

Dry Eyes, Dry Mouth, and Amyloidosis – A Case Report

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Dry Eyes, Dry Mouth, and Amyloidosis – A Case Report

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INTRODUCTION

- Sjogren's syndrome can have associated pulmonary manifestations, usually presenting as airway disease or interstitial lung disease
- A rare form of Sjogren's related pulmonary disease, is known as primary nodular pulmonary amyloidosis (PNPA)
- The pathophysiology consists of amyloid deposits forming single or multiple nodules in the lung parenchyma²
- Etiology remains largely unclear, but is found in association with localized low-grade B cell lymphoma and mucosa-associated lymphoid tissue lymphoma of the lung. Typical presentation is in the 5th–6th decade of life and often has a benign clinical course.² Presenting symptoms include cough, dyspnea, fatigue, weakness, hemoptysis, and pleuritic chest pain.

CASE HISTORY

- A 76-year-old female with a history of Sjogren's syndrome (positive ANA, SSA, SSB, elevated rheumatoid factor, sicca symptoms and recurrent parotitis) presented for a CC of syncope
- She denied symptoms of cough, dyspnea, fatigue, weakness, hemoptysis, or pleuritic chest pain

EXAMINATION

- Afebrile and hemodynamically stable
- Lungs were clear to auscultation bilaterally and there was no sign of respiratory distress.
- No nail clubbing
- CXR and confirmatory CT scan of the chest (Figure 1) revealed numerous bilateral pulmonary nodules throughout the lung and peribronchial distribution
- CT-guided biopsy was negative for malignancy and infection

- Open thoracotomy with left upper lobe wedge resection was performed, and the biopsy revealed nodule, acellular, and waxy pink lesions with associated scattered plasma cells and occasional lymphoid aggregates
- Congo red stain produced apple-green birefringence under polarized light consistent with amyloid deposition (Figure A and B)

DIAGNOSIS

- Ultimately, the leading diagnosis is Sjogren's associated organ-limited nodule pulmonary amyloidosis and monoclonal gammopathy of undetermined significance
- The patient was managed with routine CT scans of the chest to monitor for progression

DISCUSSION

- PNPA is a rare manifestation of Sjogren's syndrome; currently, there are fewer than 200 cases reported in the literature
- Diagnosis requires lung biopsy revealing amyloid deposition and the exclusion of systemic amyloidosis
- It is important to maintain a broad differential in regards to the pulmonary manifestations of Sjogren's syndrome and the etiology of pulmonary nodules
- It is essential to exclude an underlying malignant or infectious etiology
- Other rare pulmonary manifestations of Sjogren's syndrome include pulmonary embolism, pulmonary arterial hypertension, and pulmonary lymphoma³

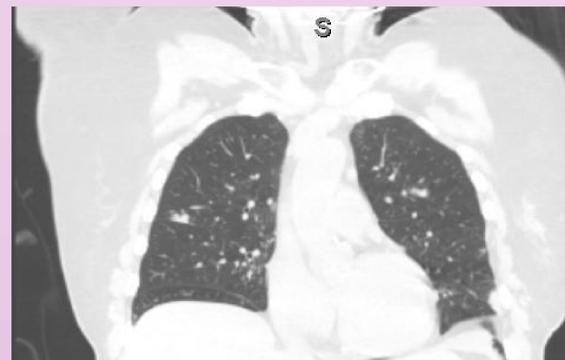


Figure 1: CT of the chest, abdomen, and pelvis without contrast confirming numerous bilateral pulmonary nodules throughout the lung and in the peribronchial vascular distribution.

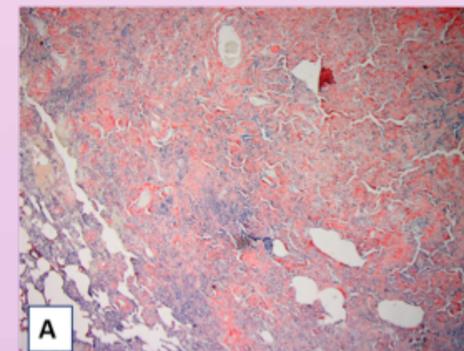


Figure A: Congo red staining the amyloid nodule [200X magnification].

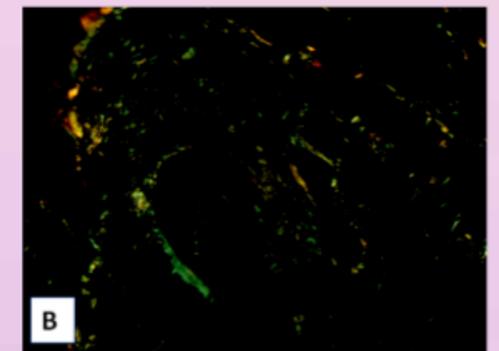


Figure B: Congo red stain producing apple-green birefringence under polarized light [400 X magnification].

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² Xian, Jonathan Z, et al. "A 45-Year-Old Woman With Multiple Pulmonary Nodules and Sjögren Syndrome." *Chest*, Elsevier, 4 Feb. 2019, www.sciencedirect.com/science/article/pii/S0012369218324395.

³ National Library of Medicine, or "Vancouver style" (International Committee of Medical Journal Editors): DynaMed Plus [Internet]. Ipswich (MA): EBSCO Information Services. 1995 -. Record No. T920750, Pulmonary manifestations of Sjogren disease; [updated 2018 Nov 30, cited 7/15/2019]. Available from https://www.dynamed.com/topics dmp~AN~T920750. Registration and login required.