Am J Transl Res 2026;18(6):4925-4928
www.ajtr.org /ISSN:1943-8141/AJTR0169204

Case Report

Tracheobronchial amyloidosis in a 64-year-old woman with type 2 diabetes mellitus and Sjögren's syndrome: a case report

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Received October 9, 2025; Accepted April 22, 2026; Epub June 15, 2026; Published June 30, 2026

Abstract: Tracheobronchial amyloidosis (TBA) is an extremely rare and localized form of amyloidosis. Effective curative therapies for amyloidosis are lacking, and no standardized guidelines exist. Here, we report a 64-year-old female with a 20-year history of Sjögren's syndrome (SS) and 30-year history of type 2 diabetes mellitus (T2DM) who presented with progressive dyspnea. Chest computed tomography (CT) revealed tracheal and bronchial stenosis. Bronchoscopy showed submucosal nodular shadows and luminal narrowing, initially suspected as lung cancer. Biopsy confirmed TBA. Bronchoscopic interventions including argon plasma coagulation (APC) and mechanical debulking alleviated symptoms. Three-month follow-up showed significant improvement. This case highlights diagnostic challenges of TBA due to nonspecific manifestations and underscores the importance of considering TBA in SS patients with airway lesions. Congo red staining remains the diagnostic gold standard.

Keywords: Tracheobronchial amyloidosis, bronchoscopy, Sjögren's syndrome, type 2 diabetes mellitus, case report

Introduction

Tracheobronchial amyloidosis (TBA) is a rare disease accounting for <1% of symptomatic tracheobronchial lesions [1]. TBA associated with Sjögren's syndrome (SS) is extremely rare, with only sporadic reports [2]. SS is an autoimmune disease characterized by exocrine gland dysfunction, with potential for secondary amyloidosis due to chronic B-cell activation [3]. We describe a unique case of TBA with concurrent long-standing SS and type 2 diabetes mellitus (T2DM), highlighting diagnostic challenges.

Case presentation

A 64-year-old woman was admitted with progressive dyspnea and wheezing over 4 months. She had a 20-year history of SS and 30-year history of T2DM with optimal glycemic control. The symptoms of SS remained clinically manageable by administrating a traditional Chinese medicine (TCM) decoction for 2 months each year, incorporating 10-15 g Radix Glehniae, 10-15 g *Ophiopogon japonicus*, 6-12 g *Angelica sinensis*, 10-15 g *Lycium barbarum*, and 10-30 g *Radix Rehmanniae*. Physical examination revealed xerostomia, xerophthalmia, wheezing, and dry crackles. Laboratory tests showed positive anti-SSA, anti-SSB, and antinuclear antibodies (ANAs) at 1:320. Arterial blood gas analysis showed mildly reduced partial pressure of oxygen (PO_2) at 71.7 mmHg.

Chest computed tomography (CT) demonstrated soft tissue nodular shadows in the distal left main bronchus with luminal stenosis, suggesting malignancy (**Figure 1A**, **1B**). However, tumor markers were normal. Bronchoscopy revealed severe mucosal congestion with irregular granulation-like protrusions causing significant luminal narrowing (**Figure 2A**). Biopsy and argon plasma coagulation (APC) debulking were performed to restore airway patency (**Figure 2B-G**). Histopathology showed amorphous eosinophilic material with Congo red staining demonstrating apple-green birefringence under polarized light, confirming amyloid deposits (**Figure 2H-K**).

