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Case Report

Internal Medicine Section

Pulmonary Hypertension and Arrhythmogenic Right Ventricular Cardiomyopathy in Atypical Sjögren's Syndrome: A Rare Case Report

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ABSTRACT

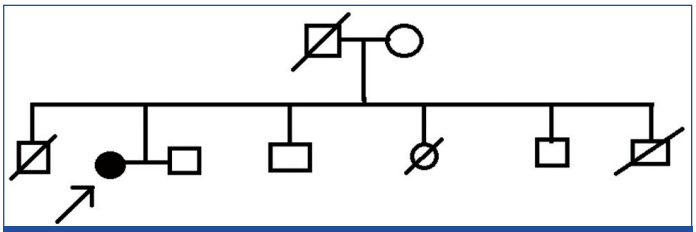
Sjögren's syndrome is a chronic autoimmune condition in which the majority of patients present with sicca symptoms. About 20% of patients, however, do not have sicca symptoms and instead present predominantly with systemic or extraglandular involvement, posing a diagnostic challenge. Pulmonary Hypertension (PH) manifests as elevated pulmonary arterial pressure due to remodelling of the pulmonary vasculature as a result of various factors, including systemic involvement, infectious agents, connective tissue diseases, or toxin/drug exposure. PH in Sjögren's syndrome is a dangerous complication that can significantly affect patient outcomes. Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a genetic cardiomyopathy characterised by fibrofatty replacement of the right ventricular myocardium, predisposing patients to arrhythmias which may cause abrupt collapse or sudden death. Sjögren's syndrome and ARVC are two distinct disorders that may lead to grave outcomes when they co-exist. Present case is a rare case of atypical Sjögren's syndrome with PH and ARVC in a patient who suffered a cardiac arrest during her initial evaluation. Further work-up revealed ARVC, following which appropriate treatment was initiated. This case underlines the need for clinicians to consider Sjögren's syndrome even in the absence of sicca symptoms, especially when unexplained systemic or cardiovascular findings are present. The co-existence of ARVC added further diagnostic complexity. Overlapping pathophysiology—right ventricular strain from PH combined with intrinsic ARVC-related myocardial damage—may accelerate clinical deterioration.

Keywords: Atypical presentation, Connective tissue disorder, Hypothyroidism, Right heart failure

CASE REPORT

A 55-year-old female presented with complaints of breathlessness associated with orthopnoea, abdominal distension, and constipation for seven days, along with reduced urine output for three days. She also reported intermittent palpitations for the past three days, cough with minimal expectoration, and abdominal distension and constipation that gradually progressed to orthopnoea. There was no history of chest pain or syncope.

She was a known case of hypothyroidism for the past 10 years and was on regular Tablet Thyroxine 75 µg. She had no other co-morbidities. A significant family history of sudden death among siblings was noted, although details were unavailable. She was the second child of a second-degree non consanguineous marriage, with five siblings, of whom two (excluding her) were alive [Table/Fig-1].



[Table/Fig-1]: Patient is second born which is shaded and 1st, 4th and 6th siblings died which has been crossed.

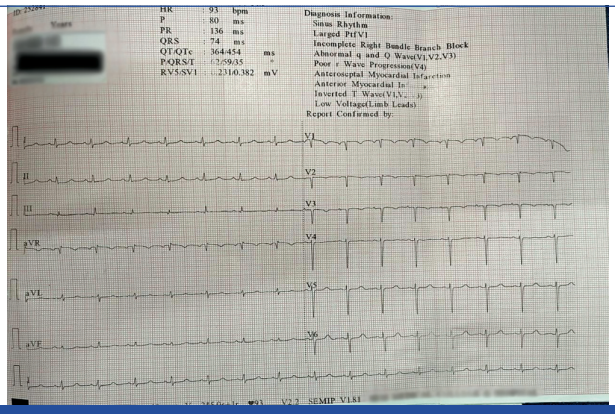
On examination, her vitals were: blood pressure 170/90 mmHg, pulse rate 98 beats per minute (regular), oxygen saturation 92% on room air, and respiratory rate 24 cycles/minute. General examination was unremarkable except for bilateral pitting pedal oedema and elevated Jugular Venous Pressure (JVP). Cardiovascular examination revealed muffled heart sounds and a loud P2. Respiratory examination revealed inspiratory crackles over the bilateral infrascapular areas. Abdominal examination showed distension but no organomegaly. Neurological examination was normal.

