Medical Image of the Week: Pulmonary Amyloidosis in Primary Sjogren’s Syndrome

Figure 1. Thoracic CT scan showing multiple pulmonary nodules and lung cysts. The lung cysts were located apart from the pulmonary nodules.

Figure 2. Follow up CT scan in one year revealing worsening of the widespread lung cysts.
A 69-year-old woman with past medical history of Sjögren’s syndrome presented with pleuritic chest pain and shortness of breath for a month. Review of systems revealed worsening dysphagia and dryness of eyes over the last one year. Physical exam was significant for a palpable left axillary node and mild rhonchi bilaterally in the lower lung bases. Laboratory work was positive for Sjögren’s Syndrome antibodies. Chest x-ray revealed multiple nodules in bilateral lung fields. HRCT showed interlobular septal thickening and multiple cystic areas throughout the lung parenchyma which had progressed over 1 year (Figure 1). Wedge resection and thorough lymph node dissection were performed via video-assisted thoracic surgery (VATS). Biopsy of the lung nodules revealed thickened alveolar septate with acellular eosinophilic homogenous materials, which took up Congo Red stain. Based on these pathological findings, the final diagnosis was diffuse septal alveolar pulmonary amyloidosis secondary to Sjogren’s Syndrome. Bortezomib. However, the patient did not want to undergo chemotherapy. She preferred the ‘wait and watch approach’ and wished to be treated with only prednisone, with the intention to switch to azathioprine in future. However, after one year thoracic CT showed worsening of the cysts (Figure 2).

Pulmonary amyloidosis of the lower respiratory tract may represent a significant clinical problem in systemic and organ-limited amyloidosis and can contribute to cardiopulmonary failure. Pulmonary amyloidosis may present as a nodular localized type, diffuse septal alveolar amyloidosis, tracheobronchial amyloidosis and even pleural amyloidosis (1). Each patient requires complete assessment and unequivocal amyloid typing to determine their optimal treatment. Diffuse alveolar-septal amyloidosis is treated according to the underlying systemic amyloidosis, with most chemotherapeutic regimes similar to that of multiple myeloma (2). Patients should be monitored very closely and physicians should frequently assess the efficacy of the chemotherapeutic regime.

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References