

Patient History:

- 48 year old female
- incidentally identified multiple pulmonary cysts ranging in diameter from 8 cm to 2 mm by CT scan (Figure 1).
- distant medical history significant for an intrauterine fetal demise in 1998 after which she tested positive for an Anti-Nuclear Antibody (ANA) titer
- No other significant medical history

Workup for pulmonary cysts:

- negative genetic test for Birt-Hogg-Dube mutation
- normal Alpha-1Antitrypsin
- normal anti-double stranded DNA titer
- positive nucleolar and speckled pattered ANA titer of 1:10,240
- positive Sjögren's antibodies (SSA and SSB) titers
- positive anti-U1 ribonucleoprotein (RNP) antibody titer

Now presenting with:

- acute bilateral proximal and distal arthralgias
- weakness accompanied with extreme fatigue
- malar rash on her face
- Auscultation revealed scattered crackles and squeaks
- Elevated ESR and CRP
- Increase in size and number of known pulmonary cysts

She was treated with a 6 week course of prednisone and ongoing hydroxychloroquine with significant resolution of her symptoms except for fatigue which persists.

Figure 1a & 1b:



Representative CT images are shown highlighting an increase in both number and size of existing lesions between September 2015 in July 2020. The right middle lobe cyst has increased in size. A posterior right lower lobe cyst has increased in diameter from 35.4 mm to 39.4 mm. A new 4 mm cyst has emerged in the mid left lower lobe



Mixed connective tissue disease (MCTD)

MCTD is defined by the presence of high titers of the autoantibody RNP.

Clinical symptoms of MCTD include features of SLE, systemic scleroderma, and polymyositis.

Patients present with overlapping features of polyarthritis, sclerodactyly, and inflammatory myositis.

Patients with MCTD commonly also present with pulmonary manifestations including interstitial lung disease and pleural effusion in up to 80% of patients. Early disease may be asymptomatic

Pulmonary cysts represent a rare pulmonary finding and are infrequently reported in MCTD.

Discussion:

The presented case documents expansion in cyst number and size associated with a moderate to severe flare of underlying disease underscoring how interstitial lung disease of MCTD can manifest as enlarging cysts. This rare pulmonary finding was incidentally found before other disease manifestations.

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Scan here for references!