Role of PET/CT in multimodality imaging in differentiating cardiac sarcoidosis from arrhythmogenic right ventricular dysplasia

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A 42-year-old otherwise healthy man was cardioverted after presenting with haemodynamically significant conscious ventricular tachycardia (VT). ECG revealed a left bundle-branch block (LBBB) pattern VT of 200/min. Baseline ECG showed PR-prolongation, Epsilon waves, and precordial T-wave inversion (Figure 1), suggestive of arrhythmogenic right ventricular dysplasia (ARVD).[1] Diagnostic coronary angiography showed normal coronary arteries. Transthoracic echocardiogram revealed normal left ventricular (LV) size with mild global systolic dysfunction, a dilated right ventricle (RV) with severe systolic dysfunction and severe right atrial dilatation with minimal tricuspid regurgitation.

Cardiac MRI confirmed the echocardiographic findings. Significant late gadolinium enhancement (LGE) was detected in the RV free wall, with mild LV involvement (Figure 2). He was discharged after insertion of an implantable cardiac defibrillator.

In light of RV dysfunction, a CTPA ruled out pulmonary embolus, but mediastinal lymphadenopathy raised the possibility of sarcoidosis. Subsequent cardiac-prepared PET/CT revealed heterogeneous cardiac FDG uptake. Incidentally, there was extensive FDG-avid lymphadenopathy (Figure 3). A Tc-99m sestamibi SPECT revealed myocardial metabolism-perfusion mismatch (Figure 4), consistent with sarcoidosis. He was commenced on prednisolone and methotrexate. Three months later, repeat PET/CT showed complete metabolic response (Figure 5).

This case illustrates the importance of multimodality imaging in differentiating between ARVD and cardiac sarcoidosis (CS). The two diagnoses will lead to different management strategies, including immunosuppressants and/or genetic testing. CS patients are normally older, more likely to have PR-prolongation, with significant LV dysfunction and mediastinal lymphadenopathy.[2]

Although our patient presented with LBBB VT and ECG suspicious of ARVD, the presence of PR-prolongation, mediastinal lymphadenopathy, and LV dysfunction with LV LGE are more consistent with CS, further supported by a positive PET/CT scan. This case demonstrates how multimodality cardiac imaging has been instrumental in altering the diagnosis of ARVD to CS and impacting on patient management.

