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Case Report

Internal Medicine Section

A Rare Case of Dilated Cardiomyopathy in Sjögren's Syndrome

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ABSTRACT

Dilated Cardiomyopathy (DCM) is a rare but significant complication in patients with primary Sjögren's Syndrome (pSS), a systemic autoimmune disease that primarily affects the exocrine glands. Cardiac involvement in pSS is uncommon, making the recognition of DCM in these patients crucial for timely intervention. This is a case of a 44-year-old female with well-controlled hypertension who developed progressively worsening shortness of breath, chest pain and bilateral pedal oedema. She also had a two-month history of dry eyes and dry mouth, indicative of sicca symptoms. Clinical examination revealed signs of fluid overload, a pansystolic murmur and sicca features. Electrocardiogram (ECG) and echocardiography demonstrated severe left ventricular dysfunction with an ejection fraction of 15%, consistent with DCM. Laboratory investigations revealed elevated N-terminal pro b-type natriuretic peptide (NT-proBNP) levels and positive anti-Ro52 and anti-La antibodies, pointing to an autoimmune aetiology. Cardiac Magnetic Resonance (CMR) Imaging confirmed non ischaemic DCM, ruling out ischaemic causes. The patient was diagnosed with DCM secondary to pSS. She was treated with diuretics, Angiotensin Converting Enzyme (ACE) inhibitors and immunosuppressive therapy, including methylprednisolone and a tapering course of prednisolone over six months. Significant clinical improvement was observed on follow-up. This case highlights the importance of recognising autoimmune aetiologies, such as pSS, in patients presenting with unexplained cardiomyopathy. Early diagnosis and interdisciplinary management are essential in preventing severe cardiac complications and improving patient outcomes in rare presentations of Sjögren-associated cardiomyopathy.

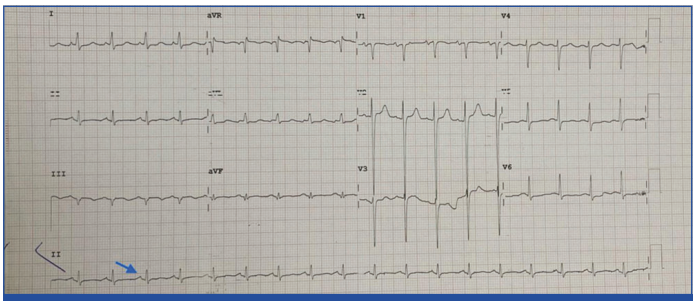
Keywords: Autoimmune disease, Cardiac magnetic resonance imaging, Heart failure, Systemic inflammatory response

CASE REPORT

A 44-year-old female presented with a one-week history of progressively worsening shortness of breath, initially experienced during exertion and later occurring even at rest. She described the shortness of breath as a sensation of tightness in her chest, which intensified while lying flat. Alongside this, she experienced intermittent chest pain, described as dull and non radiating, predominantly felt in the central chest area. This discomfort was associated with noticeable bilateral swelling in her lower extremities, which had worsened over the week. The patient also reported a two-month history of dry eyes and dry mouth, frequently needing to use artificial tears and drink water throughout the day to relieve the dryness, which had gradually worsened. Despite these complaints, she did not seek medical attention earlier. The patient denied any history of fever, cough, excessive sweating, palpitations, or light-headedness. She also had no prior episodes of similar chest pain or shortness of breath. Her medical history included well-controlled hypertension, managed with regular antihypertensive medication. There was no known history of coronary artery disease, diabetes, or thyroid dysfunction and she had no significant family history of cardiac or autoimmune diseases. She had no known allergies or recent illnesses and there were no recent changes in her medications.

On examination, her pulse rate was 90 bpm, blood pressure was 150/80 mmHg, respiratory rate was 28 breaths per minute, and oxygen saturation was 94% on room air. A general physical examination revealed dry eyes, dry mouth, a pale tongue, a butterfly-shaped rash on her face and pale palpebral conjunctiva, with no other significant findings. The cardiovascular examination showed raised jugular venous pressure, retraction of the apical impulse in the 5th intercostal space lateral to the midclavicular line, and a palpable left parasternal heave in the 3rd intercostal space. Auscultation revealed a pansystolic murmur in the left parasternal region. The respiratory examination noted percussion dullness on the right-side, reduced breath sounds, vocal fremitus and tactile fremitus on auscultation, with bilateral coarse crackles heard in the infrascapular region. Neurological and abdominal examinations were unremarkable.

