A case of cardiac sarcoidosis diagnosed after heart transplantation

Authors:
M Stratinaki, I Armenis, N Kogerakis, E Leontiadis, MJ Bonios, A Gkouziouta, A Platania, L Kaklamanis, S Adamopoulos, Onassis Cardiac Surgery Center - Athens - Greece,

Topic(s):
Acute Heart Failure: Pathophysiology, Other

Citation:
A 53-year old female Caucasian was admitted to our hospital due to presyncope and shocks delivered from her defibrillator. She had been diagnosed with non-ischemic cardiomyopathy two years ago in a tertiary hospital, where she was admitted because of worsening exertional dyspnea. In that hospitalization, reduced left ventricular ejection fraction (LVEF) and new onset conduction abnormalities [1st degree atrioventricular block (AVB) -left bundle branch block with intermittent complete AVB] were documented. A Magnetic Resonance Imaging (MRI)-scan had revealed LVEF 26%, akinetic interventricular septum (IVS) and hypokinetic anterior (AW) and inferior (IW) walls and diffuse late gadolinium enhancement mainly located at the IVS and AW and to a lesser extent to IW and anterolateral wall (with transmural/midwall distribution), a pattern indicative of old myocarditis or atypical non-ischemic cardiomyopathy. The patient had been implanted CRT-D 18 months before, but this was the first time it delivered shocks. In our hospital, she received intravenously amiodarone and esmolol and ventricular tachycardia was successfully terminated. However, the patient rapidly deteriorated. Dobutamine was initiated and the patient was transferred to the Cardiac Care Unit where an intra-aortic balloon pump (IABP) was placed. Under IABP and intravenous inotropes the patient’s haemodynamic status improved and no further arrhythmias occurred. However, the patient was unable to wean off IABP and intravenous inotropes. As a result, the patient became candidate for heart transplantation and an extensive pre-transplant evaluation, including total body Computerized Tomography (CT) was performed. Chest CT revealed few subpleural nodules mainly in upper lobes compatible with small granulomas. Successful heart transplantation was performed after three months. Post-transplantation histology of the patient's native heart revealed diffuse inflammatory reaction with noncaseating granulomas and few giant cells, findings consistent with cardiac sarcoidosis. This case highlights the need for high clinical suspicion of rare, potentially reversible causes of heart failure, especially in patients with clinical/paraclinical findings suggestive of specific causes (conduction abnormalities and MRI findings in our patient). Diagnosis of cardiac sarcoidosis in the absence of previously documented systemic sarcoidosis remains intriguing and requires high clinical suspicion. Endomyocardial biopsy, a procedure that might be diagnostic, especially if guided, was not performed in our patient, due to the relatively insidious onset of heart failure symptoms and her end-stage heart failure status. Conclusively, endomyocardial biopsy, although invasive, potentially dangerous and not always conclusive, may be considered in selected, relatively young patients with unexplained dilated cardiomyopathy and clinical features suggestive of infiltrative disease.