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(CASE REPORT)

Chronic hemoptysis leading to the diagnosis of inflammatory bronchiolitis in primary sjögren's syndrome: A case report

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Abstract

Introduction: Sjögren's syndrome (SS) is a chronic autoimmune inflammatory disease primarily affecting the exocrine glands. While airway involvement and interstitial lung disease are the most common respiratory manifestations, cellular bronchiolitis remains a rare complication of SS. We present a case of Sjögren's syndrome associated with cellular bronchiolitis, revealed by chronic hemoptysis.

Observation: Case Report. A 57-year-old patient was admitted for evaluation of cellular bronchiolitis, presenting with a chronic dry cough complicated by mild hemoptysis and progressively worsening dyspnea. The diagnosis of Sjögren's syndrome was confirmed based on objective and subjective xerostomia, xerophthalmia with an abnormal Schirmer test, positive antinuclear antibodies (1:320), and strongly positive anti-SSa antibodies. Chest Computed Tomography (CT) imaging revealed findings consistent with cellular bronchiolitis. Pulmonary function tests (PFTs) demonstrated a moderate obstructive syndrome, with a forced vital capacity of 71%. The patient was treated with inhaled and systemic corticosteroids (prednisone 1 mg/kg/day), resulting in rapid clinical improvement.

Conclusion: Bronchiolar involvement in Sjögren's syndrome is often underdiagnosed and is typically associated with follicular bronchiolitis. Its prevalence varies widely. Dry cough and dyspnea are the primary symptoms of pulmonary involvement, while hemoptysis, though rare in this context, necessitates the exclusion of other differential diagnoses. High-resolution chest CT is essential for identifying distal airway involvement. Sjögren's syndrome should be considered in all patients presenting with chronic bronchiolar diseases.

Keywords: Bronchiolitis; Sjögren's syndrome; Hemoptysis; Distal airways

1. Introduction

Sjögren's syndrome (SS) is an autoimmune disease that primarily affects the exocrine glands. However, it can also affect extra-glandular organs, especially the lungs. Histopathologically, the disease is characterized by benign lymphocytic infiltration. Pulmonary involvement is described in more than 43% of cases, typically presenting as diffuse infiltrating pneumonias or bronchiectasis. However, few studies focus on the clinical profile of Sjögren's syndrome (SS) associated with bronchiolitis. We report within the present article the case of a female patient with cellular bronchiolitis secondary to primary Gougerot-Sjögren syndrome revealed by chronic hemoptysis.

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2. Observation

A 57-year-old female patient, followed for arterial hypertension for two years, with a history of acute rheumatic fever complicated by nephritic syndrome, chronic renal failure stage IIIa, and an appendectomy at the age of 18, consulted for progressive exertional dyspnea. There was no history of occupational or environmental exposure.

The disease began one year prior with progressively worsening dyspnea associated with a dry cough, occurring in the context of apyrexia and asthenia. A morning intensification of the cough, accompanied by the onset of low-abundance hemoptysis, was noted. Extra-pulmonary signs included an incapacitating oral and ocular sicca syndrome, inflammatory arthralgia of the elbows and wrists, and diffuse myalgias reported during the anamnesis.

Clinical examination revealed hypertension (18/10 mmHg), tachypnea (26 cycles/min), and normal oxygen saturation at 97%. The pleuropulmonary examination revealed bilateral crepitant rales at the lung bases. The study of lacrimal and salivary secretions highlighted an abnormal SCHIRMER test and reduced saliva flow. The osteoarticular examination revealed pain during both passive and active mobilization of the elbows. The oral cavity showed dry mucosa and poor oral-dental health.

On the biological level, findings included an inflammatory syndrome (Erythrocyte Sedimentation Rate: 70 mm, CRP: 14 mg/L), hyper-alpha-2-globulinemia (9.3 g/L), and moderate anemia (hemoglobin: 11.2 g/dL). Renal function was stable, with creatinine at 18 mg/L and 24-hour proteinuria at 180 mg/24h. The immunological assessment revealed positive antinuclear antibodies at 1/320 (speckled pattern) and strongly positive anti-SSA (60 kDa).

