A Curious Case of Suspicious Lymphadenopathy in a Hereditary Spherocytosis Patient Reported as Extramedullary Hematopoiesis

Abstract

We present a case of a 23-year-old male patient with complaints of fever, cough, and persistent anemia for the past 6 months and with a known history of hereditary spherocytosis. Computed tomography (CT) thorax demonstrated multiple paravertebral lesions in the bilateral thoracic cavities, suggestive of lymphadenopathy; subsequently, ^{18}Flurodeoxyglucose PET/CT was done with suspicion of lymphoma, which showed no significant metabolic activity in those lesions. Thus, in view of clinical and metabolic status, lesions were considered extramedullary hematopoiesis (EMH). This case highlights the importance of considering EMH, while interpreting suspicious lymphadenopathy in cases of chronic anemia and also possible scan findings in the same.

Keywords: *Anemia, extramedullary hematopoiesis, FDG PET/computed tomography, lymphoma*

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A 23-year-old male patient presented with fever, cough, and persistent anemia for the past 6 months not subsiding with medication. On clinical examination, he had kyphoscoliosis and evidence of anemia signs. He was a known case of hereditary spherocytosis (with splenectomy done 10 years back) and type-1 diabetes mellitus (on insulin therapy). Computed tomography (CT) scan of the thorax showed multiple paravertebral lesions in bilateral thoracic cavities suggestive of lymphadenopathy [Figure 1a and b]. With a suspicion of lymphoma, the patient underwent ^{18}F-FDG PET/CT, which showed multiple lesions in the bilateral paravertebral region from D7 to D10 vertebrae (largest measuring 3 cm × 2.6 cm × 3.2 cm), with no significant metabolic activity [Figure 1c-e]. In view of the metabolic status and clinical history, lesions were considered extramedullary hematopoiesis (EMH).

EMH refers to the production of blood cells outside the bone marrow, commonly in liver, spleen, kidney, and lymph nodes, while intrathoracic cavity is less frequently involved. It is commonly seen in myelofibrosis (most common), leukemia, sickle cell disease, diffuse osseous metastatic disease replacing the bone marrow, and thalassemia.[1,2] The most commonly used diagnostic methods for EMH are magnetic resonance imaging and CT scans.[3,4] On CT, EMH typically appears as heterogeneous, hypovascular soft tissue masses, often interspersed with areas of fat attenuation.[4] In PET/CT, EMH lesions demonstrate mild-to-moderate metabolic activity when compared with malignant lesions, SUVmax value is generally low, and the tissue appears normal. Underlying hematopoietic disorder may suggest EMH, and a sampling of this tissue confirms the diagnosis.[5] However, risk of hemorrhage cannot be avoided during sampling, particularly in EMH, as it is an invasive procedure. An adequate noninvasive workup may be clinically useful in the evaluation of the paravertebral masses.[6] There have been only limited data records of PET/CT detecting EMH; all of these cases were in patients with different type of malignant diseases and characteristically exhibited mild FDG avidity (low SUVmax) compared to the malignancies with high FDG uptake.[5,7-10] Hence, EMH has to be considered as differential while reporting paraveretebral masses in patients with a history of chronic anemia or myeloproliferative disorders. Our case demonstrated incidental detection of EMH in a chronic anemia patient while investigating for suspicious lymphadenopathy on anatomic imaging.

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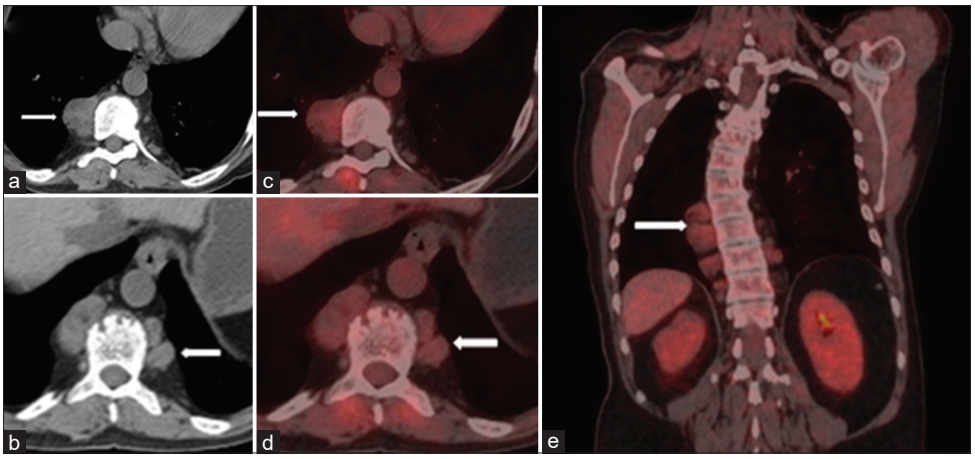

Figure 1: (a and b) With a suspicion of lymphoma, the patient underwent ^{18}F-FDG PET/computed tomography, which showed multiple lesions in bilateral paravertebral region from D7 to D10 vertebrae (largest measuring 3 cm × 2.6 cm × 3.2 cm) with no significant metabolic activity (c-e). In view of the metabolic status and clinical history, lesions were considered extramedullary hematopoiesis

These patients are usually asymptomatic but can cause symptoms on compression of neighboring structures, in which case radiation therapy or surgical interventions are required; otherwise, EMH usually regresses after treatment with blood transfusions and hydroxyurea.[11]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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