production and unbalanced accumulation of hemoglobin subunits. While inadequate hemoglobin production causes hypochromia and microcytosis, abnormal globulin accumulation causes hemolysis and ineffective erythropoiesis. According to the type of the underlying genetic defect, symptoms can be mild or very severe.

Extramedullary hematopoiesis is the production of normal blood cells out of the bone morrow. It appears as

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compensatory response in chronic hemolytic cases. It is encountered mostly in liver, spleen, lymph nodes, and less frequently in other tissues (kidney, adrenal glands, thymus, breast, heart, epididymis and peritoneum).

EMH in thalassemia was first described in 1954 by Gatto [1]. EMH is seen in hematological diseases such as thalassemia, polistemia vera, Myleofibrozis and hemolytic anemia. It is seen less frequently in school age or adolescent period possibly due to better bone quality. Recurrent traumas and surgical interventions trigger EMH. Splenectomy increases the risk [2].

Paraspinal extramedullary hematopoiesis is seen frequently in thalassemias. Although intraspinal formation is seen in more than 11% beta thalassemias, clinical signs due to compression are rarely seen [3]. No spinal cord compression was seen in a series in which 138 thalassemia patients were studied [4]. Generally, it has tendency of involving middle and lower thoracic vertebrae, leading to movement restriction [5]. It usually manifests as paraparesis, loss of sense and sphincter disorder. Complete paraparesis is rarely seen.

For diagnosis, MRI is the first choice, showing isointensity to spinal cord on T1-weighted (T1W) sequences, and hiperintensity on T2-weighted (T2W) sequences [6]. CT myelography is the ideal diagnostic tool in detecting the localization and size of epidural masses [7]. Most authors do not recommend tissue biopsy [8].

Treatment options include surgery, radiotherapy, blood transfusion and hydroxyurea. In many cases, it has been reported that when paraplegia was developed, radiation therapy was not preferred as the choice of treatment because of edema, only surgical decompression (laminectomy, hemilaminectomy) was performed. However, since hematopoietic tissues are highly sensitive to radiation, and since concurrent steroid therapy can easily prevent possible edema, radiotherapy is a proper treatment alternative [9] 10--30- Gy is considered as effective dose [10]. Although a nearly complete regression is seen after radiotherapy, a 19% of recurrence has been reported [6,10].

In extramedullary hematopoiesis, treating the anemia by blood transfusion leads to inhibiton of erythropoietin release and to a partial shrinkage in focuses of hematopoiesis. However, this is generally a transient improvement [11] and this is usually recommended in mild extramedullary hematopoiesis cases today [12] or may be implemented in the condition of pregnancy for the purpose of delaying surgical intervention [13].

Some authors have reported beneficial outcomes in using hydroxyurea in combination with transfusion [14]. Hydroxyurea is an agent that has been used as a cytotoxic agent in myeloproliperative diseases. It has a positive effect on erythropoiesis by stimulating fetal hemoglobin production. The possibility of acute hepatoxicity and myelotoxicity development during treatment and triggering leukemic transformation in long-term restricts its use [15].

In conclusion; EMH is seen as compensatory response in patients with hemoglobinopathy. Although paraspinal masses associated with EMH are frequently encountered in thalassemias, neurologic symptoms are rare. MRI is the basic diagnostic tool. Optimal treatment approach varies according to the localization and size of the mass, and to its clinical acute status and severity.

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Information Presentation

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