Thoracic computed tomography angiogram revealed multiple foci of bronchocentric micronodules, predominantly affecting the right lung field and the left lower lobe, along with a focus of atelectasis in the right middle lobe. These findings were suggestive of cellular bronchiolitis (***Figure*** 1).

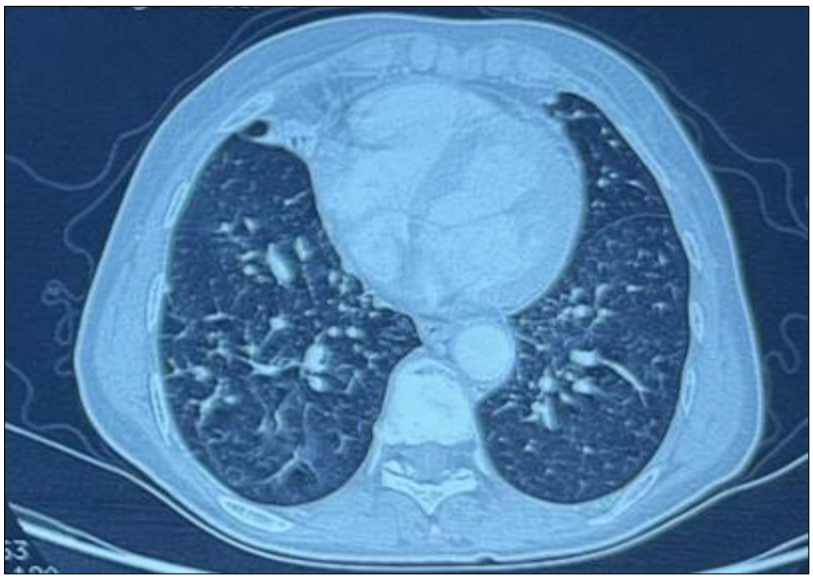
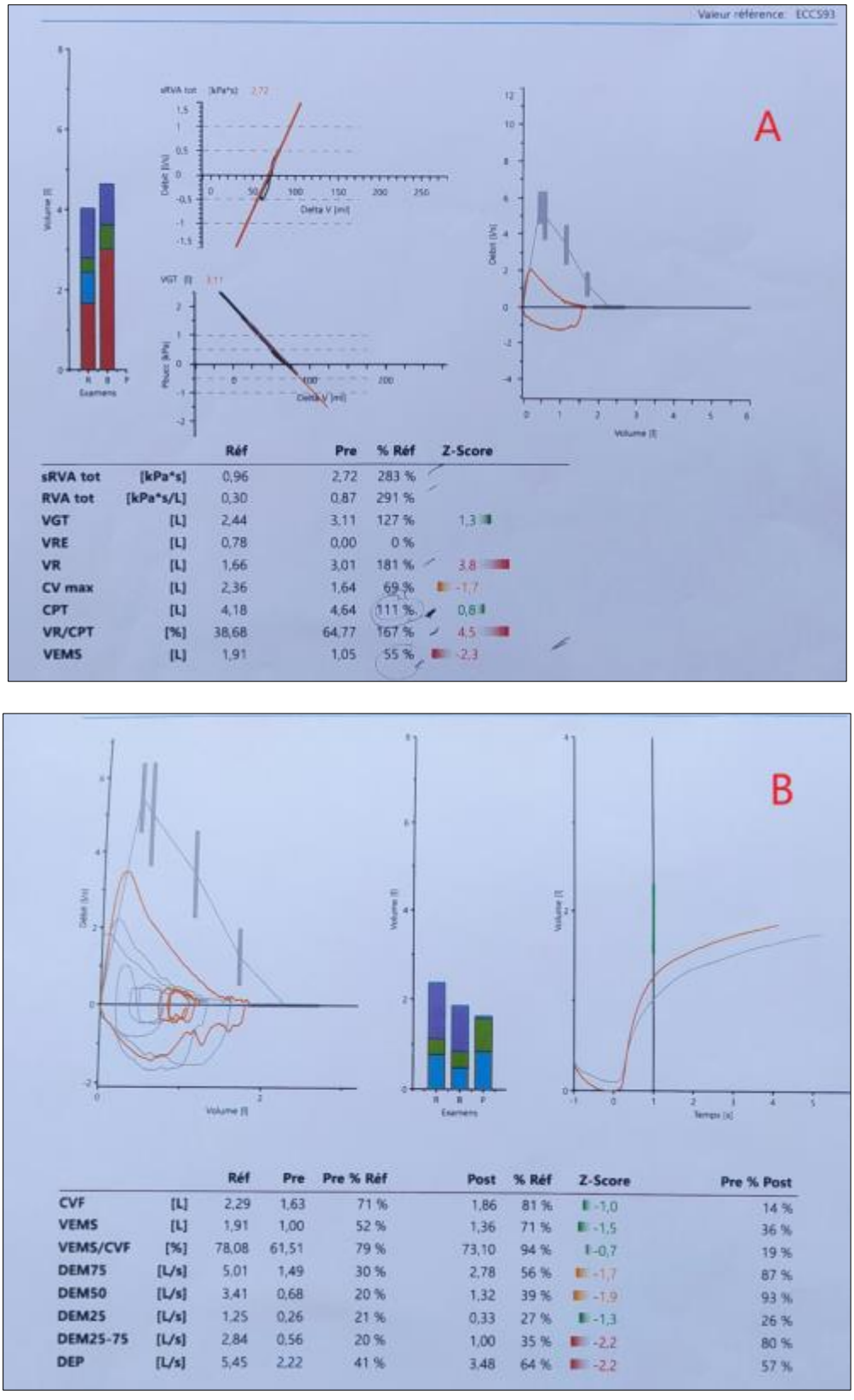


