Case Report

Usefulness of ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography in dermatofibrosarcoma protuberans on treatment with imatinib

Raghava Kashyap, Vamshi Krishna Muddu[1], Sameera Anantamakula, Satya Sri[2]

Departments of Nuclear Medicine and Positron Emission Tomography and [1]Medical Oncology, Mahatma Gandhi Cancer Hospital, [2]Department of Pathology, Indus Hospital, Visakhapatnam, Andhra Pradesh, India

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare locally aggressive tumor with distant metastases being unusual. We present a case of metastatic DFSP treated with imatinib showing complete metabolic response to treatment.

Keywords: ^{18}F-fluorodeoxyglucose, dermatofibrosarcoma protuberans, fluorodeoxyglucose positron emission tomography/computed tomography, imatinib, response evaluation

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare tumor involving the dermis of the skin with the incidence of 3–4/million.[1,2] These tumors are locally aggressive; however, distant metastases are unusual.[3-5] The hallmark of this cancer is a specific translocation of COL1A1 and PDGFB (around 90% of the lesions).[6,7] This makes the tumors susceptible to treatment with tyrosine kinase inhibitor imatinib.[8] F-18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) may be a useful modality in treatment response evaluation of metastatic DFSP.

CASE REPORT

The index case is a 62-year-old male patient. He presented for the 1st time 17 years back with a lesion in the left thigh which was completely excised and diagnosed as DFSP. He presented with local recurrence after 15 years and again underwent wide local resection with negative margins of excision. Imaging with CT scan did not show any distant metastases. There was no adjuvant therapy. There was a local recurrence a 2nd time after 1 year. An excision biopsy was repeated and revealed fibrosarcomatous components within the DFSP [Figure 1a and b]. An ^{18}F-FDG PET/CT scan done to evaluate the disease extent showed extensive metastases in the both lungs [Figure 2a, c and e] along with bone metastasis involving the left acetabulum.

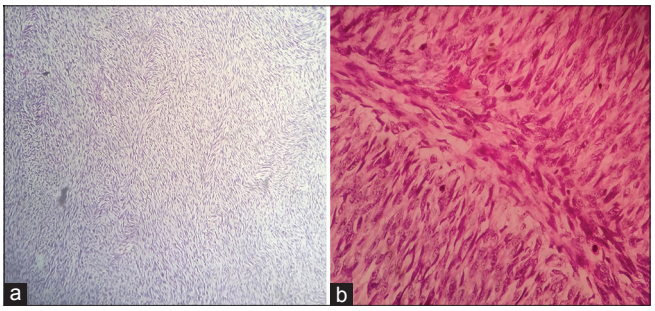

Figure 1: (a) The low power view of the tumor showing herring bone pattern (b) high power view showing spindle shaped tumor cells arranged in fascicles and exhibiting moderate pleomorphism

Address for correspondence:
Dr. Raghava Kashyap, Department of Nuclear Medicine and Positron Emission Tomography, Mahatma Gandhi Cancer Hospital, Visakhapatnam, Andhra Pradesh, India.
E-mail: kashyapkr@gmail.com

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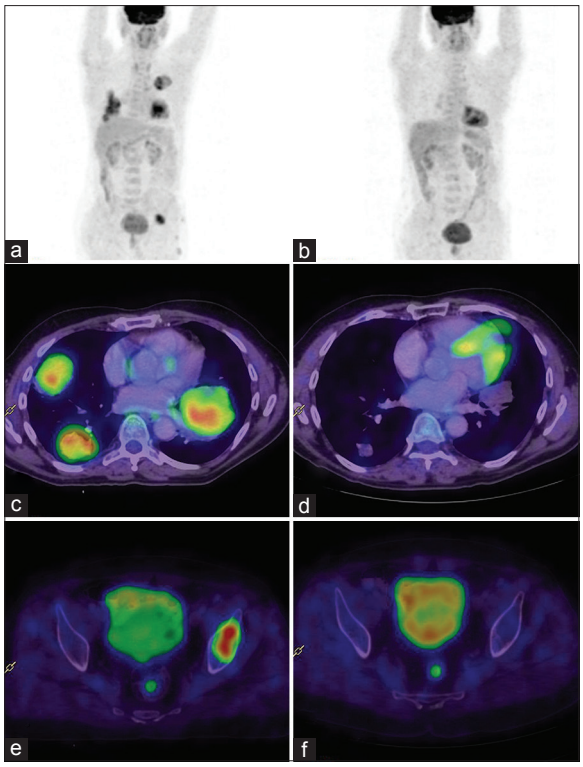

Figure 2: (a) Whole body ^{18}F-fluorodeoxyglucose positron emission tomography image prior to treatment showing lesions in the left thigh, left iliac bone and lungs. (b) Whole body ^{18}F-fluorodeoxyglucose positron emission tomography image posttreatment showing complete metabolic response of all lesions. (c) Pretreatment transaxial image of the lung showing intensely hypermetabolic lung metastases. (d) Posttreatment transaxial image of the lung showing complete metabolic resolution and partial anatomical resolution of the lung lesions. (e) Pretreatment transaxial image showing hypermetabolic left iliac bone lesion. (f) Posttreatment transaxial image showing complete metabolic resolution of the iliac lesion

A review of the previous chest CT scan done at the time of the first recurrence a year prior confirmed that the lung lesions were a new development. The patient was started on imatinib 400 mg once a day. The therapy was well tolerated without any major toxicity. A repeat PET scan was done 5 months after initiation of treatment and shows complete metabolic resolution of all the lesions while the CT reveals only partial response [Figure 2b, d and f]. The patient is currently on follow-up and has completed 7 months of therapy with imatinib.

DISCUSSION

Our case report has several noteworthy features.

DFSP is a rare tumor of the skin with intermediate malignant potential. As exemplified in our case, the tumor typically presents with multiple local recurrences despite adequate wide local excision. The best surgical outcomes have been achieved with Moh's microsurgical procedure which is practiced only at specialized centers.[9]

The tumor in our patient demonstrated fibrosarcomatous transformation within the classical DFSP which is an unusual feature.[10,11] The patient had rapid development of extensive pulmonary and skeletal metastases within a span on 1 year and the role of the transformation as a contributory factor is an intriguing question. Though pulmonary and lymph nodal metastases have been described previously in DFSP, bone metastasis are rare [Figure 2e].

The third noteworthy feature of the case is the radiological response to treatment with imatinib. More than 90% of cases of DFSP harbor translocation of 17 and 22 genes.[5,12] Imatinib being a tyrosine kinase inhibitor has been shown to have definite role in DFSP in the following situations:[5,12]

- locally advanced disease that is inoperable,
- locally advanced disease where a reduction in the size of the tumor would aid in surgical resection, and
- Metastatic disease not amenable to surgical resection.

There are very few case reports on the role of ^{18}F-FDG PET in detection and follow-up of DFSP in the literature.[13-16] Our patient demonstrated a drastic metabolic response on ^{18}F-FDG PET with the morphological response lagging behind [Figure 2e]. It is well-known that all cases of DFSP may not harbor the same genetic translocation[17] and hence might not respond to imatinib. We would like to propose that demonstration of response on ^{18}F-FDG PET/CT can act as a surrogate marker for patients who harbor gene rearrangement in DFSP and similar to the situation in nonsmall cell lung cancer, an early response evaluation PET may be able to predict the outcome in these patients.

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Conflicts of interest

There are no conflicts of interest.

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