Initial blood investigations are summarised in [Table/Fig-2]. An electrocardiogram showed T-wave inversion in V1, V2, and V3 as illustrated in [Table/Fig-3].

Investigations	Interpretation	Normal range
Hb (g/dL)	10.2	12-15
Total WBC (cells/cu.mm)	7400	4000-11000
Platelet (cells/cu.mm)	1,22,400	1,50,000-4,50,000
Urea (mg/dL)	31	17-43
Creatinine (mg/dL)	0.8	0.6-1.2
Sodium (mmol/L)	134	136-145
Potassium (mmol/L)	4.4	3.5-5.1
Chloride (mmol/L)	105	98-107
Bicarbonate (mmol/L)	24	21-31
Calcium (mg/dL)	8.6	8.8-10.6
Phosphorus (mg/dL)	4.7	2.5-4.5
Magnesium (mg/dL)	1.8	1.9-2.5
Total bilirubin (mg/dL)	1.83	0.5-1.0
Indirect bilirubin (mg/dL)/Direct bilirubin (mg/dL)	1.25/0.58	0.2-0.8/0.0-0.3
SGOT (IU/L)/SGPT (IU/L)	50/33	<31/<34
ALP (IU/L)	98	30-120
GGT (U/L)	58	<38

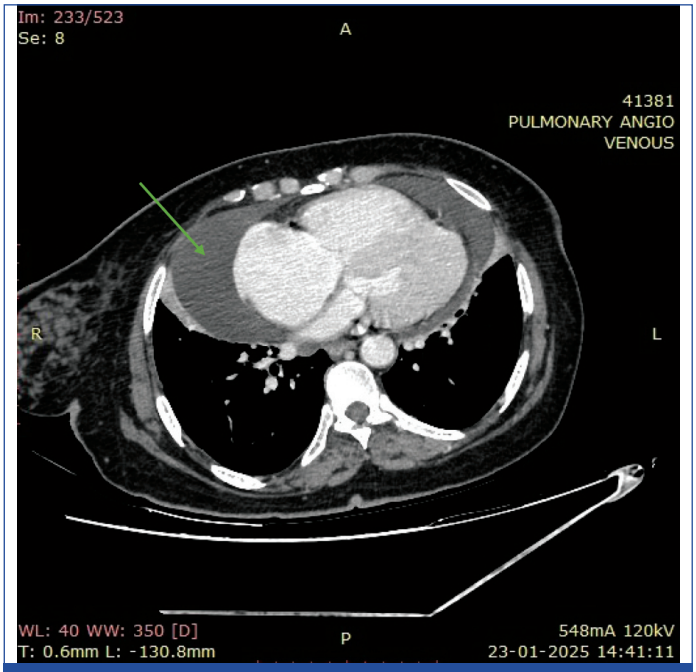
Total protein (g/dL)	6.4	6.6-8.3
Albumin (g/dL)	3.5	3.5-5.2
Globulin (g/dL)	2.9	2.5-3.0

[Table/Fig-2]: Blood investigation on initial presentation.
SGOT: Serum glutamic-oxaloacetic transaminase; SGPT: Serum glutamic-pyruvic transaminase; ALT: Alanine transaminase; GGT: Gamma-glutamyl transaminase; Hb: Haemoglobin; WBC: White blood cells



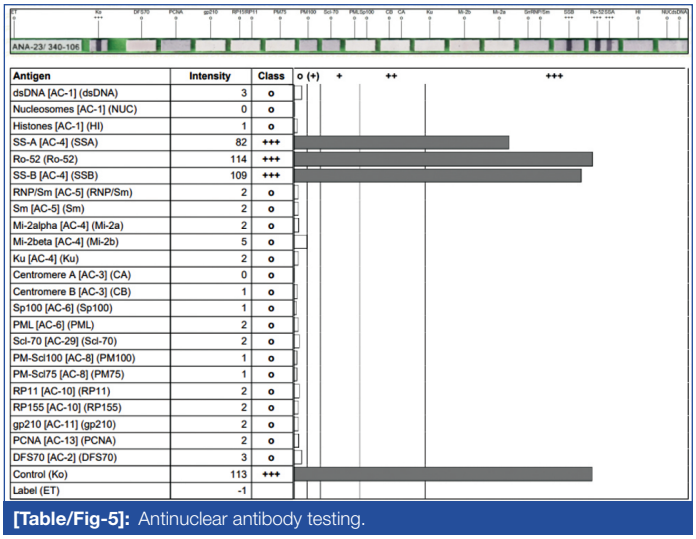
[Table/Fig-3]: Electrocardiogram revealed T wave inversion over right precordial leads V1, V2 and V3.

