

Mycosis fungoides: Positron emission tomography/computed tomography in staging and monitoring the effect of therapy

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ABSTRACT

A 58-year-old woman, diagnosed as a case of mycosis fungoides (MF), underwent [18F]-fluoro-D-glucose positron emission tomography/computed tomography (FDG PET/CT) examination. The study revealed intense FDG uptake in a large ulceroproliferative right thigh lesion, indurated plaques in the chest wall and left thigh, along with multiple sites of cutaneous involvement, axillary and inguinal lymphadenopathy. The patient underwent chemotherapy with CHOP regimen, radiotherapy for the right thigh lesion, along with topical corticosteroids and emollients for the disseminated cutaneous involvement. Repeat [18F]-FDG PET/CT study performed a year later, showed near complete disease regression specifically of the ulceroproliferative lesion and indurated cutaneous plaques, no change in lymphadenopathy, and a subtle diffuse progression of the remaining cutaneous lesions. A multidisciplinary approach to the diagnosis, staging and treatment of MF has long been suggested for optimizing outcomes from management of patients with this disease. This case highlights the potential role of incorporating PET/CT as a single modality imaging technique in the staging and assessment of response to therapy.

Keywords: [18F]-fluoro-D-glucose positron emission tomography/computed tomography, mycosis fungoides, staging

INTRODUCTION

Mycosis fungoides (MF) is a non-Hodgkin's lymphoma of T-cell origin, primarily involving the skin. It is a relatively rare disease with an annual incidence of 0.36 per 100,000.^[1] In advanced cases, it may affect extracutaneous regions such as lymph-nodes, viscera, peripheral blood and skeletal sites. Accurate delineation and staging of cutaneous and extracutaneous spread has important implications on patient management. Conventionally, a contrast enhanced computed tomography (CT) scan of the chest, abdomen and pelvis is performed, along with a lymph-node and bone marrow biopsy in advanced cases, to rule out extracutaneous involvement.^[2] This case highlights the potential role of incorporating positron emission tomography

(PET)/CT as a single modality imaging technique in the staging and assessment of response to therapy.

CASE REPORT

A 58-year-old woman, presented with an ulceroproliferative lesion in the right thigh. She had a long history of multiple pruritic patches on the skin, with gradual progression all over the body. The eruptions had been mistaken for psoriasis for which she received treatment without significant relief. Cutaneous examination revealed multiple discrete as well as confluent scaly plaques over face, trunk and extremities. She had a large noduloulcerative lesion on the right thigh with indurated plaques over the chest wall and left thigh. The clinical suspicion of MF was subsequently confirmed on repeat serial biopsies of the cutaneous lesions taken from different sites over a period.

The patient subsequently underwent a whole body [18F]-fluoro-D-glucose (FDG) PET/CT examination on a Discovery STE16 (GE Healthcare, Milwaukee, USA) camera, after administration of 10 mCi of [18F]-FDG. This revealed

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multiple foci of increased FDG uptake on the skin, which was well seen on the maximal intensity projection (MIP) image as well as the transaxial PET and fused PET/CT images [Figure 1]. An intensely FDG avid ulcerative lesion was noted on the anterior aspect of the skin of the right thigh, extending deeply into the subcutaneous tissue. There was no underlying muscle involvement. Although the lesion was obvious clinically, the depth of involvement could be appreciated only on the PET/CT image. The plaques noted in the left thigh and chest wall on clinical examination also showed increased uptake. Hypermetabolic bilateral axillary and inguinal lymph-nodes were also visualized. The metabolically active left inguinal lymph-node was subjected to biopsy which revealed dermatopathic lymphadenopathy. Bone marrow biopsy performed at the iliac crest as part of the routine work-up (despite absence of abnormal uptake) proved to be negative.

The patient subsequently underwent 6 cycles of chemotherapy with CHOP regimen and 30 sittings of external beam radiotherapy for the lesion in the right thigh. The elevated plaques noted in the chest wall, and left thigh were treated with potent topical corticosteroids (clobetasol propionate 0.5%). Over the rest of the skin, she was advised topical emollients. Oral 1st generation antihistaminics (hydroxyzine) were added to control itching.

Repeat [18F]-FDG PET/CT was performed a year after the completion of chemoradiotherapy using the same protocol. This revealed near complete resolution of the ulceroproliferative lesion in the right thigh and the lesions in the chest wall and left thigh [Figure 2]. There was no significant change in the extent of axillary and inguinal lymphadenopathy. However, there was a mild progression in the overall extent of cutaneous involvement as evidenced

by subtle areas of diffusely increased uptake on the rest of the skin surface. In view of this finding, she was advised to apply a combination of topical corticosteroid (clobetasol propionate 0.5%) with topical emollients in a ratio of 1:1 over the entire skin.

