Muscle Infiltrative Adult Multisystem Langerhans Cell Histiocytosis Detected on Fluorodeoxyglucose Positron Emission Tomography/ Computed Tomography – A Rare Case

Abstract

Langerhans cell histiocytosis (LCH) is a disease of unknown pathogenesis characterized by the accumulation of Langerhans cells which show immunopositivity for S-100 and CD1a. LCH with skeletal muscle involvement has been rarely described in literature. ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) is an important tool in identifying the sites of involvement in LCH. We present a rare case of muscle invasive LCH where ^{18}F-FDG PET/CT showed involvement of multiple other sites such as the liver, bones, bone marrow, and possibly the thyroid gland in our case. Further, the current case also shows that liver involvement by LCH (possibly fibrotic phase) can be negative on PET but show lesions on CT.

Keywords: *Fluorodeoxyglucose, Langerhans cell histiocytosis, positron emission tomography*

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Langerhans cell histiocytosis (LCH) is a disease of unknown pathogenesis characterized by the accumulation of Langerhans cells which show immunopositivity for S-100 and CD1a. The disease spectrum ranges from solitary organ involvement to multisystem disease. The incidence is higher in children as compared to adults.

Skeletal muscle involvement is extremely rare, and only few cases have been described in literature.[1-3] Liver involvement is also considered rare in adults, a finding seen in extensive LCH and indicates bad prognosis.[4] We report a rare case of multisystemic adult LCH, presenting with multiple skeletal muscle lesions along with liver, bone, diffuse bone marrow, and possibly thyroid involvement.

^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) is an established tool in identifying the extent of disease involvement in LCH, which could potentially change management.[5,6] Our patient was a 41-year-old female who initially presented with progressive discomfort of the right neck and shoulder region. Biopsy and immunohistochemistry of the right neck mass revealed immunopositivity for S-100 and CD1a. Subsequently, she underwent PET/CT from the vertex to the mid-thigh with intravenous contrast.

There was extensive FDG-avid mass-like diffuse enlargement of the right subscapularis muscle along with focally FDG-avid areas involving the right anterior and middle scalene, infraspinatus, trapezius, and right intercostal muscles [Figure 1a-e]. Some of the FDG-avid foci in the muscles involved did not have changes on CT.

There was lytic destruction of the right scapula [Figure 1d and e] and FDG-avid lytic lesions in the left femoral head and left scapula [Figure 2].

Diffuse confluent nonenhancing hypodense lesions were seen in both the lobes of the liver, which were normal in size with some of the lesions appearing as band along the portal tracts [Figure 3], compatible with hepatic involvement of LCH.[4] Bilirubin, alanine transaminase, and aspartate transaminase were normal. Alkaline phosphatase was significantly elevated reaching 1492 (normal: 20–140 IU/L), representing severe cholestasis. Liver involvement in LCH is considered to progress through the following four histopathological stages:

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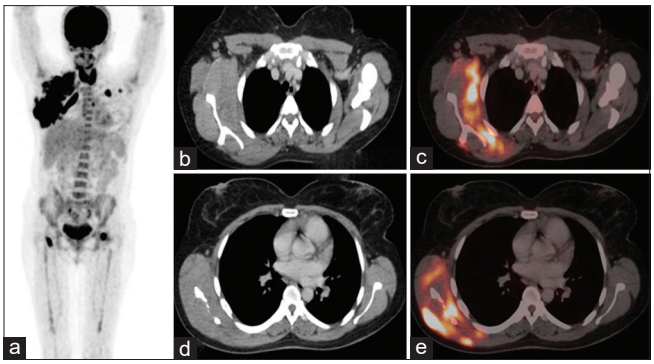

Figure 1: Maximum-intensity projection image (a), demonstrating fluorodeoxyglucose-avid lesions in the muscles, bones, thyroid gland, and diffusely increased fluorodeoxyglucose uptake in the bone marrow. Axial computed tomography (b) showing mass-like enlargement of the right subscapularis muscle and fused positron emission tomography/computed tomography (c) showing fluorodeoxyglucose-avid lesions in the right subscapularis muscle along with lytic changes in the scapula. Axial computed tomography (d) showing mass – slightly bulky right infraspinatus muscle and fused positron emission tomography/computed tomography (e) showing fluorodeoxyglucose-avid lesions in the right infraspinatus muscle

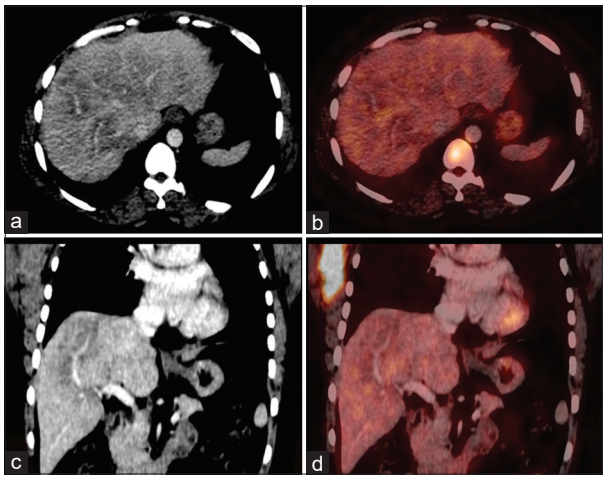

Figure 3: Axial computed tomography (a) showing avid confluent hypodense lesions without corresponding increased fluorodeoxyglucose uptake in the fused positron emission tomography/computed tomography (b). Coronal computed tomography (c) and fused positron emission tomography/computed tomography (d) showing these hypodense lesions along the portal tract

proliferative, granulomatous, xanthomatous, and fibrous phases.[7] The liver lesions in our case did not reveal high FDG uptake, which may possibly reflect advanced fibrotic phase [Figure 3b and d].

The entire bone marrow showed diffusely increased FDG uptake, reflecting bone marrow involvement [Figure 1a, maximum-intensity projection image]. There was diffusely increased FDG uptake in the enlarged thyroid gland [Figure 1a] with ill-defined hypodense nodules, possibly be due to LCH involvement which is considered rare.[8,9] Interestingly, the lungs revealed no nodular or cystic changes. In addition, other known manifestations of LCH such as cutaneous lesions and CNS lesions were not seen.

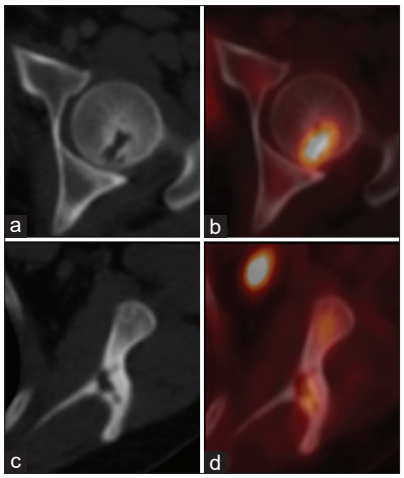

Figure 2: Axial computed tomography (a) and fused positron emission tomography/computed tomography (b) showing fluorodeoxyglucose-avid lytic lesion in the left femoral head and axial computed tomography (c) and fused positron emission tomography/computed tomography (d) showing lytic lesion in the left scapula

In conclusion, we describe a rare case of adult LCH involving the skeletal muscles, some of them without changes on CT but seen on fused PET/CT images. Second, the current case also shows that liver involvement by LCH (possibly fibrotic phase) can be negative on PET but show lesions on CT.

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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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