Case Report

Rare Case of Primary Leiomyosarcoma of Bone – Findings on 18F-fluorodeoxyglucose Positron Emission Tomography/Computed Tomography

Abstract

Primary leiomyosarcoma of bone (PLB) is a rare tumor, constituting <0.7% of all primary bone malignancies. It is clinically aggressive with heterogeneous presentation and a dismal prognosis. The most common presentation is pain with swelling and pathological fracture at times. Limited literature is available on PLB and only about 150 cases have been reported to date with only a few case reports defining the utility of 18F-fluorodeoxyglucose (18-F FDG) positron emission tomography/computed tomography (PET-CT) in its management. We hereby present a case of primary leiomyosarcoma of the right distal femur and the role of FDG-PET-CT in its management.

Keywords: *18F-fluorodeoxyglucose positron emission tomography/computed tomography, pathological fracture, primary leiomyosarcoma of bone*

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Introduction

Primary leiomyosarcoma of bone (PLB) is a rare tumor constituting <0.7% of all primary bone malignancies.[1,2] It was first described by Evans in 1965 and till date only <150 cases have been described.[2,3] PLB mainly presents with pain and swelling, but in about 20%–40% of cases, it presents with pathological fracture. The most common site of PLB includes the ends of long bones of extremities, predominantly bones around the knee joint.[4] At presentation, as high as 40% of cases have distant metastasis and approximately 39%–58% of patients develop metastasis on follow-up.[5] We present such a rare case of PLB.

Case Report

A 61-year-old male presented with right thigh pain and swelling for 2 months with a sudden increase in pain for 1 week. Contrast-enhanced computed tomography (CT) revealed an expansile lytic lesion with a soft-tissue component involving the right distal femur. The pain further progressed with time and led to an inability to bear any weight. Magnetic resonance imaging (MRI) of the leg was performed and revealed a large T2 hyperintense soft-tissue component showing patchy diffusion restriction with associated altered marrow T2 hyperintensity/T1 hypointensity measuring 10.1 cm × 10.1 cm × 13.2 cm. The lesion involved anterior and posterior compartments of the thigh with evidence of comminuted displaced fracture of the distal shaft of femur likely pathological fracture. The patient was then referred for 18F-fluorodeoxyglucose (18-F FDG) positron emission tomography/CT (PET/CT) for a whole-body survey to characterize the lesion and to look for any other site of primary causing bone metastasis. PET/CT revealed a hypermetabolic expansile lytic lesion with soft-tissue component and comminuted displaced pathological fracture involving the distal shaft of the right femur as well as extensive skeletal metastasis [Figure 1]. Trucut biopsy was taken from the swelling and histopathological examination was suggestive of leiomyosarcoma of bone which subsequently was confirmed by positive smooth muscle actin (SMA) in an immunohistochemical panel study [Figure 2]. The fracture was managed with distal femoral nail placement and screw fixation followed by local radiotherapy. Then, the patient was started on chemotherapy. Follow-up PET/CT after

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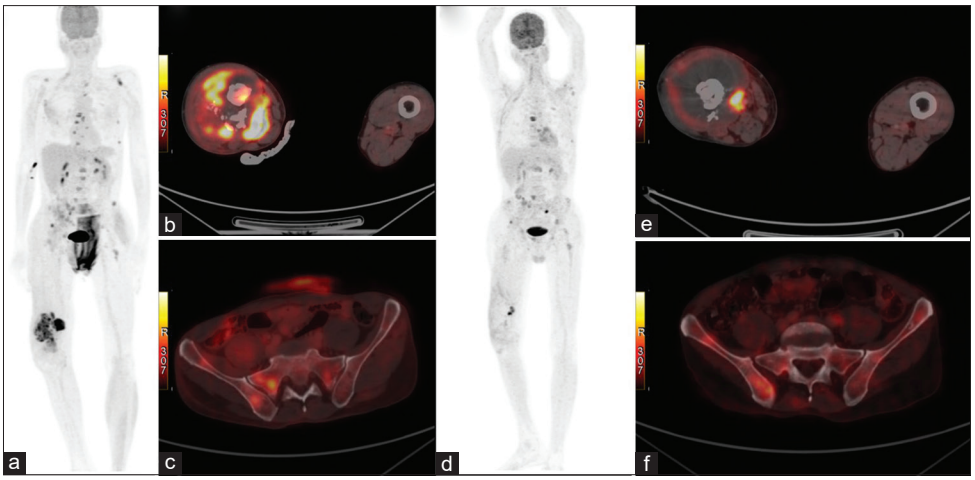

