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Moderate Transudate Pericardial Effusion and Systemic Sclerosis [CREST Syndrome]: A Rare Association

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Abstract | **Case Report**

CREST syndrome is a limited form of systemic sclerosis, characterized by five main symptoms: calcium deposits under the skin, blood vessel problems causing discoloration of the fingers [Raynaud's phenomenon], esophageal insufficiency, hardening of the fingers, and dilated blood vessels in the skin. In Mexico, 1.7 to 2.6 cases are registered per 10,000 new patients with skin diseases. It is more common in women, with an average ratio of 2 to 4, and the most frequent age of onset is between 30 and 50 years. Pericardial involvement in systemic sclerosis is usually mild and asymptomatic, with symptomatic pericardial disease reported in only 7% to 20% of patients. Echocardiography is the primary technique for the diagnosis, monitoring, and treatment of pericardial effusion. However, large pericardial effusions and cardiac tamponade are rare and associated with a poor prognosis, with a mortality rate as high as 55%. For patients deemed non-responders to first-line dual therapy, corticosteroids are added to the treatment regimen; in cases of medical treatment failure and pericardial constriction without pericarditis, pericardiectomy should be performed.

Keywords: CREST, systemic sclerosis, women, pericardium, effusion, corticosteroids.

INTRODUCTION

CREST syndrome is a limited form of systemic sclerosis, a disease that primarily affects the skin and other tissues of the body. It is called CREST because of five main symptoms: calcium deposits under the skin, blood vessel problems that cause discoloration of the fingers [Raynaud's phenomenon], difficulty moving the esophagus, stiffening of the fingers, and dilated blood vessels in the skin. It is a rare but slowly progressing disease that can significantly impact the daily lives of those affected.

The exact cause of CREST syndrome is unknown, although a combination of genetic and environmental factors is thought to play a role. This disease is more common in women than in men and usually begins between the ages of 30 and 50. Diagnosing CREST syndrome can be challenging because symptoms can vary widely and overlap with those of other autoimmune diseases [1].

The etiology of CREST syndrome is not fully understood, but it is considered an autoimmune disease in which the immune system mistakenly attacks the body's healthy tissues. A combination of genetic, environmental, and immunological factors is believed to contribute to its development.

Genetic factors: Genetic predisposition plays a key role in the development of CREST syndrome. Individuals with a family history of autoimmune diseases have been observed to have a higher risk of developing this condition. Furthermore, genetic variants, such as those in the HLA-DRB1 gene, have been identified as being associated with an increased risk of developing scleroderma, including CREST syndrome.

Environmental factors: Exposure to certain environmental factors can also contribute to the development of CREST syndrome. Exposure to chemicals and toxins, such as silica, silica dust, and organic solvents, has been associated with an increased risk of developing scleroderma, including CREST

syndrome. In addition, viral infections, such as Epstein-Barr virus, and bacterial infections, such as Mycoplasma pneumoniae infection, have also been implicated in the development of this disease.

Immunological factors: In CREST syndrome, the immune system produces autoantibodies, such as anticentromere antibodies, that attack connective tissue. This aberrant immune response causes inflammation and damage to the affected tissues, leading to fibrosis, a thickening and hardening of the tissue [2].

The global prevalence is approximately 0.3 to 24 per 1,000,000 patients. In Mexico, 1.7 to 2.6 cases are registered per 10,000 new patients with skin diseases. It predominates in women, with an average ratio of 2 to 4 people. The most frequent age of onset is 30 to 50 years. [3]

Interstitial lung disease and pulmonary arterial hypertension are the two most common complications and the main causes of morbidity and mortality. Cardiac tamponade is rare [4].

