Diagnostic Dilemma: Puzzling Coexistence of Eosinophilic Pneumonia with Sjogren's Syndrome

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Abbreviation:
Primary Sjogren's Syndrome interstitial lung disease – pSS-ILD, Interstitial lung disease – ILD, Gastroesophageal reflux disease –GERD, Bronchoalveolar lavage - BAL

Background:

Sjogren's syndrome can lead to multisystem pathologies with ILD being a considerable contributor to mortality with the mean survival time of 9 years after confirmation of pSS-ILD [1]. ILD commonly presents as the nonspecific interstitial pneumonia in its fibrosing variant. Other forms are rarer including organizing pneumonia, usual interstitial pneumonia and lymphocytic interstitial pneumonitis [2]. One form of pneumonia that is not often seen with sjogren's is eosinophilic pneumonia. We present a case where a patient with no significant pulmonary nor autoimmune medical history presents with acute hypoxic respiratory failure and simultaneously gets diagnosed with sjogren's syndrome and eosinophilic pneumonia.

Clinical Presentation:

76-year-old female with GERD comes to the hospital for a one week history of shortness of breath upon ambulation. She was recently at an outside facility hospital couple weeks ago for the same symptoms. Per patient, the autoimmune and infectious workup (including fungal, viral, and atypical organisms) were negative. CT thorax showed severe bilateral lower lobe patchy infiltrate. They diagnosed her with cryptogenic organizing pneumonia treated her with Solu Medrol 40mg, Rocephin, and azithromycin during her admission. She was then discharged with a 10-day course of prednisone 50mg and home oxygen as needed. Steroids helped her breathe, but the exertional dyspnea reoccurred once she finished her outpatient course of steroids. She brought herself to the hospital once she started requiring 4-5L nasal cannula at home upon exertion.

Upon admission, labs are detailed as below:

Elevated markers:
Anti-SSA 111.3, IgM 477, CRP 134.4, ANA slightly elevated 1: 160, Mycoplasma Igm 2117.

Normal autoimmune labs:
C3 108, C4 22, RF 12, CPK 55, ANCA IFA nondetected, myeloperoxidase AB 5.65 (normal), serine protease 3 antibody 2 (normal), anti-Jo1 0.7, anti-RNP 1.7, anti-SCL 72.7, anti-SSB 3.2

Normal infectious disease labs:
Mycoplasma PCR negative, CMV negative DNA assay

Given her significantly elevated anti-SSA, rheumatology was consulted for possible sjogren's syndrome causing interstitial lung disease. Interestingly, patient denied any sicca symptoms. Rheumatology performed bedside schirmer's test and it was weakly positive in the left eye. Nevertheless, she met diagnositc criteria for sjogren's and was planned to start rituximab infusion. Bronchoalveolar lavage was also performed, and cell count showed 70% eosinophils. This raised suspicion for eosinophilic pneumonia. This shifted the diagnosis to eosinophilic pneumonia which directed treatment with mepolizumab instead.

Patient ultimately was started on mepolizumab 100mg subcutaneous injection every 4 weeks. 4 months later, repeat CT thorax was performed which showed marked improvement in fibrosis.

Conclusion:
This patient did not have the typical sicca symptoms that Sjogren's syndrome normally presents with, however she did have hypoxic respiratory failure that was refractory to steroids. Although it is rare to have ILD as the initial manifestation of primary Sjogren's syndrome, there was a case control study where they explored how sicca and non-sicca onset affected the trajectory of primary Sjogren's syndrome with interstitial lung disease. They found that about 51% of primary Sjogren's syndrome with ILD [pSS-ILD] presented with nonsicca onset. They also stated that pulmonary complications were more progressive and severe in non-sicca onset pSS-ILD when compared to sicca onset patients [3].

Interestingly, patient also met criteria for acute eosinophilic pneumonia with eosinophils >25% in BAL. Common associations of acute eosinophilic pneumonia are chronic myelogenous leukemia (CML), HIV infection, and smoking whereas chronic eosinophilic pneumonia is associated with asthma [4]. The patient did not have any of these medical

histories. The association between Sjogren's syndrome and eosinophilic pneumonia is uncommon. From a diagnostic standpoint, it may be tricky to determine which disease process is driving the symptoms when labs are convincing for both. We directed treated towards eosinophilic pneumonia since the patient had primarily respiratory symptoms rather than Sjogren's symptoms. Ultimately, treatment modality should be determined between pulmonology and rheumatology.

Sources:

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No disclosures