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Sjögren-related cardiomyopathy presenting with cardiogenic shock

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Sjögren-related cardiomyopathy presenting with cardiogenic shock

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SUMMARY

Previous reports have described non-ischaemic cardiomyopathy related to a variety of autoimmune diseases. However, very few case reports describe Sjögren disease as a contributing factor to cardiomyopathy. We report the case of a 69-year-old woman with a history of Sjögren disease who presented with cardiogenic shock. Laboratory testing and cardiac MRI revealing apical septal late gadolinium enhancement were consistent with an autoimmune aetiology. After ruling out ischaemic, infectious and other possible causes, the patient's clinical presentation was thought to be related to underlying Sjögren disease. She was treated with intravenous steroids and evidence-based heart failure therapy, but she eventually died after having declined heart transplantation. Given the rarity of Sjögren disease, no diagnostic criteria or standard treatment has been established for cardiomyopathy related to this disease. Diagnosis should be considered in patients who show evidence of autoimmune processes after other possible causes are ruled out.

BACKGROUND

Non-ischaemic cardiomyopathy has multiple potential aetiologies, including autoimmune disease. Sjögren syndrome is a systemic autoimmune disease that primarily affects the exocrine glands and typically presents as persistent dryness of the mouth and eyes due to functional impairment of the salivary and lachrymal glands.[1] A variety of other disease manifestations affecting multiple organs and organ systems may occur, and the clinical features of Sjögren syndrome can be divided into the two broad categories of exocrine glandular features and extraglandular features.[2] Cardiovascular involvement has multiple manifestations and is most commonly seen as pericarditis[3] or autonomic dysfunction.[4] Acute systolic heart failure due to primary Sjögren syndrome is extremely rare, with only a few reported cases in the literature[5–9] and only one case reporting Sjögren disease flare-up as a possible cause of Takotsubo cardiomyopathy.[9]

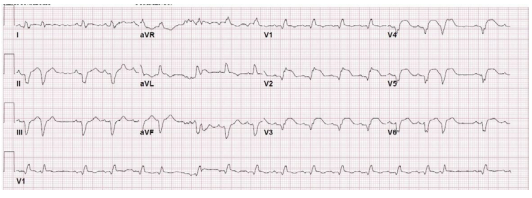


Figure 1 ECG on presentation.

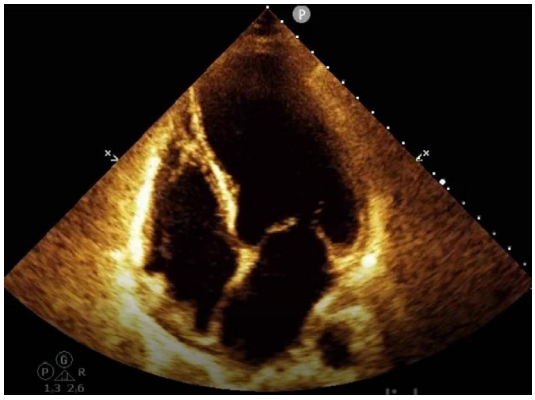
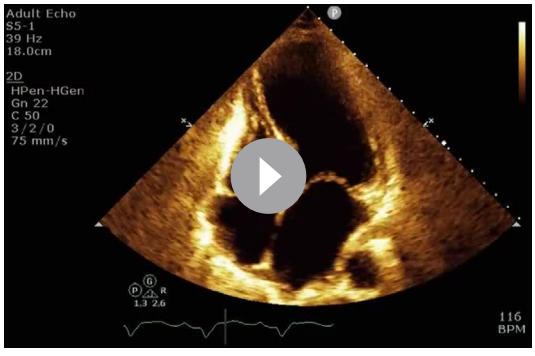


Figure 2 Transthoracic echocardiogram.