DISCUSSION

Mycosis fungoides is a non-Hodgkin's lymphoma of T-cell origin, primarily involving the skin. It is a relatively rare disease with an annual incidence of 0.36/100,000.^[1] Three classical cutaneous phases of MF-patches, infiltrated plaques and tumors have been described. The disease may progress through each of these phases, which frequently overlap or occur simultaneously.^[3] In advanced cases, it may be associated with lymph-node, visceral, peripheral blood or skeletal involvement. The staging of MF is based on the extent of skin disease, type of skin lesion and extracutaneous involvement.^[4] This has important implications on patient management and prognosis. In patients with large cell transformation, tumors, erythroderma or lymphadenopathy, a contrast enhanced CT scan of chest, abdomen and pelvis is performed, along with a lymph-node and bone marrow biopsy to rule out extracutaneous involvement.^[2]

Overall, [18F]-FDG PET/CT has been found to be more sensitive and specific than CT alone in identifying involved lymph-nodes and detecting sites of extranodal involvement in lymphomas.^[5] The superiority of PET/CT over CT alone, in providing more accurate staging and prognostic information in MF has also been recently demonstrated.^[2] The study by Valencak *et al.* on thirteen patients with primary cutaneous lymphoma (of which 9 had MF), revealed that PET was able to provide valuable clinical information beyond that provided by CECT in all cases with advanced disease.^[6] The study by Kumar *et al.* on 19 patients with primary cutaneous

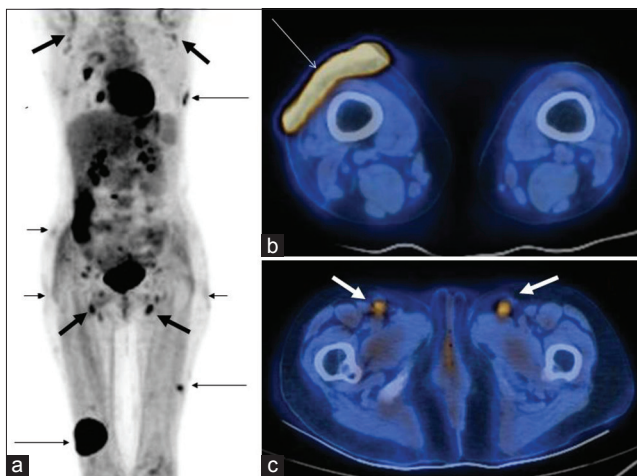


Figure 1: Baseline positron emission tomography/computed tomography: Increased fluoro-D-glucose (FDG) uptake noted at multiple cutaneous sites (short thin arrow) and bilateral axillary and inguinal lymph nodes (thick arrows) seen on the maximal intensity projection (a) and transaxial image (c). An intensely FDG avid ulcerative lesion noted on the anterior aspect of the right thigh and on indurated plaques in chest wall and left thigh (long thin arrow - a, b)

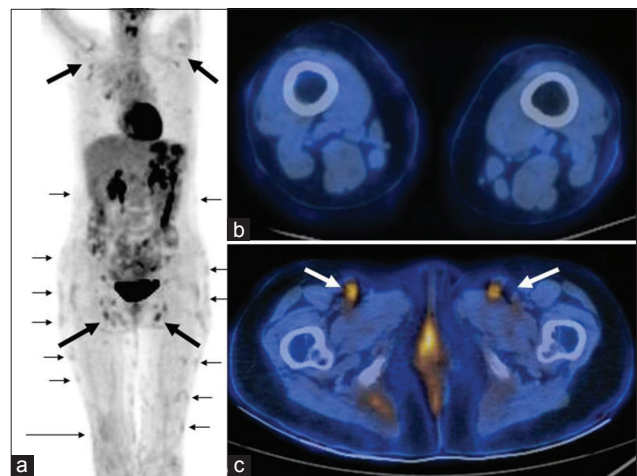


Figure 2: Posttherapy positron emission tomography/computed tomography: Near complete resolution of the ulceroproliferative lesion in the right thigh (long thin arrow - a) and the lesions in the chest wall and left thigh. No change noted in the extent of axillary and inguinal lymphadenopathy (thick arrows - a, c). However, mild progression is seen in overall extent of cutaneous involvement on the maximal intensity projection image (short thin arrows - a)

lymphoma (of which 5 had MF) showed that for restaging of cutaneous lymphoma, FDG-PET had a sensitivity of 86% and specificity of 92% for local recurrence/residual disease and a sensitivity of 100% and specificity of 100% for distant metastasis. The corresponding values for CT were 50% and 83% for local recurrence/residual disease and 100% and 67% for distant metastasis. Thus, PET clearly had a higher diagnostic value than CT for the detection of local recurrence/residual disease and distant metastases.^[7] This is largely owing to the higher sensitivity of PET in the detection of lymph-node and a bone-marrow involvement.

One of the potential pitfalls reported in the [18F]-FDG PET/CT evaluation of cutaneous lymphomas, has been the increased uptake noted in dermatopathic lymphadenitis, which may be seen in several chronic skin diseases.^[8] It was also observed in the present case. However, several studies have concluded that patients with adenopathy, even dermatopathic adenopathy, have a worse prognosis.^[9] DNA analysis of these nodes have demonstrated a high frequency of clonal rearrangements of beta T-cell receptor genes that match rearrangements in the patients cutaneous MF lesions, demonstrating molecular evidence of MF, when no histological evidence was present.^[10] Thus, the PET/CT detection of lymphadenopathy would be of significance. Although it has been found very useful for baseline evaluation and restaging of plaque, tumor or lymph-node, it may be inferior to clinical examination in mapping the extent of cutaneous involvement, especially macules or thin plaques.^[11]

Patients with generalized patch/plaque disease have a relatively high likelihood of disease progression (24%), highlighting the importance of monitoring and staging.^[12] A multidisciplinary approach to the diagnosis, staging, and treatment of MF has long been suggested for optimizing outcomes from management of patients with this disease.^[3] The possibility of incorporating PET/CT as a single modality imaging technique in the staging and assessment of response to therapy needs to be explored. Even if the initial PET/CT study does not alter the initial staging,

it could serve as a useful baseline for monitoring the effect of therapy as exemplified in our case.

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