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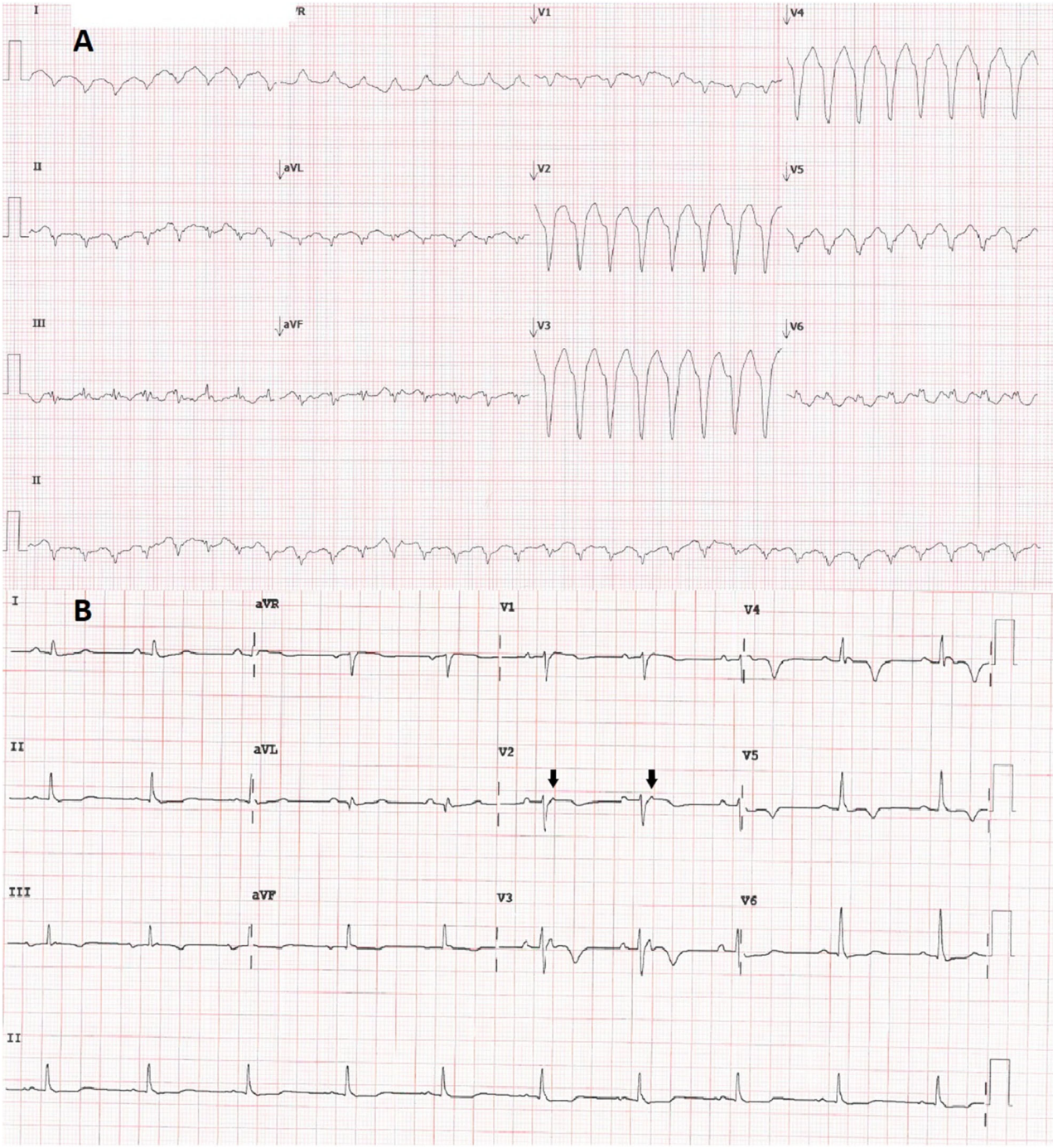

Figure 1. ECG. **A** Electrocardiogram (ECG) in a 42-year-old man presenting with palpitations and presyncope with blood pressure of 70/40 mmHg. This ECG revealed broad complex tachycardia of left bundle-branch block morphology, rate approximately 200/min with an equivocal axis. **B** Baseline ECG after successful electrical cardioversion. This shows sinus bradycardia and PR-prolongation (first degree heart block). The presence of Epsilon waves (arrow) and precordial T-wave inversion is suspicious of ARVD.

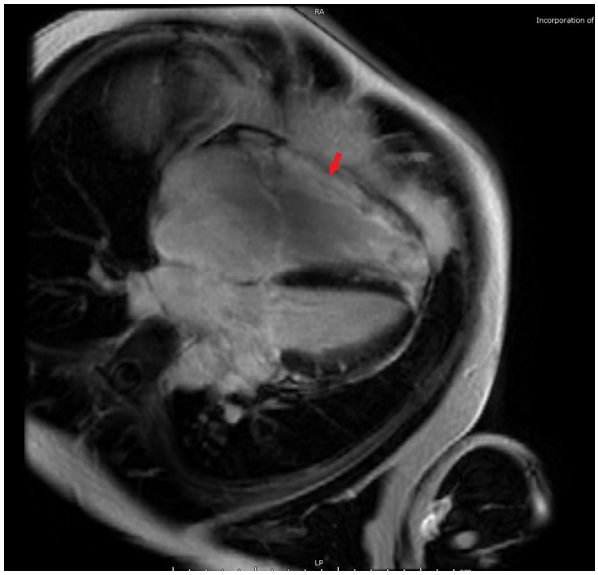

Figure 2. Cardiac MRI. Cardiac MRI demonstrated dilated right ventricle (RV) with thickened wall and reduced RV systolic function. Delayed gadolinium imaging showed prominent transmural RV hyperenhancement, particularly in the RV free wall (arrow). Additional hyperenhancement was also noted in the LV wall, predominantly in a subepicardial distribution in the LV apex, with speckled mid wall involvement.

