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A DIAGNOSTIC DILEMMA: ATYPICAL MULTISYSTEM PRESENTATION OF SJÖGREN'S SYNDROME WITH USUAL INTERSTITIAL PNEUMONIA AND A LARGE SACCULAR AORTIC ANEURYSM

Medicine

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ABSTRACT

We report a diagnostically challenging case of a 86 year old woman with established Sjögren's syndrome who presented with respiratory failure, constitutional symptoms, and profound weight loss. High-resolution imaging revealed a UIP-pattern interstitial lung disease and a large, partially thrombosed saccular aortic aneurysm. This unique systemic manifestation complicated clinical decision-making. Endovascular intervention was considered but deferred due to the patient's age and functional decline. The case highlights the need to remain alert to multisystem autoimmune complications and the diagnostic dilemma posed by overlapping vascular and pulmonary pathology.

KEYWORDS

Sjögren's Syndrome, Usual Interstitial Pneumonia (UIP), Aortic Aneurysm, Autoimmune Vasculopathy, Connective Tissue Disease

INTRODUCTION:

Sjögren's syndrome is an autoimmune disease commonly affecting exocrine glands, but systemic manifestations including interstitial lung disease (ILD) are increasingly recognized.

UIP pattern ILD has been associated with poor prognosis in connective tissue disease.

Large-vessel vasculopathy is rare in Sjögren's, and the coexistence of ILD and an aortic aneurysm in a single patient poses a complex diagnostic and therapeutic challenge.

Case Study:
86-year-old woman presented with:
- Progressive dyspnea (3 days)
- Dry cough (1 week)
- Watery stools
- Severe anorexia and weight loss (40 kg over 2 months)

Medical History:
- Sjögren's syndrome (anti-SSA/Ro52 positive)
- ILD (UIP pattern)
- Atrial fibrillation (on amiodarone)
- Diabetes mellitus, hypertension

On Examination:
- HR 116 (irregular), RR 26, SpO_2 88%, BP 132/84 mmHg
- Bibasilar crackles, cachexia, no edema or lymphadenopathy

Investigations:
Laboratory Results:

Test	Value
CRP	26 mg/L
Albumin	2.74 g/dL
Creatinine	0.77 mg/dL
ABG	pH 7.28, pCOâ,, 60.6, pOâ,, 25.4
HCOâ,*f*â »	22.1 mmol/L
Lactate	1.7 mmol/L
CBC	13.8|6.33|170
Uric Acid	3.1
Relevant Liver Profile	SGOT 27.8 SGPT 9.1 T Bilirubin 0.43 D Bilirubin 0.12

HRCT Chest: Basal-predominant subpleural reticulations, honeycombing, traction bronchiectasis — consistent with UIP pattern ILD.

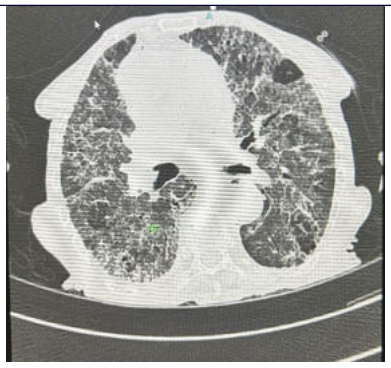
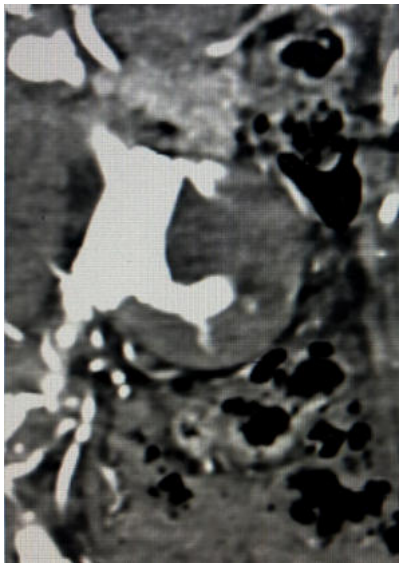


Figure 1: HRCT Chest – axial view showing basal honeycombing with traction bronchiectasis

CT Abdomen (Arterial & Delayed):
- Large saccular aortic aneurysm (5.5 × 4.4 cm), partially thrombose
- Mild delayed contrast enhancement, no rupture or perianeurysmal fat stranding
- Bilateral renal cortical/parapelvic cysts



