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Case Report

Systemic Scleroderma Revealed by Cardiac Tamponade: A Case Report

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Abstract

In systemic scleroderma, pericardial involvement is usually silent and benign. Symptomatic pericarditis is rare and cardiac tamponade even more so. We report the case of a 56-year-old woman presenting with cardiac tamponade and whose etiologic investigation concluded to a final diagnosis of limited cutaneous systemic scleroderma. Clinicians should be wary of this diagnosis when dealing with acute-onset pericarditis.

Keywords: Pericardial effusion; Tamponade; Systemic scleroderma; Case report.

INTRODUCTION

Systemic scleroderma (SSc) is a connective tissue disease characterized by autoimmunity, fibrosis, and vasculitis. Pericardial involvement is common in these patients and can take many forms, both acute and chronic. They are often (but not always) linked to some degree of pulmonary hypertension [1]. Scleroderma pericardial disease is usually asymptomatic and benign. Mild to moderate pericardial effusion has been found in 15 to 43% of SSc patients in echocardiographic studies [2, 3] and 78% based on autoptic studies [4]. Clinically significant effusion is rarer; its prevalence ranges from 5 to 16% [5].

Large effusions with significant hemodynamical consequences are exceptional and have been associated with a poorer overall prognosis. They can represent the first manifestation of the disease, before noticeable skin involvement [6-9]. In this paper, we provide a case report of a woman whose SSc diagnosis was revealed by severe pericardial involvement.

PATIENT AND OBSERVATION

A 56-year-old Moroccan woman was admitted in the Emergency Department for sudden onset dyspnea and atypical chest pain. Her medical history included recently found dyslipidemia. Her two sons had been treated for pulmonary tuberculosis twelve years ago. She also complained of polyarthralgia (mostly in the hands) starting four months ago, as well as esophageal dysphagia starting the year before. The patient also described symptoms similar to Raynaud's disease affecting her fingers, occurring intermittently for fifteen years. Asthenia and anorexia had plagued her life for the better part of two years, and she lost about 20 kilograms of weight in the last three months. She was not on any chronic medication and had never undergone surgery.

Initial physical examination showed a fatigued woman. Body temperature was normal. Her heart rate was 120 beats per minute and blood pressure 111/75 mmHg. She had a labored respiration, with a rate of 26 breaths per minute at rest, but normal oxygen saturation at 96%. Auscultation found muffled heart sounds but no rales or murmur. There was mild jugular vein distension, as well as pedal edema reaching the ankles. Pulsus paradoxus and Kussmaul's sign were absent. Abdominal examination revealed no tenderness, hepatomegaly, or ascites. Examination of the skin found sclerodactyly involving the interphalangeal and metacarpophalangeal joints. There was local thickening over the face, forehead, and hands with loss of normal skin folds, with a Rodnan skin score of 20. No digital ulcers or pitting scars were found. Motion of fingers, wrists and ankles was limited but other joints were pain-free. Palpation showed the presence of axillary lymphadenopathy. Electrocardiogram at admission showed sinus tachycardia with low QRS voltage. Chest radiograph found an enlargement of the cardiac silhouette.

Biological findings included normochromic anemia (hemoglobin level of 10.9 g/dL) as well as

inflammatory syndrome, with white blood cell count reaching 13.6×103/µL and high levels of C-reactive protein (initially 16.1 mg/L but quickly rising to 163 mg/L). High-sensitive troponin was slightly elevated (4 times the upper limit of normal). Renal and hepatic functions were normal. Standard tumor markers levels (CEA, CA 125 and CA 19-9) were normal. Sputum culture was negative and Interferon gamma test for tuberculosis were negative, but urine culture was positive to amoxicillin-sensitive Escherichia coli. Immunological tests showed the presence antinuclear antibodies.

Echocardiographic evaluation demonstrated a large pericardial effusion with collapse of the right atrium as well as marked respiratory variation in transmitral Doppler flow suggesting cardiac tamponade. Inferior vena cava was dilated and did not collapse with inspiration. The right ventricular systolic function was depressed, with an estimated systolic pressure of 65 mmHg, with mild tricuspid regurgitation. Left ventricular function was normal (Figure 1).

