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CASE REPORT OPEN ACCESS

The Cardiac Conundrum Recurrent Pericardial Tamponade in Limited Systemic Sclerosis a Case Report

Priya Sunkara1 | Agha Amad Nabi2 | Maruf Sarwar[3*]

[3]Section of Internal Medicine, White River Health, Batesville, AR, USA

***Correspondence:** Maruf Sarwar, Section of Internal Medicine, White River Health, Batesville, AR, USA, E-mail: marufsarwar2011@gmail.com

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ABSTRACT

Recurrent and symptomatic pericardial tamponade secondary to limited cutaneous systemic sclerosis (lcSSc) is an atypical presentation. This is a case of a 67-year-old female with a history of tobacco use, chronic GERD, and two decades of Raynaud's phenomenon, who presented with increased dyspnea, fatigue, and pre-syncopal episodes. Clinical examination revealed low blood pressure, sclerodactyly, digital ulcers, calcinosis cutis, and telangiectasias. Diagnostic tests showed positive ANA at 1:2560 dilution, anti-centromere antibodies (>8), and normal anti-Scl-70, anti-dsDNA, anti-Smith, and anti-RNP antibodies, with normal complement levels and a negative antiphospholipid panel. A transthoracic echocardiogram (TTE) revealed a large pericardial effusion with tamponade physiology. Right heart catheterization (RHC) indicated normal filling pressures and pulmonary artery pressure. The patient underwent emergency pericardiocentesis, however 16 days later presented with recurrent tamponade, requiring balloon pericardiotomy, followed by a pericardial window. This case underscores the rarity and complexity of pericardial tamponade in lcSSc. Comprehensive evaluation ruled out common causes such as overlap syndrome, paraneoplastic syndrome processes, viral infections, hypothyroidism, and pulmonary hypertension. Pericardial tamponade in lcSSc is infrequent and suggests a poor prognosis with limited specific treatment options. Although lcSSc typically does not present manifest significant pericardial disease symptoms, this case highlights the need for prompt recognition and investigation of internal organ involvement.

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Introduction

Limited cutaneous systemic sclerosis (lcSSc) can occasionally lead to recurrent cardiac tamponade, a rare and severe complication. While lcSSc primarily affects the skin and can involve internal organs, significant pericardial involvement is uncommon. This case report highlights a rare instance of recurrent pericardial tamponade in lcSSc, an unusual complication that deviates from the typical clinical course of the disease. Pericardial effusions with tamponade physiology in systemic sclerosis patients are frequently linked to a poorer prognosis and increased mortality rates [1]. We detail an intriguing case of recurrent early pericardial tamponade in a patient with LcSSc. Case Presentation A 67-year-old Caucasian woman with a past medical history of current tobacco use since the age of 16, lower esophageal dysphagia and dysmotility (for which she had balloon dilation), 2 decades of Raynaud's phenomenon, digital ulcerations, and a prior left fifth finger amputation [Figures 1, 2], as well as telangiectasias around the lips and on the palms [Figures 1-3], and limited cutaneous systemic sclerosis for the past 2 decades which was untreated, presented to the clinic with symptoms of increased dyspnea, fatigue, and pre-syncopal sensations over the past several months.