Pericardial involvement in systemic sclerosis is usually mild and asymptomatic, with symptomatic pericardial disease reported to occur in only 7% to 20% of patients. However, pericardial involvement is common, found in 33% to 72% of patients in autopsy studies and in 41% of patients on echocardiography; these results imply that we underestimate this situation [5]. Echocardiography is the main technique for the diagnosis, monitoring and treatment of pericardial effusion [6]

The pericardium, through the interaction of its fibrous parietal layer and its elastic visceral layer, plays a fundamental role in modulating cardiac pressure-volume dynamics, ensuring optimal cavity constriction and preventing overexpansion during cardiac cycles. The parietal pericardium is an external fibrous sac lined by a single layer of mesothelial cells, which envelops the proximal large arteries, pulmonary veins, and venae cavae to form the pericardial sinuses and recesses. The visceral pericardium is a serous membrane that directly covers and protects the cardiac surface. Cardiac motion is enhanced by 20 to 50 ml of serum ultrafiltrate within the pericardium, which is drained by lymphatic vessels on the epicardial and parietal surfaces into the mediastinal, peribronchial, and tracheobronchial lymph nodes [7].

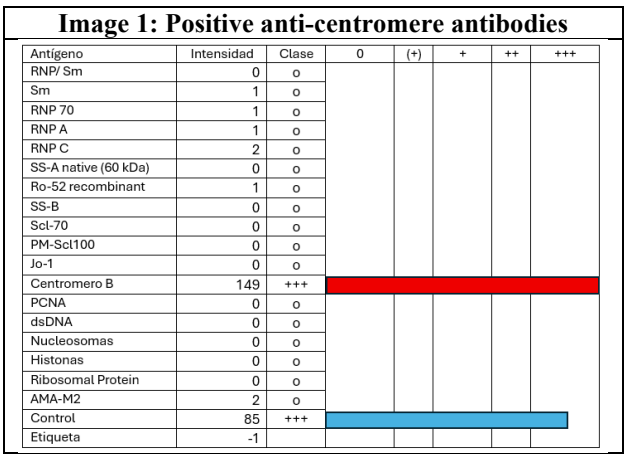
A previously undiagnosed patient with early skin changes of systemic sclerosis who develops cardiac tamponade presents a potential complication in diagnostic considerations. Up to 16% of patients will present with clinical symptoms of pericardial disease at some point during the course of their illness. This is much lower than the pericardial involvement demonstrated at autopsy, which reaches up to 70% [4].

Non-specific antibodies, such as antinuclear antibodies [ANA], are present in 95% of cases. Specific antibodies also exist: anticentromere antibodies are present in 21% of patients with diffuse systemic sclerosis and 71% of patients with CREST syndrome. Antibodies against DNA topoisomerase I [Scl-70] are found in 33% of patients with diffuse systemic sclerosis and in 18% of those with CREST syndrome [8].

Clinical Case

A 62-year-old female patient with an 8-year history of hypertension, managed with captopril 25 mg every 12 hours, had no known diabetes mellitus or dyslipidemia. However, she presented with lower extremity ulcers that began on her left leg 9 years prior, along with triphasic Raynaud's phenomenon for the past 5 years, mechanical knee pain, dysphagia to solids, and dyspnea on moderate exertion. She was evaluated by rheumatology in October 2025 and found to have distal scleroderma with absence of the distal phalanx of the second finger of her left hand, a Rodnan score of 6, and laboratory studies showing anti-centromere +++ [Image 1]. A diagnosis of limited systemic sclerosis was made, and her treatment was adjusted to celecoxib 100 mg orally every 12 hours, mycophenolic acid 500 mg orally every 8 hours, and sildenafil 25 mg orally every 24 hours.

Image 1: Positive anti-centromere antibodies



Antígeno	Intensidad	Clase
RNP/ Sm	0	o
Sm	1	o
RNP 70	1	o
RNP A	1	o
RNP C	2	o
SS-A native (60 kDa)	0	o
Ro-52 recombinant	1	o
SS-B	0	o
Scl-70	0	o
PM-Scl100	0	o
Jo-1	0	o
Centromero B	149	+++
PCNA	0	o
dsDNA	0	o
Nucleosomas	0	o
Histonas	0	o
Ribosomal Protein	0	o
AMA-M2	2	o
Control	85	+++
Etiqueta	-1	

