Scleroderma Cardiac Disease

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A 42 years Hindu female had progressive thickening and tightening of skin of both upper and lower extremities, face and trunk for eight years. She had flexion contracture of fingers for same duration. She also had arthralgia and Raynaud’s phenomenon on exposure to cold. She had heartburn and dysphagia to liquid. She developed shortness of breath and palpitation on exertion for last two years. She had mild pedal oedema. Pulse was 104/min, irregularly irregular. BP was 112/80 mm Hg. S1 and S2 were audible with varying intensity. Her blood picture revealed Hb 13.1 gm%, TLC 4700 /cumm with neutrophil 50%, lymphocyte 44%, monocyte 3%, eosinophil 3% and adequate platelet count. C-reactive protein was 3.6 mg/L. Her blood sugar, urea, creatinine and liver function tests revealed no abnormality. HRCT scan of thorax did not reveal any lung parenchymal abnormality but showed mild dilatation of lower third of esophagus. ECG revealed atrial ectopics and anterolateral ischemia. Echocardiography revealed normal sized left atrium and enlarged right atrium (Fig. 1). Tricuspid valve was displaced towards right ventricle. Grade 2+ tricuspid regurgitation (TR) was present (Figs. 2 and 3). Mitral valve morphology was normal. Right ventricular walls were thickened, more so at the apex with reduced cavity size. Restrictive pattern of right ventricular diastolic filling was present. Normal sized left ventricle with good systolic function was evident. Final diagnosis of the case was restrictive cardiomyopathy with myocardial and endomyocardial fibrosis predominantly involving the right heart in a patient of progressive systemic sclerosis (PSS).

Myocardial ischemia, fibrosis and contraction band necrosis have been reported in literature in association with PSS. Underlying mechanism appears to be microcirculatory impairment with abnormal vasoreactivity, with or without structural vascular abnormalities. Micro vascular coronary vasospasm is known as “myocardial Raynaud’s phenomenon”. Repeated focal ischemic injury causes irreversible myocardial fibrosis. Fibrosis tends to be patchy, involving all levels of the myocardium unpredictably. Right ventricle is involved as often as the left. Pericardial involvement (effusion) is frequent but usually asymptomatic. Conduction system abnormalities are common. Arrhythmias may be life-threatening. No significant valvular involvement appears to be associated with PSS (but the presently reported case had TR). Treatment for myocardial involvement includes long-term use of vasodilators; e.g. calcium channel blockers and ACE inhibitors, for improvement of myocardial perfusion and function abnormalities.

References


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