Figure 1 A radiological image demonstrating centrilobular micronodules (tree-in-bud pattern) visualized on a thoracic CT scan using a parenchymal window

Pulmonary function tests (PFTs) demonstrated a moderate reversible obstructive ventilatory disorder post-bronchodilator administration, with an FEV1 of 52%, FEF25-75 of 20%, and a residual volume (RV) of 181% (*Figure* 2 A,B).



		Réf	Pre	% Réf	Z-Score
sRVA tot	[kPa*s]	0,96	2,72	283 %	
RVA tot	[kPa*s/L]	0,30	0,87	291 %	
VGT	[L]	2,44	3,11	127 %	1,3
VRE	[L]	0,78	0,00	0 %	
VR	[L]	1,66	3,01	181 %	3,8
CV max	[L]	2,36	1,64	69 %	-1,7
CPT	[L]	4,18	4,64	111 %	0,8
VR/CPT	[%]	38,68	64,77	167 %	4,5
VEMS	[L]	1,91	1,05	55 %	-2,3

		Réf	Pre	Pre % Réf	Post	% Réf	Z-Score	Pre % Post
CVF	[L]	2,29	1,63	71 %	1,86	81 %	-1,0	14 %
VEMS	[L]	1,91	1,00	52 %	1,36	71 %	-1,5	36 %
VEMS/CVF	[%]	78,08	61,51	79 %	73,10	94 %	-0,7	19 %
DEM75	[L/s]	5,01	1,49	30 %	2,78	56 %	-1,7	87 %
DEM50	[L/s]	3,41	0,68	20 %	1,32	39 %	-1,9	93 %
DEM25	[L/s]	1,25	0,26	21 %	0,33	27 %	-1,3	26 %
DEM25-75	[L/s]	2,84	0,56	20 %	1,00	35 %	-2,2	80 %
DEP	[L/s]	5,45	2,22	41 %	3,48	64 %	-2,2	57 %

Figure 2 Respiratory function tests (RFT) reveal a reversible obstructive ventilatory disorder. [A] Elevated residual volume (RV) with a Z-score of +3.8. [B] Baseline flow-volume curve demonstrating a significant reduction in FEF25-75, FEF50, and PEF, which are reversible with salbutamol

- PEF: peak expiratory flow (= DEP: débit expiratoire de pointe)
- FEFs: the forced expiratory flows (= DEM: *débit* expiratoire moyen)
- FEV1 : Forced expiratory volume in 1 second (= VEMS : Volume expiratoire maximal seconde)

Other Complementary Investigations revealed the following: Bronchoscopy findings were unremarkable. Histopathological examination of the bronchial biopsy showed lymphocytic infiltration, with no evidence of granulomas or malignancy. Bronchoalveolar lavage analysis indicated lymphocytic alveolitis, with negative results for acid-fast bacilli. Additional tests, including eosinophil count, ANCA, rheumatoid factor, anti-CCP antibodies, and Aspergillus serology, were all within normal limits.