CASE PRESENTATION

A 69-year-old woman with a history of chronic kidney disease, anaemia, asthma and Sjögren disease presented to an outside hospital with dyspnoea on exertion, palpitations and intermittent chest pain of several weeks' duration. On presentation, she was afebrile and her initial vital signs showed a blood pressure of 80/52 mm Hg, heart rate of 95 beats per minute, respiratory rate of 18 breaths per minute and room air oxygen saturation of 99%. Her ECG showed normal sinus rhythm with ST elevations in V3–V6, Q waves in leads II, III and aVF, and a right bundle branch block (figure 1). She underwent emergent left heart catheterisation, which revealed only non-obstructive coronary artery disease. During her hospitalisation, transthoracic echocardiogram showed a new decreased left ventricular ejection fraction (EF) of 25% with apical, anterior, anteroseptum and inferoseptum wall akinesis, moderate mitral regurgitation, and a small pericardial effusion (figure 2 and video 1). Of note, she had had a similar presentation 1 month prior to this hospitalisation. Transthoracic echocardiogram



Video 1 Transthoracic echocardiogram.

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Table 1 Laboratory results

Laboratory assay	Patient's results	Reference range
Bicarbonate, mmol/L	16	21–35
Anion gap	14	3–13
Blood urea nitrogen, mg/dL	40	10–25
Creatinine, mg/dL	2.99	<1.16
Glomerular filtration rate, mL/min/1.73 m^2	18	>60
Aspartate aminotransferase, IU/L	27	<35
Alanine transaminase, IU/L	11	<52
High-sensitivity troponin, ng/L	2579	<19
Brain natriuretic peptide, pg/mL	2176	<50
Venous lactate, mmol/L	1.1	<2.1
Sedimentation rate, mm/hour	103	<30
C reactive protein, mg/dL	6.4	<0.5
Rheumatoid factor, U/mL	55	<15
Antinuclear antibody titre	>1:1280 with speckled pattern	<80
Anti-Ro/SS-A antibody, U/mL	443	0–99
Anti-La/SS-B antibody, U/mL	589	0–99
C3 complement, mg/dL	117	90–230
C4 complement, mg/dL	29	10–51
D-dimer, ng/mL	4360	0–499
Cryoglobulin	Not detected	
EBV capsid antibody, IgM	Not detected	
EBV capsid antibody, IgG	Positive	
EBV nuclear antibody	Positive	
Influenza A/B, PCR	Not detected	
Measles antibody, IgG, ELISA units	6.9	<1.0
Mumps antibody, IgG	Positive	
Varicella antibody, IgG, units	0.4	<1.0
Respiratory syncytial virus PCR	Not detected	
Treponemal IgG/IgM	Non-reactive	
Enterovirus (nucleic acid amplification)	Not detected	
EBV capsid antibody, IgM	Negative	
EBV capsid antibody, IgG	Positive	
EBV nuclear antibody	Positive	
Herpes simplex virus 1 and 2 PCR, blood	Not detected	
Toxoplasma IgM antibody, AU/mL	<3.0	<8.0
QuantiFERON-TB Gold	Negative	
CMV antibody, IgM	Negative	
CMV antibody, IgG, ELISA units	3.7	<1.00
HIV fourth-generation antigen/antibody	Non-reactive	
Hepatitis A antibody, total	Negative	
Hepatitis B antigen	Negative	
Hepatitis B core antibody, IgM	Negative	
Hepatitis C antibody	Negative	
2019-nCoV RNA (nasopharyngeal)	Negative	

AU/mL, absorbance units per millilitre; CMV, cytomegalovirus; EBV, Epstein-Barr virus; 2019-nCoV RNA, 2019 novel coronavirus RNA; SS, Sjögren syndrome; TB, tuberculosis.

showed a normal EF (60%) and mild hypokinesis of her apical and anteroseptal walls at that time.

