Abstract: P114

Cardiac transplantation in systemic sclerosis and life threatening primary cardiac involvement

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Introduction: We present the case of two patients undergoing heart transplantation (HT) for systemic sclerosis (SS). SS is rare and characterized by vasculopathy and fibrosis. The pathogenesis of cardiac damage is controversial and understood. Heart failure is uncommon, carries a poor prognosis and evidence on HT is scarce.

Objective: Analyze features of SS patients undergoing a successful HT.
Case 1: A 57-year-old female affected from cutaneous ANA-positive SS was admitted to hospital on January 2014 for predominant heart failure. An ETT showed a dilated myocardiopathy with severe biventricular systolic and diastolic dysfunction, severe functional MR and no pericardial effusion. Cardiac catheterization excluded pulmonary hypertension (PAH) and coronary atherosclerosis. CRM showed patchy edema and subendocardial delayed enhancement suggestive of a subacute myocarditis. During 4 months she was treated with Cyclophosphamide, high dose methylprednisolone and Hydroxychloroquine. Later on worsening came up with NYHA III-IV/IV, NTproBNP levels >30.000 pg/ml. A CRT-CDI was implanted with no response due to severe hemodynamic deterioration. Failing inotropic and mechanical heart assistance, she underwent orthotopic HT 11 months after heart failure diagnosis was made. During follow up time a paroxysmal AF episode was observed in the early post-transplant period.

Case 2: A 58-year-old male admitted to hospital on February 2017 with Scl 70-positive SS diagnosis: active myositis, mild interstitial lung disease without PAH and progressive cardiac biventricular severe dilatation and dysfunction (confirmed by ETT and CRM). Patchy subendocardial late enhancement was also seen. 2 months later, non-cardiac SS-related organ involvement was controlled with intensive immunosuppressive treatment (steroids, cyclophosphamide, mycofenolato mofetil and rituximab) but predominant symptoms of RV dysfunction and low cardiac output appeared. Severe recurrent ventricular arrhythmic episodes justified HT 5 months after cardiac affection was diagnosed. The post-operative period was complicated by a urinary infection by E.Coli BLEE.

Discussion: There is no specific treatment for primary cardiac involvement in SS. Even if HT is a life-saving procedure option, few cases have been reported. None of our patients had major severe and uncontrolled SS-related organ involvement at the time of HT. Extensive evaluation confirmed the absence of PAH, gastrointestinal involvement and scleroderma renal crisis. In both cases severe myocardial biventricular affection was seen and HT occurred in less than a year since the diagnosis of heart failure was made. No SS-related complications, neither ischemic or graft rejection occurred during follow up period (46 and 15 months respectively).

Conclusions: HT for SS-related end-stage cardiac involvement (heart failure and/or arrhythmic complications) is feasible and can be considered in selected patients, especially if non cardiac involvement is discarded/controlled.