Case Report

18F-fluorodeoxyglucose positron emission tomography/computed tomography in a case of malignant peripheral nerve sheath tumor: An unusual presentation

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ABSTRACT

Malignant peripheral nerve sheath tumors (MPNSTs) are rare tumors, with an estimated incidence of 0.1/100,000/year. They are regarded as a rare variety of soft-tissue sarcomas that derive from peripheral nerves or from cells associated with the nerve sheath. Until 50% of observed MPNSTs occur in patients with neurofibromatosis 1 (NF1). The typical presenting signs and symptoms of a PNST are a palpable mass involving a peripheral nerve, loss of nerve function and/or pain. Recently, positron emission tomography (PET) has been used to detect 18F-fluorodeoxyglucose uptake in these tumors. Most of the PET studies have been reported in patients with NF1. We report a case of sporadic MPNST masquerading as infectious dermatoses, with an unusual PET/computed tomography presentation.

Keywords: Fluorodeoxyglucose, positron emission tomography/computed tomography, malignant peripheral nerve sheath tumors

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are a rare variety of soft-tissue sarcoma of ectomesenchymal origin,[1] with an estimated incidence of 0.1/100,000/year.[2] Recently, fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) has been employed in differentiating the malignant from the benign peripheral nerve sheath tumors. The current case highlights an unusual PET/CT presentation of MPNST masquerading clinically as infected dermatoses.

CASE REPORT

A 64-year-old male, working as a gardener, presented with multiple rapidly progressive asymptomatic skin colored raised lesions 2-4 cm in diameter, over the medial border of the right foot and lower part of the right leg since 1 month. The lesion was subjected to punch biopsy, with clinical differential diagnosis of sporotrichosis, actinomycetoma, linear cutaneous leishmaniasis and atypical mycobacterial infection. Histopathology revealed a malignant mesenchymal tumor, which was confirmed to be a MPNST on immunohistochemistry. Soon thereafter, the lesions rapidly increased in size and number and began to ulcerate and become painful [Figure 1a]. Magnetic resonance imaging (MRI) of the right lower limb revealed multiple enhancing nodular lesions seen involving the skin and subcutaneous tissue. Subsequently, a whole body 18F-FDG PET/contrast-enhanced computed tomography (CECT) scan was performed on a whole body full ring PET/CT camera (Discovery STE 16, GE). 370 MBq of 18F-FDG was administered intravenously subsequent to a 6 h fast. Whole body CECT scan was performed after intravenous instillation of non-ionic contrast medium. After the CT scan, an emission scan was performed from head to thigh for 2 min per frame. Images were reconstructed by 3D VUE algorithm (GE) and viewed on a Xeleris workstation (GE) using the volumetric protocol. The study revealed multiple FDG avid nodular lesions (maximum standardized uptake value 18.6) in the right lower limb [Figure 1b], which were many

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Quick Response Code:

Website: www.ijnm.in

DOI: 10.4103/0972-3919.119551

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more in number compared with those picked up on cutaneous examination.

MPNSTs arise from a peripheral nerve or its branches or from the nerve sheath.[3] Although they may arise spontaneously, up to 50% arise in patients of neurofibromatosis 1 (NF1).[4] Histopathological and immunohistochemical studies play an important role in the diagnosis of these tumors. MRI is the imaging modality of choice because it can reveal the nerve of origin and its relationship to adjacent structures.[5] Although it is well-known that these tumors can extend for considerable distances along nerves, it may not always be possible to delineate the origin from the nerve. A recent study by Bilgic *et al.* Has shown that the nerve origin could be identified only in 45-56% cases.[6] There have been recent reports of FDG PET/CT in MPNST, most of which have been performed in patients with NF1. It has been documented as a useful tool in monitoring clinically stable NF1 patients with plexiform neurofibromas as it could predict which were more likely to subsequently grow rapidly.[7] It has also been found that in patients with NF1 harboring MPNSTs; higher FDG uptake is associated with poorer survival rates.[8] It has proved efficacious in distinguishing benign from malignant nerve sheath tumors. In addition, it can assist in guiding targeted needle core biopsies and may provide critical information in tumors that are not amenable to biopsy.[9] It thus plays an important role in the staging, restaging and post-therapy follow-up of MPNST in NF1.[10] A high FDG uptake has been documented in sporadic MPNSTs also, as is well seen in the coronal maximum intensity projection image [Figure 2] in the present case.

MPNSTs usually present as discrete masses, which may be multifocal, especially in cases of NF1. The present case was unusual in its presentation, owing to the linear arrangement of multiple nodules over the lower extremity in a gardener, raising the possibility of infectious dermatoses. These multiple hypermetabolic mildly enhancing nodular lesions were restricted to the skin and subcutaneous tissue with no extension into the underlying muscles [Figure 3]. The additional lesions detected on PET/CT were also proven to be MPNST based on histopathology with immunocytochemistry [Figure 4]. Radical surgical resection is the treatment of choice in MPNST.[11] They are generally considered chemotherapy and radiotherapy resistant tumors. Though multimodality therapy, including surgical resection and adjuvant radiotherapy, is available, the prognosis remains dismal.