The final diagnosis was cellular bronchiolitis secondary to primary Gougerot-Sjögren syndrome, based on the integration of clinical, immunological, and radiological findings. The patient was initiated on a treatment regimen combining oral corticosteroids at a dose of 1 mg/kg/day and inhaled corticosteroids. This led to rapid clinical improvement, with the disappearance of hemoptysis and a reduction in dyspnea. However, follow-up imaging showed that the radiological lesions remained stable (***Figure*** 3).

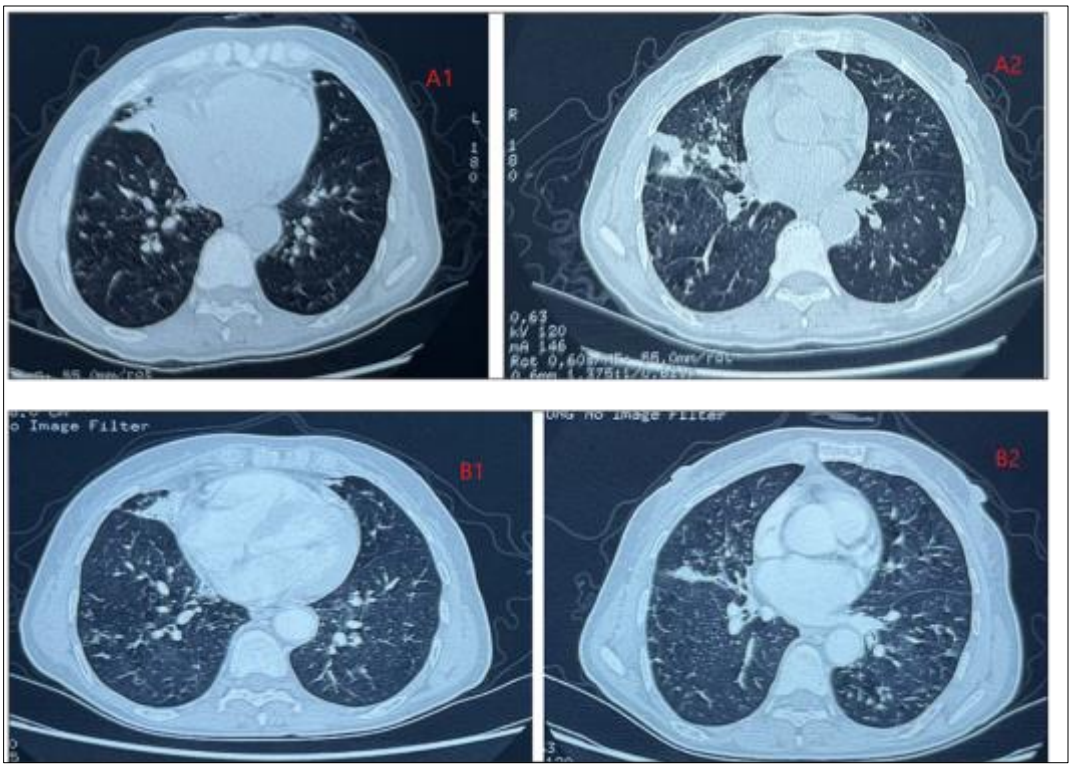


Figure 3 Images (A1 and A2) correspond to the initial investigations. A CT scan (B1 and B2) performed one year later revealed that the radiological lesions of bronchiolitis remained stable following therapeutic treatment

3. Discussion

Sjögren's syndrome is a connective tissue disease characterized by lymphocytic infiltration of the exocrine glands. Although the salivary and lacrimal glands are most frequently involved, respiratory manifestations are common, affecting between 43% and 75% of patients, potentially involving the entire respiratory system. [1]

Interstitial pneumonia was previously considered the most common respiratory manifestation, but bronchiolitis prevalence is not insignificant, and may affect up to 25% of Sjögren's cases. [2,3]

