Use of multimodality imaging to diagnose cardiac sarcoidosis as well as identify recurrence following heart transplantation

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A middle-aged man presented with a ventricular fibrillation arrest and five months later developed complete heart block requiring placement of permanent pacemaker and defibrillator. Coronary angiography showed no evidence of coronary artery disease. MRI demonstrated inflammatory lesions within the liver and spleen and subsequent mediastinoscopy with lymph node biopsy was diagnostic for sarcoidosis. The patient developed worsening symptoms of heart failure and was subsequently referred for a cardiac PET study to evaluate for cardiac sarcoidosis. Rest perfusion images using rubidium-82 identified the presence of a regional perfusion defect along the basal and mid inferior wall (Figure 1), while the fluorodeoxyglucose (FDG) images (acquired after high fat and low carbohydrate diet to suppress FDG uptake by normal myocardium without heparin infusion[1]) showed heterogeneous and patchy FDG uptake involving the entire myocardium, with notable focal areas of more intense FDG uptake corresponding to focal perfusion defects.

Despite steroid therapy, the patient's heart failure progressed and, ultimately, he underwent an orthotopic heart transplant approximately seven months later. Gross pathology of the explanted heart (Figure 2, gross) demonstrated moderate biventricular dilatation with multiple foci of dense fibrosis involving the myocardium of both ventricles and the interventricular septum. Microscopic evaluation revealed multiple non-necrotizing granulomata with mild chronic inflammation and associated serpiginous fibrosis (Figure 2, microscopy). The imaging and gross pathological findings were concordant in identifying the presence and the location of perfusion defects due to fibrosis, while microscopic evaluation demonstrated the presence of active inflammation.

Four months after transplantation, the patient developed reactivation of his sarcoidosis within the native pericardium and required pericardiectomy due to constrictive physiology. Focal granulomatous infiltration of the pericardium was noted on pathology. Subsequently, his cardiac function remained stable for 3 years. During this interval, his immunosuppressive medications and steroids were adjusted in light of other medical problems, including opportunistic infections of his skin and extremities. During a surveillance PET scan performed three years post OHT, he was found to have a new medium-sized perfusion defect of moderate intensity throughout the inferolateral and anterolateral walls with associated increased FDG uptake that is indicative of perfusion metabolic mismatch (Figure 3, panels A and B), and is suggestive of possible recurrence of cardiac sarcoidosis. This finding also correlated with a cardiac MRI, which identified small amount of fibrosis along the inferolateral wall (Figure 3, panel C). A subsequent right and left heart catheterization showed no evidence of coronary disease. A biopsy done on the same day as his positive PET scan did not demonstrate any evidence of rejection, granulomatous disease, infection, or ischemic changes.

This article illustrates the role of non-invasive imaging in the evaluation of patients with suspected cardiac sarcoidosis and provides a unique comparison between PET/CT findings and pathology. In this case, the PET/CT study was used to identify the presence of myocardial inflammation, and subsequently led to the diagnosis of cardiac sarcoidosis, which was confirmed with pathology. In addition to diagnosis of cardiac sarcoidosis, PET/CT can be used to identify extracardiac