The patient was given intravenous Furosemide 40 mg for symptomatic relief and was subsequently subjected to echocardiography, which revealed severe PH (68 mmHg), a TAPSE of 20 mm, normal left ventricular systolic function, and an ejection fraction of 56%, along with a moderate pericardial effusion. A comprehensive medical history excluded alternative causes of PH. The scheduled pulmonary angiographic study was interrupted by an unexpected cardiac arrest. The patient was revived with cardiopulmonary resuscitation and transferred to the critical care unit. After stabilisation, she was shifted again for the pulmonary angiographic study, which revealed cardiomegaly and moderate to severe pericardial effusion as shown in [Table/Fig-4].



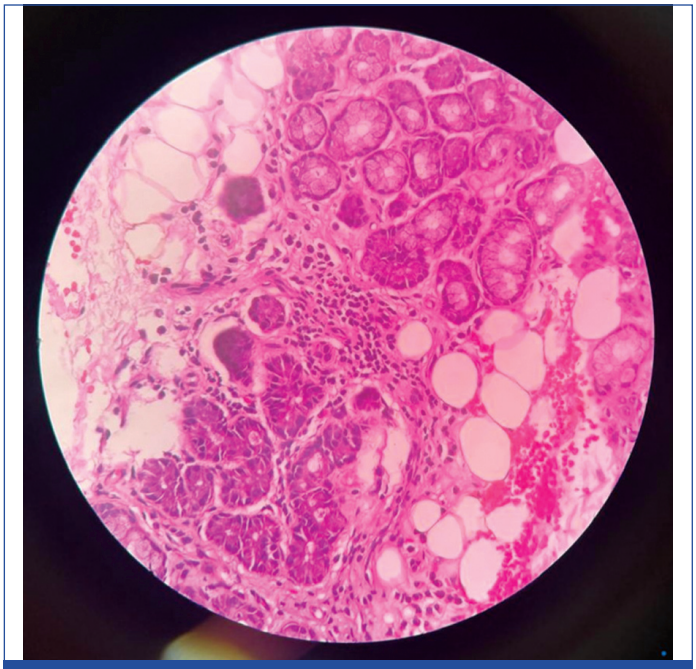
[Table/Fig-4]: CT pulmonary angiography revealed pericardial effusion (green arrow).

As no definitive cause for PH could be identified, an antinuclear antibody test was performed, which was positive with a speckled pattern (3+). To assess specific antibody levels, an Antinuclear Antibody (ANA) panel was done and revealed positivity for SS-A, Ro-52, and SS-B antibodies, as shown in [Table/Fig-5]. A lip biopsy was performed as part of the diagnostic criteria, which demonstrated focal lymphocytic sialadenitis with a FOCUS score [1] of three, as shown in [Table/Fig-6]. Schirmer's test was negative.