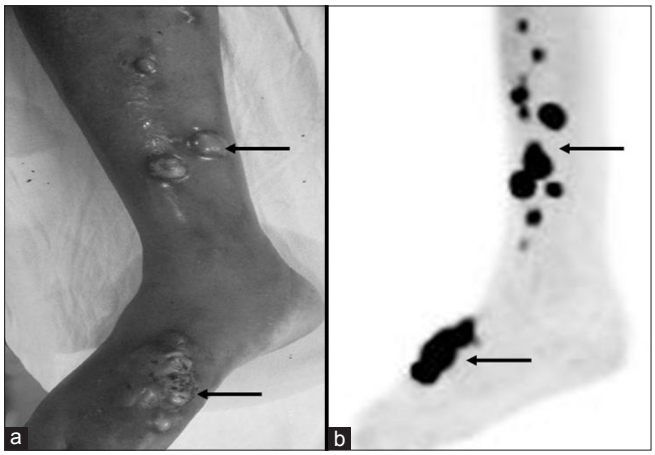

Figure 1: Multiple nodular lesions (arrows) with evidence of ulceration on the medial aspect of the right leg and foot (a), which show increased fluorodeoxyglucose avidity (b)

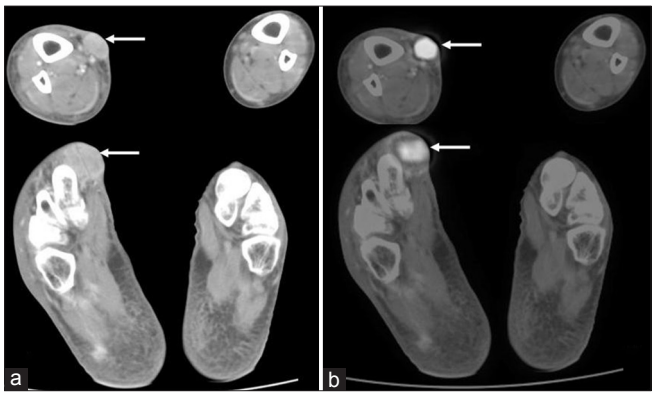

Figure 3: Axial contrast-enhanced computed tomography (a) and fused positron emission tomography/computed tomography (b) images of the leg (upper row) and foot (lower row) demonstrate multiple hypermetabolic mildly enhancing nodular lesions (arrows) involving the skin and subcutaneous tissue with no extension into the underlying muscles

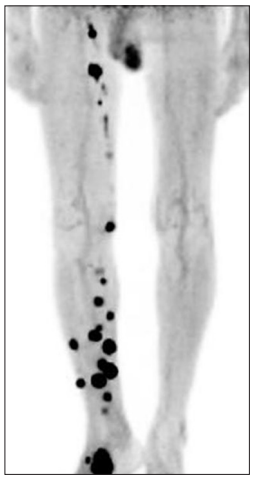

Figure 2: Coronal maximum intensity projection image showing high fluorodeoxyglucose uptake in multiple nodular lesions on the right lower limb, predominantly on the medial aspect

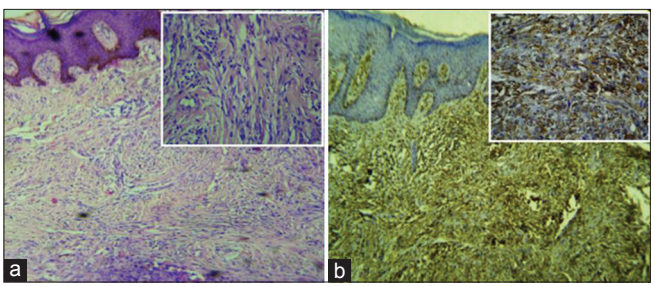

Figure 4: (a) H and E stain (×10 [inset × 40]) shows a tumor in the dermis with multiple spindle-shaped cells in whorls with pleomorphic nuclei forming a storiform pattern. (b) Vimentin stain (×10 [inset × 40]) for mesenchymal structures is diffusely positive throughout the dermis

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How to cite this article: D'souza MM, Jaimini A, Dhali TK, D'souza P, Saw S, Sharma R, Mondal A. 18F-fluorodeoxyglucose positron emission tomography/computed tomography in a case of malignant peripheral nerve sheath tumor: An unusual presentation. Indian J Nucl Med 2013;28:168-70.

Source of Support: Nil. **Conflict of Interest:** None declared.