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Heart transplantation in female recipient with cardiac sarcoidosis

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There is a challenge to diagnose cardiac sarcoidosis (CS) which is extremely rare multisystem disease. Often it is diagnosed only in patients with end-stage chronic heart failure (CHF). A 30-yr-old female patient from her early childhood coped with arrhythmias, such as paroxysmal frequent supraventricular tachycardia. At the age of 9 WPW was found, and she underwent epicardial catheter ablation (CA) of accessory atrioventricular (AV) pathways. Since 13-yr-old frequent syncope-associated ventricular extrasystoles were diagnosed. When she was 17-yr-old, CA of left posterior accessory connections of AV pathway was performed. Then cardiac pacemaker was implanted due to complete AV block. Moreover, LVEF dropped to 30%. To prevent sudden cardiovascular death we implanted CRT+D. Less in 1 yr patient was admitted to our hospital with CHF NYHA class III. According to TTE, LVEF was 20%, mitral regurgitation grade 4. Due to she was hemodynamically unstable we could not perform endomyocardial biopsy (EMB) prior heart transplantation

(HTx). Patient's examination did not show any mediastinal lymphadenopathy or lung lesions. Despite the treatment, patient's condition deteriorated. She was heart transplanted less than in 6 months. Time in ICU was complicated by severe right heart failure. Two weeks after HTx we performed Batista procedure with mitral valve repair. Explanted heart biopsy revealed typical sarcoidosis signs: specific myocarditis, non-necrotic granulomas, fibrosis fields. Patient was treated with triple-drug therapy (steroids, tacrolimus, mycophenolic acid) plus the induction (thymoglobulin). After HTx EMB did not reveal any signs of myocardial cellular rejection or specific granulomatosis. In long-term follow-up there was no signs of CHF. In fact, 5 yrs after HTx according to EMB results there was no signs of CS. In conclusion, HTx can be considered as an effective treatment for patients with CS complicated by end-stage CHF. Post-HTx immunosuppressive therapy may prevent sarcoidosis relapse.

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