CASE REPORT

New development of cardiac tamponade on underlying effusive–constrictive pericarditis: an uncommon initial presentation of scleroderma

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SUMMARY

A 40-year-old man with a medical history of hypertension was admitted for weight loss, generalised weakness, joint pains and mottling of fingertips. The initial laboratory data revealed microangiopathic haemolytic anaemia, thrombocytopenia and acute renal failure. Intravenous steroids were started for possible diagnosis of systemic lupus erythematosus based on admission assessment. Intravenous immunoglobulin and plasmapheresis were subsequently added to the treatment plan to cover thrombotic thrombocytopenic purpura while his autoimmune panel was pending. The echocardiogram study on day 2 revealed cardiac tamponade for which he underwent pericardiocentesis and right heart catheterisation. The atrial waveforms postpericardiocentesis demonstrated effusive–constrictive pericarditis. His clinical condition kept on deteriorating with reaccumulation of pericardial effusion and further complicated by hemoperitoneum and colonic obstruction. He had cardiorespiratory arrest on his fourth admission day and was not revived. Anti-Scl-70 antibody came back positive. Autopsy findings confirmed the presence of fibrinous pericarditis and hemoperitoneum.

BACKGROUND

Scleroderma is a chronic connective tissue disorder characterised by vascular dysfunction and excessive fibrosis. Cardiac involvement in scleroderma can be manifested as direct myocardial, pericardial or conduction system abnormalities or can be secondary to scleroderma renal crisis or pulmonary arterial hypertension. While pericardial effusion is frequently documented in the literature, cardiac tamponade requiring pericardiocentesis is a rare finding.¹ Calcific constrictive pericarditis has also been reported in calcinosis, raynaud phenomenon, esophageal dysmotility, sclerodactyly and telangiectasia (CREST) syndrome.² Treatment of underlying visceral pericardial constriction by pericardiectomy remains the mainstay of management in effusive-constrictive pericarditis.³ The role of emergent corticosteroid therapy remains controversial, as there were a few case reports suggestive of deterioration of clinical condition after starting steroids.⁴

We describe a case of a 40-year-old African-American man with undiagnosed scleroderma who developed an acute pericardial effusion with tamponade within 24 h of his admission after he was started on steroids. His right heart catheterisation revealed almost equal cardiac filling pressures in all the four chambers. In cardiac catheterisation tracing, the

attenuated y-descent prior to drainage indicated cardiac tamponade but development of a steep y-descent after the drainage, unmasked the constrictive nature of the pathology. He had a rapidly worsening clinical course with reaccumulation of pericardial effusion, despite being on corticosteroids. As effusive–constrictive pericarditis secondary to scleroderma is a very rare clinical entity and sudden development of tamponade within 24 h has never been reported in the literature. We are presenting this case to highlight this unusual clinical presentation, discuss the possible causes of sudden decompensation and diagnostic and management challenges encountered in similar clinical scenarios.

CASE PRESENTATION

A 40-year-old African-American man with medical history of hypertension was brought to emergency room (ER) with 3–4 months history of gradually progressive generalised weakness. His symptoms worsened to the point of limiting his ambulation to less than a block. On further questioning, he was also found to have a 20 pounds weight loss during the past 2 months associated with anorexia, decreased vision, on and off dizziness and photosensitivity. Review of systems was positive for 6 months history of joint pain in both the knees and small joints of the hands and recently bluish discoloration of the fingertips. Family history and social history were unremarkable.

Physical examination revealed a cachectic, African-American man who was tachycardiac (HR 106 bpm), hypertensive (BP: 183/110 mm Hg) and febrile (101°F) at the time of presentation. He was noted to have conjunctival pallor, hyperpigmented lesions on the face and trunk, minimally retractable, firm skin on distal part of extremities, bluish discoloration of finger tips. Visual acuity was decreased to finger-counting at 4 ft distance. Deep tendon reflexes were bilaterally decreased and stiffness of the small joints of the hands with inability to make a full fist noted. Cardiac auscultation revealed normal S1 and S2 with an early diastolic pericardial knock but no rub or murmur. There was mild jugular venous distension but no Kussmaul's sign or pulsus paradoxus were noted. He was found not to have any other significant abnormal findings on chest auscultation. There were no palpable visceromegaly, no signs of fluid overload, no lymphadenopathy, no focal neurological deficits or signs of meningeal irritation.