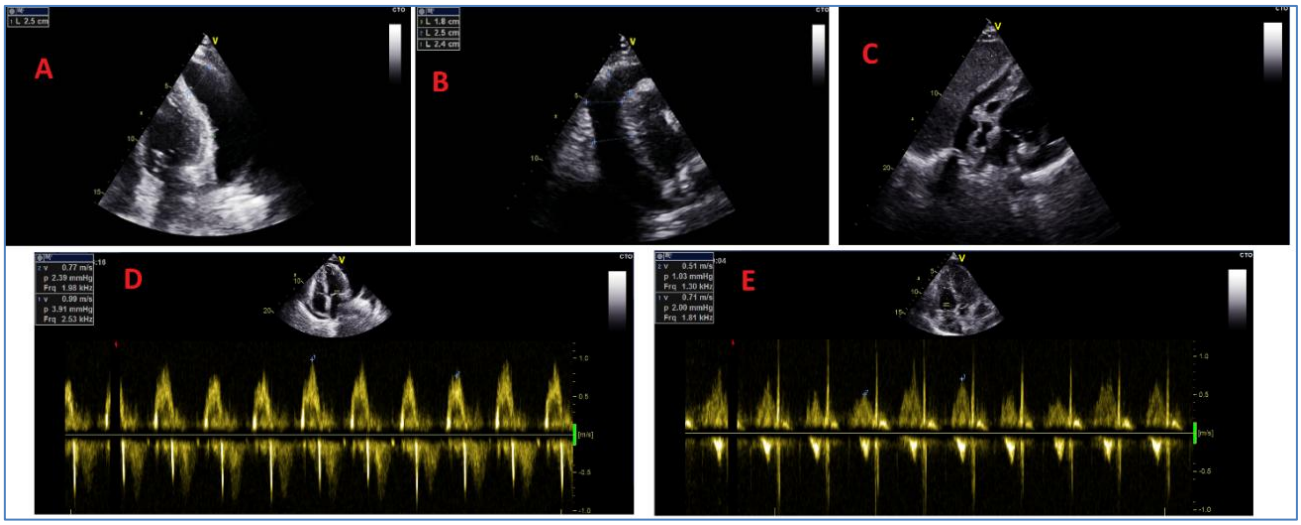

Fig-1: Transthoracic echocardiogram done at admission. Apical (A) and subcostal (B) views showing a large circular pericardial effusion with end-diastolic compression of right cavities (C) and respiratory variation of transmitral (D) and transtricuspid (E) flows

This patient urgently underwent an ultrasound-guided pericardiocentesis removing about 800 cc of clear pericardial fluid. Analysis showed an exudative pattern based on Light's criteria and the presence of reactive mesothelial cells, but no malignant cells were found. Cultures isolated a probable contamination of Aerococcus viridans. Mycobacterium tuberculosis culture and PCR were negative.

The patient felt better and was admitted three days in the Cardiology Department where an etiologic investigation was conducted. Internal Medicine and Rheumatology specialists were consulted, leading to the diagnosis of limited cutaneous SSc based on a 2013 ACR/EULAR score of 18. Treatment regimen included loop diuretics, aspirin, colchicine, and amoxicillin. A week after admission, the patient complained of escalating dyspnea, prompting another transthoracic echocardiography. It demonstrated the persistence of a large pericardial effusion responsible for the collapsus of the right atrium and part of the right ventricle and marked respiratory variation in transmitral and transtricuspid Doppler flows. The day after, the patient underwent surgical drainage using a pericardial window. Analysis of the fluid showed findings similar to the first one. The patient felt much better and had no rest dyspnea after the surgery. She was later transferred to the Internal Medicine Department for further diagnosis and treatment optimization. A transthoracic echocardiography performed a month later showed minimal pericardial effusion (Figure 2).

