Journal of Taibah University Medical Sciences (2018) 13(4), 395–397

Taibah University

Journal of Taibah University Medical Sciences

www.sciencedirect.com

Case Report

Lung masses in a thalassemia patient: A diagnostic dilemma

Rudrajit Paul, MRCP * and Biplab K. Gayen, MD

Department of Medicine, Midnapore Medical College, West Bengal, India

Received 11 December 2017; revised 9 February 2018; accepted 11 February 2018; Available online 13 March 2018

الملخص

الثلاسيميا هي فقر الدم الإنحلالي الخلقي الشائع في جنوب شبه القارة الآسيوية. ويرتبط المرض بالعديد من المضاعفات، التي تتراكم كلما تقدم عمر المريض. ويعتبر تكوّن الدم خارج النقي آلية فسيولوجية تعويضية لفقر الدم المزمن. يمكن أن يحدث تكوّن الدم خارج النقي في أجزاء مختلفة من الجسم ويؤدي إلى صفات تصويرية متنوعة، قد يكون بعضها مضللا. نشرح هنا حالة مريض ثلاسيميا ذكر عمره ٢٢ عاما أظهرت الأشعة السينية للصدر وجود ورم مفصص مجاور-للقلب. وكان التفكير حول أنه ورم في الرئة، ولكن أظهرت الأشعة المقطعية للصدر لاحقا صفات تقليدية لتكوّن الدم خارج النقي شملت الفقرات، والأضلاع والقص. تم الشرح للمريض عما ظهر بأشعة الصدر السينية، والصفات التصويرية لتكوّن الدم خارج النقي في الصدر بالتفصيل.

الكلمات المفتاحية: الأشعة المقطعية؛ تكون الدم خارج النقي؛ الثلاسيميا؛ ورم صدري

Abstract

Thalassemia is a common congenital hemolytic anemia in the South Asian subcontinent. This disease is associated with multiple complications, which accumulate as the age of the patient progresses. Extramedullary hematopoiesis (EMH) is a compensatory physiological mechanism for chronic anemia. EMH can occur in different parts of the body and present with varied imaging features, some of which may be misleading. We here describe the case of a 22-year-old male patient with thalassemia who presented with paracardiac lobulated masses on chest X-ray. They were thought to be lung tumors; however, subsequent computed tomography scan of the thorax showed typical features of EMH involving the vertebrae, ribs and sternum. The patient was assured about the appearance of the masses on the chest X-ray. Imaging features of EMH in the thorax has been discussed in detail.

Keywords: CT scan; Extramedullary hematopoiesis; Thalassemia; Thoracic mass

Introduction

Thalassemia, a form of congenital hemolytic anemia due to faulty globin chain synthesis, is the most common congenital hemolytic anemia in India. Common presentations of thalassemia depend on the degree of severity of the genetic defect. In very mild cases, the patient may be completely asymptomatic with abnormalities only detected by blood tests.[1] In moderate and severe cases, common presentations include repeated episodes of anemia, icterus, protuberant abdomen due to hepatosplenomegaly and stunted growth. Skeletal deformities (detailed later) and typical abnormal facies are also found. Thalassemia is associated with many complications.

The complications can be due to repeated transfusions (including iron overload from repeated infusion of packed red blood cells (RBC)), extramedullary hematopoiesis, thrombophilia or affection of specific systems such as liver and synovial joints.[1] As the life span of thalassemia patients is increasing, such long-term complications are becoming even more visible.

Skeletal changes in thalassemia patients can take different forms. Common skeletal pathologies in thalassemia include generalized osteoporosis, high frequency of fractures, stunted growth, limb-length discrepancy, spinal deformities and

* Corresponding address: 15/5 Bose Pukur Road, Kolkata, 700039, West Bengal, India.
E-mail: docr89@gmail.com (R. Paul)

Peer review under responsibility of Taibah University.