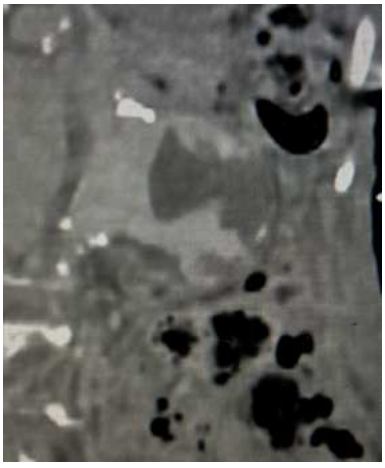


Figure 2 & 3: CT Abdomen – saccular aneurysm with partial thrombosis and delayed phase showing partial contrast filling

Diagnostic Dilemma

The simultaneous presentation of ILD and aortic aneurysm in Sjögren's syndrome is unusual. The aneurysm may represent:

- A rare autoimmune vasculopathy (large-vessel involvement)
- A degenerative atherosclerotic process

Due to overlapping features and absence of rupture or systemic vasculitis, definitive etiology could not be determined without surgical intervention, which was deemed high risk.

Intervention And Outcome:

- Evaluated by interventional radiology for possible EVAR
- Stenting deferred due to age, frailty, and pulmonary compromise
- Managed conservatively with oxygen therapy and corticosteroids
- Discharged with supportive care and outpatient follow-up

DISCUSSION:

Sjögren's syndrome is traditionally characterized by immune-mediated destruction of the exocrine glands, but it can manifest as a multisystem disease involving the lungs, kidneys, nervous system, and, in rare cases, blood vessels. Approximately 9–24% of patients develop interstitial lung disease (ILD), most commonly with a nonspecific interstitial pneumonia (NSIP) pattern. However, UIP-pattern ILD, as observed in this case, is increasingly recognized in primary Sjögren's syndrome and is associated with a worse prognosis, greater functional decline, and less responsiveness to immunosuppressive therapy compared to NSIP [1,2].

The diagnostic dilemma in this case stems from the coexistence of two significant systemic pathologies:

- A UIP-pattern ILD, confirmed on HRCT.
- A partially thrombosed, saccular abdominal aortic aneurysm, discovered incidentally on imaging.

While ILD is a well-established extraglandular manifestation of Sjögren's syndrome, large-vessel involvement is exceptionally rare. Most vasculitic manifestations of Sjögren's involve small to medium-sized vessels, often presenting as palpable purpura or mononeuritis multiplex [3]. The involvement of large vessels, such as the aorta, is sparsely documented in literature, mostly in case reports [4].

The etiology of the aneurysm in this case was unclear — raising the possibility of:

- A degenerative atherosclerotic aneurysm, more common in elderly males with vascular risk factors (e.g., diabetes, hypertension, age).
- Or, a rare autoimmune vasculopathy resulting from chronic inflammation, endothelial dysfunction, and immune complex deposition.

Autoimmune diseases like Takayasu arteritis, Behçet's disease, and systemic lupus erythematosus (SLE) are better known for large-vessel involvement [5]. In contrast, aortic aneurysm formation in Sjögren's remains largely anecdotal. In one case, Sjögren's was associated with an aortic dissection, raising the possibility that immune-mediated vascular fragility, perhaps exacerbated by steroid exposure and systemic inflammation, may contribute to aneurysm formation or progression [6].

Furthermore, this case demonstrates how systemic autoimmune diseases may evolve atypically in elderly patients, where the clinical picture is often complicated by:

- Comorbid atherosclerosis
- Degenerative organ decline
- Reduced procedural fitness, limiting invasive interventions

In this patient, the decision to forgo aneurysm repair was based on a careful balance between procedural risk and potential benefit. The patient had significant ILD-related respiratory compromise and frailty, factors known to predict poor outcomes in endovascular aneurysm repair (EVAR) [7].

This case underscores three critical clinical messages:

1. UIP-pattern ILD can occur in Sjögren's syndrome and should prompt aggressive pulmonary monitoring.
2. Large-vessel aneurysms, although rare, should be considered in Sjögren's patients presenting with systemic symptoms and weight loss.
3. Multidisciplinary decision-making, including interventional radiology consultation, is essential in balancing treatment options for medically fragile patients.

Ultimately, this rare presentation highlights the need for heightened clinical suspicion and individualized risk stratification in patients with connective tissue diseases exhibiting systemic deterioration.

CONCLUSION:

This case presents a rare overlap of Sjögren's-associated ILD and a large aortic aneurysm. The diagnostic uncertainty regarding vascular involvement underscores the need for high suspicion and multidisciplinary evaluation—even when intervention is deferred.

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