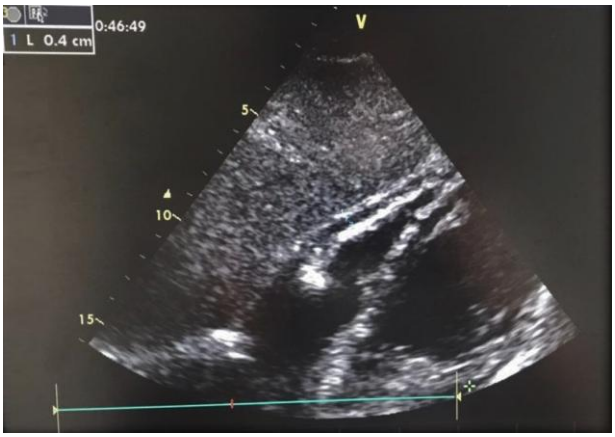

Fig-2: Transthoracic echocardiogram done at discharge, showing minimal effusion

DISCUSSION

Overall, large pericardial effusion due to scleroderma remains rare. In a prospective study performed in 322 patients with moderate-to-large pericardial effusion, acute idiopathic pericarditis was the most common diagnosis (20%), followed by iatrogenic (16%), neoplastic (13%) and chronic idiopathic effusions (9%). Connective tissue diseases were involved only 5% of patients, mostly systemic lupus erythematosus. SSc was diagnosed in only 0.6% of cases [10].

In 2016, Fernández Morales *et al*. attempted to describe the epidemiology of this rare phenomenon. Affected patients are mostly women with diffuse cutaneous SSc, and in a third of the cases, pericardial involvement preceded or happened simultaneously to SSc onset. Moreover, 10% of them presented with severe pericardial complications prior to SSc onset with a median time of 6 months; an initial screening for SSc is thus strongly advised in any patient with severe pericardial effusion. It is important to note that the majority (63%) presented with signs of tamponade [8].

From a clinical point of view, diagnosis of SSc-associated pericardial disease does not differ significantly from other causes; dyspnea and chest pain are the most common chief complaints, as in our case report. ECG and chest-X ray contribute to the diagnosis but in a non-specific manner. In Fernández Morales *et al*., pericardial fluid is mostly serohemorrhagic or straw-colored. It was an exudate in about 88% of the cases, with marked cellular reaction [8].

The particularity of SSc-associated pericardial disease is the frequent association with scleroderma renal crisis, which was absent in our case report. It was diagnosed in 12.5% of cases in Fernández Morales *et al*. and was strongly associated with mortality. One possible explanation of this association is diminished renal perfusion due to heart failure or diuretic use [7]. Some series have reported that patients diagnosed with tamponade are more likely to develop renal failure in the following 6 months [11].

There is no specific treatment for scleroderma pericardial disease; therefore medical management should align with current international guidelines [12]. Nonsteroidal anti-inflammatory therapy is indicated in the setting of acute pericarditis, with careful monitoring of renal function. Corticosteroids can be of benefit; it was the most common treatment in Fernández Morales *et al*. Pericardiocentesis or surgical intervention is considered in case of tamponade or pericardial constriction. In Fernández Morales *et al*., pericardiocentesis was performed on 20% of the patients and 35% underwent surgical treatment such as pericardial window or pericardiectomy [8]. Routine pericardiocentesis in the absence of tamponade is unadvised; it has low diagnostic yield in scleroderma patients and does not appear to affect the outcome [13].

Overall, severe pericardial disease induced by scleroderma has a poor prognosis. In Fernández Morales *et al*., 32.4% of patients died (13% in the acute phase). Due to the low number of cases, prognostic factors remain statistically unproven for the most part, but cardiac tamponade is a strong contender [8].

CONCLUSION

Cardiac tamponade or severe pericardial effusion are rare complications of both limited cutaneous and diffuse cutaneous SSc and may be the presenting manifestation of the disease. Diagnosis is similar to other causes of pericardial effusion. Prognosis remains poor and no specific treatment exists yet.

Competing interests

The authors declare no competing interest.

HC: main author and managed the patient; HO and WB: managed the patient; NL: helped drafting the work and managed the patient; ZH, NM and AB: supervised the management of the patient. All the authors have read and agreed to the final manuscript.

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Figure 1: Transthoracic echocardiogram done at admission. Apical (A) and subcostal (B) views showing a large circular pericardial effusion with end-diastolic compression of right cavities (C) and respiratory variation of transmitral (D) and transtricuspid (E) flows, Figure 2: Transthoracic echocardiogram done at discharge, showing minimal effusion.

