

# Survival of an elderly patient with limited cutaneous systemic sclerosis, pretamponade and pulmonary hypertension

**Objective:** Conservative management of systemic sclerosis-induced pretamponade in the setting of pulmonary hypertension. **Methods:** A case report with a review of the literature. **Results:** An 81-year-old Caucasian woman with a history of limited cutaneous systemic sclerosis was admitted to the hospital for mechanical fall and shortness of breath. In the emergency department, she was cyanotic, in severe respiratory distress, tachycardic and hypertensive with oxygen desaturation. A chest x-ray showed cardiomegaly with bilateral infiltrates. An echocardiogram showed large pericardial effusion compressing mainly on the left ventricle, and right atrial collapse at the end of the diastole, with peak pulmonary pressure of 65 mmHg. The patient was managed conservatively with labetalol, intravenous fluids and furosemide until hemodynamically stabilized. The next day she had an elective pericardiotomy with a window. A total of 700 ml of serous exudative fluid was removed. Following the pericardial fluid drainage, the patient's symptoms resolved and a repeat echocardiogram showed no pericardial effusion and better left ventricular filling. The patient continued to be asymptomatic after 6 months of follow-up. **Conclusion:** Conservative management of systemic sclerosis-induced pretamponade is effective and may be superior to emergent pericardiocentesis.

**KEYWORDS:** anticentromere antibody ■ diffuse cutaneous systemic sclerosis ■ ECG ■ limited cutaneous systemic sclerosis ■ pericardial effusion ■ pericardiocentesis ■ pulmonary arterial hypertension ■ Raynaud's phenomenon ■ systemic sclerosis ■ tamponade

Ali Naboush\*<sup>1</sup>,  
Mokhtar Abdallah<sup>1</sup>,  
Deepak Asti<sup>1</sup>  
& Mark Goldstein<sup>1</sup>

<sup>1</sup>Staten Island University Hospital, NY, USA

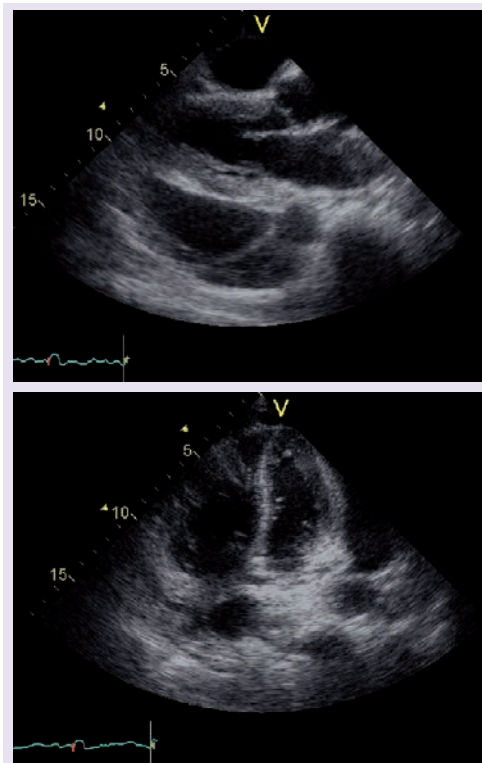
\*Author for correspondence:  
naboush@hotmail.com

Systemic sclerosis (SSc) is a connective tissue disease that involves changes in the skin, blood vessels, muscles and internal organs. It is divided into diffuse cutaneous SSc and limited cutaneous SSc (lcSSc) on the basis of the extent and distribution of skin involvement [1]. Involvement of the heart in SSc is common and has been estimated to occur in up to 100% of patients when sensitive tools are used [2]. When those patients have a symptomatic cardiac involvement, their prognosis is poor with 2- and 5-year mortality rates of approximately 60 and 75%, respectively [3]. Pulmonary arterial hypertension (PAH) is an important clinical complication of SSc and is a leading cause of mortality in this disease [4,5], and this factor may be independent of interstitial lung disease [6]. Symptomatic pericarditis also occurs in 7–20% of patients with SSc, with autopsy results showing pericardial involvement in up to 70–80% of patients with SSc [7]. Pericardial effusions are also common in SSc and can be small causing no symptoms or large causing hemodynamic instability that can lead to cardiac tamponade [8–24]. Hemodynamically significant pericardial effusions are rare in SSc and usually have a poor outcome [8–24]. This outcome is even worse in the setting of PAH [8–12].