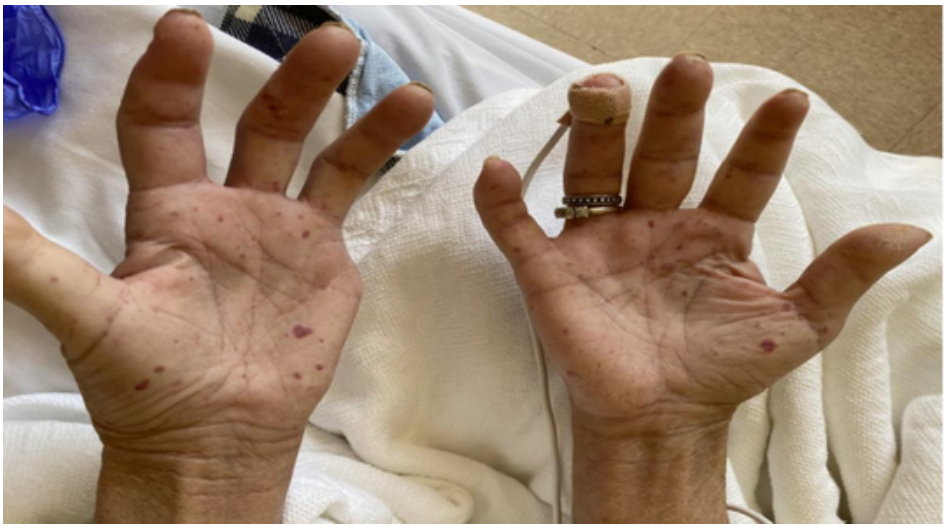

Figure 1: Showing palmar telangectasias and prior left 5th finger amputation

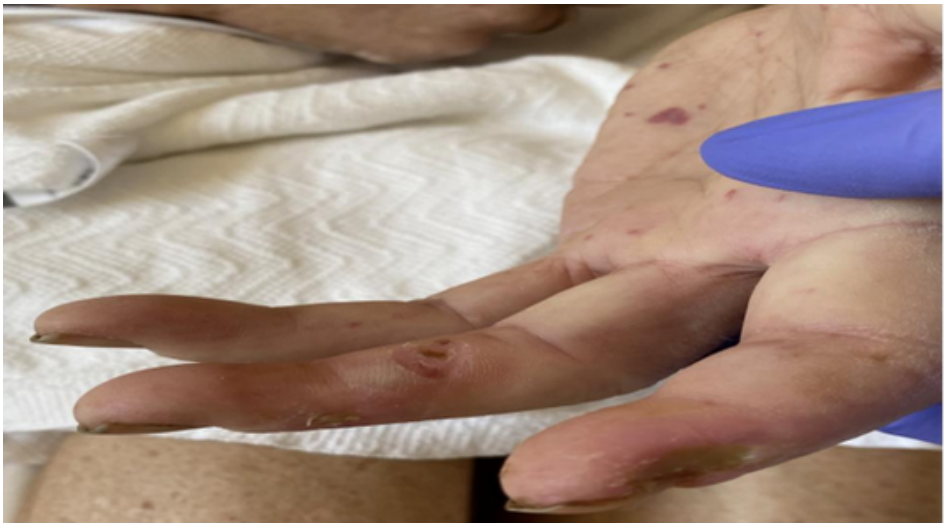

Figure 2: showing digital ulcers

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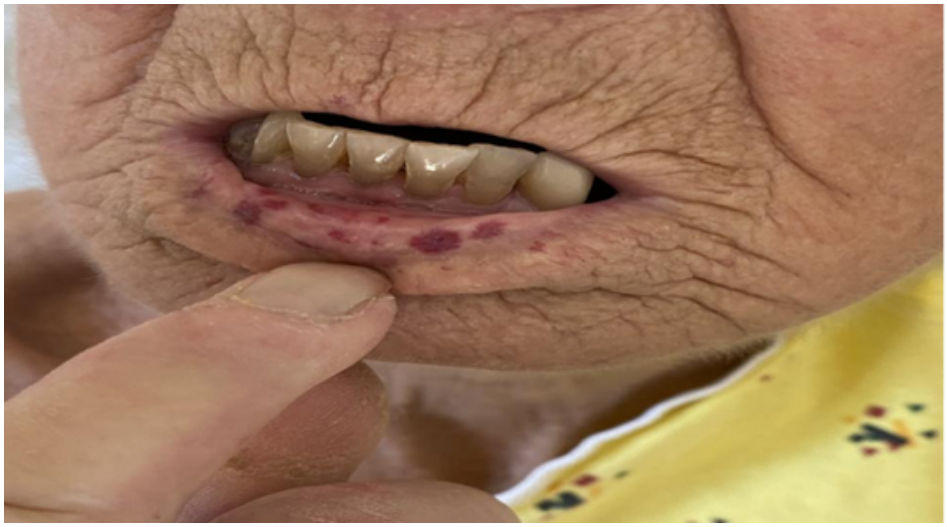

Figure 3: Showing telangectacias around the lip

The exam was notable for a blood pressure of 100/60 mm Hg, normal oxygenation, sclerodactyly, digital ulcers on the left hand (2nd and 3rd fingers), calcinosis cutis on the elbows, telangiectasias scattered throughout the palms, lips, and forehead, and clear lungs with mildly distant heart sounds. The patient was immediately transferred to the emergency department. Due to concern for underlying pulmonary hypertension, a transthoracic echocardiogram (TTE) and right heart catheterization (RHC) were performed. The TTE revealed an ejection fraction of 60-65% with a large pericardial effusion and significant tamponade physiology [Figure 4].

The RHC indicated a pulmonary artery pressure of 42/10/21 mm Hg and a pulmonary capillary wedge pressure of mean 5/4/1 mm Hg. The patient was admitted for emergent pericardiocentesis, with 1170 cc of fluid removed. Fluid analysis revealed a white blood cell count of 586 cells/uL with lymphocytes predominating at 88%, a protein concentration of 3.7 g/dL, a lactate dehydrogenase level of 159 U/L, and negative cytology findings. A CT chest scan showed no pulmonary or renal involvement. Autoimmune workup revealed positive antibodies as detailed in [Table 1]