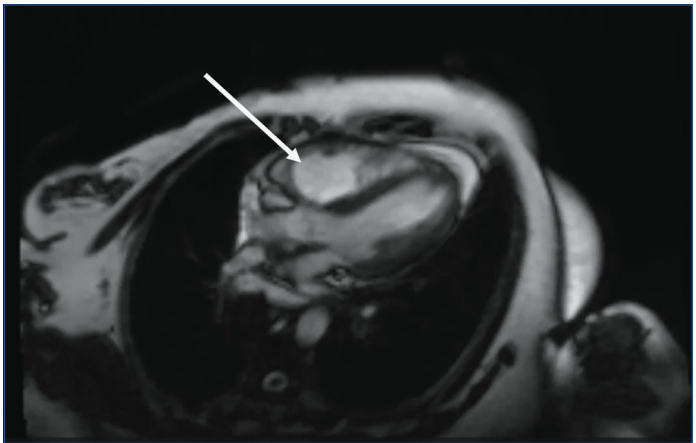
Antigen	Intensity	Class
dsDNA [AC-1] (dsDNA)	3	o
Nucleosomes [AC-1] (NUC)	0	o
Histones [AC-1] (HI)	1	o
SS-A [AC-4] (SSA)	82	+++
Ro-52 (Ro-52)	114	+++
SS-B [AC-4] (SSB)	109	+++
RNP/Sm [AC-5] (RNP/Sm)	2	o
Sm [AC-5] (Sm)	2	o
Mi-2alpha [AC-4] (Mi-2a)	2	o
Mi-2beta [AC-4] (Mi-2b)	5	o
Ku [AC-4] (Ku)	2	o
Centromere A [AC-3] (CA)	0	o
Centromere B [AC-3] (CB)	1	o
Sp100 [AC-6] (Sp100)	1	o
PML [AC-6] (PML)	2	o
Scl-70 [AC-29] (Scl-70)	2	o
PM-Scl100 [AC-8] (PM100)	1	o
PM-Scl75 [AC-8] (PM75)	1	o
RP11 [AC-10] (RP11)	2	o
RP155 [AC-10] (RP155)	2	o
gp210 [AC-11] (gp210)	2	o
PCNA [AC-13] (PCNA)	2	o
DFS70 [AC-2] (DFS70)	3	o
Control (Ko)	113	+++
Label (ET)	-1	

[Table/Fig-5]: Antinuclear antibody testing.



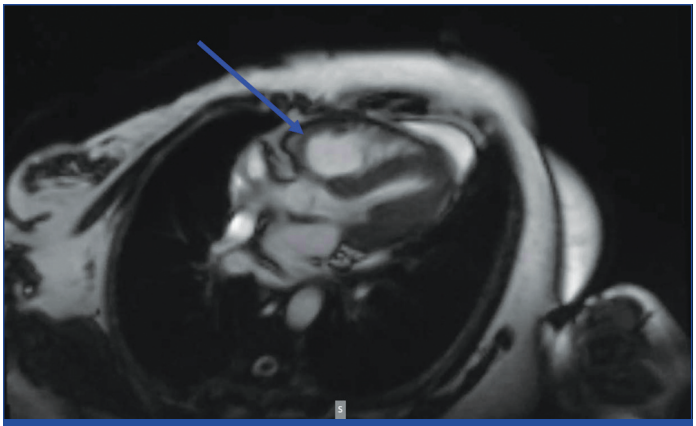
[Table/Fig-6]: Lip biopsy showing focal lymphocytic sialadenitis (H&E,40x).

Cardiac magnetic resonance imaging revealed right ventricular flow hypokinesia, a right ventricular ejection fraction of 9.7%, a right ventricular end-diastolic volume of 124 mL, and mild pericardial effusion, with features suggestive of ARVC, as shown in [Table/Fig-7,8].



[Table/Fig-7]: Cardiac MRI showing right ventricle during diastole (white arrow).

Based on all available evidence, a diagnosis of Sjögren's syndrome with an atypical presentation was confirmed. A rheumatology opinion was obtained, and the patient was started on Tablet Prednisolone 60 mg, which was tapered and stopped, along with



[Table/Fig-8]: Cardiac MRI showing right ventricle hypokinesia during systole (blue arrow).

Tablet Hydroxychloroquine 200 mg, Tablet Sildenafil 20 mg, and Tablet Metoprolol 25 mg twice daily. The patient is currently on regular follow-up, and her symptoms have decreased significantly.

