Abstract: P1688

Rare cause of acute heart failure in patient with scleroderma

Authors:
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Topic(s):
Echocardiography: Systolic and Diastolic Function

Citation:
A 32-year old female patient, known to have scleroderma, presented with symptoms of recurrent attacks of palpitations, dyspnea (NYHA class III) & dizziness for 3 days before presentation. No chest pain or syncope. Physical examination at presentation revealed hemodynamic instability (BP: 80/50 mmHg, HR: 200 bpm/ regular thready pulse with fine bilateral basal chest crackles. ECG revealed ventricular tachycardia (figure 1), so electrical cardioversion was immediately done upon which the reverted to multifocal atrial tachycardia with RBBB and restoration of hemodynamics and disappearance of chest crackles.

She also had thickening of the fingertips’ skin, amputated tip of the right middle finger with severe associated Raynaud’s phenomenon, skin tightness of both upper and lower limbs, Buccal buckling with limited mouth opening (figure 2 and 3). Renal function and serum electrolytes were normal with elevated troponin I, creatine kinase and CK-MB serum levels.

Echocardiogram showed mild concentric LV hypertrophy with normal internal dimensions and severely impaired systolic function (EF~ 20%) with severe global hypokinesia. The RV was also dilated and impaired (TAPSE~8 mm). Mild pericardial effusion with no chamber collapse (Figure 4, 5 and 6). Coronary angiography revealed normal epicardial coronaries.

Echocardiography one month prior to presentation revealed normal LV dimensions, preserved systolic function and mild diastolic dysfunction with no resting wall motion abnormalities, upper normal RV size with preserved RV systolic function and mild elevation of pulmonary artery systolic pressure.

The presence of new acute biventricular failure in the absence of CAD and elevated cardiac biomarkers are highly suggestive of acute myocarditis.

Medical therapy included loop diuretics, angiotensin-converting-enzyme inhibitor, beta-blocker and aldosterone receptor antagonist. She was discharged in NYHA I-II, with normal cardiac enzymes and left ventricle ejection fraction of 30% and resolution of the pericardial effusion.

Discussion: Scleroderma is rarely present with impaired LV contractility but more commonly presented with diastolic dysfunction (in up to 40% of cases) (1,2). This case was presented with acute myopericarditis with severe biventricular systolic dysfunction which is very rare with scleroderma and only mentioned in few case studies, so no consensus on specific treatment. Few patients with myocarditis and scleroderma were treated with immunosuppressants along with diuretics, beta-blockers, and ACE inhibitors with most of them showed improvement clinically with recovery of systolic function after 12 months of follow up (3). We noted improvement of patient symptoms within few days of diuretics, beta-blockers, Aldosterone antagonists and ACE inhibitors. This case had baseline conduction abnormalities (RBBB with multifocal atrial tachycardia) which could be present in up to 20% of cases (4).
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