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Atypical Muscular Sarcoidosis Involvement Revealed by ^{18}F-FDG PET/CT

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Abstract

We present the case of a 65-year-old woman with known pulmonary and muscular sarcoidosis who presented with dyspnea. FDG-PET/CT revealed unsuspected active myositis of multiple muscle groups, including the neck, and was useful in monitoring treatment response.

Keywords

- sarcoidosis
- myositis
- PET
- FDG
- fluoro-deoxyglucose

Introduction

While subclinical muscle involvement in sarcoidosis is frequent, symptomatic sarcoid myopathy is a rare presentation of systemic sarcoidosis with multiple clinical forms described in the literature. Fluoro-deoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) has been shown to be able to detect chronic myopathy, the most common presentation that typically affects proximal muscle groups.

Case Report

A 65-year-old woman presented with increasing dyspnea for 2 months following tapering of prednisone dosage to 10 mg per day. She was otherwise being treated with oral methotrexate 25 mg administered once a week and intravenous infliximab 5 mg/kg administered every 8 weeks. She had been previously diagnosed with pulmonary and muscular sarcoidosis, histologically confirmed by deltoid muscle and supraclavicular lymph node biopsies, both demonstrating noncaseating granulomas. Electromyography of all four limbs was classified as "irritable", consistent with inflammatory myopathy. CT pulmonary angiography was negative for pulmonary embolism. Gadolinium enhancement was absent on cardiac magnetic resonance imaging (MRI). Echocardiogram was remarkable for an increased pulmonary artery systolic pressure of 49 mmHg. Whole-body FDG-PET/CT was performed following a myocardial suppression protocol (24 hour high-fat, low-carbohydrate diet, 12 hour fasting, intravenous heparin) to exclude cardiac sarcoidosis (►**Fig. 1A**). Incomplete myocardial suppression due to nonadherence to the preparation protocol limited cardiac assessment. Uptake was absent at the thoracic and abdomen level. Intense FDG uptake was noted in multiple upper body muscle groups, notably the scalene and sternocleidomastoid ($SUV_{max} = 18.0$) (►**Fig. 1B**), trapezius, biceps,

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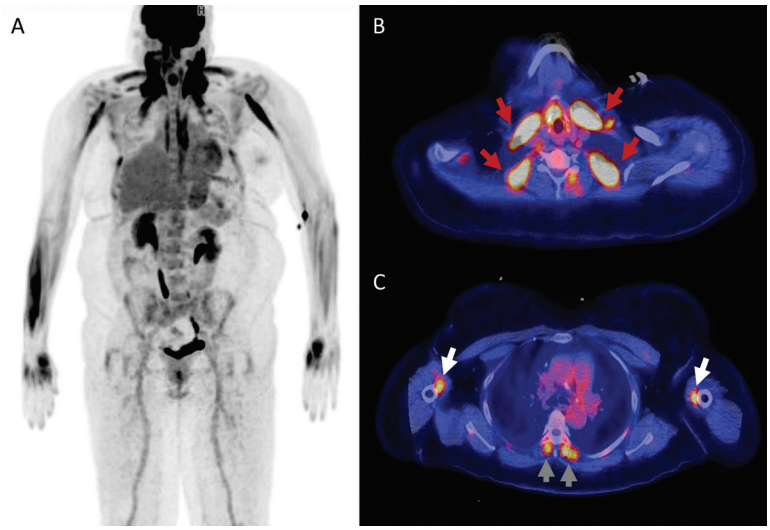

Fig. 1 (A) ^{18}F-fluoro-deoxyglucose (FDG) positron emission tomography (PET) showing intense FDG uptake in multiple upper body muscle groups (**B** and **C**) in a patient with histologically proven muscle sarcoidosis.

forearm, and paravertebral muscles (►**Fig. 1C**). Patient denied any strenuous physical activity or trauma in the days preceding the study. The patient's dyspnea was gradually relieved following prednisone dosage increase, a switch from oral to subcutaneous methotrexate, and decreased dosing interval of infliximab. Repeat FDG-PET/CT was performed a week later following strict adherence to the preparation protocol, yielding a complete myocardial suppression (►**Fig. 2A**). Pathological myocardial uptake was absent. Overall, muscle FDG uptake was significantly reduced, with scalene and sternocleidomastoid SUV_{max} at 4.3 compared with 18.0 previously (►**Fig. 2B**). Paravertebral muscle uptake intensity was reduced ($SUV_{max} = 4.4$) (►**Fig. 2C**). Trapezius and biceps abnormal uptake was resolved. New FDG uptake was noted in the right masseter muscle ($SUV_{max} = 9.9$) (►**Fig. 2D**).

