April 2017 Pulmonary Case of the Month

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History of Present Illness

A 63-year-old woman with a prior diagnosis of possible rheumatoid arthritis was referred for dyspnea with more vigorous activities in Prescott where she now lives (elevation 5367 ft.). She is receiving hydroxychloroquine 400 mg/day.

Past Medical History, Social History and Family History

She has a past medical history of hypertension. She smoked about a pack per day from age 20 to 40. There is a history of colon cancer in her mother and lung cancer in a sister.

Physical Examination

- Vitals: BP 155/102, SpO2 93% on room air
- Chest: slightly decreased breath sounds but clear
- Cardiovascular: regular rhythm without murmur
- Extremities: no cyanosis, clubbing or edema
- The remainder of the physical examination is normal

What **testing** would you perform at this time?

1. Chest X-ray
2. Pulmonary function testing
3. Rheumatoid factor
4. 1 and 3
5. All of the above

Correct!
5. All of the above

Her rheumatoid factor was positive at 45 IU/mL (normal <15). Her pulmonary function tests are below:

- FVC 94% predicted
- FEV1 80% predicted
- FEV1/FVC 65%
- TLC 113% predicted
- Dlco 75% predicted
- No significant change in FEV1 or FVC after bronchodilator

Her chest X-ray is shown in Figure 1.

Figure 1. Chest radiography.

Which of the following are **true**?

1. Her chest x-ray shows increased interstitial markings
2. Her positive rheumatoid factor is diagnostic of rheumatoid arthritis
3. The pulmonary function tests are normal
4. 1 and 3
5. None of the above

Correct!

5. None of the above

A positive rheumatoid factor occurs when antibodies are formed against the Fc portion of IgG (also an antibody) (1). Although a rheumatoid factor is elevated in rheumatoid arthritis, it can be seen in a variety of other rheumatologic diseases and nonrheumatologic disorders including cancer, chronic infections, inflammatory lung diseases (sarcoidosis, tuberculosis, idiopathic pulmonary fibrosis), mixed connective tissue disease, Sjögren's syndrome and systemic lupus erythematosus. To add even more confusion, some healthy individuals also have a positive rheumatoid factor (5% increasing to 20% over the age of 65 years).

Her pulmonary function tests show mild obstruction with a FEV1/FVC ration of 65% despite her FEV1 being within the normal range. Her diffusing limit for carbon monoxide (DLco) is also slightly decreased 75% of predicted (normal >80% predicted). Although these abnormalities might be dismissed in an otherwise normal healthy subject, they may be significant in a patient complaining of dyspnea.

Lastly, her chest x-ray does not show increased interstitial markings but does show hyperinflation (> 10 posterior ribs in the midclavicular line above the diaphragm).

Which of the following should be done next?

1. Rheumatology consultation
2. Bronchoscopy
3. Thoracic CT scan
4. 1 and 3
5. All of the above

Correct!
4. 1 and 3

The picture is sufficiently complicated that rheumatology consultation is appropriate. The rheumatologist examined the patient and ordered some basic laboratory including an anti SSA and anti SSB which were both positive. Anti Scl 70 and Jo1 were negative. Based on these findings the rheumatologist made a tentative diagnosis of Sjögren's syndrome.

At the present there are no indications for bronchoscopy.

A thoracic CT scan was ordered and is shown in Figure 2.

Figure 2. Representative images from high resolution thoracic CT scan in lung windows.

Based on the CT scan, the pulmonary function tests, and complaint of dyspnea, what is the **most likely diagnosis?**

1. Bronchiectasis
2. Emphysema
3. Small (< 2 cm) pulmonary nodules
4. 1 and 3
5. All of the above

Correct!
2. Emphysema

The complaint of dyspnea, the mild obstruction on spirometry, the mild decrease in the diffusing capacity, and the centrilobular emphysema seen on the CT scan are all consistent with emphysema. Emphysema, bronchiectasis, and small pulmonary nodules have been reported with Sjögren's syndrome (2,3). However, there are no nodules on the CT scan. Bronchiectasis usually presents with productive cough and is associated with central airways larger than the accompanied pulmonary artery.

A review of the high-resolution CT scan findings in Sjögren's syndrome found an incidence of emphysema of 37% (9 of 24 patients) (2). A variety of other abnormalities were seen mostly commonly bronchiectasis (11 of 24), thin-walled parenchymal cysts (11 of 24), and small pulmonary nodules (11 of 24). Our patient's smoking may have contributed to her emphysema but the diffuse involvement seen in our patient is more consistent with the previous reports rather than the apical predominance seen with smoking-associated emphysema. Emphysema has also been reported in nonsmokers with Sjögren's (3).

References

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2. Lohrmann C, Uhl M, Warnatz K, Ghanem N, Kotter E, Schaefer O, Langer M. High-resolution CT imaging of the lung for patients with primary Sjogren's syndrome. Eur J Radiol. 2004 Nov;52(2):137-43. [\[CrossRef\]](#) [\[PubMed\]](#)
3. Batra K, Wadhwa V, Torrealba J, Abbara S, Glazer C. Sjögren syndrome and emphysema: an atypical association. Clin Pulm Med. 2014;21:193-5. [\[CrossRef\]](#)