Initial laboratory data on admission to ER revealed severe thrombocytopenia (platelet count of

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Figure 1 Initial electrocardiogram with normal sinus rhythm, normal voltage with no electrical alternans or evidence of ischaemia.

25 000/ μ L), microcytic, hypochromic anaemia and acute renal failure. Initial chest X-ray (CXR) showed cardiomegaly. CT head was unremarkable. Electrocardiogram showed a normal sinus rhythm, normal voltage with no electrical alternans or evidence of ischaemia (figure 1).

Considering his clinical presentation and initial laboratory findings suggestive of the possibility of an immune-mediated disease process, basic autoimmune work up was sent (table 1). He was started on steroids immediately on the same day of admission. On day 2 as there was no clinical improvement; haematology and rheumatology services were consulted. Peripheral blood smear showed evidence of microangiopathic haemolytic anaemia with 1+ schistocytes. Despite considering a SLE, CREST syndrome and Scleroderma as main differential diagnosis, the possibility of thrombotic thrombocytopenic purpura (TTP) could not be ruled out. Subsequently, the patient was started on IVIG and plasmapheresis later on day 2.

Also on day 2 of hospitalisation, two-dimensional (2D) echocardiogram (requested originally due to evidence of cardiomegaly on CXR, diastolic click and elevated JVD) revealed 3–4 cm of pericardial effusion with echocardiographic evidence of late diastolic right ventricular collapse and early pericardial tamponade (video 1). As the patient became haemodynamically unstable, emergent pericardiocentesis with right heart catheterisation was performed and 800 mL of exudative pericardial fluid was drained with only transient clinical improvement. On right heart catheterisation, the venous waveform prior to drainage of effusion showed an amputated 'y' descent indicating cardiac tamponade, while following the drainage, a steep 'y' descent along with a steep 'x' descent appeared. This finding was suggestive of presence of effusive constrictive pericarditis (figure 2).

In the meantime, his autoimmune panel came back as reactive for both rheumatoid factor and antinuclear antibodies. His clinical condition further deteriorated with reaccumulation of pericardial fluid (moderate-to-large pericardial effusion on the repeat 2D echocardiograms with no echocardiographic evidence of tamponade), respiratory distress and worsening renal failure with severe metabolic acidosis. A CT surgery was consulted for a pericardial window for persistent pericardial effusion but before he could go for any surgical procedure, his clinical course was complicated by hemoperitoneum and colonic obstruction (figure 3). He was being conservatively managed for his bowel obstruction for almost 24 h when he went into acute respiratory distress followed by cardiac arrest. An emergent bedside pericardiocentesis revealed 300 mL of haemorrhagic pericardial

fluid but he did not survive. A few hours after death, anti-Scl-70 antibody was reported reactive making scleroderma the most likely cause of his multiorgan system involvement (table 1 for all the serology results).

INVESTIGATIONS

Initial laboratory data on admission to ER revealed severe thrombocytopenia, microcytic hypochromic anaemia and acute renal failure (table 1).

Two-dimensional echocardiogram revealed 3–4 cm of pericardial effusion with echocardiographic evidence of late diastolic right ventricular collapse and early pericardial tamponade (video 1).

On right heart catheterisation, the venous waveform prior to drainage of effusion showed an amputated 'y' descent indicating cardiac tamponade while following the drainage, a steep 'y' descent along with a steep 'x' descent appeared (figure 2).

DIFFERENTIAL DIAGNOSIS

Systemic lupus erythematosus, mixed connective tissue disease, scleroderma/CREST syndrome, TTP

TREATMENT

Patient was started on oral corticosteroids for possible autoimmune disorder, possibly SLE, on the day of admission. Since the possibility of TTP could not be ruled out based on his peripheral blood smear, thrombocytopenia and renal involvement, IVIG, plasmapheresis and intravenous corticosteroids were initiated on his second admission day. As he needed to undergo emergent pericardiocentesis, he received packed red blood cells and platelet transfusion for anaemia and thrombocytopenia prior to the procedure. His clinical condition continued to deteriorate and got intubated with mechanical ventilator support, vasopressors, continuation of plasmapheresis and steroids along with supportive care.

