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MODERATE PERICARDIAL EFFUSION WITH PULMONARY ARTERY HYPERTENSION IN A CASE OF SYSTEMIC SCLEROSIS: A CASE STUDY

General Medicine

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ABSTRACT

A case of a 40 years old woman who was a known case of systemic sclerosis, presented with cardiac menifestations of moderate pericardial effusion with pulmonary hypertension. Cardiac involvement in systemic sclerosis is a poor prognostic factor and pulmonary arterial hypertension (PAH) is an important clinical complication and a leading cause of mortality in this disease. Although, there is no specific treatment for pericardial diseases in systemic sclerosis, but pericardial effusion is associated with PAH or renal crisis, once pericardial effusion is diagnosed, echocardiography is vital to evaluate whether the effusion is progressive, to assess the hemodynamic effects, and to monitor the response to anti-inflammatory therapy in acute setting. Cautious pericardial drainage should be considered.

KEYWORDS

Pericardial effusion, PAH, Systemic sclerosis, DMARDs, etc

INTRODUCTION :

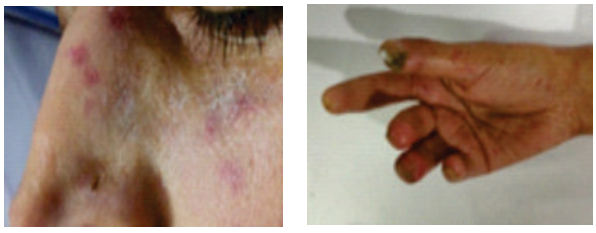
Systemic sclerosis is a multi-system connective tissue disease whose hallmarks are autoimmunity, vasculopathy and fibrosis. Cardiac involvement is a poor prognostic factor, but diagnosis may be late or missing because of the frequent discrepancy between clinical manifestation and cardiac involvement. Hence, all available diagnostic investigations should be recommended to achieve an early diagnosis. Cardiac manifestations include myocardial fibrosis, myocardial ischemia, conduction abnormalities, systolic and diastolic dysfunction, pericarditis and pericardial effusion [1], [2]. It can involve the pulmonary vasculature causing pulmonary hypertension. Pulmonary arterial hypertension (PAH) is an important clinical complication and a leading cause of mortality in this disease & the reported prevalence is ranging from 7% to 29%. [3]

Case Report :

A case of a 40 years old woman who was a known case of systemic sclerosis, presented with dyspnea and easy fatiguability. Past history of skin discolouration of her fingers and thickening of skin, dysphagia, hysterectomy in the past.

The patient's vitals were stable including temperature 37.0 °C; blood pressure 108/74 mm Hg, pulse 78 beats per minute, normal character; and respiratory rate 16 breaths per minute. The jugular veins were distended and vy descent appreciated. The carotid pulses were normal without bruits. There was no palpable lymphadenopathy. No dependent edema. The patient's chest was without abnormalities. On pulmonary examination bilateral course crepitations heard lower one third of chest and cardiac auscultation pan systolic murmur heard at tricuspid area . Abdomen was soft and nontender, no organomegaly, & bowel sounds were normal.

The objective examination revealed Raynaud's phenomenon, the thickening around the nail bed and a more extensive thicker area of the skin on the hands & forearms , sclerodactyle and talengiectasia over nose & cheeks.



A B

Image 1 : (A)Talengiectasia over nose and cheeks, (B) Sclerodactyle

Laboratory examinations showed leukocytosis, elevation of inflammatory markers and albuminuria. Chest X-ray showed bilateral reticulonodular shadows and pleural effusion, cardiothoracic index was normal and a normal ECG. Echocardiographic examination which revealed a moderate pericardial effusion without any signs of a cardiac tamponade, and signs of severe pulmonary hypertension.

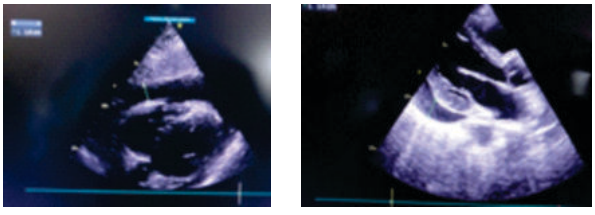


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Image 2 : 2D-Echocardiography (A) Subcoastal view, (B) Parasternal long axis view showing moderate pericardial effusion.

HRCT thorax showing fibrotic changes suggestive of scleroderma lung involvement alongwith consolidation and pleural effusion, suggestive of recent infective etiology. CT pulmonary angiography suggestive of dilated main pulmonary, right and left pulmonary arteries and features suggestive of pulmonary hypertension. Also moderate pericardial effusion was observed.