https://doi.org/10.62347/MEYD3605

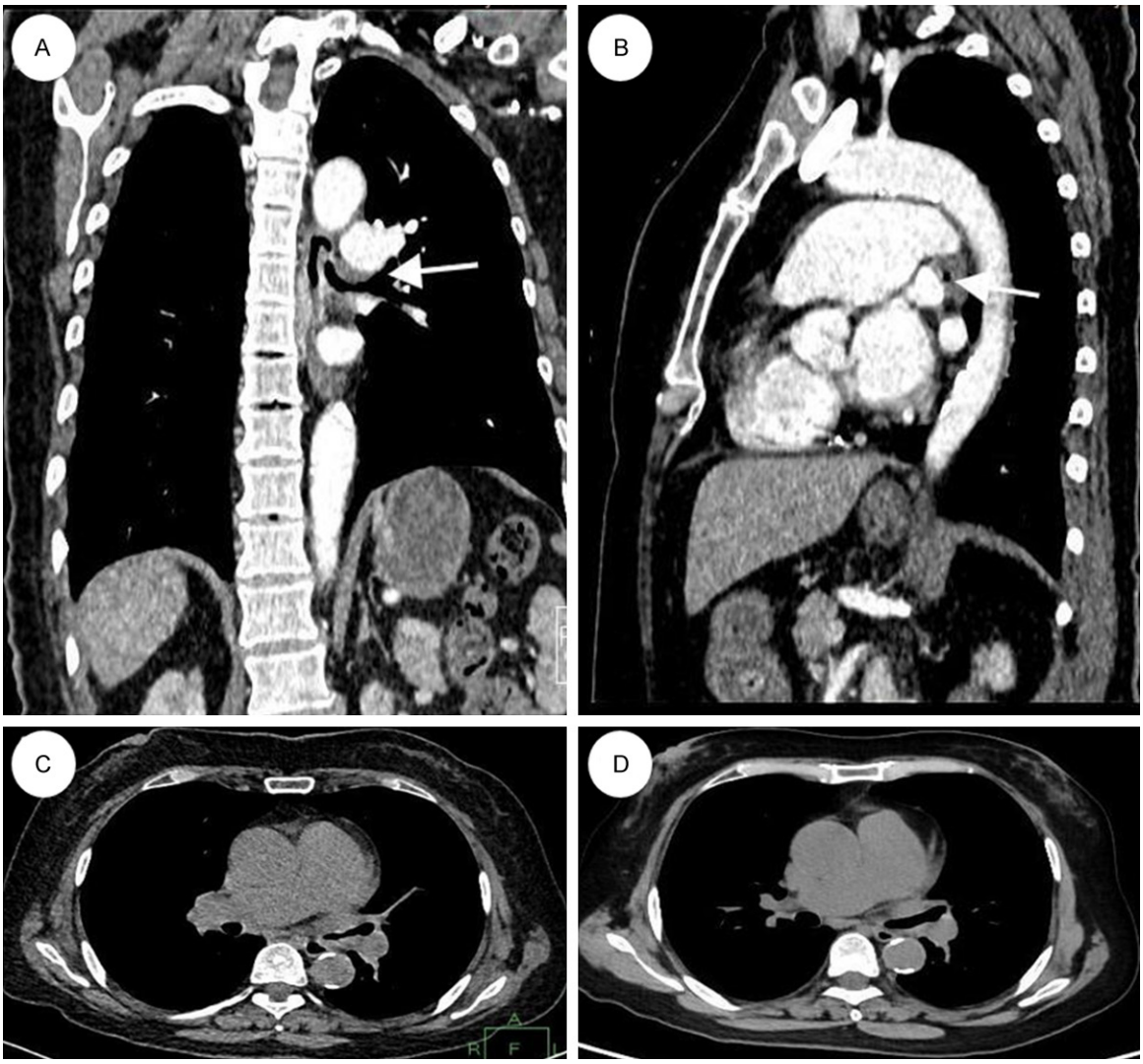

Figure 1. Chest contrast-enhanced computed tomography (CT) scans of a 64-year-old female patient with tracheobronchial amyloidosis (TBA) associated with Sjögren's syndrome (SS) and type 2 diabetes mellitus (T2DM). A: Coronal reconstruction at admission demonstrating irregular circumferential thickening of the airway wall involving the trachea and left main bronchus (white arrow) with associated luminal narrowing. B: Axial view at the level of the carina showing severe stenosis of the distal left main bronchus (approximately 70% reduction in diameter) and soft tissue nodular shadows obstructing the proximal segments of the upper and lower lobar bronchi (white arrow). C: Initial presentation at admission: irregular circumferential thickening of the left main bronchus wall with severe luminal narrowing and nodular soft tissue densities obstructing the bronchial tree. D: Three-month follow-up contrast-enhanced CT scan showing significant improvement with reduced airway wall thickness, restored luminal patency (>80% recovery in diameter), and resolution of previously observed nodular shadows. No new lesions or progression of stenosis was evident; the patient remained asymptomatic with stable disease status. CT parameters: 256-slice multidetector CT, slice thickness 1.0 mm, intravenous contrast enhancement (100 mL iodinated contrast, 30-second delay).