ELSEVIER Production and hosting by Elsevier

 https://doi.org/10.1016/j.jtumed.2018.02.005

thalassemic osteoarthropathy. Extramedullary hematopoiesis can cause striking bone changes.[2] Furthermore, defective epiphyseal closure is common in these patients. Since thalassemia is a common disease in India, physicians should be aware of the common X-ray imaging features of the disease, so as to avoid unnecessary investigations and diagnostic traps. We here present such an imaging feature of thalassemia.

The case report

A 22-year-old male, a known case of thalassemia major, presented to the general clinic with cough for five days. As part of his evaluation, a chest X-ray was performed, which showed several lobulated masses in the paracardiac region on both sides (Figure 1). He was referred to our medical outpatient clinic as a possible case of lung tumor.

In our outpatient clinic, on examination, the patient was found to have severe pallor, massive splenomegaly and moderate hepatomegaly. His family members said that he needed frequent blood transfusion. However, his particular blood group was often scarce in the nearby rural hospital and suitable donors could not be arranged. Hence, he often missed his regular transfusion schedule. His cough had subsided in the meantime. He was dyspneic and had mild pedal edema. There was no bone tenderness over the vertebrae. Clinical examination of the chest was normal, except for the presence of sternal tenderness. The patient stated that he previously needed one unit of packed RBC transfusion per month. However, for the last two years, his transfusion requirement had increased and he presently needed one unit per week. He was not on iron chelation therapy as he could not afford it.

Laboratory tests revealed a hemoglobin level of 3.6 g/dL. His serum bilirubin was 3.9 mg/dL (indirect 3.4). He was immediately given blood transfusion to relieve his dyspnea. For the chest masses, a contrast-enhanced (CE) CT scan of the thorax was performed. It revealed lobulated soft tissue masses along the anterior and posterior ends of the ribs and also around the transverse process of the vertebral bodies (Figure 2). Lytic areas were also found in the ribs and sternum. There were scattered soft tissue masses, enhanced slightly with contrast, around vertebral bodies. This radiological appearance is typical of extramedullary hematopoiesis in the axial skeleton, as found in congenital hemolytic anemia. The lung "masses" as seen on the chest X-ray were thus masses of hematopoietic tissue arising from bones. The patient was reassured regarding the appearance of his chest X-ray and referred to the hematology department of a higher center.

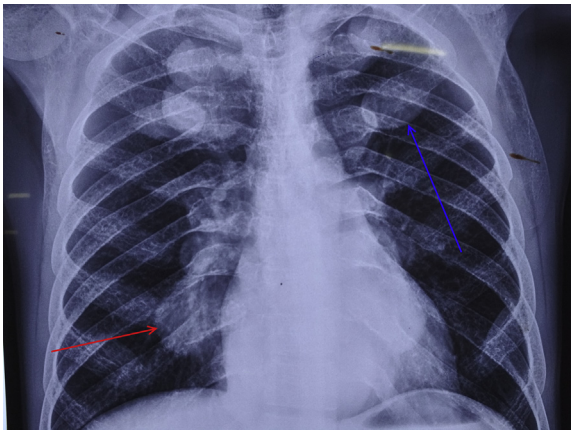

Figure 1: Chest X-ray showing paracardiac lobulated masses not silhouetting the cardiac border (red arrow) and expansile anterior ends of the ribs (blue arrow).

Discussion

In thalassemia, extramedullary hematopoiesis (EMH) occurs due to ineffective production of RBCs. The marrow of the axial skeleton expands in order to compensate for the ineffective erythropoiesis. Thus, the areas of axial bones with active marrow will expand and the marrow tissue often bursts through the cortex. Furthermore, emboli of hematopoietic tissue from the spleen may sometimes lodge in the thorax and give rise to centers of EMH.[3] The extent of EMH depends on the severity of anemia.

The most common sites of EMH are the liver and spleen. However, the skeletal structures are also commonly involved.[2] Radiological appearances of skeletal EMH sites are varied. On chest X-ray, the usual appearance is lobulated retrocardiac or paracardiac masses.[3] These masses are in the posterior mediastinum and do not silhouette the cardiac border.[3] The appearance was similar in our patient as well. Sometimes there are expansile lesions of the ribs, either along the entire length or in its anterior and posterior ends.[2,3] These EMH sites are usually asymptomatic and diagnosed incidentally during imaging for other purposes.