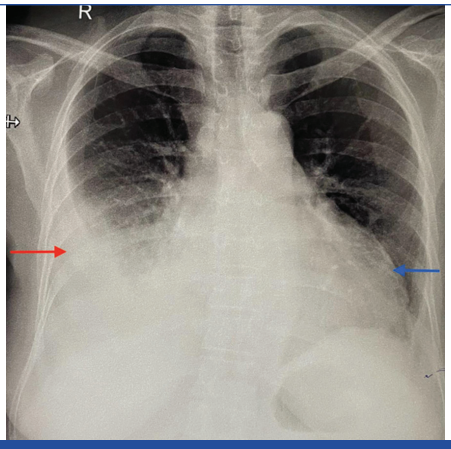
An ECG showed sinus tachycardia [Table/Fig-1], while a chest X-ray revealed cardiomegaly and right-sided pleural effusion [Table/Fig-2]. A 2D Echocardiogram (ECHO) demonstrated depressed Left Ventricular Systolic Function with an Ejection Fraction (LVEF) of 15%, dilation of all four chambers, grade II diastolic dysfunction, severe tricuspid regurgitation and no evidence of clots or vegetations [Table/Fig-3].



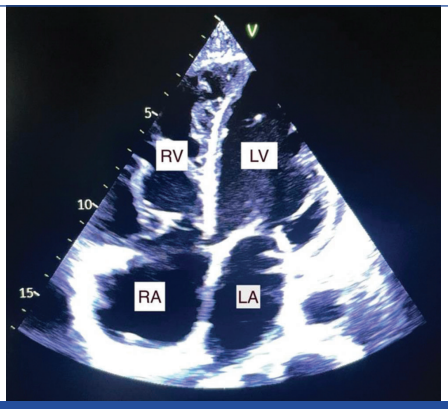
[Table/Fig-1]: Electrocardiograph (ECG) showing sinus tachycardia.

Laboratory investigations revealed positive results for anti-Ro52 and anti-La antibodies on an Antinuclear Antibody (ANA) blot, an NT-proBNP level greater than 35,000 pg/mL, and a haemoglobin level of 9 g/dL [Table/Fig-4] [1].

Based on the clinical examination, laboratory investigations and imaging findings, the provisional diagnosis included ischaemic cardiomyopathy, Systemic Lupus Erythematosus (SLE), Takotsubo cardiomyopathy and Dilated Cardiomyopathy (DCM) secondary to Sjögren's syndrome. The patient underwent coronary angiography, which revealed no significant coronary stenosis, effectively ruling out ischaemic heart disease as the cause of the cardiomyopathy. Subsequent CMR further supported the diagnosis, demonstrating



[Table/Fig-2]: Chest X-ray shows cardiomegaly (blue arrow) and right-sided pleural effusion (red arrow).

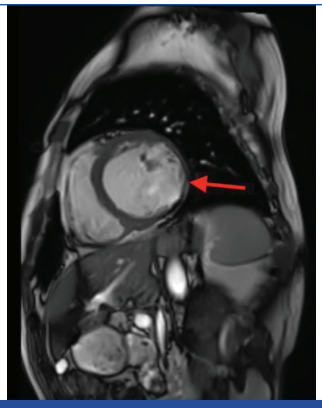


[Table/Fig-3]: A 2D-Echocardiography showing dilated LA/LV/RA/RV.

Investigation	Normal values [1]	Value on presentation	Value on follow-up after 2 weeks
Haemoglobin (g/dL)	11.6-15.0	9	9.2
Total leukocyte count (/µL)	4000-10,000	8200	8000
Platelet (/µL)	150,000-410,000	277,000	300,000
Total bilirubin (mg/dL)	0.22-1.20	0.77	0.9
Direct bilirubin (mg/dL)	Upto 0.5	0.51	0.50
Urea (mg/dL)	17 to 49	20	16
Creatinine (mg/dL)	0.6 to 1.2	1.0	0.8
Serum sodium (mmol/L)	136 to 145	137	140
Serum potassium (mmol/L)	3.5 to 5.10	3.8	3.9
Trop I (pg/mL)	Female: Upto 15.60	<10	-
CK-MB (U/L)	Upto 24	09	-
NTProBNP (pg/mL)	Age </=75 Years: </=125 Age >75 years: </=450	35,000	12000
ANA by IF	Negative	3+ Positivity; Titre- 1:640; Speckled Pattern	-
ANA BLOT	Negative	Anti Ro-52 Positive Anti SS-B Positive	-

[Table/Fig-4]: Laboratory investigations on presentation and on follow-up after two weeks.

an increased indexed Left Ventricular End-Diastolic Volume (LVEDV) and Left Ventricular End-Systolic Volume (LVESV), along with a markedly reduced LVEF of 20%. The absence of left ventricular hypertrophy, combined with global hypokinesia of the left ventricle and viability in 15 of the 17 segments, was consistent with non ischaemic cardiomyopathy. These findings confirmed the diagnosis of DCM likely secondary to Sjögren's syndrome [Table/Fig-5].



