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The place of ^{18}F FDG PET/CT in the management of patients with eosinophilic fasciitis: a case report

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SUMMARY

Eosinophilic fasciitis is a rare connective tissue disease with a clinical presentation of scleroderma-like disease. We report a case of a 36-year-old female patient with a 6-month history of progressive stiffness involving her forearms and legs with joint pain. Laboratory examinations showed hypereosinophilia and elevated C-reactive protein. ^{18}F FDG PET/CT showed diffuse and symmetrical increased uptake in the fasciae of the upper and lower limbs, sparing both muscles and fat tissues. Guided biopsy and histologic examination confirmed the diagnosis of eosinophilic fasciitis. 18F FDG PET/CT is of great help in the diagnosis of eosinophilic fasciitis, as it can guide the biopsy where FDG uptake is strongest and also help rule out possible associated neoplasms.

Key words: Eosinophilic fasciitis, FDG, PET/CT.

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INTRODUCTION

Eosinophilic fasciitis (EF) is a rare disorder with a clinical presentation of scleroderma-like illness. Magnetic resonance imaging (MRI) provides a useful aid for diagnosis and as a marker for disease activity (1). ^{18}F FDG PET/CT has been previously reported to be a helpful test in the diagnosis of various autoimmune diseases: we report the case of a patient with EF, showing that ^{18}F FDG PET/CT may be a useful test for diagnosis but also important to rule out possible associated neoplasms.

CASE REPORT

A 36-year-old woman without significant past medical history presented with a 6-month history of fatigue, progressive stiffness affecting the distal extremities, hyperpigmentation of the upper extremities, and pitting edema with induration of both legs. Physical examination showed provoked myalgia on both forearms and legs, bilateral knee effusion, wrists synovitis, and flexor tendon retraction assessed by the prayer sign (Figure 1). Cardiac and pulmonary auscultations were normal. There was neither lymph node nor liver

Figure 1 - Forearm tendon retraction assessed by the prayer sign in patient with eosinophilic fasciitis.

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or spleen enlargement on abdominal and lymph node area palpation. Urine strips were normal. Routine laboratory examinations showed peripheral eosinophilia at 6 G/L, high C-reactive protein of 100 mg/dL, polyclonal hypergammaglobulinemia at 18 g/L, and elevated serum creatinine kinase at 500 UI/L. Auto-immune investigations including antinuclear antibodies, anti-extractable nuclear antigen antibodies, systemic sclerosis and myositis dot-blot assay, and anti-neutrophil cytoplasmic antibodies were negative. ^{18}F FDG-PET/CT showed diffuse and symmetrical high uptake in the fasciae of the upper and lower limbs, sparing both muscles and fat tissues (Figure 2). Besides, ^{18}F FDG-PET/CT discarded an occult neoplasia. MRI of forearms and legs showed prominent superficial and deep fascial thickening on T1-weighted (Figure 3A), diffusion weighted (Figure 3B), and on STIR images (Figure 3C). A biopsy specimen from the leg, in which the FDG uptake was strongest, showed an inflammatory infiltrate composed of lymphocytes and eosinophils, with perivascular infiltrates of lymphocytes mainly CD8+. Moreover, there was no muscular involvement and skin biopsy showed no images of morphea or cutaneous T-cell lymphoma (Figure 4). The diagnosis of idiopathic EF was retained and the patient received 1 mg/kg/day oral prednisone with progressive tapering in 24 months. The outcome was favorable with progressive reduction of stiffness and edema of the limbs. We ob-

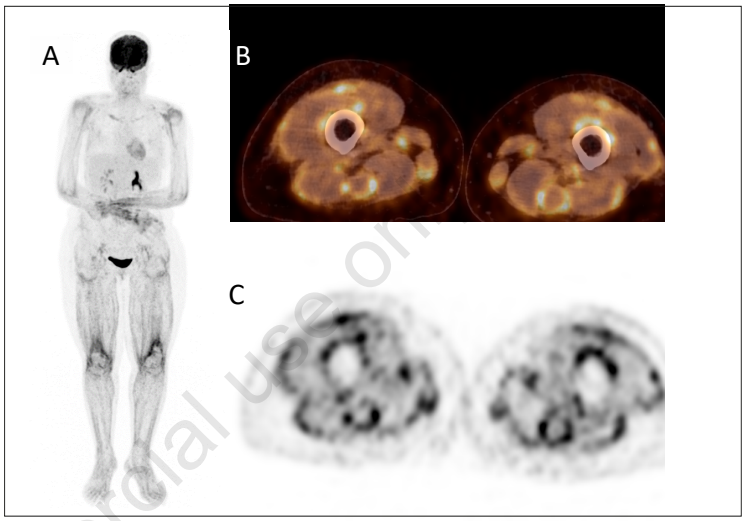

Figure 2 - ^{18}F FDG-PET/CT shows diffuse and symmetrical increased uptake in the fasciae of the upper and lower limbs, sparing both muscles and fat tissues on ^{18}F-FDG MIP images (A) as well as in ^{18}F-FDG PET-CT fusion (B) and ^{18}F-FDG PET alone (C).

