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Short Communication

Whole Body FDG PET-CT in the Evaluation and Management of Cardiac Sarcoidosis and Isolated Cardiac Sarcoidosis

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Introduction

Sarcoidosis is a multisystem inflammatory disease defined histologically by the formation of non-caseating granulomas. Cardiac involvement can be seen in up to 5% of cases [1]. These patients are at a high risk for major cardiac events [1]. Diagnosing and monitoring Cardiac Sarcoidosis (CS) is not trivial but can be currently accomplished by a combination of cardiac MRI and cardiac FDG PET-CT scanning [2]. With appropriate patient preparation, cardiac FDG PET has a high sensitivity for detecting cardiac lesions but also extra-cardiac lesions and for monitoring the efficacy of treatment.

Discussion

Current standard clinical practice consists of acquiring dedicated, limited field of view cardiac FDG PET scans. This would cover the heart and immediately adjacent thoracic structures. Although this is of great value and used with great success to diagnose and monitor cardiac sarcoidosis, it fails to assess the patient's disease in a holistic manner. Still with this limited field of view, several reports including ours have described a high rate of extra-cardiac findings. We reviewed a series of sixty-five PET-CT scans of 54 patients referred for CS evaluation. These were performed between September 2010 and April 2013 at Yale New Haven Hospital and we found extra-cardiac findings were present in 92.3% of scans (n=60). Overall disease distribution is summarized in (Table 1). Highlight some of these extra-cardiac findings (Figures 1-4). 52.3% of these patients were asymptomatic at the time of the scan. CS was diagnosed in 50.8% of patients (n=33). All the patients (100%) with CS had extra-cardiac findings. Additionally, extra-cardiac findings were present in 48.3% of the scans when there was no cardiac involvement. In our cohort, we had no cases of Isolated Cardiac Sarcoidosis (ICS).

Baughman et al., reported on findings from the A Case Control Etiologic Study of Sarcoidosis (ACCESS) [3]. This multicenter study evaluated distribution of disease in 736 incident cases with sarcoidosis. One should note that this preceded the widespread use of FDG PET in clinical practice and the true distribution of disease is likely different when highly sensitive techniques such as FDG PET are used. Although ICS is described in the literature with an expected

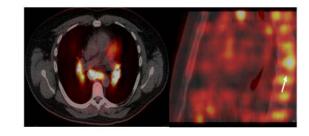


Figure 1: 52 year old asymptomatic gentleman. FDG PET was negative for CS, however FDG avid mediastinal and hilar lymphadenopathy was seen in addition to focal skeletal uptake (white arrow).

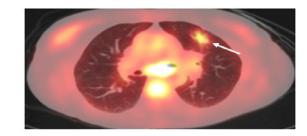


Figure 2: 55 year old symptomatic lady with palpitations. FDG PET negative for CS. FDG-avid subcarinal lymph node and left upper lobe nodule seen (white arrow).

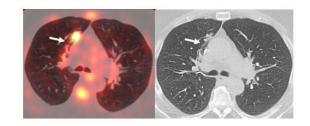


Figure 3: 69 year old lady presenting with ventricular tachycardia and near syncope. CS was suspected. However, there was no evidence of CS on FDG PET. An FDG-avid lung nodule seen incidentally found and it was only partially visualized (white arrows). The patient underwent a diagnostic chest CT, which demonstrated a nodule with suspicious features for primary lung cancer (red arrow). A wedge resection was performed and the pathologic evaluation revealed organizing pneumonia.

high prevalence ranging from 29-52% of cases of CS [4]. It seems that in the absence of proper staging during the CS workup one may overestimate the cases of ICS. Giudicatti et al., looked into the impact of whole body FDG PET in assessing the true prevalence of ICS [4]. They found it to be much lower at about 9.4%. Juneau et al in their study on the other hand found the prevalence to be even lower at 3.2% [5]. This was much closer to our cohort where we actually found

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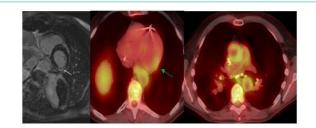


Figure 4: 73 year old asymptomatic gentleman with known sarcoidosis. Cardiac MRI study performed 1 year ago showed infero-lateral wall enhancement (white arrow). FDG PET showed increased uptake in the same region (green arrow). In addition, he had diffuse FDG uptake along the thoracic aorta.

Table 1: Distribution of extra-cardiac findings on cardiac sarcoid FDG PET scans.

	Percentage
Mediastinal/Hilar lymphadenopathy	66.10%
Proximal stomach/esophagus	43.10%
Bone marrow uptake	35.40%
Thoracic aorta	16.90%
FDG avid lung nodules	16.90%
Focal skeletal uptake	10.80%
Non-FDG avid lung nodules	3.10%

no cases of ICS.

Ishiyama et al., also noted in their study of 16 positive CS cases a high propensity for extra-cardiac disease (81.3%) [1]. Their study also hinted at the potential prognostic value of extra-cardiac disease. Furthermore, extra-cardiac uptake was reported to increase the likelihood of finding CS by Tuominen et al., [6]. Considering that in our cohort all CS patients had extra-cardiac findings it is possible to miss extra-cardiac disease due to a limited field of view or incomplete staging or improper interpretation of the scans. In order not to miss any disease that would increase the diagnostic certainty of having CS as suggested by Tuominen et al., [6], one should strongly consider doing a whole body PET in cases of suspected CS. In these cases, the patient is already being injected with FDG to perform the cardiac study and the additional whole body acquisition mostly adds some additional time to the patient's acquisition. With newer scanners, this should not be a limiting factor. Higushi et al., [7] reported on a case of large vessel aortitis in a patient with CS as we have also encountered and demonstrated in (Figure 4). Extra-cardiac findings such as this one may have a significant prognostic implication.

Conclusion

Considering the high incidence of extra-cardiac findings in patients undergoing a cardiac sarcoid FDG PET scan, it may seem advisable to perform a whole body FDG PET acquisition in order to properly assess these patients. This would have a high impact for accurate initial staging, monitoring of treatment, assessing differential treatment responses in different aspects of the disease. This also allows for more adequate biopsy sites selection and a more holistic evaluation of the patient.

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