DISCUSSION

The World Symposium on Pulmonary Hypertension classifies PH into five major groups, of which index patient fits into Group 1: Pulmonary Arterial Hypertension (PAH). PAH associated with connective tissue diseases accounts for 25% of all conditions in Group 1 PH [2]. The prevalence of PH in Sjögren's syndrome patients is approximately 1.6%, with male patients showing a greater predisposition. Evidence of altered respiratory function and periodic echocardiography to detect early diastolic dysfunction is recommended for male SS patients [3]. Clinically significant primary Sjögren's syndrome (pSS) lung disease has a reported prevalence of 9–20%, which increases to 43-75% on comprehensive evaluation, suggesting frequent subclinical presentation [4]. The prevalence of PH in connective tissue disorders has been reported as 6–28.3% in Asians with systemic sclerosis, 35.3-49% in Asians with systemic lupus erythematosus, 1-16% in pSS, 5.9-8% in mixed connective tissue disease, and 0.9-4.4% in dermatomyositis–polymyositis [5]. Kobak S et al., reported a prevalence of PH of 23.4% in pSS patients [6]. Although the mechanism of PH in pSS is not fully understood, several proposed aetiologies in primary Sjögren's syndrome are attributed to immune-mediated endothelial injury involving immune complex deposition, necrotising vasculitis, and altered endothelial vasoactive pathways. Sustained vasoconstriction triggers vascular remodelling, characterised by medial hypertrophy, intimal proliferation, thrombotic obstruction, and plexiform lesions, ultimately leading to progressive arterial obliteration and elevated pulmonary arterial pressures [6].

Dryness or sicca symptoms involving the eyes and oral mucosa occur due to lymphocytic infiltration of the exocrine glands, which is a hallmark feature and is reported in over 90% of pSS cases, with a female preponderance. Non dryness (atypical) pSS patients constitute a clinical subset who share the same immunopathological mechanisms as classic sicca pSS patients. Atypical pSS is rare and characterised by younger age, extraglandular involvement, parotid enlargement, and positive anti-Ro/SSA antibodies [7]. However, pSS can involve any organ system, and patients with pSS and interstitial lung disease have a higher risk of cardiovascular co-morbidities and progressive lung function decline. Echocardiography can predict early cardiac deterioration through periodic monitoring of parameters such as TAPSE and sPAP, allowing for timely and structured management in these patients [8].

The FOCUS score is a histopathological scoring system used to quantify lymphocytic infiltration in minor salivary glands. The inflammatory infiltrate is assessed, and an aggregation of ≥50 lymphocytes, plasma cells, or histiocytes within 4 mm^2 is defined as a "focus." A score of ≥1 is suggestive of pSS and is considered highly specific for Sjögren's syndrome [9]. ARVC is a genetic cardiomyopathy characterised by structural alterations of the right ventricular myocardium. It is defined by right ventricular dilation due to fibrofatty infiltration and manifests clinically as cardiac arrhythmias ranging from non sustained ventricular tachycardia to sudden cardiac death [10]. ARVC has an estimated prevalence of 1:2000 to 1:5000 [11]. There is an asymptomatic phase commonly seen in younger individuals or in family members of affected patients. Close monitoring of these individuals is warranted, as the disease may become symptomatic over time.

Follow-up annually or semiannually is recommended for patients diagnosed with ARVC. Screening modalities include annual ECG monitoring, echocardiography every 1-3 years, and cardiac magnetic resonance imaging every 3-5 years [12]. Current pharmacological management of ARVC focuses on controlling arrhythmias and managing heart failure. Beta blockers such as sotalol or metoprolol are commonly used, along with guideline-directed heart failure therapy when required [13]. Asian PAH patients have demonstrated significant improvement with Sildenafil at a dose of 20 mg thrice daily, as measured through exercise capacity, cardiopulmonary function, and haemodynamic indices [14]. Hydroxychloroquine, although older than more modern immunomodulatory drugs, is still widely used due to its cost-effectiveness and has shown moderate efficacy in improving unstimulated salivary flow rate and other inflammatory markers [15]. Glucocorticoids are preferred in typical SS with glandular, joint, cutaneous, haematological, renal, or neurological involvement as induction therapy, and can be replaced with immunomodulators if Cushingoid features develop [16].

CONCLUSION(S)

Although PH is not common in Sjögren's syndrome, it is a significant complication that can adversely affect patient prognosis and quality of life. ARVC, a severe co-morbidity, can worsen right heart failure and reduce survival; therefore, early detection and timely treatment are crucial. Research on targeted therapies and improved diagnostic strategies can help enhance outcomes in such cases. This case highlights an atypical presentation of Sjögren's syndrome with overlapping arrhythmogenic pathology, managed effectively with appropriate therapy.

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