## Case report

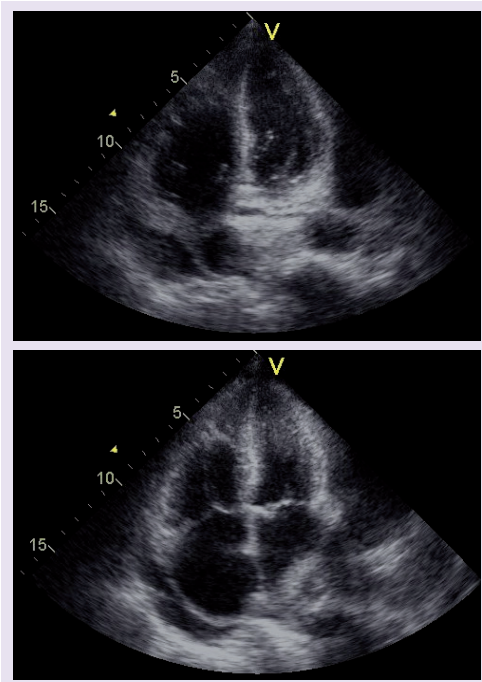
We report a case of lcSSc with hemodynamically significant pericardial effusion in the setting of PAH. An 81-year-old Caucasian woman with a history of hypothyroidism and lcSSc was admitted to the hospital for mechanical fall and shortness of breath. She was diagnosed with lcSSc 2 years before presentation after symptoms of fatigue, arthralgias and Raynaud's phenomenon. A blood workup was positive for antinuclear antibodies: anticentromere antibodies (1:640) and was negative for SM/RNP, SM, SS-A, SS-B, SCL-70, DNA DS, antiphospholipid, anticardiolipin and myeloperoxidase antibodies as well as CK, aldolase, cryoglobulin TSH, C3 and C4. The patient failed symptomatic treatment for her Raynaud's phenomenon, so she had partial autoamputation of the left second and third fingers 1 month prior to admission.

On presentation to the emergency department she complained of mechanical fall, after missing a step and hitting her head on the wall. She had no palpitations, chest pain, syncope or loss of consciousness. She also complained of shortness of breath on exertion over the last 1 month, which was increasing in intensity. She reported no fever, chills or other associated symptoms. On physical



**Figure 1.** Echocardiogram showing left ventricular collapse and enlarged right ventricle at the end of diastole.

examination; she was cyanotic, in severe respiratory distress, with a heart rate of 120 beats per minute, blood pressure (BP) of 184/110 mmHg



**Figure 2.** Echocardiogram showing right atrium collapse at the end of diastole.

and oxygen saturation in the low 80s. Chest auscultation showed basal crackles, a pericardial rub and no murmurs. A chest x-ray showed cardiomegaly, bilateral interstitial infiltrates and small pleural effusion. An ECG showed sinus tachycardia at 125 beats per minute with right bundle branch block. The initial blood workup was benign, including creatinine, except for elevated brain natriuretic peptide. Urine analysis was negative except for very minimal proteinuria. The patient was put on oxygen treatment and received one dose of intravenous (iv.) labetalol to decrease her BP. Computed tomography of the head was negative and computed tomography of the chest showed cardiomegaly, with interstitial infiltrates suggestive of congestive heart failure and large pericardial effusion mainly compressing the left side of the heart. A bedside echocardiogram was carried out showing normal ejection fraction, paradoxical motion of septum, moderate-to-large pericardial effusion compressing on left ventricle with decrease diastolic filling, right atrial collapse at the end of diastole with systolic pulmonary peak pressure of 65 mmHg (FIGURES 1 & 2). The patient's BP dropped significantly to below 90 mmHg so she was started gently on iv. fluids. After 6 h of that, the patient had an acute episode of shortness of breath, high BP, increased crackles on lung examination and oxygen desaturation. She was given one dose of iv. furosemide and transferred to the intensive care unit for close monitoring. She also had one episode of supraventricular tachycardia managed with adenosine. The next day, she underwent an elective pericardiotomy with pericardial window placement. In total, 500 ml of serous pericardial effusion was removed and a drain was left in place (total yield of 200 ml). Pericardial fluid was amber in color, hazy in appearance with analysis showing an exudative fluid as per the Lights Criteria, was negative for acid fast bacilli and culture. The cytology was negative for malignant cells showing mixed inflammatory and mesothelial cells. Repeat ECG after stopping the drainage showed no pericardial effusion and improvement of left ventricle diastolic filling. The patient's dyspnea improved following the drainage. She was maintained on oral furosemide and diltiazem until she was discharged with no further events after 6 months of follow-up.