On CT scan, the "masses" are better characterized. They are usually lobulated, smoothly marginated, bilateral soft tissue masses which may or may not be enhanced with contrast.[3,4] They usually do not have mass effect.[4] However, rarely, the EMH tissue can compress the spinal cord, leading to paraplegia.[5] The masses are usually multiple and involve all thoracic vertebrae, especially the lower ones.[4] Rarely, the EMH may occur entirely in the lung parenchyma, appearing as scattered nodules on CT scan.[6]

MRI scans can also be used to image these EMH masses. Since the EMH tissue contains iron, T1 and T2 relaxation times can shorten.[5] The MRI appearances of the EMH masses are distinctive. They show intermediate signal intensity in both T1 and T2 sequences.[7] Gadolinium enhancement is minimal, differentiating the masses of EMH from metastases or abscesses.[7] However, older lesions can show either high or low signal intensity, depending on the amount of fatty infiltration or iron deposition, respectively.[7] The MRI appearance may also change with blood transfusion. For skeletal masses, usually CT scan is adequate for diagnosis. However, for EMH sites in the liver or spleen, MRI is the imaging of choice.[8]

EMH in thalassemia can be an incidental finding; however, it can sometimes present with symptoms such as paraplegia.[9] At times they can simulate retrocardiac neural tumors.[10] A case similar to ours was reported from Israel where the patient had thoracic EMH masses.[11] However, the skeletal EMH masses in their case were much more florid, with enlarged ribs almost obliterating the lung fields on the X-ray.[11] In our case, the EMH masses were confined to a few areas only.

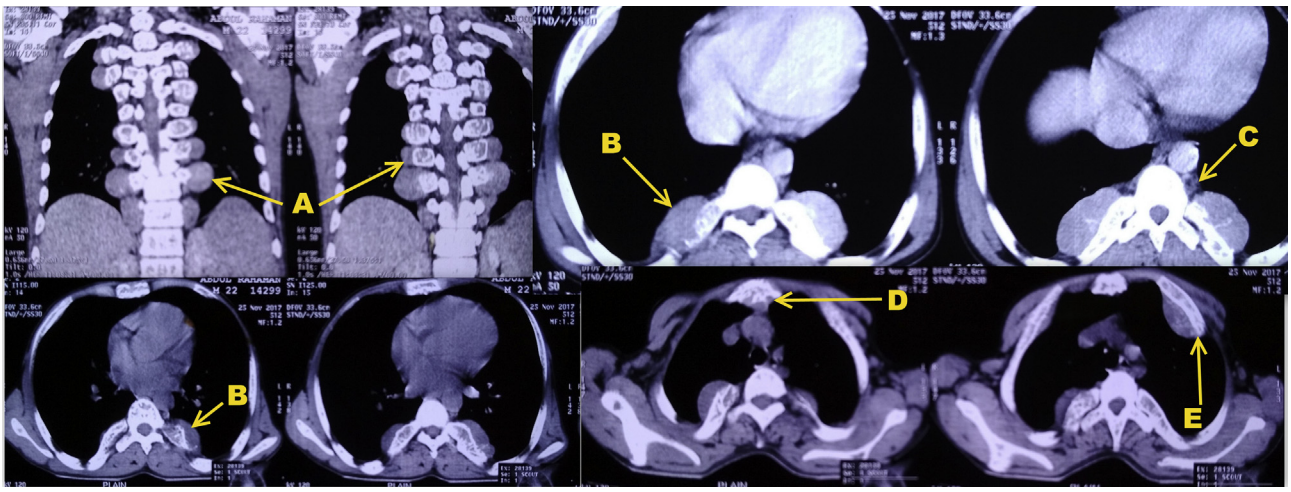

Figure 2: Computed tomography scan of the thorax showing extramedullary hematopoiesis (EMH) tissue in the transverse process of the vertebrae (A), EMH tissue around the posterior end of the ribs with lytic expansile lesions (B), masses of EMH tissue around the aorta enhanced slightly with contrast (C), expansile lesion of the sternum (D) and EMH at the anterior end of the ribs (E).