The patient was admitted with dyspnea on minimal exertion. Control laboratory tests were performed on March 28, 2026, with hemoglobin 13.6, platelets 258, leukocytes 6300, glucose 90, BUN 21, urea 44, creatinine 1.0, chloride 107, potassium 3.5, sodium 141, phosphorus 2.7, calcium 8.7, magnesium 1.9, procalcitonin negative, BNP 2853, and D-dimer 767. Based on clinical and biochemical data suggestive of heart failure, treatment was initiated with spironolactone, atorvastatin, dapagliflozin, and supplemental oxygen via nasal cannula at 2 liters [FiO2 28%], resulting in partial improvement of symptoms.

On physical examination, we found a female of age consistent with the reported age, a four-point score of 16, alert, oriented, without characteristic facies, normocephalic, oral opening of 4 cm, neck with grade I jugular plethora, without organomegaly, symmetrical thorax with telangiectasias on the anterior face, lung fields with bibasilar hypoventilation, rhythmic precordium, heart sounds of low tone and intensity, with the presence of pleural friction rub, soft abdomen, normoperistalsis of 0.6 per minute, without signs of peritoneal irritation, upper extremities with distal scleroderma with absence of the distal phalanx of the second finger of the left hand, with telangiectasias on the palms of the hands bilaterally [Image 2] and distal sclerodactyly, moderate thickening Rodnan 6, pelvic extremities with preserved strength and sensation, REMS ++, without edema, capillary refill of 2 seconds, negative meningeal signs.

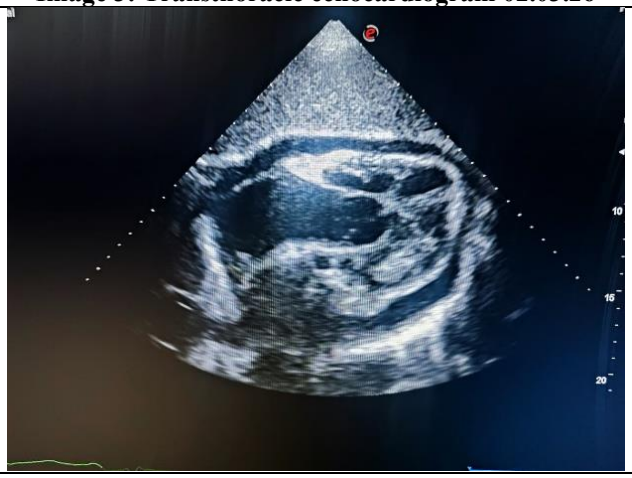
Image 2: Photograph of hands, palmar region.

Multiple disseminated telangiectasias, moderate skin thickening, and distal sclerodactyly are observed.

She was evaluated by cardiology on 02.03.26 for heart failure, documented by transthoracic echocardiogram [image 3] as right heart failure with moderate pericardial effusion, preserved LVEF of 75% with the following report:

1. Left ventricle of normal size, with concentric remodeling with relative wall thickness of 0.53 and ventricular mass index of 67 GR/M2 SC, end-diastolic volume 30 mls, end-systolic volume 7.43 mls, normal systolic function, LVEF of 75%.
2. Dilated right ventricle: basal diameter 46.4 x mid-diameter 37.3 x longitudinal diameter 64.8 mms, Tapse systolic dysfunction of 9.6 mm and shortening fraction of 18%.
3. Normal-sized left atrium: volume of 30 mls, dimensions of 30.3 mms x 49 mms.
4. Dilated right atrium, area of 25.95 cm2, dimensions 51.9 mms x 61.8 mms.
5. Trivalved aortic valve, with sclerosis of its leaflets, without insufficiency or stenosis, maximum gradient of 7.6 mm Hg, mean of 4.1 mm Hg and maximum speed 1.38 m/seconds.
6. Mitral valve with posterior valve sclerosis with mild insufficiency, maximum speed of 3.9 meters/seconds without stenosis.
7. Tricuspid valve with annular dilation and presents severe insufficiency, regurgitant volume of 141.8 mls, regurgitant orifice of 1.02 cm2, maximum gradient 63.2 mm Hg, maximum speed 3.97 m/sec with high probability for HP with estimated mean gradient of 54.8 mm HG.
8. Pulmonary artery with root of 21.7 mms, trunk of 26.4 mms, right branch of 12.5 mms, left branch of 12.5 mms, maximum gradient of 1.7 mm Hg, speed of 0.65 ms/sec, a mild central regurgitation jet is observed.
9. Pericardium with posteroinferior thickening of 8 mm, serosal separation of 18.1 mms is observed. Effusion volume of 518-741 CC by cube formula, estimated by volumetric method at 633 CC.
10. Dilated inferior vena cava, diameter of 24 mm, with less than 50% collapse