During admission at an outside hospital, she was unable to tolerate guideline-directed medical therapy due to hypotension,

Table 2 Right heart catheterisation results

Right heart catheterisation	Measurement
Right atrium, mm Hg	8
Right ventricle, mm Hg	45/6 (12)
Pulmonary artery main, mm Hg	50/28 (36)
Pulmonary capillary wedge, mm Hg	30
Cardiac output, L/min	3.29
Cardiac index, L/min/m^2	1.83
Pulmonary arterial pulsatility index	2.75

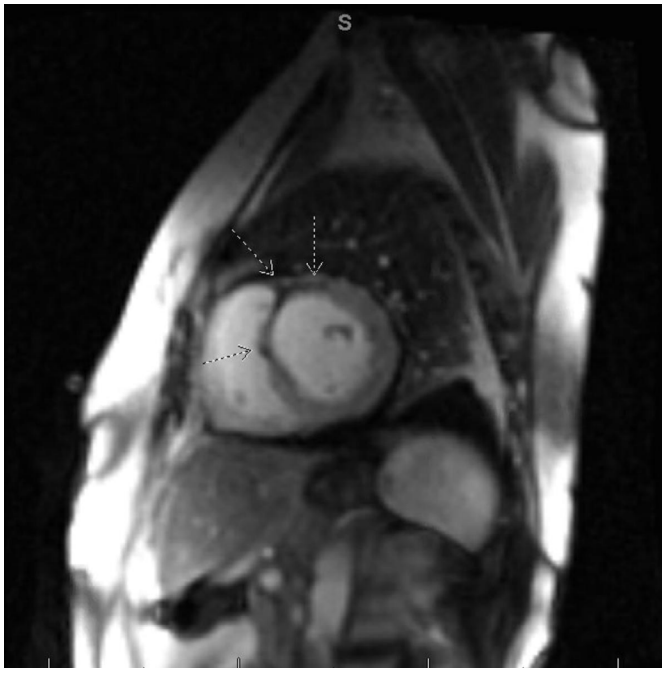
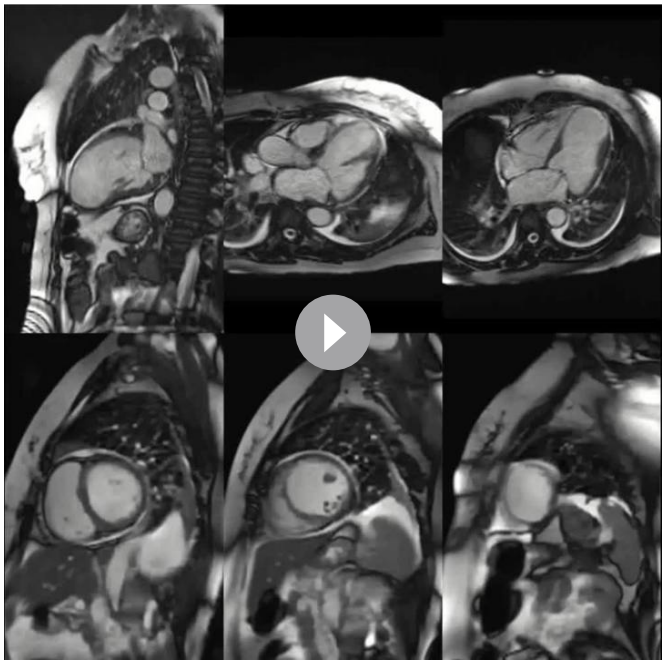


Figure 3 Late gadolinium enhancement cardiac MRI.



Video 2 Cardiac magnetic resonance.

for which she initially received norepinephrine and then dobutamine. She was transferred to our facility for advanced heart failure evaluation where she was found to have acute kidney injury (creatinine: 2.99 mg/dL; baseline 1.94) and elevated cardiac markers (high-sensitivity troponin of 2579 ng/L and B-type natriuretic peptide of 2176 pg/mL). Her venous lactate levels were within reference range, but her erythrocyte sedimentation rate and C reactive protein results were elevated. She also had elevated levels in the following laboratory assays: antinuclear antibody titre, anti-Sjögren syndrome type A antibodies, anti-Sjögren syndrome type B antibodies and rheumatoid factor (table 1). These elevated findings were consistent with her known diagnosis of Sjögren disease. An extensive infectious work-up was unremarkable (table 1). The patient underwent right heart catheterisation, which showed elevated filling pressures and a cardiac output of 3.29 L/min (table 2).