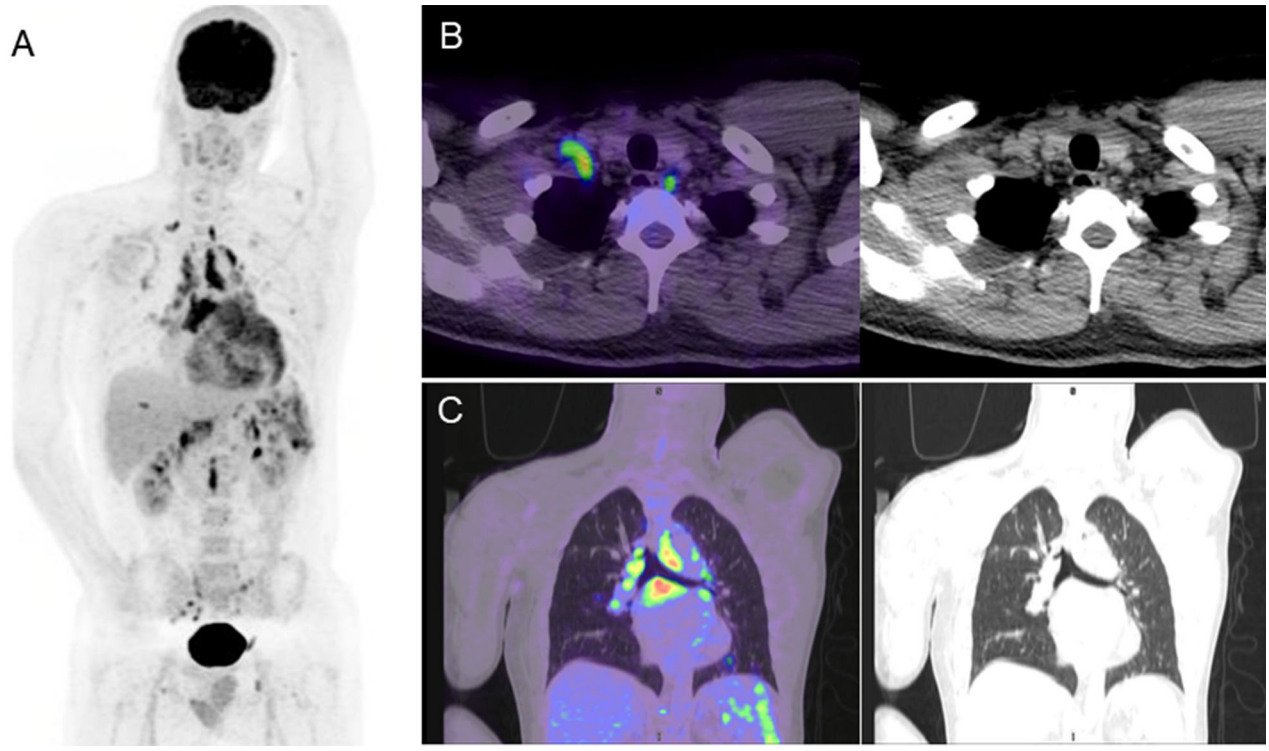

Figure 3. Whole body PET/CT. The patient underwent a 48-hour high-fat, low-carbohydrate diet, followed by 12 hours of fasting and 2500 units of IV heparin prior to Fluorodeoxyglucose (FDG) administration. The maximum intensity projection (**A**) demonstrated abnormalities in a biodistribution characteristic of active sarcoidosis including high-intensity metabolic activity in supraclavicular, mediastinal, and paraaortic nodes. There was also focal uptake in pulmonary opacities and multifocal abnormalities in the spleen and myocardium. **B**, **C** demonstrate representative axial (**B**) and coronal (**C**) PET-CT slices.

