An Unusual Presentation of Richter's Transformation of Chronic Lymphocytic Leukemia in Liver and Lung on ^{18}F-Labeled Fluoro-2-Deoxyglucose Positron Emission Tomography/Computed Tomography

Abstract

Richter's transformation (RT) is a rare complication of chronic lymphocytic leukemia (CLL) or small lymphocytic lymphoma (SLL) with unfavorable prognosis. The clinical and laboratory findings of RT are nonspecific and requires histopathological confirmation for the diagnosis. ^{18}F-labeled fluoro-2-deoxyglucose positron-emission tomography/computed tomography (^{18}F-FDG PET/CT) has shown higher diagnostic values for the detection of transformation of CLL/SLL to aggressive lymphoma. We present a case of CLL in remission for 6 years presenting with clinical features suggestive of RT. ^{18}F-FDG PET/CT done in our case showed liver and lung involvement with no lymphadenopathy, which is an unusual presentation of RT.

Keywords: *^{18}F-labeled fluoro-2-deoxyglucose positron emission tomography/computed tomography, chronic lymphocytic leukemia, liver and lung, Richter's transformation*

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A 67-year-old male, known case of chronic lymphocytic leukemia (CLL) in remission for 6 years, presented with fever and loss of weight. On physical examination, apart from fever, he had hepatosplenomegaly. Routine laboratory examination revealed low hemoglobin: 9 g/dL (12–15.5 g/dL), increased white blood cell counts: 13 × 10 9/L (4.5–11 × 10 9/L), and raised serum lactate dehydrogenase levels 312 U/L (100–190 U/L). In view of clinical suspicion of Richter's transformation (RT), he underwent whole-body ^{18}F-labeled fluoro-2-deoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) to guide the site of biopsy [Figure 1]. PET/CT showed intensely ^{18}F-FDG avid hypodense lesions in liver and solitary nodule in the lung. In view of high metabolic activity on ^{18}F-FDG PET/CT, a PET/CT-directed biopsy was done from one of the liver lesions, which revealed diffuse large B-cell lymphoma (DLBCL), suggestive of RT. The patient received rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy regimen with a significant clinical improvement.

CLL is a low-grade lymphoproliferative disorder with a generally indolent course.[1] However, in 2%–10% of the cases, the patient might develop an aggressive lymphoma by undergoing RT. Although DLBCL is the most common histology seen in RT, plasmablastic lymphoma, B-lymphoblastic lymphoma, and Hodgkin lymphoma might also be seen.[2-4] The risk of RT is independent of the duration, stage, and response to prior treatment and has poor prognosis with median survival of <6 months.[5] The clinical features of RT are nonspecific and include fever, night sweats, weight loss, rapid enlargement of lymph nodes, elevated lactate dehydrogenase, and beta-2 microglobulin levels.[2] Histopathology is the gold standard for the diagnosis but is not always possible.[5] FDG has been increasingly used to detect RT due to its high sensitivity to detect increased metabolic activity in the large cell/aggressive lymphomas. Furthermore, PET/CT also guides in identifying intensely metabolically active sites that are most likely to demonstrate RT on histopathology.[6] RT most commonly occurs in the lymph nodes, but in

How to cite this article: Sood A, Parihar AS, Lad D, Kumar R, Singh H, Mittal BR. An unusual presentation of Richter's transformation of chronic lymphocytic leukemia in liver and lung on ^{18}F-labeled fluoro-2-deoxyglucose positron emission tomography/computed tomography. Indian J Nucl Med 2020;35:70-1.

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Received: 22-09-2019,
Accepted: 14-10-2019,
Published: 31-12-2019.

Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.IJNM_175_19

Quick Response Code:

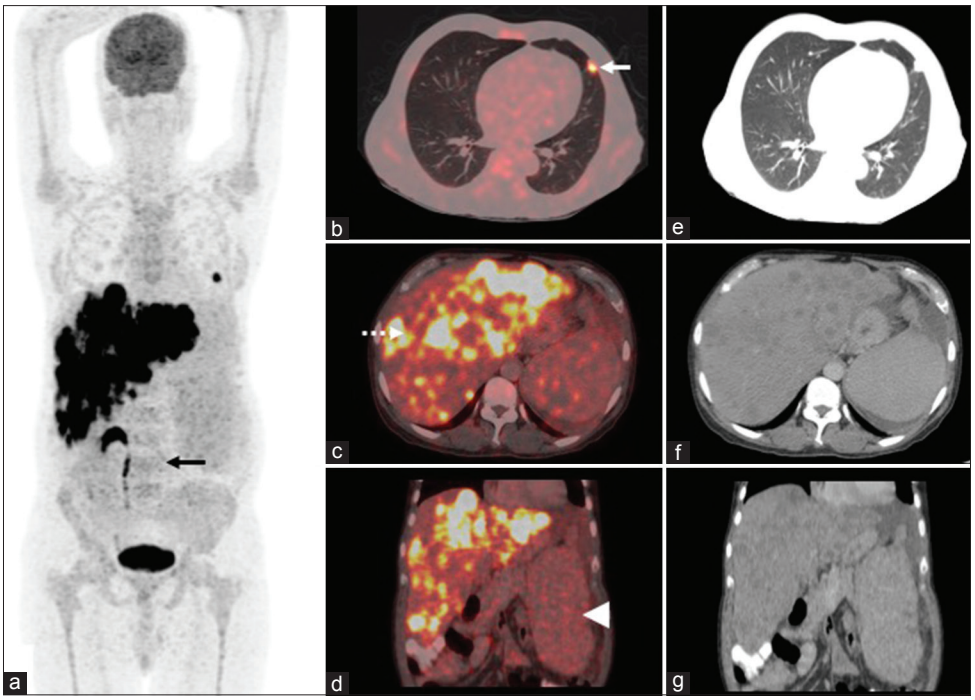

Figure 1: 18F-labeled fluoro-2-deoxyglucose PET/CT; Maximum intensity projection image (a) of positron-emission tomography shows focal increased fluoro-2-deoxyglucose uptake in the left hemithorax and multiple areas of increased tracer uptake in the liver. PET/CT fused (trans-axial [b and c] and coronal [d]) and CT-images (transaxial [e and f] and coronal [g]) revealed intense fluoro-2-deoxyglucose uptake in pleural-based nodule in the left lung lower lobe ([b]: arrow) and intensely fluoro-2-deoxyglucose-avid multiple hypodense lesions in enlarged liver ([c]: dashed arrow). Note was also made of mild heterogeneous tracer uptake in the marrow ([a]: black arrow) and enlarged spleen with no abnormal fluoro-2-deoxyglucose uptake ([d]: arrowhead)

rare cases, involvement of extranodal sites such as marrow, central nervous system, pleura, lung, skin, and gastrointestinal tract might also be seen.[3,7] The presence of isolated or prominent hepatic involvement, without lymphadenopathy is an unusual presentation of RT.[8] ^{18}F-FDG PET/CT done in our case showed prominent liver involvement and lung involvement with no lymphadenopathy which is an unusual presentation of RT. Therefore, ^{18}F-FDG PET/CT can help in the detection of transformation of CLL to RT, guide in the identification of site for biopsy and for staging of the disease.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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