TREATMENT

The patient was started on a furosemide drip; however, her lactate levels increased the next day and her dobutamine was increased to 7.5 μg/kg/min. Cardiac MRI was done, which showed an EF of 20%, severely dilated left ventricle with left ventricular apical ballooning, and late gadolinium enhancement in the mid-myocardium of the mid to apical anterior and anteroseptal segments (figure 3 and video 2). Rheumatology specialists were consulted for concern of autoimmune myocarditis. At their recommendation, she was started on intravenous methylprednisolone sodium succinate 500 mg daily for 5 days followed by prednisone of 1 mg/kg, with a plan of reducing the oral dose by 10 mg every 4 weeks over a period of 6 months. Treatment with azathioprine or mycophenolate mofetil after discharge was to be considered. The patient was not started on any corticosteroid-sparing immunosuppressants during her hospitalisation.

OUTCOME AND FOLLOW-UP

She had worsening kidney function (creatinine 4.62 mg/dL) and she was started on norepinephrine because her systemic vascular resistance was low. An intra-aortic balloon pump was placed for haemodynamic support and a heart and kidney transplantation evaluation was initiated; however, the patient declined transplant evaluation. The intra-aortic balloon pump was removed the next day and her kidney function remained stable. Five days later, the patient had a pulseless electrical activity arrest without return of spontaneous circulation and she died.

DISCUSSION

We report the case of a woman with Sjögren syndrome new-onset cardiomyopathy. The patient had elevated acute inflammatory markers and autoimmune serologies, normal coronary arteries, and cardiac MRI that demonstrated late gadolinium enhancement in the mid-myocardium of the mid to apical anterior and anteroseptal segments. Five previous case reports[5–9] have shown Sjögren-related congestive heart failure diagnosed in patients presenting with new systolic dysfunction found on echocardiogram along with elevated inflammatory markers after other aetiologies, including ischaemic heart disease and viral infections, were ruled out. Golan *et al*[6] made the diagnosis within a setting of leucoplastic vasculitis seen on skin biopsy. Llanos-Chea *et al*[8] reported a similar case of a patient with primary Sjögren syndrome who presented with new-onset systolic heart failure and who showed similar MRI findings to our patient, suggestive of autoimmune myocarditis. Most recently in 2018, Vindhyal *et al*[9] published the case of a patient who had a similar clinical presentation and echocardiographic findings as our patient. They made the diagnosis of Takotsubo cardiomyopathy, and the aetiology was thought to be adrenal crisis or Sjögren syndrome flare, as the patient was on a steroid taper on initial presentation.

Previous case reports have described patients with near to complete normalisation of EF after 1.5–6 months of steroid therapy in conjunction with standard heart failure medical therapy.[5–9] Levin *et al*[5] reported therapy that included intravenous cyclophosphamide with steroids, and the patient reached normalisation of EF after 5 months of treatment. Our patient was started on high-dose intravenous steroids followed by oral prednisone with minimal improvement of her heart function. Heart transplant work-up was offered, but the patient declined. Unfortunately, she died 2 weeks after the diagnosis was made. The patient's family declined having an autopsy performed and a cardiac biopsy was not done.

Given the rarity of Sjögren syndrome, no diagnostic criteria or standard treatment has been established for cardiomyopathy related to this disease. Sjögren disease association should be considered in patients who show evidence of autoimmune processes after other possible causes are ruled out. Apical septal late gadolinium enhancement should also raise concern for autoimmune aetiology in clinical settings as reported previously.[10 11] A number of published case reports have shown that initiation of high-dose intravenous steroids followed by oral steroids for 3–6 months can successfully improve systolic EF. Conventional heart failure and advanced treatments, such as heart transplant or left ventricular assist device implantation, can also be considered in the appropriate setting. We report this case to raise awareness of cardiomyopathy as a potential complication of Sjögren syndrome that may be secondary to myocarditis as suggested by cardiac MRI.

Learning points

- Recognize autoimmune processes, including Sjögren disease, as a potential etiology for non-ischemic cardiomyopathy.
- Describe the role of late gadolinium enhancement as seen on cardiac magnetic resonance imaging when investigating etiology of cardiomyopathy
- Acknowledge the importance of a multidisplinary team approach in the diagnosis and management of autoimmune-related cardiac disease.

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