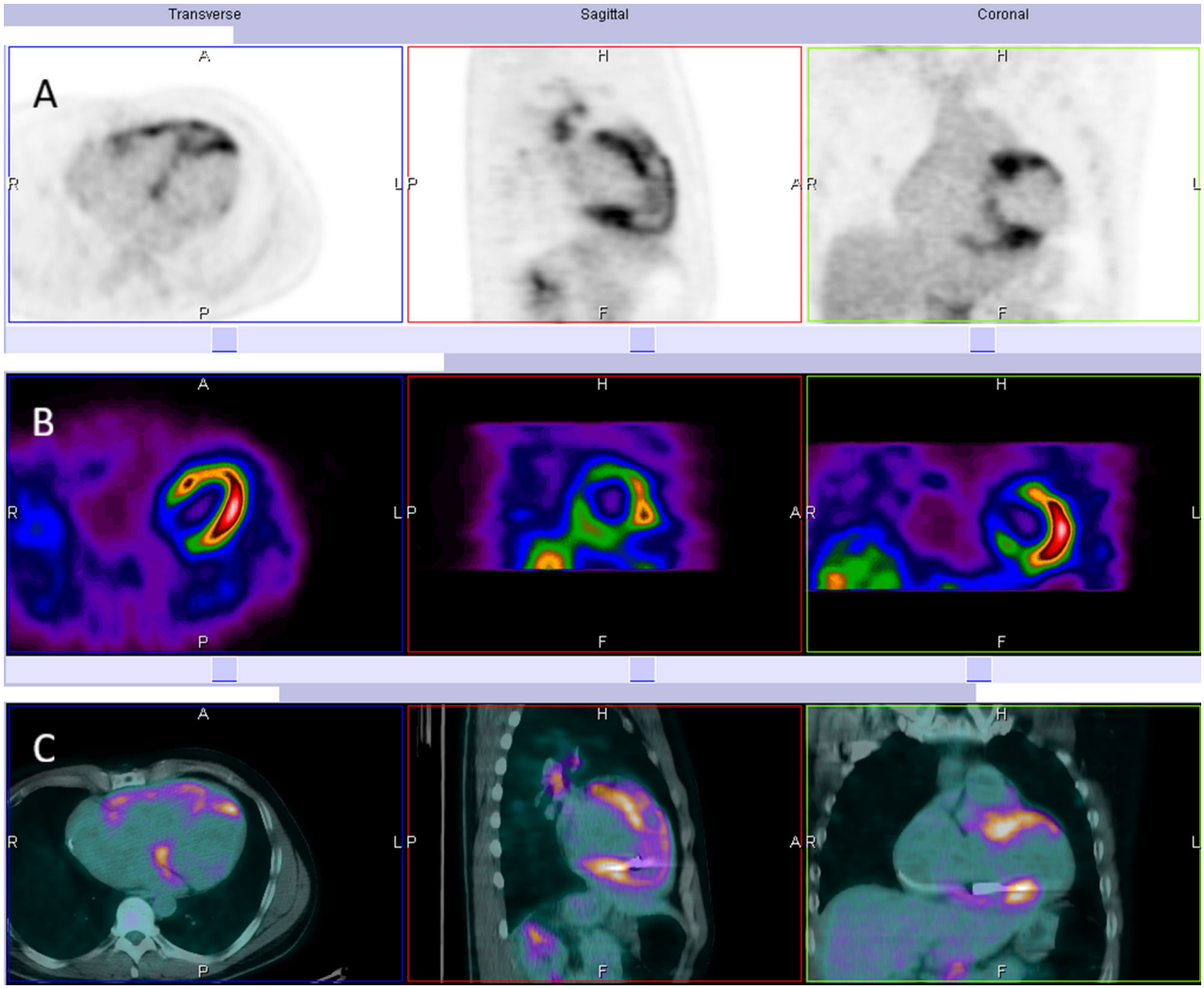

Figure 4. PET/Tc-99m sestamibi SPECT fused images. PET scan (**A**) showed FDG uptake in the LV apex, interventricular septum and RV free wall. SPECT images (**B**) showed a reduction in myocardial perfusion in the corresponding regions. In the setting of normal coronary angiography (excluding hibernating myocardium as a question), the metabolism-perfusion mismatch is suggestive of cardiac sarcoidosis. **C** Shows the PET-CT fused images.