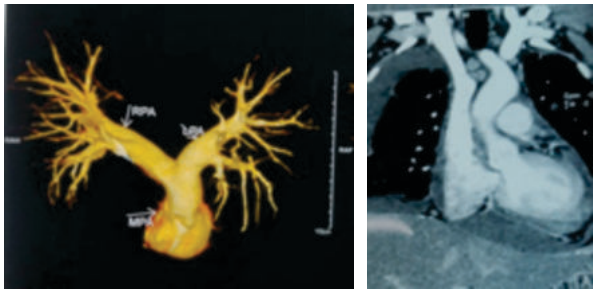

Image 3: CT pulmonary angiography suggestive of dilated main pulmonary, right and left pulmonary arteries (features suggestive of pulmonary hypertension) and moderate pericardial effusion.

Past treatment history of DMARDs (Disease-modifying anti-rheumatic drugs) including methotrexate, leflunomide, hydroxychloroquine and other symptomatic management, but patient was not compliance to medications. On present admission patient was treated with steroids, drugs for pulmonary hypertension, angiotensin converting enzyme inhibitors, antibiotics and symptomatic management. Patient reviewed during hospital stay, improved clinically and pericardial effusion also. Started on DMARDs as per rheumatologist consultation.

DISCUSSION :

The incidence of pericardial involvement in scleroderma is about 50% according to autopsy results, but symptomatic pericarditis manifests in about 16% of patients with diffuse scleroderma and in about 30% of patients with limited scleroderma. The clinically evident pericardial effusion is rare in scleroderma, although it can be detected in about 41% of patients with echocardiography [4]. However, clinically

symptomatic pericardial disease (5–16%) is much less frequent than autopsy-demonstrated pericardial involvement (33–72%) [5] There is no specific treatment for pericardial diseases in systemic sclerosis, but non-steroidal anti-inflammatory therapy with careful monitoring of renal function can be used in acute pericarditis. In cases of cardiac tamponade or pericardial constriction, pericardiocentesis and surgical intervention must be considered. Corticosteroids are useful in the setting of associated myocarditis.[6] Since studies certified that pericardial effusion is associated with PAH or renal crisis, once pericardial effusion is diagnosed, echocardiography is vital to evaluate whether the effusion is progressive, to assess the hemodynamic effects, and to monitor the response to therapy. With large significant pericardial effusions and PAH, the first step is to use vasoactive therapy to stabilize pulmonary artery pressure and right heart function. Moreover, cautious pericardial drainage should be considered. [7] It may need surgical intervention in recurrent pericardial effusions.

CONCLUSION :

Cardiac involvement in systemic sclerosis is a poor prognostic factor and pulmonary arterial hypertension (PAH) is an important clinical complication and a leading cause of mortality in this disease. Although, there is no specific treatment for pericardial diseases in systemic sclerosis, but pericardial effusion is associated with PAH or renal crisis, once pericardial effusion is diagnosed, echocardiography is vital to evaluate whether the effusion is progressive, to assess the hemodynamic effects, and to monitor the response to anti-inflammatory therapy in acute setting & DMARDs as a long term tharapy. With large significant pericardial effusions and PAH, the first step is to use vasoactive therapy to stabilize pulmonary artery pressure and right heart function. Cautious pericardial drainage should be considered. Moreover, patient education, counselling, strict use of medical & physiotherapy and regular follow should be considered as essential parts of management.

REFERENCES:

1. A. Deswal, W.P. Follansbee. Cardiac involvement in scleroderma; Rheumatic Disease Clinics of North America, 22 (1996), pp. 841-860
2. S. Generini, G. Fiori, A. Moggi Piqnone, et al. Systemic sclerosis. A clinical overview; Advances in Experimental Medicine and Biology, 455 (1999), pp. 73-83
3. R.W. Battle, M.A. Davitt, S.M. Cooper, et al. Prevalence of pulmonary hypertension in limited and diffuse scleroderma; Chest, 110 (1996), pp. 1515-1519
4. R. Gowda, I. Khan, T. Sacchi, et al. Scleroderma pericardial disease presented with a large pericardial effusion—a case report; Angiology, 52 (2001), pp. 59-62
5. Byers RJ, Marshall DA, Freemont AJ. Pericardial involvement in systemic sclerosis. Ann Rheum Dis 1997; 56:393–394. doi: 10.1136/ard.56.6.393.
6. Vacca A, Meune C, Gordon J, Chung L, Proudman S, Assassi S, et al. Cardiac arrhythmias and conduction defects in systemic sclerosis. Rheumatology (Oxford) 2014; 53:1172–1177.
7. Gowda RM, Khan IA, Sacchi TJ, Vasavada BC. Scleroderma pericardial disease presented with a large pericardial effusion--a case report. Angiology 2001; 52:59–62.