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CASE REPORT

Coronary disease in scleroderma; report of two cases

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ABSTRACT

Scleroderma (SSC) is a systemic inflammatory disease characterized by vascular dysfunction and excessive fibrosis which may involve many organs particularly the heart, lungs and kidney. In scleroderma cardiac disease and pulmonary arterial hypertension (PAH) are common findings where as coronary diseases have been reported with a similar frequency or as rare in several studies. Atypical presentation, wide range of differential diagnosis and co morbidities can result in certain difficulties in diagnosing coronary disease in these patients. Hereby, we report two cases with scleroderma and PAH and a mild increased filling pressure in which significant coronary disease was discovered incidentally. Therefore, it seems crucial to perform further studies on the preferred time for evaluating CAD in such patients.

Key words: pulmonary arterial hypertension (PAP), scleroderma; coronary artery disease (CAD), pulmonary capillary wedge pressure (PCWP)

INTRODUCTION

Scleroderma (SSC) is a systemic inflammatory disease characterized by vascular dysfunction and excessive fibrosis which may involve many organs particularly the heart, lungs and kidney.[1] In such cases interstitial lung fibrosis and pulmonary vascular disease can lead to pulmonary arterial hypertension. Myocardial fibrosis, valvular disease, microcirculation and conduction abnormalities have also been reported. Coronary involvement is not so common, reported as 22% in one study.[2] Herein, we present two cases of scleroderma with coronary involvement in which the presentation is unusual and coronary disease in diagnosed incidentally.

CASE REPORT

Case 1

A 65-year-old woman presented to our Heart Failure clinic with dyspnea and leg edema. She was a known case of scleroderma since 3 years before and was on prednisolone medication. She had an unremarkable medical history. She complained of dyspnea NYHA class III which had exacerbated in the previous 2 months. On physical examination fine basilar rales and pitting leg edema were diagnosed. Cardiac exam revealed loud P2 and systolic murmur II/VI in left sternal border with inverted T waves in V1-V4. Serum ProBNP level was 1784 pg/ml (normal less than 125 pg/ml). Echocardiography demonstrated preserved left ventricular ejection fraction (EF: 55%) and mild LV diastolic dysfunction with enlargement of the right atrium and ventricle and severe pulmonary arterial hypertension (sysPAP: 65 mmHg) (nl less than 30 mmHg).[2]

Pulmonary spirometry showed a mild restrictive pattern (FEV 1/FVC: 83%) while lung HRCT (high resolution chest scan) revealed mild lung fibrosis. Lung perfusion scan was not in favor of pulmonary emboli. Therefore, she was scheduled for right heart catheterization (RHC) by which the following data were obtained: mean PAP: 45 mmHg (nl less than 25), RA pressure: 15 mmHg (nl less than 5), RVP: 70/0-10 mmHg (nl less than 25), mean pulmonary capillary wedge pressure (PCWP): 15 mmHg (nl less than 8) (Trans pulmonary gradient (TPG): 30 mmHg(nl less than 12) and her Cardiac index (CI): 2.7ml/kg/m2 (nl

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more than 4).
According to cath data the patient had post capillary pulmonary hypertension with arterial obstructive disease. In order to precisely evaluate the left ventricle we decided to measure the left ventricular end diastolic pressure (LVEDP) and coronary artery disease which showed significant stenosis (75-90%) in both the left anterior descending and left circumflex arteries (LAD and LCX). Percutaneous coronary intervention (PCI) was unsuccessful and surgery was ignored due to severe PAH and a high mortality risk.

Case 2
A 55-year-old lady, known case of scleroderma since 2 years before, was referred to our Heart Failure clinic with exertional dyspnea and chest pain which had exacerbated since 4 months ago. She had an elevated jugular vein (JVP), rales and pitting edema but no clear finding in heart exam. Electrocardiogram, revealed a normal sinus rhythm with incomplete right bundle branch block (RBBB) and no significant ST changes. TnI level was 0.01 ng/ml (nl:negative) while her serum PRoBNP level was measured as 793 pg/ml .Echocardiography demonstrated mild LV systolic and diastolic dysfunction (LVEF: 45%), RA and RV enlargement and sysPAP: 65-70 mmHg. PFT, Lung HRCT and V/Q scan had no significant findings. Her RHC data were as follows: RAP: 18mmHg, RVP: 90/0^{-15} mmHg, mean PAP: 58 mmHg, PCWP: 18 mmHg and CI: 2.5 ml/kg/m2 and TPG: 40 mmHg. Due to chest pain, mild LV systolic dysfunction and elevated PCWP, we decided to do coronary angiography. It revealed significant (90%) stenosis in the LAD and right coronary arteries (RCA) for which PCI was done successfully.

DISCUSSION
Coronary disease is not a common presentation in SSC but due to inflammatory process, CAD is 6 fold more frequent and SSC is associated with coronary calcification.[3] Akram et al in 2006 implied the frequency of coronary disease in scleroderma patients is fairly similar to general population matched for age and sex.[3] In general, chest pain can be due to PAH, pulmonary disease, gastro esophageal reflux, chest wall arthritis or pericardial disease.[5] Co morbidities can limit patient's ability to exercise and imaging modalities can have some false positive results due to microvascular dysfunction.[6, 7] The low prevalence of CAD reported in several studies may be due to the atypical presentation and low frequency of angiographic studies conducted in these patients.[8]
The increased left ventricular filling pressures (LVEDP) in PAH can be due to ischemia and coronary disease.[9] The incidental finding of CAD in SSC patients can raise the question of when to evaluate coronary disease in SSC patients?
Nevertheless, whether the detection of PAH or some RHC data like elevated PCWP can be a clue to consider left ventricular ischemia and CAD needs to be further clarified.
Prescription of certain drugs such as nitrates in combination with pulmonary vasodilators like sildenafil can be problematic.[2] Patient Follow up and repeated RHC can be crucial whereas prostanoids and bosentan can be harmful in post capillary PAH. However, PWCP may decrease after treating the ischemia with evascularization.

CONCLUSION
Cardiac disease is common in SSC. The incidental finding of CAD has been demonstrated in some SSC patients with PAH and elevated PCWP. Due to the atypical presentation of CAD in these patients, further studies should be performed focusing on the preferred time to evaluate coronary disease in SSC cases.

CONFLICT OF INTEREST
None.

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