Figure 1: A 61-year-old male, case of primary leiomyosarcoma of right femur. (a) Whole-body maximum intensity projection (MIP) image shows heterogeneously increased fluorodeoxyglucose (FDG) uptake in the right distal femur and multiple other foci of increased FDG uptake, (b) Axial fused positron emission tomography/computed tomography (PET-CT) image showing comminuted displaced pathological fracture of the distal shaft of the right femur with increased FDG uptake and associated intensely FDG avid soft-tissue component, consistent with the primary leiomyosarcoma in this particular case, (c) Axial fused PET-CT image of the pelvis showing FDG avid subtle sclerotic lesion in the right ala of the sacrum and right ilium suggestive of metastasis. Follow-up 18-F FDG-PET-CT study performed post 6 cycles of chemotherapy, (d) Whole body MIP image, (e) Axial fused PET-CT image showing reduction of fracture with distal femur nail and regression in the extent of soft-tissue component associated with the primary lesion, (f) Axial fused PET-CT image of pelvis showing no significant interval change in FDG avidity of lesions in right ala of the sacrum and right ilium overall suggestive of stable disease

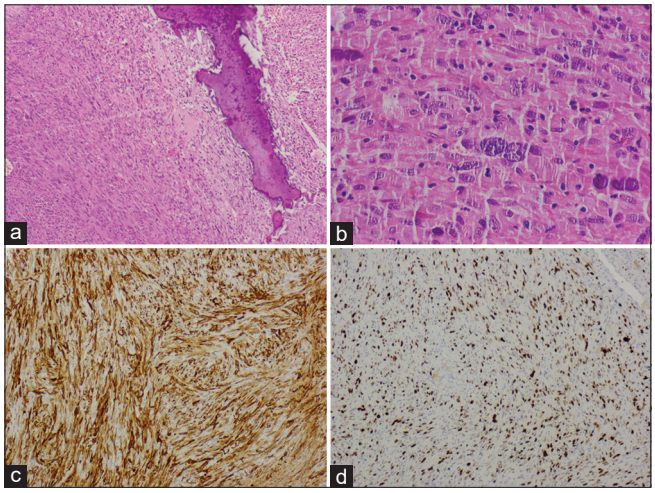

Figure 2: (a) An intersecting fascicular pattern of spindle-shaped tumor cells with infiltration into bony trabeculae (H and E, ×100), (b) Individual tumor cells showing a moderate amount of eosinophilic cytoplasm, oval nucleus, vesicular chromatin, prominent nucleoli, and moderate to marked nuclear pleomorphism and occasional multinucleation (H and E, ×400), (c) These tumor cells are diffuse and strongly positive for smooth muscle actin (IHC, ×400) (d) Mib1 labeling index was 30% in the highest proliferative area (IHC, ×400)

six cycles of chemotherapy revealed regression in the extent of soft-tissue component associated with the primary lesion; however, there was no significant change in FDG avidity of metastatic disease, overall suggestive of stable disease.

Primary leiomyosarcoma of bone (PLB)

Discussion

PLB is a rare spindle cell neoplasm and typically originates from smooth muscles. The origin of PLB in bones remains undefined. However, it has been suggested that it develops from undifferentiated mesenchymal stem cells or smooth muscle cells within the marrow cavity. The diagnosis is usually based on imaging to rule out the possibility of other primaries with bony metastasis as well as histopathological confirmation of leiomyosarcoma.[4,6]

PLB lacks any specific imaging manifestation and is difficult to differentiate from other aggressive sarcomas. Standard radiography has limited utility in PLB. However, CT and MRI examinations provide a more detailed anatomical evaluation of the local disease extent. They can clearly display alterations in the bone and the invasion of surrounding soft tissues. MRI can detect the signal alteration in the marrow as well as surrounding soft tissues. CT is particularly valuable in identifying changes in bone structure, including bone destruction and residual bone trabeculae. Functional imaging with 18-F FDG can evaluate the whole body in a single sitting and thus can confirm the lesion as the bone from primary rather than metastasis with high accuracy. It can also be used for the detection of metastatic sites and evaluation of response to chemotherapy. In addition, semiquantitative parameters such as maximum value of the standard uptake value, metabolic tumor volume, and total lesion glycolysis have been shown to strongly correlate with histopathology and prognosis.[7]

The histopathological examination typically demonstrates spindle cells with smooth muscle differentiation, expression of SMA, muscle-specific actin, and other muscle markers on tumor cells (desmin, h-caldesmon, etc.). In the present case, 18-F-FDG confirmed the primary nature of the histopathologically proven case of leiomyosarcoma,

demonstrated pathological fracture at the primary site, all the metastatic sites, and its response to therapy, highlighting its pivotal role in overall patient management.

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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Conflicts of interest

There are no conflicts of interest.

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