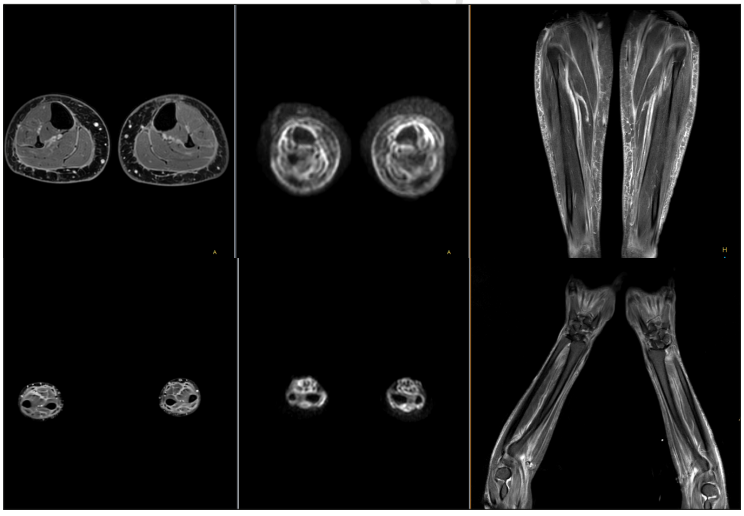

Figure 3 - Forearms and legs MRI findings in patient with eosinophilic fasciitis showing prominent superficial and deep fascial thickening on axial T1-weighted images (A), diffusion contrast-enhanced weighted images (B), and coronal short STIR images (C).

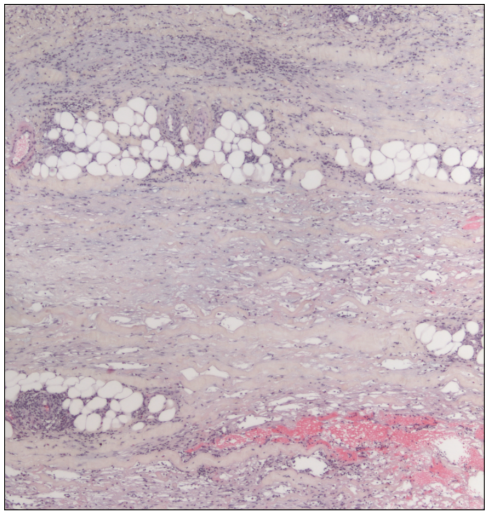

Figure 4 - Fascio-muscular biopsy of our patient with eosinophilic fasciitis. The hematoxylin eosin staining shows dense, diffuse and perivascular inflammatory infiltrates in the fascia, composed mainly of lymphocytes, but also of eosinophils.

served a rapid normalization of peripheral eosinophils and C-reactive protein after 10 days of treatment.

DISCUSSION AND CONCLUSIONS

EF is a rare connective tissue disease defined by scleroderma-like skin changes characterized by symmetrical and painful swelling with a progressive induration and thickening of the skin and soft tissues (2). EF suffers from a lack of diagnostic criteria and the diagnosis is often based on the association of skin abnormalities and thickened fascia, with an inflammatory infiltration composed of lymphocytes and eosinophils (3). Muscle magnetic resonance imaging is considered the best procedure for EF diagnosis (4). As illustrated in our case, MRI showed increased signal intensity within the fascia on fluid sensitive sequences and marked fascia enhancement after gadolinium administration at the acute phase. Little is known about the contribution of ^{18}F FDG-PET/CT in EF. As shown in this case, it can be a useful imaging tool to conduct systemic screening for inflammation and thus to determine the optimal site to perform the biopsy (5-7). This case deserves attention because EF is a rather rare rheumatic condition that may not be easy to diagnose, especially when eosinophilia is absent. On the basis of the FDG PET/CT findings described here, we think that this technique should have a role in the differential diagnosis of soft tissue pain. In fact, it shows specific hypermetabolism of fascia and highlights the normal aspect of muscle and skin. It guides the biopsy in the area where FDG uptake is higher. Moreover, EF is associated with hematological malignancy in 10% of cases (8). Thus, this imaging technique is helpful to rule out malignancy associated with EF, an advantage, when compared with MRI. However, no definite conclusion can be drawn from our findings, and our data warrant further investigation to assess the usefulness of FDG PET/CT in EF.

Conflict of interests

The authors declare no potential conflict of interests.

REFERENCES

1. Moulton SJ, Kransdorf MJ, Ginsburg W, et al. Eosinophilic fasciitis: spectrum of MRI findings. AJR Am J Roentgenol. 2005; 184: 975-8.
2. Pinal-Fernandez I, Selva O, Callaghan A, et al. Diagnosis and classification of eosinophilic fasciitis. Autoimmun Rev. 2014; 13: 379382.
3. Lebeaux D, Sène D. Eosinophilic fasciitis (Shulman disease). Best Pract Res Clin Rheumatol. 2012; 26: 449-58.
4. Lebeaux D, Frances C, Barete S, et al. Eosinophilic fasciitis (Shulman disease): new insights into the therapeutic management from a series of 34 patients. Rheumatology (Oxford). 2012; 51: 557-61.
5. Cheriet S, Chastan M, Levesque H, et al. Positron emission tomography in the diagnosis of eosinophilic fasciitis. QJM. 2011; 104: 987-8.
6. Kim HJ, Lee SW, Kim GJ, et al. Usefulness of FDG PET/CT in the diagnosis of eosinophilic fasciitis. Clin Nucl Med. 2014; 39: 801-2.
7. Marie I, Sauvetre G. Fluorodeoxyglucose positron emission tomography in eosinophilic fasciitis. Joint Bone Spine. 2014; 81: 541.
8. Mazori DR, Femia AN, Vleugels RA. Eosinophilic fasciitis: an updated review on diagnosis and treatment. Curr Rheumatol Rep. 2017; 19: 74.