### Discussion

SSc associated with hemodynamically significant pericardial effusion requiring drainage is rare with only 29 cases reported in the medical literature [8-24]. Those cases can be divided into:

eight cases associated with severe PAH, with a mortality rate of 62.5% [8–12]; five cases with no PAH or renal disease, with a mortality rate of 0% [8,13–16]; seven cases with renal disease with a mortality rate of 42.8 [17–21]; and the rest with unreported comorbidities [23–26].

In the group of patients with PAH (TABLE 1), the average age was 40.8 years, most of them were females (seven out of eight), having either limited disease (three cases) or diffuse disease (five cases), with a range of duration of 1–18 years of SSc. Of those who died, all the patients had a systolic BP below 100 mmHg and average systolic pulmonary BP of 87.2 mmHg [8,10–12] compared with higher BP in the group who survived, and relatively lower systolic pulmonary BP except for

one case [9]. Interesting is the echocardiographic finding seen in some of those patients where the left atrium and left ventricle collapse [9,11,12] as opposed to the pathognomonic signs of tamponade, which usually involves collapse of the right side of the heart. This can be explained by the elevated right-sided pressure that can often exceed those in the left side. The left ventricular collapse can possibly be exaggerated following pericardiocentesis, leading to shock. All the mentioned deaths occurred within a short period of time after pericardiocentesis probably from severe right ventricular failure leading to shock and arrhythmia [8,10–12]. On the other hand, the patients without PAH survived despite frank tamponade [15,16]. The mechanism of death in

**Table 1. Cases of systemic sclerosis with hemodynamically significant pericardial effusion requiring drainage in the setting of pulmonary hypertension.**

Study (year)	Age (years)	Sex	SSc duration (years)	SSc type	SBP (mmHg)	Chest x-ray	ECG features	PBP (mmHg)	PF volume (cc)	Outcome	Ref.
Uhl <i>et al.</i> (1979)	18	F	3	dcSSc	Not detected	Cardiomegaly, bilateral pleural effusions	Large effusion, paradoxical septal motion	NK	500	Died	[8]
Sattar <i>et al.</i> (1990)	45	M	10	lcSSc	95/60	Cardiomegaly	Large pericardial effusion	98	850	Died	[10]
Hemnes <i>et al.</i> (2008)	55	F	1	SSc	89/62	Cardiomegaly	Large pericardial effusion, RV hypertrophy, respiratory variation, LV collapse	83	600	Died	[11]
Dunne <i>et al.</i> (2011)	27	F	6	lcSSc	100/77	Cardiomegaly	Large pericardial effusion, RA collapse, respiratory variation, IVC plethora	81	400	Died	[12]
Dunne <i>et al.</i> (2011)	46	F	15	lcSSc	100/60	NK	Large effusion, LA, LV collapse	87	400	Died	[12]
Uhl <i>et al.</i> (1979)	48	F	6	dcSSc	100/60	Cardiomegaly, bilateral pleural effusions	Large effusion, paradoxical septal motion	NK	550	Survived	[8]
Lee <i>et al.</i> (1989)	46	F	15	SSc	124/86	Cardiomegaly, bilateral interstitial infiltrates	Diastolic LV collapse, severe TR, RV hypertrophic	80	500	Survived	[9]
Dunne <i>et al.</i> (2011)	42	F	18	dcSSc	NK	NK	Large effusion, RA, RV collapse	44	2300	Survived	[12]
This case report (2013)	81	F	2	lcSSc	181/100	Cardiomegaly, bilateral interstitial infiltrates	Large effusion, RA collapse	65	700	Survived	