OUTCOME AND FOLLOW-UP

Patient had a rapidly worsening clinical course with multiorgan system involvement and could not survive beyond his fourth admission day. His autopsy report showed moderate amount of haemorrhagic effusion in pericardial cavity (200 mL) and presence of acute and chronic inflammation of the pericardium along with fibrinous exudates. Myocardium was hypertrophied with left ventricular thickness of 2.1 cm. There was evidence of necrotising arteriolitis and necrotising glomerulitis of the kidneys. Large amount of haemorrhagic peritoneal effusion was

Table 1 Laboratory values

Value	Day 0	Day 4	Normal range	Units
Haematology				
WBC	4.7	7.0	4.00–11.00	10 ⁹ /L
RBC	4.09	1.69	4.20–5.80	10 ¹² /L
HGB	10.0	4.8	12.3–17.3	g/dL
HCT	30.3	14.1	37.0–51.0	%
MCV	74.3	83.2	80.0–101.0	fL
RDW	19.1	22.9	11.5–14.5	%
PLT	25	20	130–450	10 ⁹ /L
Segs	81	83.6	40.0–75.0	%
Lymphs	8.3	11	19.0–48.0	%
Mono	8.6	5.3	3.4–9.0	%
Eos	1.6	0.1	1.0–3.0	%
Basos	0.0	0.0	0.0–0.8	%
Coagulation				
PTT	27.1	41.6	10.4–14.5	s
PT	12.1	16.0	9.2–12.8	s
INR	1.1	1.5	0.7–1.2	
Chemistry				
Glucose	118	298	65–100	mg/dL
BUN	55.0	64	8.0–25	mg/dL
Creatine	3.0	3.4	0.66–1.25	mg/dL
Sodium	128	141	133–145	mEq/dL
Potassium	3.2	5.1	3.5–5.5	mEq/dL
Chloride	86	93	96–109	mEq/dL
CO ₂	26	25	22–28	mEq/dL
Calcium	7.9	6.1	8.5–10.5	mg/dL
Phosphorus	12.0	12.0	2.5–4.5	mg/dL
Total protein	8.5	3.4	5.7–8.0	g/dL
Albumin	3.8	1.7	3.8–5.0	g/dL
Bilirubin	1.3	0.7	0.0–1.0	mg/dL
ALT	35	46	21–72	U/L
AST	41	38	17–59	U/L
Alkaline phos	72	34	38–126	U/L
Lactate	8.1	>12	0.7–2.1	mmol/L
Trop1	0.238	0.18	30–135	IU/L
Ferritin	490		17–464	ng/mL
Transferrin	209		215–365	mg/dL
TIBC	254		240–450	µg/dL
Iron	54		49–180	µg/dL
CRP	1.2		0–1	mg/dL
D-dimer	1659	2735	0–278	ng/mL
ANA	1:2560			
ANA pattern	Homogeneous			
RA factor	1:2			
Scl-70	1:8			
Anti-Sm Ab	Non-reactive			
C3	72		90–180	mg/dL
C4	14		16–47	mg/dL
LDH	813		313–618	IU/L
Haptoglobin	7		43–212	mg/dL
Anticentromere B Ab	<1			
Hepatitis B serology	Negative			
Hepatitis C serology	Negative			
Fluid analysis				
WBC: 20				
RBC: 7000				
Polys: 72%				

ANA, antinuclear antibody; AST, aspartate aminotransferase; ALT, alanine transaminase; BUN, blood urea nitrogen; CRP, C reactive protein; HCT, haematocrit; HGB, haemoglobin; INR, international normalized ratio; LDH, lactate dehydrogenase; MCV, mean corpuscular volume; PLT, platelets; PTT, partial thromboplastin time; PT, prothrombin time; RA, rheumatoid arthritis; RBC, red blood cell; RDW, red cell width; TIBC, total iron binding capacity; WBC, white blood cell.

Video 1 2D echocardiogram revealed 3–4 cm of pericardial effusion with echocardiographic evidence of late diastolic right ventricular collapse and early pericardial tamponade.

found but there was no evidence vascular injury or bowel perforation. Honey combing of bilateral lower lung lobes was also noted.