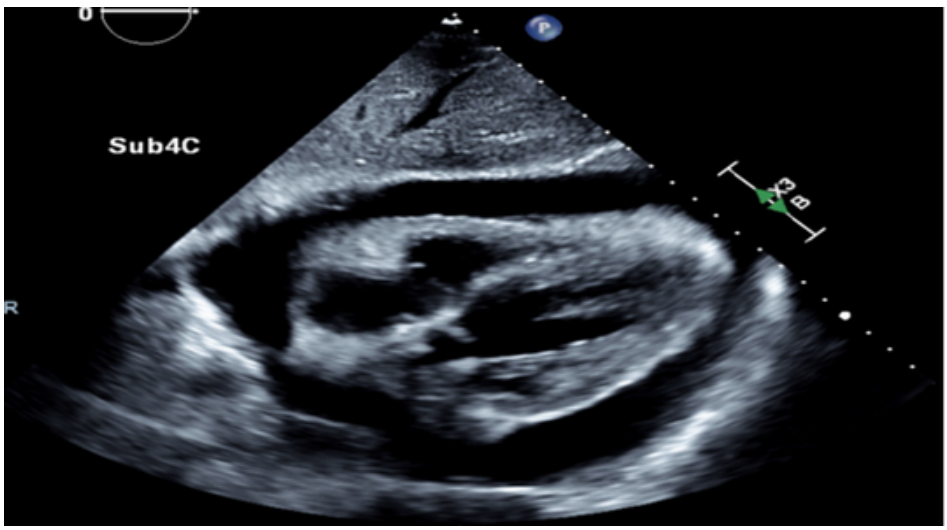

Figure 4: Showing massive pericardial effusion

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Antibodies	Titers
ANA	1:2560
Anti centromere Ab	>8
Anti-Scl-70 Ab	NEGATIVE
Anti-Smith Ab	NEGATIVE
Anti-RNP Ab	NEGATIVE
Complement levels	Normal
Antiphospholipid panel	NEGATIVE

ANA: Anti-nuclear antibody; Ab: Antibody; Scl-70: DNA-Topoisomerase-1; RNP: Ribonucleoproteins

Table 1: Antibody panel results

Sixteen days later, the patient was readmitted with recurrent cardiac tamponade and underwent balloon pericardiotomy, ultimately requiring a pericardial window. The patient was started on a tapering dose of steroids and colchicine, and at follow-up in the clinic, mycophenolate mofetil was added to the treatment regimen.

Discussion

Pericardial effusion occurs in systemic sclerosis (SSc) patients, though it is usually mild and asymptomatic. SHowever, symptomatic recurrent pericardial effusion leading to pericardial tamponade is exceptionally rare, particularly in patients with the limited cutaneous form of systemic sclerosis (lcSSc) subtype. Vlach Yiannopoulos with the limited cutaneous, PG et al. l states that retrospective studies reviewing the clinical records of 254 patients over a four-year period estimated an annual mortality rate of 2%. They found that the incidence of cardiac disease ranged from 7% in lcSSc patients to 21% in diffuse cutaneous SSc (dcSSc) patients [2]. LcSSc is a complex autoimmune disorder that infrequently affects the cardiovascular system; when it does, cardiac manifestations are often secondary to pulmonary hypertension. Autopsy studies have indicated that pericardial involvement is present in 52-73% of SSc patients, but it is often asymptomatic [3]. Recurrent pericardial tamponade is an unusual and severe complication. In the case described, a thorough evaluation excluded other potential causes such as overlap syndrome with systemic lupus erythematosus (SLE), paraneoplastic processes, infectious diseases, and pulmonary hypertension. The absence of these common causes highlights the rarity and complexity of this presentation.

The precise etiology of pericardial effusion in SSc remains unclear. Cardiac and pulmonary complications are significant sources of morbidity and mortality in SSc sclerosis [4]. Cardiac manifestations in SSc can include conduction abnormalities, early coronary artery disease, valvular issues, left ventricular diastolic dysfunction, and pericardial complications/effusions/conditions? [5]. Small pericardial effusions, less than 50 mL in volume, generally do not affect prognosis, but those exceeding 200 mLl are associated with a higher morbidity/mortality? [6]. Pericardial tamponade is infrequent, but has a high mortality rate of up to 55%, and is linked to a poor prognosis [7]. Few cases of lcSSc leading to pericardial tamponade have been reported, and specific treatment strategies for this complication are not well established [8]. Management typically involves medications such as corticosteroids, aspirin, NSAIDs, and colchicine, or surgical interventions like pericardiectomy or pericardial window. Despite the significant morbidity pericardial disease, data on treatment outcomes and decision-making for specific therapies remain limited.

Conclusion

LcSSc usually does not exhibit significant clinical symptoms of pericardial disease. It is crucial to identify symptoms as was done in this case and promptly investigate internal organ involvement.

Patient's consent

Consent was obtained or waived by all participants in this study.

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