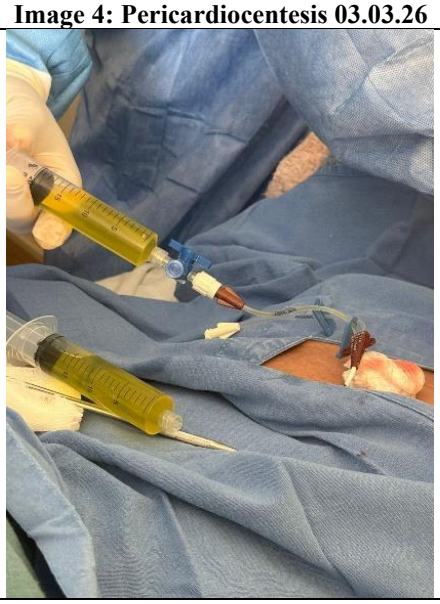
Image 3: Transthoracic echocardiogram 02.03.26



Data on pericarditis are observed with thickening of the visceral pericardium of 8 MM in the posteroinferior region [yellow arrow] and pericardial effusion of 518-741 CC by cube formula, estimated by volumetric method at 633 CC, without tamponade data.

Based on this data and prior informed consent, diagnostic and evacuation pericardiocentesis was performed [Image 4], draining on day 03.03.36 initially 340 ml of hyaline-serous characteristics, which were sent for cytological, cytochemical, Gram, culture, and India ink studies.

Image 4: Pericardiocentesis 03.03.26



Ultrasound-guided puncture was performed using a subxiphoid approach, obtaining 17 syringes of 20 ml each with a total of 340 cc.

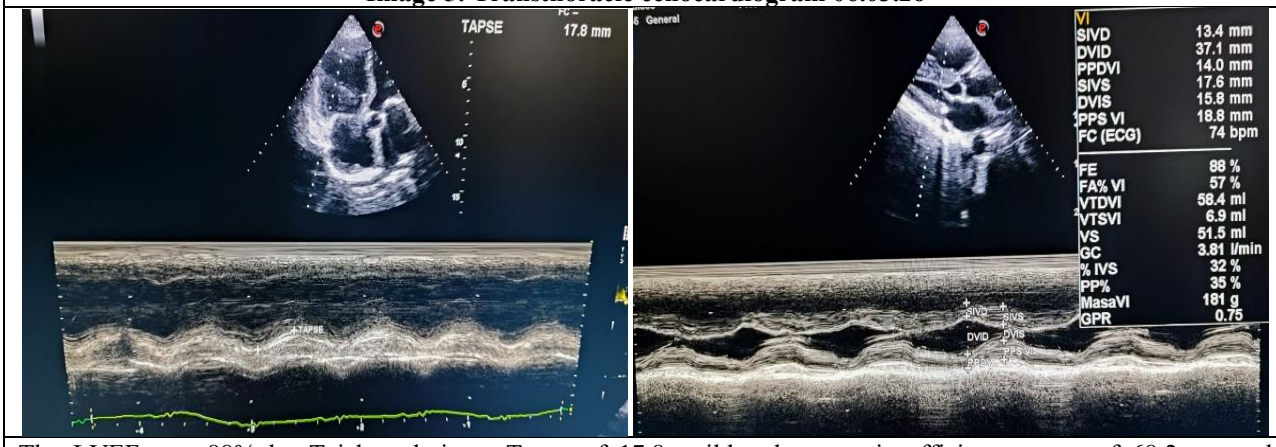
Laboratory studies of pericardial fluid with cytology are collected: turbid, yellow, pH 8, density 1010, glucose 96, LDH 167, cholesterol 50, uric acid 6.7, triglycerides 6, proteins more than 500, cell count of 21 leukocytes with differential of 89% with lymphocytes, Gram: negative, no bacteria observed, AFB negative, India ink negative.

