KANSAS JOURNAL of MEDICINE

Scleroderma as an Uncommon Cause of Pericardial Effusion

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Received March 21, 2022; Accepted for publication May 16, 2022; Published online July 21, 2022
https://doi.org/10.17161/kjm.vol15.17026

INTRODUCTION

Autoimmune diseases are a rare but important cause of recurrent pericardial effusions, and patients with scleroderma often present with pericardial effusion as their initial symptom.[1] Many patients with a rheumatologic condition go undiagnosed for years,[2] as with the presented case. With recurrent effusion, one must investigate the cause to prevent worsening morbidity, as simple drainage will not prevent reaccumulation of the fluid. Recurrent effusion in an elderly patient regularly indicates a workup for possible malignancy, but if tissue and cytology are negative for malignant cells, then autoimmune diagnosis should be ruled out. During the patient's hospital stay, she was found to have physical and laboratory findings consistent with scleroderma. Given her decompensated state after years of undiagnosed disease, little could be offered other than symptomatic relief. We stress the importance of considering rheumatologic causes of pericardial effusions, as early detection may change the clinical course of a patient significantly.

CASE REPORT

A 70-year-old female with recurrent pericardial and pleural effusions presented to the emergency department with bilateral lower extremity swelling. She recently was admitted for a similar presentation at an outside facility, treated with diuretics, and discharged. Notably, she had regular outpatient pulmonology appointments for recurrent pleural effusions with no known cause and negative malignancy workup.

On presentation, her temperature was 98.1° F, blood pressure 61/52 mmHg, heart rate 98 bpm, and respiratory rate 23 bpm saturating 95% on room air. Her physical exam was significant for cachexia with bitemporal wasting and bilateral elbow and distal interphalangeal joint edema without erythema. She exhibited jugular venous distension and distant heart sounds.

Initial lab findings were significant for mildly elevated B-type natriuretic peptide (450 pg/mL); troponins, creatinine, and lactic acid were within normal limits. An echocardiogram showed an ejection fraction > 55%, right ventricular systolic pressure > 60 mmHg, a moderate-sized pericardial effusion with right ventricular collapse during diastole, and mildly dilated right ventricle and bilateral atria (Figure 1).

Given her clinical deterioration with tamponade physiology, a decision was made to perform a fluoroscopy-guided pericardiocentesis. Fluid cytology and culture were negative, and cell count was significant for white blood cells with polymorphonuclear predominance (Table 1). Given her negative cytology and recurrent effusions with joint edema, further workup resulted in a normal ESR, elevated CRP (36), positive ANA antibody with a homogenous staining pattern (1:80), and positive scleroderma antibody (Table 2).

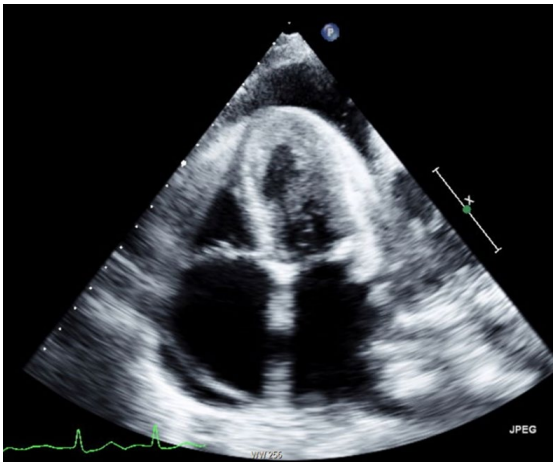

Figure 1. Transthoracic echocardiography showed fluid in the pericardium at the apex of the heart and right ventricular collapse.

Table 1. Laboratory analysis breakdown of contents in pericardial fluid.

White Blood Cell count	160 mm^3
Red Blood Cell count	< 2,000 mm^3
Polymononuclear Neutrophils	58%*
Mononuclear cells	42%*

*Values provided for informational purposes only, as there is no generally accepted reference interval.

Table 2. Laboratory result breakdown of autoantibodies present in patient's blood.

Antibody	Reference Range & Units	Patient Results
Antinuclear Antibody Screen	Negative < 1:80 Borderline 1:80 Positive > 1:80	Positive
Anti-DNA Antibody, double strand	0-9 IU/ml Negative < 5 Equivocal 5-9 Positive > 9	< 1
Jo-1 Antibody	0.0-0.9 AI	< 0.2
Anti-RNP	0.0-0.9 AI	< 0.2
Scleroderma Antibody	0.0-0.9 AI	2.8
SSA Antibody	0.0-0.9 AI	< 0.2
SSB Antibody	0.0-0.9 AI	< 0.2
Anti-Smith Antibody	0.0-0.9 AI	< 0.2

DISCUSSION

Autoimmune diseases are rare, and it is common for rheumatologic conditions to be either misdiagnosed or undiagnosed. Up to 25% of patients with rheumatologic diseases were unable to receive a definitive diagnosis, while others were undiagnosed for an average of 5 to 10 years.[2] The rheumatologic disorder diagnosed in this case study, scleroderma, has a prevalence of 135 million to 184 million cases in the U.S.[3] The majority of patients are female,[4,5] and those particularly with scleroderma present with a complication of pericardial effusion at an average age 52.2 ± 10.8 years.[1]

In scleroderma patients, pericardial effusions occur at a frequency of 17%, and pleural effusions occur less frequently at a rate of 7%.[6] Cardiac involvement at presentation (pericardial effusion or cardiac tamponade) was shown to be either prior to or simultaneous with a diagnosis of scleroderma in 32.5% of cases.[5] Cardiac symptoms of scleroderma were associated with an increase in mortality by 2.8 fold,[7] with a large portion due to arrhythmias or severe heart failure.[3]

Treatment options for pericardial effusion in scleroderma include medical management with steroids, non-steroidal anti-inflammatory drugs, colchicine, or surgical intervention through pericardiocentesis or pericardial window.[4] The majority of cases may be treated with medical therapy, reserving surgical intervention for more severe cases. Patients with pulmonary hypertension or an effusion causing hemodynamic compromise are advised to be optimized medically and managed cautiously to prevent cardiovascular collapse, reserving surgical intervention in cases that are absolutely necessary.

With our patient's advanced age, significant cachexia, and recurrent pleural effusions, clinical signs suggested the more common explanation of malignancy, and past encounters treated her accordingly. Her deviation from the typical age range of scleroderma and lack of other cardinal findings aside from her effusions did not fit the typical sclerodermal picture, possibly resulting in misdiagnosis and a likely more severe presentation during our encounter.

Because autoimmune diseases are rare, it is often the last etiology pursued, if at all. Yet, knowing that a significant percentage of scleroderma patients initially present with pericardial and/or pleural effusions was essential, and our decision to seek less common explanations led to her definitive diagnosis. Although our patient and family elected hospice, if her etiology was found earlier, it may have reduced her long-term morbidity and a more promising outcome may have been reached. We stress the importance of considering rheumatologic causes of pericardial effusions, as early detection can change the clinical course of a patient significantly.

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Keywords: systemic scleroderma, pleural effusion, autoimmune diseases, case report