Article published online: 2022-05-18

Case report

Fluorine-18-2-fluoro-2-deoxy-D-glucose Positron Emission Tomography/Computed Tomography Masquerading as a Case of Sporadic Malignant Peripheral Nerve Sheath Tumor of Lower Extremity Presenting as Massive Lower Limb Edema

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Abstract

Malignant peripheral nerve sheath tumors (MPNSTs) are rare neuroectodermal tumors resulting from the malignant transformation of benign plexiform neurofibromas. The sporadic form of these tumors is rare than familial variants (seen in neurofibromatosis Type 1) and making the diagnosis difficult. We are presenting a case of 40–-year-old female with the complaint of progressive swelling of lower limb with initial suspicion of lymphedema and underwent lymphoscintigraphy, magnetic resonance imaging, and finally fluorine-18-2-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography scans were done to rule out mitotic etiology and extent of the disease. The patient underwent below-knee amputation, and histopathological examination confirmed the diagnosis of sporadic MPNST.

Keywords: Fluorine-18-2-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography, malignant peripheral nerve sheath tumors, neurofibromatosis Type I

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are rare neuroectodermal tumors resulting from the malignant transformation of benign peripheral nerves tumors. They are usually associated with neurofibromatosis Type 1 (NF-1), and sporadic form of the tumor is less common than the familial variant.[1] We are presenting a case of incidentally detected sporadic MPNST, in which fluorine-18-2-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography (F-18 FDG PET/CT) scan revealed heterogeneously increased tracer uptake involving left distal lower limb. The patient underwent below-knee amputation of left limb and proven histopathologically.

Case Report

A 40-year-old female presented with slowly progressive below-knee swelling of left lower limb. It was associated with mild pain, but there was no associated motor weakness, sensory loss, tingling, or numbness of limb. There was no history of any trauma/fever/chronic disease and no any other swelling elsewhere in the

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Access this article online

Quick Response Code:

Website: www.wjnm.org

DOI: 10.4103/1450-1147.215494

How to cite this article: Singh D, Kumar R, Das A, Varma SC, Mittal BR. Fluorine-18-2-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography masquerading as a case of sporadic malignant peripheral nerve sheath tumor of lower extremity presenting as massive lower limb edema. World J Nucl Med 2017;16:324-7.

body. There was no family history of neurocutaneous disorder. On local examination, the swelling was noticed in the anterior, medial, and lateral aspect of left leg with multiple firm nodular lesions on medial aspect. The swelling was fluctuant, and local temperature was raised. To rule out lymphedema, she underwent lymphoscintigraphy which revealed relatively slow lymphatic drainage with no definite E/O lymphatic obstruction in left lower limb [Figure 1]. Fine-needle aspiration cytology (FNAC) from left leg swelling was suspicious for sarcoma. Subsequently, magnetic resonance (MR) revealed nonencapsulated enhancing soft tissue mass infiltrating subcutaneous and cutaneous planes of left leg from below the knee to forefoot. MR imaging also showed the suspicious extension of lesion along the superficial peroneal neurovascular bundle and intermuscular fascial planes in the medial aspect of the leg [Figure 2]. Repeat FNAC from the swelling was suspicious for malignant myxoid fibrous histiocytoma, and skin biopsy was suspicious of T-cell lymphoma panniculitis. To characterize the disease and to know the extent, she underwent F-18 FDG PET/CT, which showed heterogeneously increased FDG uptake (standardized uptake value [SUV] max 4.5) in enlarged soft tissue mass involving left leg extending from left knee joint up to forefoot with areas of necrosis and a few intensely FDG avid lesions (~1.4 cm × 1.6 cm; SUVmax 10.6) in the intermuscular planes. In addition, enlarged left inguinal and external iliac (largest measuring ~ 2.1 cm × 1.1 cm; SUVmax 4.5) lymph nodes were noted [Figure 3].

She underwent below-knee amputation of left lower limb in view of metabolically active extensive soft tissue mass suspicious for malignancy and histopathology confirmed the diagnosis of MPNST [Figure 4]. Subsequently, she received 4 cycles of chemotherapy (cyclophosphamide, vincristine, and adriamycin) at 3 weeks interval and planned for next cycle.

