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, known as primary nodular pulmonary amyloidosis (PNPA).
- The pathophysiology consists of amyloid deposits forming single or multiple nodules in the lung parenchyma.
- 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.
- 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 in the peribronchial vascular 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 lymphoma.

REFERENCES

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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