Given studies compatible with transudate, steroid pulses were initiated with Methylprednisolone at 1 gram intravenously for 3 days [from 04 to 06.03.26], continuing drainage every 24 hours through an intracardiac catheter with a total of 552 mls: on 03.03.25 [340 mls], 04.03.25 [130 mls], 05.03.26 [55 mls], 06.03.26 [27 mls], ending the intravenous pulses and removing the intracardiac catheter without incident, with improvement of the clinical condition, with withdrawal of supplemental oxygen.

Follow-up lab results showed Hb 14.5, Hct 47.6, platelets 266, Leukocytes 12.6, glucose 118, BUN 26, urea 55, creatinine 0.9, Cl 106, K 4.52, Na 140, P 4.1, Ca 8.55, Mg 2, BNP: 1830, Trop: 1.5, Myoglobin 20. A new follow-up transthoracic echocardiogram [TTE] was performed on March 6, 2026 [Image 5], which showed an LVEF of 88% by Teich technique, right ventricular

systolic function with Tapse of 17.8, transtricuspid gradient of 49.43 mmHg, velocity of 3.5 m/s, mild pulmonary insufficiency, PSAP of 68.2, and half-pressure time of 48.2. m/sec, with an average velocity of 44.22, inferior vena cava thickness of 20 mms. There is no hemodynamic compromise, normal contractility of the ventricles, visceral pericardium without effusion, posteroinferior region with thickening and hyperechogenicity.

Image 5: Transthoracic echocardiogram 06.03.26



The LVEF was 88% by Teich technique, Tapse of 17.8, mild pulmonary insufficiency, psap of 68.2, normal contractility of the ventricles, visceral pericardium without effusion, posteroinferior region with thickening and hyperechogenicity.

The results of pericardial fluid culture from 03.03.26 were collected, showing no microbial growth at 168 hours; given the clinical improvement, echocardiographic study showing improvement in right heart failure, catheter removal without incident, and compliance with antibiotic prophylaxis with cephalosporin for 5 days due to the invasive procedure; It is concluded that it is a pericardial effusion secondary to an underlying rheumatological disease, with type 5 pulmonary hypertension and right heart failure, continuing treatment with mycophenolic acid 1 tablet orally every 8 hours, with oral prednisone calculated at 1 mg/kg with weekly decreases of 10 mgs until obtaining a maintenance dose of 5 mgs per day, spironolactone 50 mg orally every 12 hours, atorvastatin 40 mg orally every 24 hours, sildenafil 25 mg orally every 8 hours and apixaban 5 mg orally every 12 hours, with a cardiology appointment for a follow-up echocardiogram for evaluation of the effusion and to the rheumatology service for follow-up for the underlying pathology.

Results and Discussion

The two most serious complications of pericarditis are cardiac tamponade and constrictive pericarditis. Pericarditis with cardiac tamponade is a life-threatening condition in which inflammation of the pericardium leads to the compressive accumulation of pericardial fluid. Symptoms include chest pain, fatigue, and shortness of breath. Clinical signs include hypotension, tachycardia, elevated jugular venous pressure, pulsus paradoxus, muffled heart sounds, electrical alternans with decreased electrocardiographic voltage, and increased cardiac silhouette on chest radiography. The extent of clinical and hemodynamic abnormalities depends on the amount of pericardial fluid and the rate of accumulation, the distensibility and compliance of the pericardium, and the filling pressures of the cardiac chambers.