Post-procedure, wheezing improved significantly.

At 3-month follow-up, the patient reported no dyspnea, and CT showed restored luminal patency (>80% diameter recovery) (**Figure 1C**, **1D**). Final bronchoscopy confirmed patent bronchial orifices. Considering the diagnosis of TBA at admission and the complexity of TCM, including its components and theoretical underpinnings, the patient believed that the TCM failed to control the condition and stopped using it on her own. After further rheumatology consultation, hydroxychloroquine was initiated

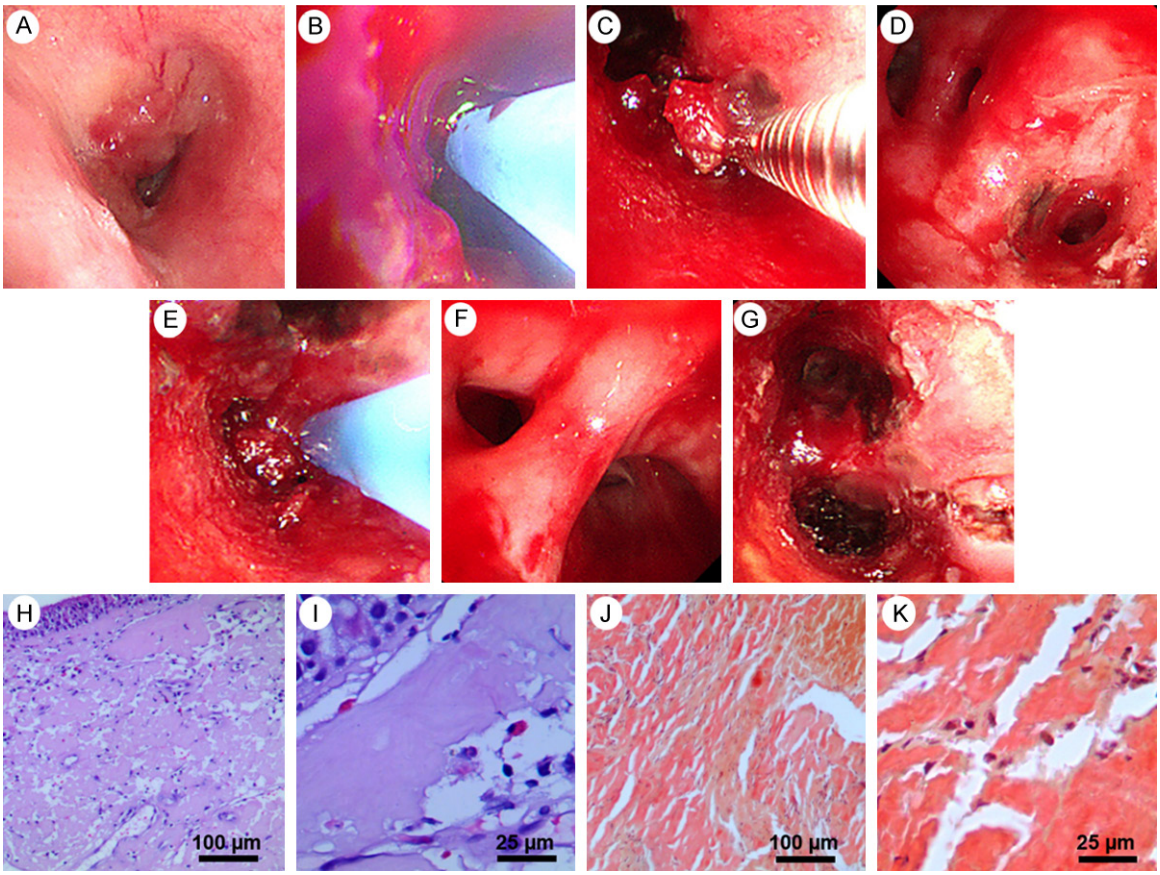

Figure 2. Flexible bronchoscopy findings and histopathological confirmation of tracheobronchial amyloidosis (TBA). A: Initial view of the distal left main bronchus showing severe mucosal congestion with diffuse irregular granulation-like protrusions causing significant luminal narrowing. B, C: Interventional sequence using argon plasma coagulation (APC; power 40 W, gas flow 0.8 L/min) and biopsy forceps for mechanical debulking of amyloid deposits. D: Post-intervention view of the left upper lobe bronchus orifice (dashed circle) after removal of obstructing tissue. E: Treatment of the interlobar ridge between left upper and lower lobes; thick, rigid granulation tissue requiring multiple APC applications. F: Final visualization of the patent left lower lobe bronchus orifice after complete debulking. G: Overview of the re-established interlobar ridge showing patent upper and lower lobe orifices (asterisks indicate bronchial openings). H: Hematoxylin and eosin (HE) staining showing amorphous eosinophilic hyaline material deposition within bronchial submucosa (×100 magnification). I: Higher magnification HE staining showing scattered lymphocytic and plasma cell infiltration with multinucleated giant cell reaction (×400 magnification). J: Congo red staining under polarized light microscopy demonstrating characteristic apple-green birefringence of amyloid deposits (×100 magnification). K: Higher magnification Congo red staining showing dense acellular eosinophilic material disrupting normal mucosal architecture (×400 magnification). Scale bars represent actual measurements. Bronchoscopic intervention under general anesthesia (BF-1TQ290; Olympus, Tokyo, Japan; 8.5 French tracheal tube). Tissue samples of histopathology were obtained from the distal left main bronchus, fixed in 10% neutral buffered formalin, and embedded in paraffin.