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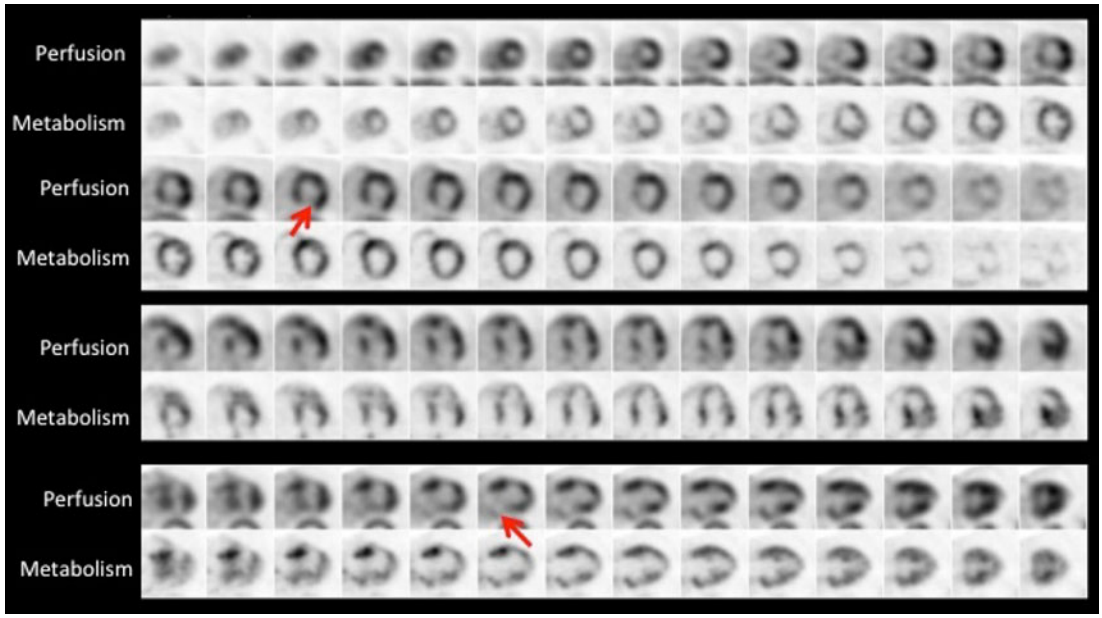

Figure 1. PET CT before heart transplantation. Rest rubidium-82 perfusion images show the presence of scar along the basal and mid inferior wall (*red arrow*) as well as the distal septum. The FDG images show heterogeneous uptake throughout the myocardium. There is FDG uptake in both areas of reduced as well as normal perfusion, consistent with the presence of active inflammation as well as scar.