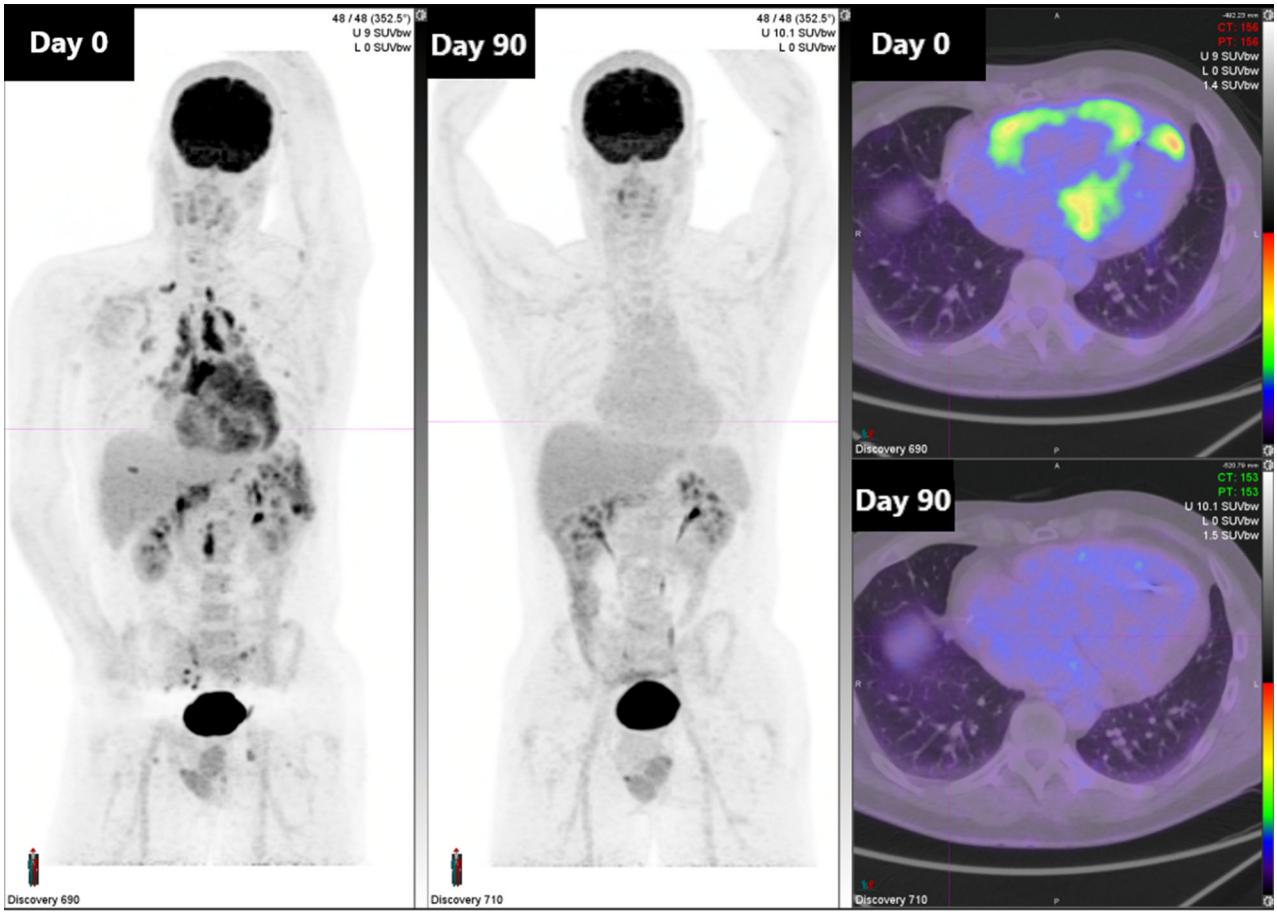

Figure 5. Comparative PET images. Comparative PET scans at 0 and 3 months. The repeat PET scan at 3 months was normal, with complete resolution of pathological FDG uptake (including cardiac).

Disclosure

C.Y. Goh, A. Gay, M.S. Hofman, C. Wong, J. Westcott and N. Better have nothing to disclose.

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