Pericardial constriction is a chronic condition characterized by thickening, fibrosis, and often calcification of the pericardium, leading to insufficient diastolic filling. It is termed constrictive pericarditis when the constriction is associated with pericarditis. Constrictive pericarditis is a consequence of pericardial inflammation and is characterized by scarring, fibrosis, and loss of pericardial elasticity. The main symptoms are dyspnea, edema, orthopnea, and fatigue. The typical clinical picture is characterized by isolated right heart failure with normal natriuretic peptide titers. Venous congestion, hepatomegaly, pleural effusions, and ascites may also be observed. Constrictive pericarditis should be treated empirically with anti-inflammatory therapy. In cases of medical treatment failure and pericardial constriction without pericarditis, pericardiectomy should be performed as soon as possible [9].

Large pericardial effusions and cardiac tamponade are rare and are associated with a poor prognosis with a mortality rate of up to 55% [10].

When clinical manifestations of cardiac damage become evident, the prognosis for patients is poor, with a mortality risk 2.8 times higher than that of those who do not suffer from them [11].

Traditionally, for patients considered non-responders to first-line dual therapy [who experience recurrence or intolerance to treatment with first-line

agents], corticosteroids were added to the treatment regimen. Low to moderate doses of corticosteroids [e.g., prednisone 0.2–0.5 mg/kg/day] were initiated and maintained until clinical remission, followed by a gradual reduction over several months [12].

On the other hand, the treatment of CREST syndrome is based on an organ-directed approach, as there is no curative therapy. Vasodilators such as calcium channel blockers are used for Raynaud's phenomenon; esophageal dysmotility is managed with proton pump inhibitors and prokinetics; while sclerodactyly and skin involvement may require immunomodulators in selected cases. In the presence of vascular or pulmonary complications, specific therapies such as endothelin antagonists or phosphodiesterase-5 inhibitors are employed. [13]

Regarding pericardial involvement, pericardial effusion in systemic sclerosis is usually mild and asymptomatic, so in many cases it only requires clinical monitoring and follow-up with echocardiography. However, in patients with moderate to severe or symptomatic effusions, management should be individualized. The use of anti-inflammatory drugs and corticosteroids may be considered, although the latter should be used with caution due to the risk of scleroderma renal crisis.

In cases of progression to cardiac tamponade, urgent pericardiocentesis is indicated. Likewise, in patients with persistent or recurrent effusions, or progression to pericardial constriction, surgical management via pericardiectomy may be required. It is important to emphasize that the presence of large pericardial effusions or tamponade in patients with systemic sclerosis is associated with a poor prognosis and high mortality rates, underscoring the importance of timely diagnosis and multidisciplinary management. [14]

CONCLUSIONS

The patient initially presented with signs of cardiac tamponade with right heart failure. Transthoracic echocardiography revealed pulmonary artery hypertension staged as type 5 secondary to systemic sclerosis, as well as a moderate pericardial effusion with thickening of the posterior region.

Given the lack of improvement despite management with anti-inflammatory treatment by rheumatology and the presence of data of cardiac tamponade, it was decided to perform diagnostic and therapeutic paracentesis with results of fluid compatible with transudate, an infectious process was ruled out, methylprednisolone pulses were administered with reduction of the effusion, improvement of right heart failure and remission of dyspnea.

This patient is an atypical case, with a rare and often underdiagnosed incidence due to the lack of routine cardiac screening tests to stage the type of pulmonary hypertension, estimate the type of effusion, and measure pleural thickness. To complete the diagnosis, we must request a right heart catheterization with pulmonary artery pressure measurement, periodic echocardiogram follow-up, and continued treatment with anti-inflammatory medications, diuretics, phosphodiesterase-5 inhibitors, statins, and anticoagulation.

Conflicts of Interest: The authors declare no conflicts of interest.

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ACCOUNTINGS

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