Fine needle aspiration cytology or percutaneous biopsy should never be attempted in these "masses" as there are highly friable and vascular and can cause severe bleeding.[3] Thus, a lung mass in a thalassemia patient must be investigated thoroughly by imaging before attempting diagnostic interventions. Common differential diagnoses of these masses are neurogenic tumors, paravertebral abscesses, metastatic tumors and lymph nodes.[3]

EMH usually needs no therapy. However, if there is a serious complication such as paraplegia, it may be treated successfully by radiotherapy, surgery or a combination of these two.[12]

We present this case to sensitize clinicians about this imaging appearance in thalassemia patients. Such masses on chest X-ray are mostly innocuous and patients should be assured about their nature. Furthermore, unnecessary interventions such as biopsy of these masses should be avoided.

Source of funding

None.

Conflict of interest

The authors have no conflict of interest to declare.

Consent

The patient consented.

Authors' contributions

RP and BG were both involved in clinical care of the patient; RP was responsible for data collection and writing of the manuscript. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

References

1. Borgna-Pignatti C, Gamberini MR. Complications of thalassemia major and their treatment. **Expert Rev Hematol 2011**; 4: 353–366.
2. Cullu N, Deveer M, Yeniceri O, Kalemci S. Changes in the skeletal system and extramedullary hematopoiesis in a patient with thalassemia. **Quant Imag Med Surg 2015**; 5: 626–627.
3. Marchiori E, Escuissato DL, Irion KL, Zanetti G, Rodrigues RS, de Souza G, et al. Extramedullary hematopoiesis: findings on computed tomography scans of the chest in 6 patients. **J Bras Pneumol 2008**; 34: 812–816.
4. Kwak HS, Lee JM. CT findings of extramedullary hematopoiesis in the thorax, liver and kidneys, in a patient with idiopathic myelofibrosis. **J Kor Med Sci 2000**; 15: 460–462.
5. Hashmi MA, Guha S, Sengupta P, Basu D, Baboo S. Neha. Thoracic cord compression by extramedullary hematopoiesis in thalassemia. **Asian J Neurosurg 2014**; 9: 102–104.
6. Chunduri S, Gaitonde S, Ciurea SO, Hoffman R, Rondelli D. Pulmonary extramedullary hematopoiesis in patients with myelofibrosis undergoing allogeneic stem cell transplantation. **Haematologica 2008**; 93: 1593–1595.
7. Haidar R, Mhaidli H, Taher AT. Paraspinal extramedullary hematopoiesis in patients with thalassemia intermedia. **Eur Spine J 2010**; 19: 871–878.
8. Albrizio M, Peroni C, Reduzzi C, Piazza V, la Fianza A. *Unusual extramedullary hematopoiesis sites shown by CT and MRI-scan*. Poster: C-069; ECR 2009.
9. Bukhari SS, Junaid M, Rashid MU. Thalassemia, extramedullary hematopoiesis, and spinal cord compression: a case report. **Surg Neurol Int 2016**; 7: S148–S152.
10. Hassanzadeh M. Extramedullary hematopoiesis in thalassemia. **N Engl J Med 2013**; 369: 1252.
11. Kalchiem-Dekel O, Greenbaum U. Extramedullary hematopoiesis in β-thalassemia. **Mayo Clin Proc 2015**; 90: 1591–1592.
12. Malik M, Pillai LS, Gogia N, Puri T, Mahapatra M, Sharma DN, et al. Paraplegia due to extramedullary hematopoiesis in thalassemia treated successfully with radiation therapy. **Haematologica 2007**; 92: e28–e30.

How to cite this article: Paul R, Gayen BK. Lung masses in a thalassemia patient: A diagnostic dilemma. J Taibah Univ Med Sc 2018;13(4):395–397.