Discussion

Sarcoidosis is a multisystem granulomatous disease of unknown origin. While asymptomatic muscle involvement is common in systemic sarcoidosis (50–80%), symptomatic myositis is a rare entity (< 0.5%).[1] Three patterns of sarcoid myositis have been reported: nodular, chronic, and acute myositis.[1] Chronic myopathy is the most frequent type and typically presents in older women as progressive proximal weakness and atrophy.[2,3] While sarcoidosis muscle involvement requires confirmation with a biopsy, many imaging tests can aid in the diagnosis. Ultrasonography and CT scan can detect nodules but are not useful for chronic myopathy and acute myositis. Atrophy of the muscles and a high-signal intensity on T2-weighted imaging on MRI can suggest sarcoid muscle involvement.[1–3] However, FDG-PET/CT appears to be

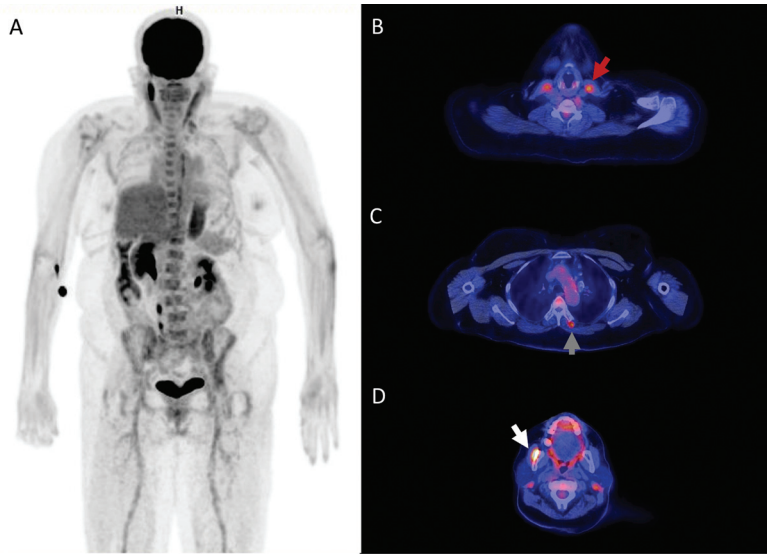

Fig. 2 (A)^{18}F-fluoro-deoxyglucose (FDG) positron emission tomography (PET) performed following treatment optimization showing important regression of FDG uptake in the upper body muscle groups (**B** and **C**), while new FDG uptake was noted in the right masseter muscle (**D**).

able to accurately detect chronic myopathy and acute myositis in addition to being helpful for the initial diagnosis of sarcoidosis, identifying extrapulmonary involvement, guiding biopsy, and monitoring treatment efficacy.[1–3] A variety of imaging findings on FDG-PET/CT have been described in patients with myositis, ranging from focal symmetrical limb uptake to the presence of multiple linear and patchy hypermetabolic lesions referred to as the "Tiger man sign."[4–6] To the best of our knowledge, sarcoid myositis involving the neck muscles has not been previously reported. In this case, the patient's presentation was most consistent with chronic myopathy. Although active sarcoid myositis was not clinically suspected, as in most cases because of nonspecific symptoms, FDG-PET/CT scan revealed active muscle involvement. In patients with chronic myopathy, initial treatment with glucocorticoids gradually tapered over 6 to 12 months is recommended. In patients with an inadequate response following 3 months of glucocorticoids, treatment is escalated with the addition of an immunosuppressive agent such as methotrexate or azathioprine. If myopathy remains refractory, tumor necrosis factor (TNF) inhibitors such as infliximab and adalimumab have been shown to be effective in case reports.[7,8] Indeed, TNF-α is recognized to play a role in initiating and maintaining granulomas.[7,8] Literature regarding the ability of FDG-PET/CT imaging to assess treatment response in sarcoid myositis is very limited. Both Han et al and Dhomps et al reported cases of complete response of sarcoid myositis on follow-up FDG-PET/CT scans, 18 and 4 months following initiation of glucocorticoids, respectively.[5,9] Marie et al reported a case of sarcoid myositis refractory to prednisone and methotrexate with favorable outcome 6 months following initiation of infliximab as evidenced by normalization of abnormal muscle FDG uptake.[7] Our case is original as our patient had refractory muscular sarcoidosis despite being treated with all three mainstays of treatment. FDG-PET/CT was shown to be useful in assessing positive response as early as a week following dosage optimization. As available data remains scarce, further investigations are necessary to determine the role of FDG-PET imaging in muscular sarcoidosis follow-up.

Conflict of Interest
None declared.

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