

A Case of Thalassemia Intermedia with Extramedullar Hematopoiesis Finging on FDG-PET CT

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Abstract

Extramedullary hematopoiesis (EMH) is a physiological compensatory event that develops in hematological diseases characterized by hemolysis such as thalassemia, sickle cell anemia, myelofibrosis, hereditary spherocytosis, as a result of the insufficiency of the bone marrow in meeting the need for circulation. Intrathoracic extramedullary hematopoiesis is a rare condition. In this study, we present a 49-year-old male patient who was followed up by the hematology clinic due to thalassemia intermedia, and who underwent PET-CT examination for metabolic characterization after detecting a mass located in the paravertebral area in the posterior mediastinum.

Keywords: FDG, PET/CT, intrathoracic extramedullary hematopoiesis, thalassemi intermedia.

Introduction

Extramedullary hematopoiesis is a physiological compensatory response that develops as a result of the insufficiency of the bone marrow in meeting the need for circulation (1). Em is more common in patients with chronic hemolytic anemia, especially in Thalassemia intermedia patients who do not require transfusion (1,2). It can be seen in almost all organs, especially the spleen, liver, lymph nodes, thymus, heart, breast tissue, prostate, wide ligaments, kidneys, adrenal glands, pleura, retroperitoneal tissues, skin, peripheral or cranial nerves, and spinal canal (3-7). These regions are thought to have an active hematopoiesis role in the fetus during gestation. While the frequency of extramedullary hematopoiesis is around 1% in thalassemia major patients, it can reach up to 20% in thalassemia intermedia patients (1,8,9). Extramedullary hematopoiesis is found in paraspinal localization in 11-15% of cases (2,10). Paraspinal extramedullary hematopoiesis presents as pseudotumors and may cause spinal compression, leading to various neurological findings. However, more than 80% of the patients are asymptomatic and are detected incidentally during radiological evaluations. The male:female ratio is 5:1 (11). Patients may present with various complaints and findings such as back pain, leg pain, paraesthesia, impaired proprioception, exaggerated or brisk deep tendon reflexes, Babinski response, Laseque sign, paraparesis, paraplegia, clonus, spastic gait, "urgency" sign and fecal incontinence (12).

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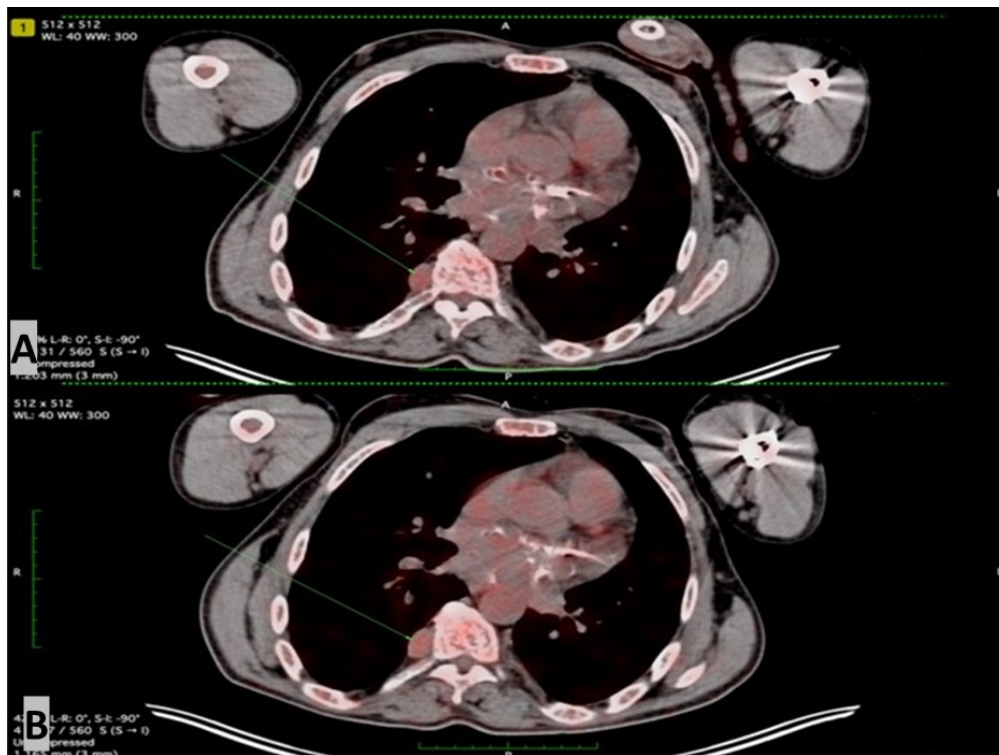
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To date, only a few cases regarding the diagnosis of EMH using PET/CT have been reported with EMH detected as a benign mass with low SUV max values and normal appearance of the tissue. The concurrent presence of an underlying hematopoietic disorder may suggest a diagnosis of EMH (13). In another study on this subject, FDG-PET/CT showed medullary expansion with soft tissue lesions medial to the ribs, paravertebral spaces, and the presacral area. All lesions were with mild to moderate FDG uptake (14).

Case Report

A 49-year-old male patient who had no active complaints and was diagnosed with thalassemia intermedia and who underwent intermittent blood transfusion was detected to have increased opacity in the right hilar region on the PA chest radiograph. In the CT examination, a smooth-circumscribed, ovoid-shaped, soft tissue density lesion of 25x15 mm in size, located extraparenchymal in the right paravertebral area, was detected. In the PET-CT examination performed for the metabolic characterization of this lesion, a well-circumscribed soft tissue lesion was visualized on the right anterolateral side of the T9 vertebra, which showed low FDG affinity and could be compatible with extramedullary hematopoiesis in Figure B. In the control PET-CT examination, the lesion remains stable (SUVmax: 3.61>>>2.93) in Figure A.



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Authorship Contributions

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