

Case Report

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Incidental Detection of Systemic Vasculitis in A Sarcoidosis Patient on FDG PET/CT scan: A Case Report

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Abstract

Sarcoidosis is a rare disease with variable clinical presentation as it can involve different organ systems. Systemic vasculitis is a rare presentation of extra pulmonary sarcoidosis that can be fatal if undiagnosed. FDG PET/CT scan is a great modality in detecting both pulmonary and extrapulmonary manifestation of sarcoidosis. In this case report, we present a case of a 47-year-old lady with underlying pulmonary sarcoidosis stage 2 with sick sinus syndrome, who underwent FDG PET/CT scan to rule out cardiac sarcoidosis. The ¹⁸F-FDG PET/CT scan showed FDG avid disease in the lungs, lymph nodes and the vessels, suggestive of systemic vasculitis. If undiagnosed and untreated, it may lead to development of aneurysm and dissection. Immunosuppressive treatment using steroids and cytotoxic therapy can be life-saving in this condition and may lead to remission in some of the patients.

Keywords: sarcoidosis; extra pulmonary sarcoidosis; systemic vasculitis; malaysia

Introduction

Sarcoidosis is a multisystem inflammatory disease of unknown etiology. It is characterized by formation of noncaseating granuloma [1]. Classic site of involvement are the lungs and thoracic lymph nodes [2]. The clinical features and imaging features are variable depending on the site of involvement and degree of inflammation. The consideration whether to start the immunosuppressive treatment or not, for sarcoidosis is depending on the presence of reversible granuloma or active inflammation [3]. Sarcoidosis may involve multiple organ system including pulmonary, skin, ocular, central nervous system, cardiovascular, gastrointestinal, genitourinary and musculoskeletal system [4]. In view of whole-body scan nature of FDG PET/CT, it has become an excellent modality to detect the presence of active sarcoidosis both within the thoracic and outside of the thoracic region. In this case report, we present an incidental finding of systemic vasculitis in sarcoidosis, for a patient that was initially planned to assess the cardiac sarcoidosis using FDG PET/CT scan.

Case Report

We report a case of a 47-year-old lady with underlying hypertension. She presented with reduced effort tolerance since 2017. Chest x-ray showed features of stage 2 pulmonary sarcoidosis. The left upper lobe lung biopsy was done on 30/11/2017 which showed granulomatous inflammation. The patient was diagnosed with pulmonary sarcoidosis stage 2. Eventually, the patient was also diagnosed with sick sinus syndrome and pacemaker was implanted in 2017. Patient was started on oral prednisolone from December 2017 until January 2019 which was then stopped in view of improving clinical symptoms. However, since early 2022, she developed reduced effort tolerance again. HRCT thorax was done on 27/05/2022 that showed stable mediastinal lymphadenopathy and bilateral upper lobe residual lung parenchymal changes. Cardiac MRI on 16/08/2022 showed no evidence of cardiac sarcoidosis. She was then referred to Department of Nuclear Medicine, National Cancer Institute for ¹⁸F-FDG PET/CT scan for assessment of intrathoracic disease activity (active inflammation vs fibrosis) and to rule out cardiac involvement of sarcoidosis. Whole body ¹⁸F-FDG PET/CT scan with cardiac protocol was done on 07/09/2022.

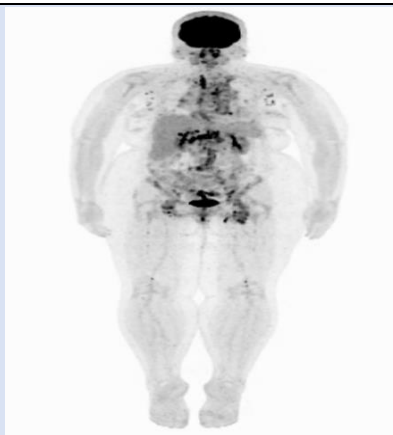


Figure 1: Maximum intensity projection image of ^{18}F -FDG PET/CT scan of this patient showed multifocal ^{18}F -FDG hypermetabolism involving bilateral lungs, vessels at mediastinum, abdominopelvic, bilateral inguinal and bilateral common femoral region. There are also multifocal ^{18}F -FDG hypermetabolism involving the lymph nodes at mediastinal, bilateral axillary, abdominopelvic and bilateral inguinal region.

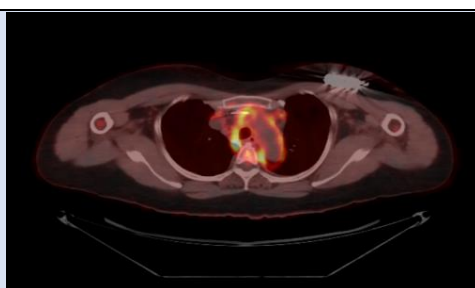


Figure 2: FDG PET/CT scan image, axial view at the aortic arch level shows FDG hypermetabolism along the wall of the aortic arch, likely representing sarcoid vasculitis.

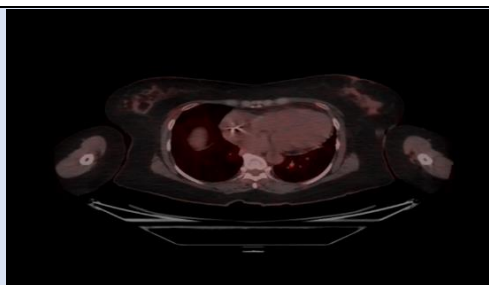


Figure 3: FDG PET/CT scan image, axial view at the heart level shows no FDG hypermetabolism in the myocardium indicating no active sarcoidosis at the heart.

