Case Report

Tiger Man Sign-F-18 FDG PET CT Scan Pattern in Muscular and Cutaneous Sarcoidosis: A Case Report and Literature Survey

Abstract

Sarcoidosis is a chronic granulomatous disorder of unknown etiology which primarily affects the respiratory system. However, 0.5%–2.5% of patients with sarcoidosis show muscle involvement, namely sarcoid myopathy. F-18 Fluorodeoxyglucose positron-emission tomography (F-18 FDG PET) has become an important component of the diagnostic algorithm of these patients, owing to its ability to assess disease extent and identify occult sites of disease involvement and guiding sites of biopsy. Awareness of pattern of FDG uptake in sarcoid myopathy not only helps in identifying muscular involvement in already known cases but also helps in the initial diagnosis of sarcoidosis as in the present case.

Keywords: *FDG PET-CT, sarcoid myopathy, sarcoidosis, Tiger man sign*

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Introduction

Sarcoidosis is a chronic granulomatous disorder of unknown etiology characterized by the accumulation of lymphocytes and monocytes in the affected organs.[1] Lungs are the most frequently involved organs followed by the lymph nodes, skin, eyes, heart, nervous system, and musculoskeletal system. The diagnosis is established on the presence of clinical symptoms, chest X-ray findings, histolopathology report and exclusion of other granulomatous infections, especially tuberculosis.[1] Owing to its ability to detect active inflammation throughout the body with a high sensitivity, FDG PET has gained a vital role in management of sarcoidosis.[2] FDG PET is useful for the assessment of reversible granuloma, occult disease, disease extent, treatment response, and guiding the biopsy site.[3] We report a case where F-18 FDG PET computed tomography (CT) scan findings helped in diagnosing sarcoidosis with rare muscular and cutaneous involvement.

Case Report

We report a case of a 65-year-old female previously diagnosed with interstitial lung disease (ILD) for the past 10–12 years, with on and off steroid treatment. She was admitted twice in our hospital in subsequent months with complaints of dyspnea, generalized weakness, significant weight loss, and myalgia. A provisional diagnosis of ILD with type II respiratory failure was made. Blood investigations revealed hypercalcemia (15.9 mg/dl), hypokalemia (3.3 mg/dl), and hyperuricemia (7.4 mg/dl). Complete blood counts picture revealed anemia (Hb ~9.6 g/dl) with low total leukocyte counts and red blood cell counts. However, there was evidence of monocytosis and increased erythrocyte sedimentation rate (ESR) (~70 mm/h). Serum phosphorous, parathyroid hormone, and Vitamin D levels were within the normal limits. A medical oncology opinion was also sought in view of suspicion of malignancy. Tumor markers such as serum lactate dehydrogenase and CA-125 were unremarkable. Transvaginal ultrasound was suggestive of atrophic vaginitis. Pap smear was suggestive of reactive cellular changes. Later, an F-18 FDG PET CT scan was requested to rule out any occult malignancy.

On the day of the scan, the patient was bedridden, appeared cachectic, and was on bi-level positive airway pressure respiratory support. We performed whole-body F-18 FDG PET contrast-enhanced CT scan

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from the vertex to the toes on Siemens Biograph Horizon Time-of-Flight PET CT using standard patient preparation and acquisition protocol. The patient's renal functions were within normal limits.

FDG PET CT scan demonstrated metabolically active symmetric peribronchovascular and perihilar ground-glass opacities and consolidative lesions in bilateral lung fields with diffuse interstitial thickening with metabolically active calcified mediastinal and abdominopelvic lymph nodes [Figure 1]. Another striking finding on FDG PET CT scan was severe atrophy of skeletal muscles predominantly of the lower half of the body with numerous metabolically active (maximal standardized uptake value – 5.3) cutaneous, subcutaneous, and intramuscular lesions at multiple sites in the body [Figure 2]. A possibility of granulomatous involvement, likely sarcoidosis, was raised and histopathological correlation from hypermetabolic skeletal muscle lesion/mediastinal lymph node was suggested. No scan evidence of mitotic disease was noted in the FDG PET CT scan. Her serum angiotensin-converting enzyme (ACE) levels were subsequently found elevated (65 U/L). Trucut biopsy performed from the hypermetabolic lesion in gluteal muscle demonstrated occasional histiocytic cell clusters and multinucleated giant cells with cells showing intracytoplasmic calcification (? Schaumann body). A diagnosis of sarcoidosis was confirmed in the histopathology report [Figure 3].

Hence, a clinical diagnosis of advanced stage sarcoidosis with muscular and cutaneous involvement was established.

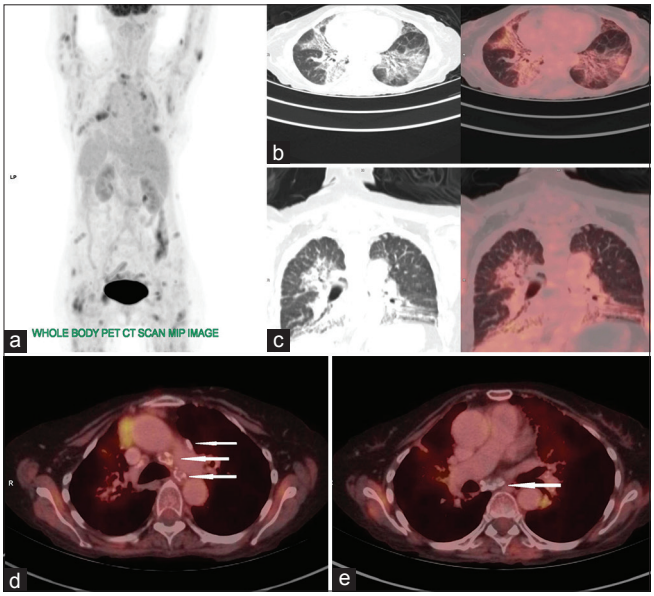

Figure 1: (a) F-18 FDG PET CT scan in maximal intensity projection (MIP) view shows pathological fluorodeoxyglucose uptake in bilateral lung fields and mediastinal lymph nodes and numerous sites of linear and patchy FDG uptake in the skeletal muscles and cutaneous tissue (b and c) are axial CT and fused PET CT images demonstrating metabolically active symmetric peribrochovascular and perihilar ground-glass opacities and consolidative lesions in bilateral lung fields. (d and e) show calcified mediastinal lymph nodes (white arrows)