DISCUSSION

Scleroderma is a chronic, multisystem connective tissue disease of unknown aetiology characterised by the involvement of small

vessels resulting in fibrosis and obliteration of vessels and fibrosis of various organs including the heart.⁵ Cardiac involvement in scleroderma can be direct (myositis, cardiac failure, myocardial fibrosis, conduction system abnormalities, coronary artery disease and pericardial disease) or indirect due to the involvement of other organs (eg, pulmonary hypertension or renal crisis).⁵ Though myocardial fibrosis and heart block have been considered the most frequent clinical presentation of cardiac involvement. Recently, pericardial disease is being increasingly reported, probably secondary to widespread availability of 2D echo.^{5 6} The characteristics of pericardial involvement include fibrinous pericarditis, chronic fibrous pericarditis, pericardial adhesions and pericardial effusions.⁷ Necropsy studies report the prevalence of pericardial disease between 33% and 72% of cases, while symptoms from pericarditis occur in only 7–20% of patients.⁵ While chronic pericardial effusion is common in patients with scleroderma, the development of cardiac tamponade is extremely rare but has been described in patients with both diffuse and limited disease.¹

It was suggested that histologically, pericarditis in scleroderma is characterised by chronic inflammation of pericardium with a significant increase in fibrous tissue. This theory was supported by the studies done by Byers *et al*,⁸ that there was increased levels of mast-cell infiltration in the pericardium and the fibrosis could be the result of mast cells' release of basic fibroblast growth factor and histamine. Autopsy findings in our patient

Figure 2 (A–B) On right heart catheterisation, the venous waveform prior to drainage of effusion showing an amputated 'y' descent indicating cardiac tamponade while following the drainage, a steep 'y' descent along with a steep 'x' descent appeared. This finding was suggestive of presence of effusive constrictive pericarditis.

Figure 3 (A) CT scan showing massive pericardial effusion; (B) CT scan showing haemorrhagic peritoneal effusion with colonic dilation.

also demonstrated acute and chronic pericardial inflammation with fibrinous exudates.

Calcific constrictive pericarditis, as a rare complication of CREST syndrome, has also been reported in the literature.² Constrictive pericarditis, a late sequela of an inflammatory condition of the visceral pericardium, occurs when healing of an acute fibrinous or serofibrinous pericarditis results in obliteration of pericardial cavity and formation of granulation tissue. This is characterised by equalisation of pressures in the four cardiac chambers and is said to be the most significant haemodynamic criterion for the diagnosis of constriction. The right atrial pressure tracings in this condition classically show a deeper and steeper 'y' descent while the 'x' descent may equalise or exceed it. Therefore, the right atrial waveform in constrictive pericarditis appears to be an exaggeration of a normal venous pulse. Cardiac tamponade, on the contrary, characteristically has an abnormal venous waveform with an attenuated 'y' descent or an upwardly sloping segment replacing it.

Cardiac tamponade is exceptional in scleroderma patients and the treatment of choice is usually pericardiocentesis.^{9–12} The majority of the reported cases of tamponade had a poor outcome due to severe pulmonary hypertension, renal or heart failure, with infrequent exceptions.⁹ It is often suggested that pericardial fibrosis in scleroderma may predispose to pericardial tamponade¹³ and tend to develop more often in patients with chronic pericardial effusions. However, Uhl *et al*¹⁴ reported three cases of systemic sclerosis with patients presenting with clinical syndrome of acute pericarditis who developed severe tamponade necessitating pericardiocentesis. It is, therefore, conceivable that with the relative incapacity for distension in scleroderma complicated by pericardial fibrosis, only a small volume of pericardial fluid may precipitate pericardial tamponade.¹⁴ Furthermore, there is obliteration of pericardial cavity in constrictive pericarditis which barely contains the normal physiological amount of pericardial fluid. In this condition, even a small amount of increase in pericardial effusion, causing an increased pressure by the scarred pericardium, generates the clinical signs suggesting cardiac tamponade. This is a unique clinical condition known as effusive constrictive pericarditis. Effusive-constrictive pericarditis most likely represents an intermediate transition from acute pericarditis with pericardial effusion to pericardial constriction.