The coexistence of parenchymal and bronchial involvement is frequently observed in Sjögren's syndrome. In a series of 33 patients with Sjögren's syndrome and biopsy-confirmed lung disease, four (4) cases presented with bronchiolitis associated with diffuse interstitial pneumonia [4]. Bronchiolar involvement in Sjögren's syndrome is primarily due to cellular bronchiolitis. Unlike tracheobronchial involvement, bronchiolitis is not caused by sicca syndrome or defective

mucociliary clearance but rather by localized or diffuse lymphoid infiltration, manifesting as atelectasis that may mimic middle lobe syndrome without proximal bronchial obstruction. [2]

Clinically, patients typically present with cough and dyspnea. The prevalence of cough is estimated between 41% and 50%, significantly impacting patients' quality of life. Dyspnea has a prevalence of around 42%. Hemoptysis is much rarer and is likely due to bronchiectasis. [1,5]

Pulmonary function tests (PFTs) play a limited role in diagnosing bronchiolitis. In symptomatic cases, the Papiris study noted a reduction in MEF 25-75 and MEF50% to less than 75% of the predicted value in 87% of patients [6] . Reported data indicate that the frequency of distal airway dysfunction ranges between 22% and 46% [2]. PFTs reveal restrictive (35%), obstructive (18%) or mixed (24%) ventilatory defects. Several studies have concluded that mixed parenchymal and airway involvement is the rule in primary Sjögren's syndrome [4,7]. In contrast, our observation identified an obstructive ventilatory defect, pointing to bronchiolar involvement.

CT scans most often show bronchiolar micronodules (80% of cases). The most frequent diagnosis is cellular or follicular bronchiolitis. Advances in axial computed tomography (CT) techniques have improved sensitivity in detecting distal airway dysfunction. It is also more sensitive than PFTs in detecting distal airway dysfunctions. [7,8]

Other features suggestive of bronchiolitis are well described in the SGS: [1,8]

- A mosaic pattern indicating significant air trapping particularly visible on expiratory chest CT scan.
- Bronchiectasis and bronchial wall thickening.
- Ground-glass opacities, which can sometimes be diffuse.
- Signs of distension, especially in cases of constrictive bronchiolitis.

In our case, the radiological appearance is that of cellular bronchiolitis, showing branching micronodules, aligning with studies reporting bilateral nodules in over 78% of cases. These nodules have a centrilobular or peribronchial distribution. [8,9]

Surgical lung biopsy is sometimes necessary to confirm the nature of pulmonary lesions. Biopsy reveals various types of bronchiolitis, primarily follicular bronchiolitis (29%). Follicular bronchiolitis is characterized by hyperplastic lymphoid follicles in the bronchiolar walls, distributed along bronchovascular bundles. Less commonly, other types of bronchiolitis, such as obliterative bronchiolitis, constrictive bronchiolitis, lymphocytic bronchiolitis and panbronchiolitis, are observed. [2,3]

Corticosteroid treatment may be considered in cases of cellular bronchiolitis. However, symptomatic treatment is recommended with the use of bronchodilators or inhaled corticosteroids. Immunosuppressive drugs are ineffective and not recommended due to the increased risk of infectious complications. In cases of recurrent infections linked to bronchiectasis, a low-dose macrolide therapy is often prescribed (e.g., 500 mg of erythromycin per day or 500 mg of azithromycin three times a week) [10]. In our observation, a short course of corticosteroids, combined with a spray containing a bronchodilator and inhaled corticosteroid, successfully controlled the symptoms and stabilized the radiological lesions.

4. Conclusion

Bronchiolar involvement in the context of Sjögren's syndrome is often underdiagnosed. In fact, estimates of its prevalence vary widely. Dry cough and dyspnea are the main symptoms indicating pulmonary involvement. Hemoptysis, although rare in this context, should lead to rule out other differential diagnoses. High-resolution chest CT scan plays a crucial role in identifying various forms of distal airway involvement.

- The manifestation of Sjögren's syndrome through bronchiolitis is already a rare occurrence.
- Haemoptysis is an exceptionally uncommon symptom of bronchiolitis and should be thoroughly evaluated to rule out more frequent pathologies such as tuberculosis, pulmonary embolism, or vasculitis before associating it with Sjögren's syndrome. It is essential to exclude these common conditions before attributing the symptom to cellular bronchiolitis in the context of Sjögren's syndrome.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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