for SS management. At the present time, the condition of the patient has remained stable.

Discussion

TBA is a localized amyloidosis characterized by β-pleated fibrillar protein deposition in the tracheobronchial wall, causing progressive luminal narrowing [1]. Its clinical presentation - including dyspnea, wheezing, and cough - is nonspecific, often mimicking asthma, chronic obstructive pulmonary disease (COPD), or malignancy, leading to diagnostic delays averaging 6-12 months [4].

The association between SS and TBA is rare; only 23.1% of Japanese SS patients with amyloidosis developed TBA [2]. Chronic B-cell activation and dysregulated immune responses in SS may predispose patients to localized amy-

loid deposition in the respiratory tract [2, 3]. Notably, SS-related amyloidosis may coexist with mucosa-associated lymphoid tissue (MALT) lymphoma, necessitating long-term surveillance [5]. While type 2 diabetes mellitus (T2DM) has no established causal link to TBA, chronic hyperglycemia-induced inflammation may exacerbate disease progression.

Definitive diagnosis requires bronchoscopy with histopathological confirmation. Congo red staining demonstrating apple-green birefringence under polarized light microscopy remains the gold standard [6]. Chest CT findings of airway wall thickening, calcification, and luminal stenosis are suggestive but nonspecific [4, 7].

No standardized treatment guidelines exist for TBA. Management focuses on maintaining airway patency through bronchoscopic interventions, including APC, laser therapy, and mechanical debulking [7, 8]. These modalities provide effective symptomatic relief, though recurrence is common and repeat procedures are often necessary. Our case demonstrates successful bronchoscopic management with sustained clinical and radiological improvement at 3 months. Hydroxychloroquine was initiated to modulate underlying SS activity, although its efficacy in preventing TBA recurrence remains unproven [9, 10].

Clinicians should maintain a high index of suspicion for TBA in SS patients presenting with unexplained airway stenosis or endobronchial lesions. Early bronchoscopic evaluation is essential to establishing diagnosis and initiating appropriate intervention.

Acknowledgements

This study was funded by Tianjin Key Medical Discipline Construction Project (Grant No. TJYXZDXK-3-032C).

Disclosure of conflict of interest

None.

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