Effusive-constrictive pericarditis is characterised by a dynamic spectrum of clinical and haemodynamic patterns varying from

pure tamponade, effusive-constrictive physiology to complete constriction based on the physical properties of the pericardial fluid accumulating as effusion and the intrinsic properties of the injured and subsequently healing pericardium. Hancock^{4 15} described it as one of the types of subacute elastic constrictive processes with physical examination and haemodynamic findings prior to pericardiocentesis resembling tamponade. Drainage of pericardial fluid results in only incomplete resolution of elevated filling pressures despite lowering the pericardial pressure to normal and development of 'deeper and steeper' atrial y-descents and ventricular 'dip and plateau', thus unveiling the characteristics of constriction. Similar haemodynamic findings were seen in our patient on right heart catheterisation, prepericardiocentesis and postpericardiocentesis (figure 2).

Rapid diagnosis and treatment is indicated in cardiac tamponade. Intravascular volume expansion and inotropic support should immediately be started if the patient is haemodynamically compromised. Echocardiography-guided pericardiocentesis remains the urgent definitive treatment of choice regardless of the underlying cause. The decision to leave an indwelling catheter after initial complete drainage depends primarily upon the possible aetiology of pericardial effusion.

- ▶ In our patient, absence of pulsus paradoxus/electrical alternans/muffled heart sounds but the presence of diastolic pericardial knock were features suggestive of possible constrictive pericarditis on admission.
- ▶ Within 24 h, the patient's sudden clinical deterioration and haemodynamical instability with documented two-dimensional echo features suggestive of cardiac tamponade emphasise the importance of understanding the dynamic spectrum of disease from constriction to tamponade.
- ▶ The postpericardiocentesis right heart catheter pressure tracings were suggestive of effusive-constrictive pericarditis.
- ▶ Even though non-steroidal anti-inflammatory drugs are the standard of care for the management of acute pericarditis and effusive-constrictive pericarditis, our patient's renal failure precluded its use in first place.
- ▶ Even though worsening of renal function after starting steroids in scleroderma patients, in the form of scleroderma renal crisis is a well-known phenomenon, there were no clear reports against steroid use in pericarditis in such patients.
- ▶ Although there are some anecdotal reports of rapid clinical deterioration of pericardial disease after starting steroids,⁴ emphasises the need for more studies in this aspect.

Learning points

- ▶ Cardiac involvement in scleroderma can be direct (myositis, cardiac failure, myocardial fibrosis, conduction system abnormalities, coronary artery disease and pericardial disease) or indirect due to the involvement of other organs (eg, pulmonary hypertension, renal crisis).⁵ Though myocardial fibrosis and heart block have been considered the most frequent clinical presentation of cardiac involvement, pericardial disease is recently being increasingly reported. Necropsy studies report the prevalence of pericardial disease is in between 33% and 72% of cases, while symptoms from pericarditis occur in only 7–20% of patients.⁵
- ▶ Rapid diagnosis and treatment is indicated in cardiac tamponade. Intravascular volume expansion and inotropic support should immediately be started if the patient is haemodynamically compromised. Echocardiography-guided pericardiocentesis remains the urgent definitive treatment of choice regardless of the underlying cause.
- ▶ Effusive–constrictive pericarditis is characterised by a dynamic spectrum of clinical and haemodynamic patterns varying from pure tamponade, effusive–constrictive physiology to complete constriction based on the physical properties of the pericardial fluid accumulating as effusion and the intrinsic properties of the injured and subsequently healing pericardium. Effusive–constrictive pericarditis most likely represents an intermediate transition from acute pericarditis with pericardial effusion to pericardial constriction.
- ▶ Even though worsening of renal function after starting steroids in scleroderma patients in the form of scleroderma renal crisis is a well-known phenomenon; there were no clear reports suggesting against its use in pericarditis in such patients. Although there are some anecdotal reports of rapid clinical deterioration of pericardial disease after starting steroids,⁴ emphasises the need for more studies in this aspect.

- ▶ We are reporting this case due to rarity, unusual initial presentation, challenges in establishing the final diagnosis and aggressive clinical course.

Contributors SRS had the original idea of writing this up and made all the efforts to collect and analyse, write it up at the end with the help of RA. AV collected all the data like CT scan, echo, labs and proof reading. HC was the mentor of the project, corrected the mistakes and guided us through.

Competing interests None.

Patient consent Obtained.

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