Severe ventricular arrhythmias in a patient with cardiac sarcoidosis: insights from MRI and PET imaging and importance of early corticosteroid therapy

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A 37-year-old male was referred to our institution for evaluation of exertional angina and non-sustained ventricular tachycardia. Physical examination and laboratory results were normal. The echocardiogram (Panel E) revealed left ventricular (LV) hypertrophy (interventricular septum 16 mm, posterior wall 13.8 mm) with normal ejection fraction. Coronary angiography was normal.

Further exploration including cardiac magnetic resonance imaging showed perfusion defects in the basal septal and apical segments (Panel A) as well as a delayed enhancement in the same segments (Panel B).

99mTc-tetrofosmin scintigraphy demonstrated basal septal and apical LV hypoperfusion (Panel C) and cardiac 18F-Fluorodeoxyglucose Positron Emission Tomography (18F-FDG-PET) scanning revealed intense glucose metabolism in the same myocardial segments (Panel D). Whole body 18F-FDG scanning showed pathological uptake in the heart, thoracic, and abdominal lymph nodes, as well as in the lungs and the spleen.

Because of recurrent episodes of ventricular tachycardia with haemodynamic instability, an implantable cardioverter defibrillator was placed and the patient was started on oral amiodarone. Boluses of steroids were administered resulting in a prompt control of ventricular arrhythmia and a rapid regression (6 days) of left ventricular hypertrophy (interventricular septum 10.7 mm, posterior wall 11.1 mm) (Panel F) and of whole-body 18FDG uptake (2 months) (Panel H).

Cardiac sarcoidosis might present in young patients as left ventricular hypertrophy associated with severe ventricular arrhythmias. Magnetic resonance imaging and 18F-FDG-PET scan are extremely helpful in the diagnostic workup, showing the granulomatous lesions as contrast enhancement after gadolinium administration and a perfusion-metabolic mismatch, respectively. Early initiation of high-dose steroid treatment might result in a rapid clinical stabilization and regression of left ventricular hypertrophy.