REFERENCES

1. Deswal, A., & Follansbee, W. P. (1996). Cardiac involvement in scleroderma. Rheumatic Disease Clinics of North America, 22(4), 841–860. https://doi.org/10.1016/s0889-857x(05)70304-5
2. Meune, C., Avouac, J., Wahbi, K., Cabanes, L., Wipff, J., Mouthon, L., Guillevin, L., Kahan, A., & Allanore, Y. (2008). Cardiac involvement in systemic sclerosis assessed by tissue-doppler echocardiography during routine care: A controlled study of 100 consecutive patients. Arthritis & Rheumatism, 58(6), 1803–1809. https://doi.org/10.1016/j.semarthrit.2004.11.001
3. Maione, S., Cuomo, G., Giunta, A., De Horatio, L. T., La Montagna, G., Manguso, F., ... & Valentini, G. (2005, April). Echocardiographic alterations in systemic sclerosis: a longitudinal study. In Seminars in arthritis and rheumatism (Vol. 34, No. 5, pp. 721-727). WB Saunders.

4. Byers, R. J., Marshall, D. A. S., & Freemont, A. J. (1997). Pericardial involvement in systemic sclerosis. Annals of the Rheumatic Diseases, 56(6), 393–394. https://doi.org/10.1136/ard.56.6.393
5. Hachulla, A. L., Launay, D., Gaxotte, V., de Groote, P., Lamblin, N., Devos, P., Hatron, P. Y., Beregi, J. P., & Hachulla, E. (2008). Cardiac magnetic resonance imaging in systemic sclerosis: a cross-sectional observational study of 52 patients. Annals of the Rheumatic Diseases, 68(12), 1878–1884. https://doi.org/10.1136/ard.2008.095836
6. Gowda, R. M., Khan, I. A., Sacchi, T. J., & Vasavada, B. C. (2001). Scleroderma Pericardial Disease Presented with a Large Pericardial Effusion. Angiology, 52(1), 59–62. https://doi.org/10.1177/000331970105200108
7. Dunne, J. V., Chou, J. P., Viswanathan, M., Wilcox, P., & Huang, S. H. (2011). Cardiac tamponade and large pericardial effusions in systemic sclerosis. Clinical Rheumatology, 30(3), 433–438. https://doi.org/10.1007/s10067-010-1667-0
8. Fernández Morales, A., Iniesta, N., Fernández-Codina, A., Vaz De Cunha, J., Pérez Romero, T., Hurtado García, R., Simeón-Aznar, C. P., Fonollosa, V., Cervera, R., & Espinosa, G. (2016). Cardiac tamponade and severe pericardial effusion in systemic sclerosis: report of nine patients and review of the literature. International Journal of Rheumatic Diseases, 20(10), 1582–1592. https://doi.org/10.1111/1756-185x.12952
9. Peronato, G., de Sandre, P., Podswiadek, M., Arfiero, S., & Bozzola, L. (2002). Cardiac tamponade preceding skin involvement in systemic sclerosis. Reumatismo, 54(3). https://doi.org/10.4081/reumatismo.2002.257
10. Sagristà-Sauleda, J., Mercé, J., Permanyer-Miralda, G., & Soler-Soler, J. (2000). Clinical clues to the causes of large pericardial effusions. The American Journal of Medicine, 109(2), 95–101. https://doi.org/10.1016/s0002-9343(00)00459-9
11. Campo, A., Mathai, S. C., le Pavec, J., Zaiman, A. L., Hummers, L. K., Boyce, D., Housten, T., Champion, H. C., Lechtzin, N., Wigley, F. M., Girgis, R. E., & Hassoun, P. M. (2010). Hemodynamic Predictors of Survival in Scleroderma-related Pulmonary Arterial Hypertension. American Journal of Respiratory and Critical Care Medicine, 182(2), 252–260. https://doi.org/10.1164/rccm.200912-1820oc
12. Adler, Y., Charron, P., Imazio, M., Badano, L., Barón-Esquivias, G., Bogaert, J., Brucato, A., Gueret, P., Klingel, K., Lionis, C., Maisch, B., Mayosi, B., Pavie, A., Ristić, A. D., Tenas, M. S., Seferovic, P., Swedberg, K., & Tomkowski, W. (2015). 2015 ESC Guidelines for the diagnosis and management of pericardial diseases. Kardiologia Polska, 73(11), 1028–1091. https://doi.org/10.5603/kp.2015.0228
13. Mercé, J., Sagristà-Sauleda, J., Permanyer-Miralda, G., & Soler-Soler, J. (1998). Should pericardial drainage be performed routinely in patients who have a large pericardial effusion without tamponade? The American Journal of Medicine, 105(2), 106–109. https://doi.org/10.1016/s0002-9343(98)00192-2