[Table/Fig-5]: Cardiac Magnetic Resonance Imaging (CMR) showing enlarged left ventricular chamber with increased Left Ventricular End-Diastolic Volume (LVEDV).

The patient was started on a furosemide drip to alleviate acute heart failure and Ramipril was initiated to slow the progression of DCM. A rheumatological opinion was sought for Sjögren's syndrome and DCM, where pulse therapy with methylprednisolone 1 gm for five days was initiated, followed by prednisolone 1 mg/kg with tapering doses every four weeks for a total of six months.

On follow-up, the patient showed significant improvement in symptoms, including reduced shortness of breath, chest pain and pedal oedema. Repeat laboratory investigations demonstrated a decline in NT-proBNP levels [Table/Fig-4]. The patient responded well to the combination of diuretics, ACE inhibitors and immunosuppressive therapy, with a gradual tapering of corticosteroids.

DISCUSSION

The pSS is a systemic autoimmune disease that primarily presents with sicca symptoms, characterised by dryness of the eyes and mouth. This dryness is a consequence of inflammation and damage to the lacrimal and salivary glands. While glandular manifestations are the hallmark of pSS, up to 50% of patients may experience extraglandular involvement, affecting various organs such as the joints, skin, lungs, gastrointestinal tract, nervous system and kidneys [2].

Interestingly, cardiac involvement in pSS is relatively rare but has recently drawn attention due to emerging evidence [3].

DCM is a non ischaemic heart disease characterised by structural and functional myocardial abnormalities. Clinically, DCM presents with left or biventricular dilation and systolic dysfunction, without underlying coronary artery disease, hypertension, or valvular abnormalities. While DCM is classified differently by the American Heart Association and the European Society of Cardiology [4], it remains a rare condition globally, with significant geographical variations in its incidence. In India, hospital-based studies suggest a higher prevalence of familial DCM, though broader epidemiological data is lacking [5].

Recent research has proposed a potential autoimmune basis for DCM, supported by findings such as cardiac immune cell infiltrates and the presence of cardiac autoantibodies in patients and their relatives. Given these immunological features, the connection between autoimmune diseases like pSS and DCM is becoming more plausible.

Cardiac involvement in pSS remains less explored compared to other rheumatic diseases. However, growing evidence suggests that patients with pSS may have an increased risk of developing heart failure. Advanced cardiac imaging techniques, such as myocardial deformation imaging and CMR, have detected significant structural abnormalities in pSS patients, even in subclinical stages. Subtle yet clinically important changes, like left ventricular diastolic dysfunction, are common and may be related to increased arterial stiffness [6]. This presents a diagnostic challenge, particularly as fatigue-a common feature of both pSS and heart failure-can complicate clinical evaluations.

Even in asymptomatic pSS patients, left ventricular dysfunction is significantly more prevalent compared to controls. Modern imaging modalities such as tissue Doppler echography and speckle-tracking echocardiography have demonstrated reduced systolic and diastolic myocardial velocities, suggesting early compromise in cardiac function. Similarly, 4D strain imaging highlights impairment in global longitudinal and area strains, likely due to the early involvement of subendocardial fibers [6].

CMR has proven particularly valuable in assessing myocardial involvement in pSS. Techniques such as T1/T2-weighted imaging and Late Gadolinium Enhancement (LGE) have identified non ischaemic inflammatory myocardial changes, often linked to higher disease activity. Additionally, CMR Feature Tracking (CMR-FT) has detected reduced left ventricular strain in pSS patients, particularly those with higher disease activity or Raynaud's phenomenon. Importantly, the presence of myocardial fibrosis in these patients correlates with higher salivary gland focus scores, suggesting that intense lymphocytic infiltration may contribute to fibrosis [6].

Due to the rarity of Sjögren's syndrome-associated cardiomyopathy, no standardised diagnostic or treatment protocols exist. Diagnosing cardiomyopathy in pSS patients requires careful consideration of its autoimmune nature, particularly when common causes such as ischaemic heart disease or hypertension have been ruled out [7]. The presence of DCM in patients with pSS, especially in the absence of traditional risk factors, warrants a deeper investigation into potential autoimmune triggers.