dcSSc: Diffuse cutaneous systemic sclerosis; F: Female; IVC: Inferior vena cava; LA: Left atrium; lcSSc: Limited cutaneous systemic sclerosis; LV: Left ventricle; M: Male; NK: Not known; PBP: Pulmonary blood pressure; PF: Pericardial fluid; RA: Right atrium; RV: Right ventricle; SBP: Systemic blood pressure; SSc: Systemic sclerosis; TR: Tricuspid regurgitation.

those patients is unclear, but is consistent with some recently published observation made at Johns Hopkins University (MD, USA) that more mortality is associated with percutaneous drainage of pericardial fluids in the setting of significantly elevated pulmonary artery pressure [27]. In this setting, the removal of pericardial fluid is like removing the pericardium allowing a volume overloaded right ventricle to enlarge. This in turn will decrease the left ventricular filling causing hypotension, heart failure, decreased coronary flow in diastole and death [21].

There is a reported case of pericardial effusion in the setting of PAH treated with fluids and diuretics without drainage of fluids [28], raising the importance of conservative treatment in those patients as opposed to emergent drainage that might cause significant mortality. Similarly, our patient had large pericardial effusion with impending tamponade and was managed conservatively with iv. fluids and diuretics, which probably decreased her pulmonary artery pressure. Then she underwent an elective pericardiectomy that, together with the initial management, decreased the deleterious effects of emergent pericardiocentesis in the context of elevated right ventricular pressure and left ventricle collapse. We believe that this should be the mainstay of treatment in cases where patients can tolerate it. Those patients in addition to the ones with frank tamponade are so critical that they should be admitted to the intensive care unit with close hemodynamic monitoring via pulmonary Swan–Ganz catheters, with cautious gradual removal of pericardial effusion [9].

Our case is unique in that this is probably the first reported case of SSc with hemodynamically significant pericardial effusion requiring drainage in the setting of severe pulmonary hypertension to be managed conservatively with iv. fluids and diuretics followed by elective pericardiectomy. In

addition, this is the first reported case of survival from cardiac tamponade in a patient with lcSSc and pulmonary hypertension who is over 60 years of age.

### Limitations

This case is unusual in that it occurred in a patient who had a diagnosis of lcSSc at an older age where diastolic dysfunction is also common. We cannot generalize the management of this case to young patients with SSc who might have different hemodynamic statuses.

### Conclusion

In summary, a large pericardial effusion in the presence of PAH in SSc represents hemodynamic instability and poor prognosis. This prognosis is largely dependent on the systemic BP at presentation and the severity of PAH irrespective of the duration of scleroderma or the amount of fluids drained. When appropriate it is better to manage this group of patients conservatively before undergoing pericardial fluid drainage.

### Financial & competing interests disclosure

*The authors have no relevant affiliations or financial involvement with any organization or entity with a financial interest in or financial conflict with the subject matter or materials discussed in the manuscript. This includes employment, consultancies, honoraria, stock ownership or options, expert testimony, grants or patents received or pending, or royalties.*

*No writing assistance was utilized in the production of this manuscript.*

### Ethical conduct of research

*The authors state that they have obtained appropriate institutional review board approval or have followed the principles outlined in the Declaration of Helsinki for all human or animal experimental investigations. In addition, for investigations involving human subjects, informed consent has been obtained from the participants involved.*

### Executive summary

- Conservative management of systemic sclerosis-induced tamponade in the setting of pulmonary arterial hypertension might be superior to emergent pericardiocentesis.
- This is the first case of limited cutaneous systemic sclerosis to survive tamponade in the setting of pulmonary arterial hypertension.
- Tamponade with low systolic blood pressure is a very poor prognostic sign in systemic sclerosis the setting of pulmonary arterial hypertension.

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