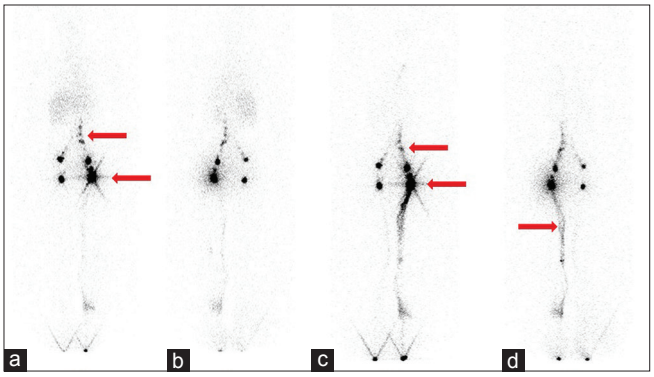

Figure 1: Lymphoscintigraphy images scan, (a and b) Early at 10 min and (c and d) delayed at 1 h anterior and posterior whole body images showing tracer avid iliac and inguinal lymph nodes bilaterally with no lymphatic obstruction

Discussion

MPNST accounts 5%–10% of soft tissue sarcoma with the incidence of 0.1/100,000 populations/year. These are highly malignant and aggressive tumors with poor prognosis and high disease-specific mortality up to 75% during the period of 5 years. The 5-year survival rate of MPNST without NF-1 is 50% but with NF-1 is 10%.[1]

Most of the tumors associated with NF-1 occur around 30 years of age, but sporadic MPNSTs are seen in the fifth decade.[2] These tumors grow along nerve with the involvement of surrounding structures and overlying skin. Distant hematogenous metastasis can occur; however, lymph nodal metastasis is rare.[1-3] In the present study, we present a 40-year-old female patient with left lower limb swelling who was diagnosed as MPNST on

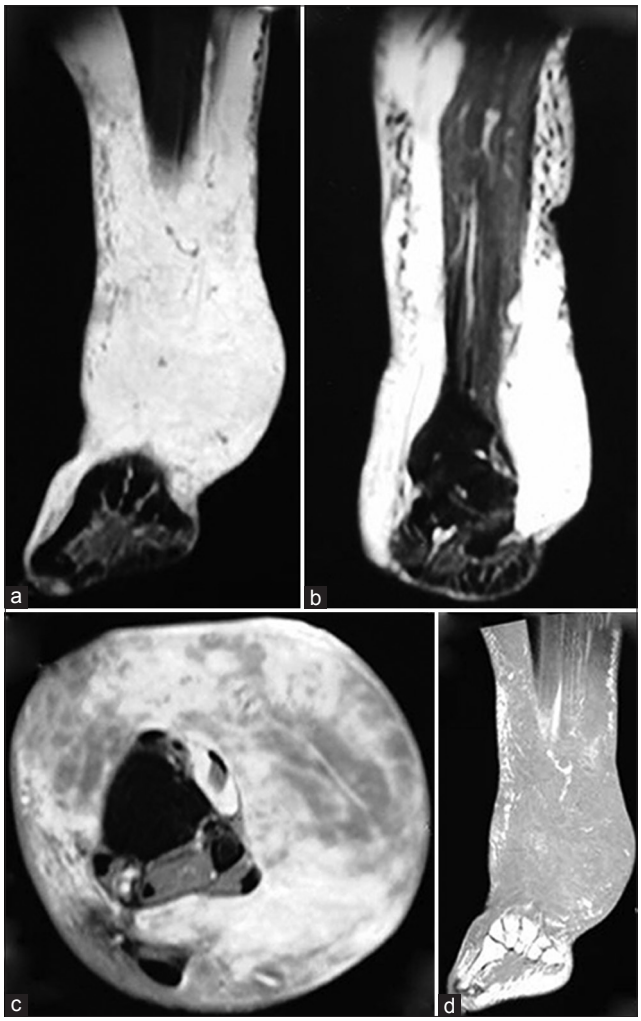

Figure 2: Magnetic resonance imaging (a and b) Short tau inversion recovery coronal, (c) postcontrast T1 axial and (d) T1 coronal images showing nonencapsulated intense homogenously enhancing soft tissue mass infiltrating within subcutaneous and cutaneous planes of left lower leg