The management of pSS and DCM involves different therapeutic approaches due to the distinct nature of each condition; however, there are shared principles, particularly in managing inflammation and preventing cardiovascular complications. In pSS, therapies such as Glucocorticoids (GCs) are commonly used for their anti-inflammatory and immunosuppressive effects. However, prolonged use of GCs is known to negatively impact cardiovascular health, a concern that is also central to managing DCM. In both diseases, systemic inflammation is a key factor and controlling it is vital in preventing further complications, including cardiovascular events [8,9].

The use of Hydroxychloroquine (HCQ) in Sjögren's syndrome for its immunomodulatory properties may also indirectly benefit cardiovascular function by reducing systemic inflammation, which could play a role in the progression of cardiomyopathy. Similarly, the immunosuppressive properties of Rituximab (RTX), used in pSS to target systemic manifestations such as arthritis, can potentially limit inflammation that might contribute to cardiovascular involvement, although more research is needed to clarify this connection [8].

For patients with DCM, therapies such as ACE inhibitors, Angiotensin Receptor Blockers (ARBs), and beta-blockers, which are crucial for improving heart function, can also be relevant in managing cardiovascular complications in pSS patients with cardiac involvement. Patients with pSS are at an increased risk of cardiovascular disease due to systemic inflammation, making these drugs beneficial in mitigating heart failure symptoms or preventing the progression of cardiovascular conditions. In both diseases, these overlapping treatments emphasise the need for comprehensive management to address both systemic and cardiovascular health concerns [9].

Finally, for patients with refractory disease in either condition, more invasive options such as cardiac transplantation or Left Ventricular Assist Devices (LVADs) may be considered, though these options are typically reserved for advanced cases of DCM [9]. The shared cardiovascular risks in pSS and DCM highlight the importance of a multidisciplinary approach to optimise treatment outcomes and prevent further complications [8,9].

[Table/Fig-6] shows that, like other cases, this patient exhibited an autoimmune mechanism with elevated anti-SSA/SSB antibodies [3,7]. This strengthens the evidence of Sjögren's syndrome playing a role in the development of DCM, which is a key takeaway for comparative analysis.

Feature	Current case (2024)	Nishinarita M et al., (2000) [3]	Al Turk Y et al., (2021) [7]
Patient demographics	44-year-old woman with a two-month history of dry eyes and dry mouth, hypertensive	33-year-old Japanese woman	69-year-old woman with chronic kidney disease and anaemia
Pathogenesis	Autoimmune mechanism suspected with positive anti-SSA/SSB antibodies	Autoimmune association with positive SSA antibody during the clinical course	Autoimmune aetiology suspected; positive anti-SSA/SSB antibodies
Cardiac manifestations	DCM with reduced Left Ventricular Ejection Fraction (LVEF) of 15%, severe tricuspid regurgitation, biventricular dilation	DCM with LVEF of 32%, severe hypokinesis and cardiomegaly	Cardiogenic shock with LVEF of 20%, apical ballooning and MR
Laboratory findings	Elevated NT-proBNP (>35,000 pg/mL), positive anti-SSA/SSB antibodies, low haemoglobin (9 g/dL)	High IgG, positive SSA antibodies, mild hypocomplementemia	High inflammatory markers (CRP, ESR), positive SSA/SSB antibodies
Imaging and testing	Echocardiogram (ECHO) showed depressed LV systolic function (LVEF 15%), MRI confirmed non ischaemic cardiomyopathy	Echocardiogram (ECHO) revealing diffuse hypokinesis	Echocardiogram (ECHO), cardiac MRI showing Late Gadolinium Enhancement (LGE)
Treatment	Diuretics, ACE inhibitors, immunosuppressive therapy (methylprednisolone, prednisolone)	High-dose corticosteroids, immunosuppressive therapy	Steroids, heart failure management, transplantation declined
Outcome	Patient's condition improved post-treatment with significant reduction in heart failure symptoms	Cardiac dysfunction progressed; patient died four months post diagnosis	Patient died after cardiogenic shock and declining transplantation
Differential diagnosis	Ruled out ischaemic heart disease with coronary angiography; viral myocarditis also excluded	Ruled out viral infection and sarcoidosis	Ruled out ischaemia, infection and adrenal crisis

[Table/Fig-6]: Various comparative analysis studies in view of diagnosis, treatment and outcomes [3,7].

CONCLUSION(S)

DCM secondary to pSS was identified, confirmed by ECG and elevated anti-Ro52/anti-La antibodies, despite the rarity of cardiac involvement in Sjögren's. This case highlights the importance of considering autoimmune aetiologies in patients with unexplained heart failure. The patient showed significant clinical improvement following the timely initiation of immunosuppressive therapy and heart failure management.

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