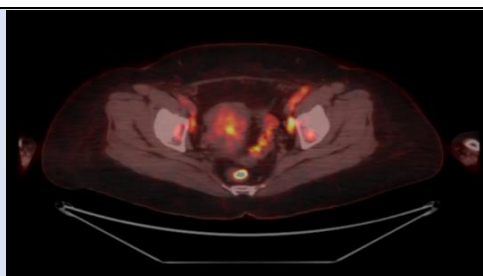


Figure 4: FDG PET/CT scan image, axial view at bilateral external iliac level shows FDG hypermetabolism within the wall of bilateral external iliac vessels, likely represent sarcoid vasculitis.

Discussion

Sarcoidosis is a relatively common disease in the West but quite uncommon in Malaysia. ACCESS study reported incidence of sarcoidosis as 35 to 80 per

100,000 African American, 15 to 20 per 100,000 Northern Europeans and 3 to 10 per 100,000 among European American. The epidemiology of sarcoidosis in Malaysia is largely unknown due to its rarity [5].

The disease remains as an imaging challenge due to its rare prevalence and due to some overlapping imaging features with more common diseases in Malaysia such as pulmonary tuberculosis and lymphoma. Chest x-ray is the first line imaging test, which are graded as stage I for hilar or mediastinal lymphadenopathy only, stage II for hilar or mediastinal lymphadenopathy plus lung parenchymal disease, stage III for lung parenchymal disease only and stage IV for pulmonary fibrosis [6].

If other organ systems are suspected to be involved, other diagnostic modalities can be used, such as cardiac MRI, contrast enhanced CT scan or ^{18}F -FDG PET/CT scan. Common indication for ^{18}F -FDG PET/CT scan includes detection of reversible granuloma (FDG avid active sarcoidosis), detection of occult active disease, assessment of treatment response, evaluation of optimal biopsy site as well as to rule out cardiac, nervous system and musculoskeletal sarcoidosis [3]. In a post treatment setting, FDG PET/CT scan is useful in disease reassessment to look for post treatment response. The resolution of previously FDG avid lesion without new FDG avid lesion is an indication of a good response to treatment [3].

The decision to start or not to start the treatment is complex. This is due to significant portion of sarcoidosis patients do not require treatment as most of the patients are asymptomatic [7]. However, some types of sarcoidosis are related with more serious complication such as in sarcoid vasculitis, where the disease involves the vascular system, as the involved vessels may become aneurysm with risk of dissection if left untreated [8]. The other type of sarcoidosis that require prompt treatment include ocular sarcoidosis which may lead to vision loss in delayed treatment. For renal sarcoidosis, delayed in treatment can lead to development of renal insufficiency [9].

The decision to start immunosuppressive treatment for sarcoidosis patients is dependent on the presence of reversible granuloma. Reversible granuloma in sarcoidosis is active sarcoidosis that showed FDG avidity on ^{18}F -FDG PET/CT scan. The presence of reversible granuloma may predict good clinical response post immunosuppressive treatment. ^{18}F -FDG PET/CT scan is an excellent tool for its detection, in view of FDG hypermetabolism in the reversible granuloma [10]. HRCT thorax cannot differentiate between lung fibrosis and reversible granuloma in stage IV sarcoidosis. Cardiac MRI is good to detect fibrosis within the myocardium with

late gadolinium contrast enhancement, however, it cannot provide the information of reversible granuloma or non-reversible granuloma (active sarcoidosis or fibrosis). Besides that, full body scan nature of ^{18}F -FDG PET/CT may detect occult active sarcoidosis disease outside the thorax, which is uncommon in sarcoidosis but important to be diagnosed and treated. For cardiac ^{18}F -FDG PET/CT scan, sarcoidosis findings are divided into four grades; Grade 1 for no perfusion and FDG uptake, grade 2 for patchy FDG uptake with normal or slightly decreased perfusion, grade 3 for FDG uptake at hypoperfusion area and grade 4 for severe perfusion defect with no or minimal FDG uptake [11].

In this case, the initial indication was to assess active sarcoidosis in the thorax and cardiac involvement. The ^{18}F -FDG PET/CT scan showed FDG avid disease in lungs, lymph nodes, and vessels with no evidence of active cardiac sarcoidosis. The differential diagnosis for these findings includes systemic vasculitis in sarcoidosis (sarcoid vasculitis), systemic lupus erythematosus, polyarteritis nodosa and Takayasu's arteritis. Clinical and HPE correlation is needed to confirm the exact diagnosis as the imaging findings of these conditions are generally similar. However, the diagnosis for this patient is more towards sarcoid vasculitis, in view of the patient's underlying pulmonary sarcoidosis that was previously confirmed by HPE. Detection of sarcoid vasculitis is important, as the untreated disease is associated with poor prognosis due to risk of aneurysm and dissection. Immunosuppressive therapy such as corticosteroid with cytotoxic agent can be lifesaving in this condition [8].

Conclusion

Although ^{18}F -FDG PET/CT scan is not the first line modality for establishing the diagnosis of sarcoidosis, it remains as a very valuable tool in suspicious active sarcoidosis that responds well to immunosuppressive therapy, extrapulmonary disease, focal cardiac involvement, and to assess the high-risk form of sarcoidosis such as sarcoid vasculitis.

Competing interests

The authors declare that they have no competing interests.

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