She was treated with oral steroids, hydroxychloroquine, and other supportive medications and was later discharged. We checked her subsequent clinical course in electronic medical records at the time of writing this case report. She reported for follow-ups with the chest physician for few months after the scan. However, she died of severe respiratory failure 2 months back.

Discussion

Sarcoid myopathy or involvement of muscles in sarcoidosis is a rare entity. Symptomatic muscle involvement has been described in approximately 0.5%–2.3% of sarcoid patients only.[4-7] Three different types of myopathy were described: chronic myopathy, palpable nodules, and acute myositis.[2,8-10] Chronic myopathy is the most frequent type of symptomatic muscle sarcoidosis.[11] Chronic myopathy occurs mainly in elderly women and is characterized by slow progressive symmetrical weakness and atrophy of the proximal muscle groups.[3] In this case, the patient presented with features similar to chronic myopathy.

Wieërs *et al*. in 2012 described FDG PET scan pattern in a patient with sarcoid myopathy. The scan demonstrated multiple linear and patchy hypermetabolic lesions in skeletal muscles, mimicking the coat of a tiger and was referred to as *"A tiger man"* sign.[12] This sign has been sparingly mentioned in few case reports published later on.[13-16]

In the present case, the patient was previously diagnosed with ILD and presented with respiratory distress, generalized weakness, and myalgia. The patient's blood investigations revealed monocytosis, elevated ESR values, ACE levels, and hypercalcemia. FDG PET scan findings showed bilateral lung lesions and calcified mediastinal

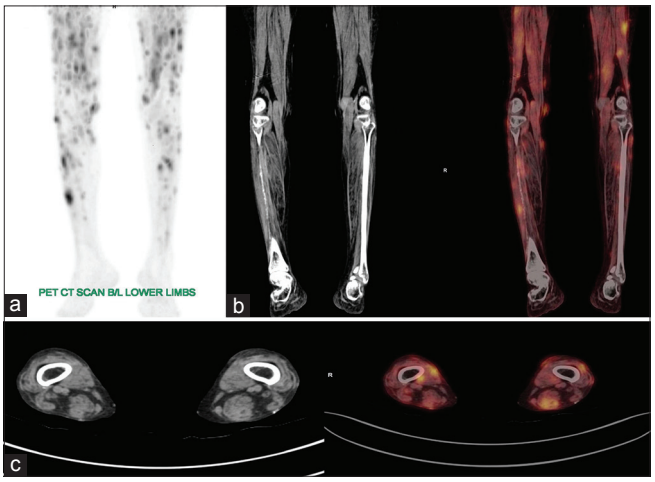

Figure 2: (a) 18FFDG PET CT scan in maximal intensity projection (MIP) view of bilateral lower limbs shows numerous sites of linear and patchy FDG uptake (maximal standardized uptake value – 5.3) in the skeletal muscles and cutaneous tissue in the bilateral lower limbs. (b and c) are coronal and axial CT and fused PET CT images demonstrating diffuse muscle atrophy with multiple metabolically active lesions in skeletal muscles and cutaneous tissue (white arrow)

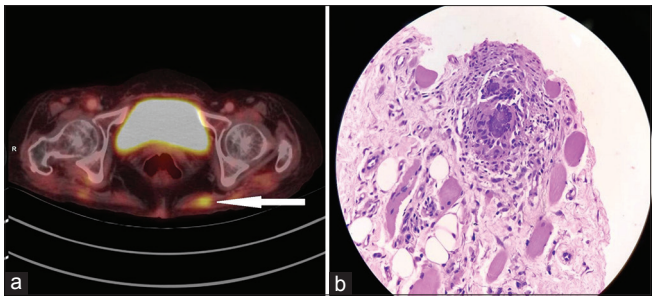

Figure 3: (a) is an axial fused positron-emission tomography computed tomography image depicting the site of biopsy from metabolically active left gluteal nodule (white arrow). (b) is the histopathology image (H and E, 40X) showing skeletal muscle bundles with occasional plump histiocytic cell clusters and multinucleated giant cells, occasional giant cells showing intracytoplasmic calcification (? Schaumann body), suggestive of sarcoidosis

lymphadenopathy along with numerous linear and patchy hypermetabolic lesions in skeletal muscles and cutaneous and subcutaneous tissue, also referred to as the "Tiger Man sign." The clinical picture, laboratory investigations in this case along with typical F-18 FDG PET CT scan pattern, helped in clinching a definitive diagnosis of sarcoidosis. Furthermore, FDG PET CT scan guided the site of muscle biopsy.

The present case highlights the value of F-18 FDG PET CT scan in the management of sarcoidosis and in detecting previously unknown sites of disease involvement. With the increased availability and utilization of F-18 FDG PET CT scan, there are higher chances of identification of such rare organ involvements such as sarcoid myopathy.

Conclusion

"Tiger man sign" is a hallmark sign of sarcoid myopathy. The prompt recognition of this sign is essential for nuclear medicine physicians to guide the treating physicians in reaching a quick and definitive diagnosis of sarcoid myopathy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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