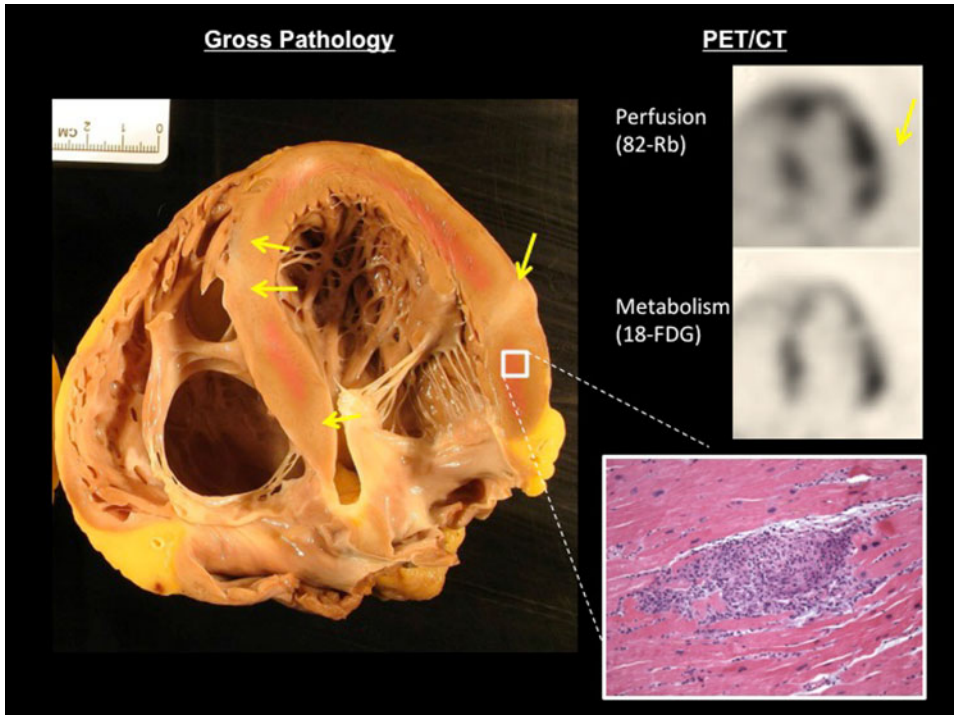

Figure 2. Pathology of explanted heart showing gross and microscopic changes associated with cardiac sarcoidosis. The gross specimen demonstrates moderate biventricular dilatation with multiple white fibrotic plaques (*arrows*), involving the midwall and epicardium of both ventricles. Microscopy shows patchy fibrosis with chronic inflammation and occasional non-necrotizing granulomata (histologic *inset*). Of note, the areas of fibrosis in the gross specimen correspond to areas of scar identified on PET/CT imaging. Hematoxylin- and eosin-stained sections, ×100 original magnification.

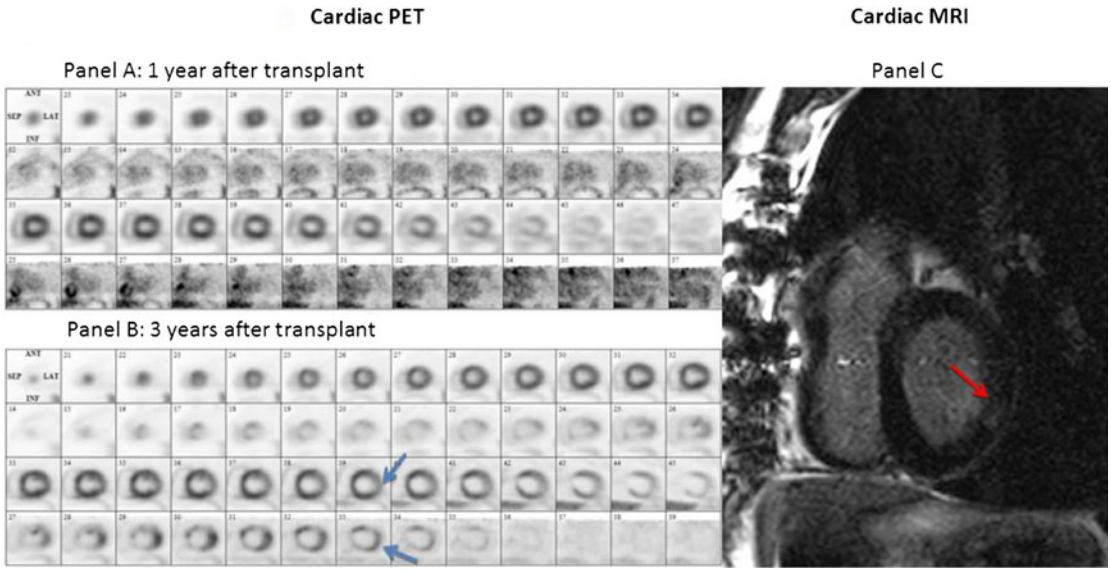

Figure 3. PET CT and cardiac MRI following heart transplantation. *Panel A* One year post transplantation, PET/CT showing normal perfusion with no FDG uptake. *Panel B* Two years later, PET/CT identified a medium-sized perfusion defect of moderate intensity (*arrow*) involving the basal inferolateral and anterolateral walls which was associated with increased FDG uptake (*arrow*). This pattern of perfusion metabolic mismatch has been described in cardiac sarcoidosis. *Panel C* Corresponding to these findings, cardiac MRI identified a small amount of late gadolinium enhancement along the basal inferolateral wall, suggesting the presence of scar.

inflammation and can also be used to assess for response to anti-inflammatory therapy.[2,3] Unlike CMR, PET/CT imaging is feasible in patients with defibrillators and pacemakers.

Three years later, findings suggestive of reactivation of cardiac sarcoidosis within the transplanted heart were identified, as visualized by both PET and CMR. To our knowledge, this is the first reported case of PET/CT imaging used to identify cardiac sarcoidosis recurrent after orthotopic heart transplantation.

Disclosures

None.

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