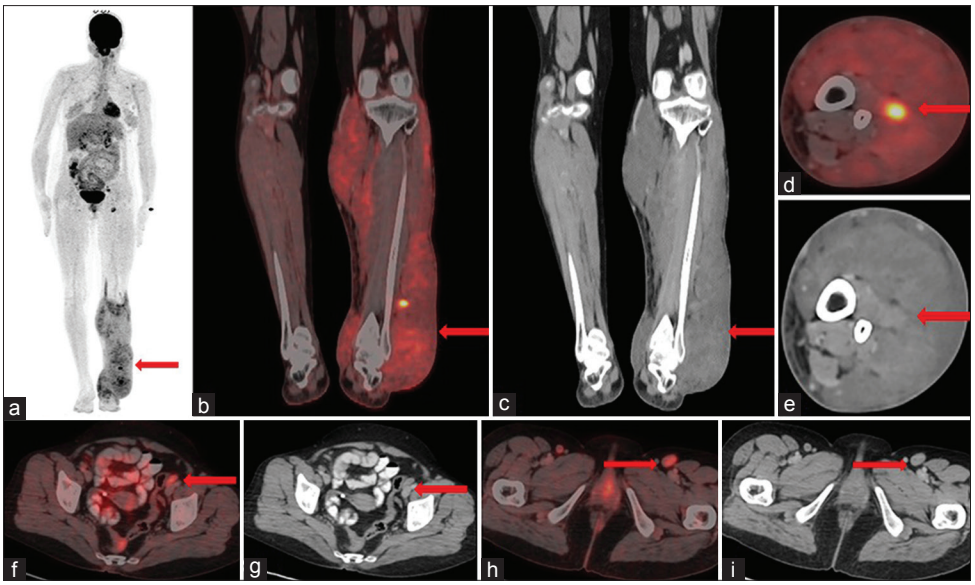

Figure 3: 2-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography, (a) Maximum intensity projection-2-fluoro-2-deoxy-D-glucose uptake in left leg, (b and c) coronal positron emission tomography/computed tomography and computed tomography-2-fluoro-2-deoxy-D-glucose uptake in primary lesion, (d and e) transaxial positron emission tomography/computed tomography and computed tomography-2-fluoro-2-deoxy-D-glucose avid focus, (f-i) positron emission tomography/computed tomography and computed tomography-2-fluoro-2-deoxy-D-glucose avid external iliac and inguinal lymph node

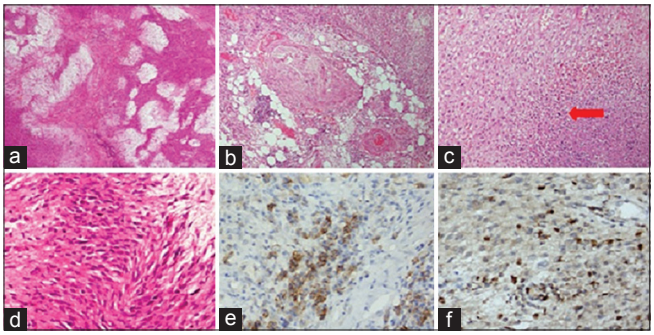

Figure 4: Histopathology, (a) Tumor cells in fascicles with myxoid changes (H and E, ×40), (b) cells infiltrating fat, nerves, vessels (×100), (c) necrosis (×200), (d) cells with kinked tapered nuclei, coarse chromatin (×400), (e) immunoperoxidase (epithelial membrane antigen) - membranous positivity, (f) immunoperoxidase (S100) - nuclear positivity

histopathological examination. The exact localization for tissue sampling in these tumors is challenging due to heterogeneous nature of tumor and difficulties in the differentiation of MPNST from benign PNST on conventional imaging. Functional PET imaging is useful to define anatomic extent, differentiate malignant from benign tumors, localize exact site for biopsy to avoid blind, painful, and unsuccessful conventional biopsies, and to rule out distant metastasis.[3-5]

After disease characterization, definitive management of MPNST is surgical resection followed by adjuvant chemotherapy or radiotherapy. In the indexed case, after PET/CT, the patient underwent below-knee amputation of left lower limb followed by 4 cycles of chemotherapy.

MPNSTs are high-grade malignancies and degree of FDG uptake correlates with histological grading.[6,7] However, in a retrospective study by Brenner *et al*. in 16 patients of NF-1 with MPNSTs with FDG-PET showed it is tumor SUVmax and not histopathological grading which acts as survival parameter. They calculated sensitivity, specificity, positive and negative predictive values, and accuracy for long-term survival as 75%, 100%, 100%, 92%, and 94%, respectively, with SUVmax cutoff of 3.0.[8]

Most of the previous studies evaluated the role of PET/CT in familial NF-1 where the probability of detection of MPNST is higher; however, in the present case, we highlight the usefulness of FDG-PET in sporadic MPNST.

In a study by Ferner *et al*. detected 116 lesions in 105 patients, 80 plexiform neurofibromas, 5 atypical neurofibromas, 29 MPNST, and 2 other malignancies. Fifty-nine tumors were histopathologically proven. Study showed FDG-PET/CT sensitivity of 0.89 (95% confidence interval (CI): 0.76–0.96] and specificity of 0.95 (CI: 0.88–0.98) in NF-1 associated tumors. They used SUVmax cutoff of 3.5 to discriminate neurofibroma from MPNST.[9] In our study, mass showed highest SUVmax of 4.5, favoring malignancy.

In the present case, we report a patient of sporadic MPNST, in which FDG-PET/CT showed heterogeneous tracer uptake in the left leg, operated subsequently, and proven histopathologically.

Conclusion

We concluded that FDG-PET/CT is helpful to see the metabolic behavior of the tumor, to guided biopsy, planning and optimizing of treatment strategies, and to predict long-term survival following definite surgery.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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