Case Report

Pericardial effusion as a first sign of systemic scleroderma

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1. Introduction

Systemic sclerosis is a connective tissue disease whose hallmarks are autoimmunity, vasculopathy and fibrosis. It involves the heart causing fibrosis and the pulmonary vasculature causing pulmonary hypertension. Pulmonary arterial hypertension (PAH) is an important clinical complication of Ssc and a leading cause of mortality in this disease, the reported prevalence ranging from 7% to 29%, depending on study methodology, populations, and definitions used [1–4]. Small pericardial effusions are not uncommon in Ssc, and pericarditis is a frequent post-mortem finding. Large hemodynamically significant pericardial effusions with cardiac tamponade are rare and have a poor outcome [5,6]. We report a case of Ssc patient who presented with moderate pericardial effusion. This case report aims to explore that pericardial effusion can be the first sign of Ssc.

2. Case report

In our study we would like to refer a case of a 49 years old Caucasian woman who was seeking medical advice for the first time due to dyspnea and intolerance of horizontal position. At the time of admission the patient reported some complaint about skin discolouration of her fingers by thickening and pale. She had also some problems with swallowing like dysphagia. She had a personal history of arterial hypertension on medication with inhibitor of angiotensin converting enzyme beta-blockator, primary hypothyroidism treated with L-thyroxin, and a state after hysterectomy in the past.

The patient's temperature was 36.8 °C; blood pressure, 100/70 mm Hg, pulse normal, heart rate, 85 beats per minute; and respirations, 16. The patient was a well-developed, well-nourished woman in no acute distress who was alert and oriented. Examination of the head, eyes, ears, nose, and throat were normocephalic, atraumatic. The nose and throat were clear, and the tongue was normal in size. Scratch purpura was absent. The jugular veins were without distention. No Kussmaul sign or x and y descent were appreciated. The carotid pulses were full without bruits. There was no palpable lymphadenopathy. The patient's chest was without abnormalities. Pulmonary and cardiac auscultation was without pathology. Her abdomen was soft and nontender. There was no splenomegaly. The bowel sounds were normal. Limbs were without edema.
The objective examination revealed Raynaud's phenomenon, the mentioned thickening around the nail bed and a more extensive thicker area of the skin on the left forearm. Laboratory examinations showed a slight leucocytosis, a mild elevation of inflammatory parameters (FW, CRP).

Chest X-ray was without pathology and she showed a normal ECG, the cardiothoracic index was also normal. According to this clinical signs we suggested a form of autoimmune disease and because of progressive dyspnea made an echocardiographic examination which revealed a moderate pericardial effusion without any signs of a cardiac tamponade, and without echocardiographic signs of severe pulmonary hypertension (Fig. 1). The results of this examinations showed, that the clinical problems the patient presented, could be of autoimmune origin and suggested scleroderma.

The biochemical results such as highly positive antinuclear antibodies/ANA/and also anti-Scl 70/antitDNA topoisomerase I antibodies/completed the definitive diagnosis of systemic scleroderma.

After making the diagnosis we administered corticosteroids and calcium channel blockers to improve the Raynauds phenomenon and took away the betablocker from the medication. The clinical status of the patient improved very quickly, now she is without any serious symptoms and the second ECHO after administration of steroids showed a significant regression of the mentioned pericardial effusion.

3. Discussion

Cardiovascular disease in patients with SS may be due to either primary involvement of the heart by sclerosing disease or a secondary involvement from disease of the kidney or lungs. 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, for this reason, resort to all available diagnostic procedures is recommended to achieve an early diagnosis. Cardiac signs include myocardial fibrosis, myocardial ischemia, conduction abnormalities such as left anterior hemiblock, systolic and diastolic dysfunction, pericarditis and pericardial effusion [5,7].

Pericardial abnormalities in scleroderma have noted fibrous pericarditis, pericardial adhesions, and pericardial effusions at the time of autopsy. 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 [8]. However, clinically symptomatic pericardial disease (5–16%) is much less frequent than autopsy-demonstrated pericardial involvement (33–72%) [9–13]. Most cases are asymptomatic, with premorbid symptoms being reported in only 7–20% of patients [14]. Moreover, there also have been large effusions causing tamponade and can even occur prior to skin thickening and the diagnosis of scleroderma [8,15].

In our case pericardial effusion was the first sign of scleroderma, without tamponade. Pericardial effusions are also frequently associated with pulmonary hypertension and may be the presenting feature of pulmonary hypertension in scleroderma. Large pericardial effusions can lead to pericardial tamponade and are a marker for poor outcome. If an inflammatory component is thought to be the cause of the effusion, immunosuppression therapy can markedly reduce the volume of the effusion. Moreover, if clinical heart failure is present, the effusion can be reduced with diuretics. Small, asymptomatic pericardial effusions may occur if there is cardiac involvement and should be observed. Oral prednisolone should be given for patients with pericardial effusions causing marked symptoms. These patients are at risk of developing cardiac tamponade. Patients with tamponade should receive a pericardial window and systemic corticosteroids [12,13].

4. Conclusion

Our case report showed that systemic scleroderma can be presented with a moderate to severe pericardial effusion with or without any other clinical symptoms or systemic scleroderma. Therefore it is necessary to think about this disease mainly in young women with progressive dyspnea without any other signs of congestive heart failure and with an echocardiographic picture that shows a mild to severe pericardial effusion.

References


