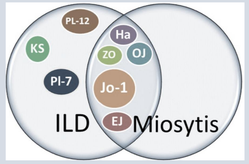
FEVER OF UNKNOWN ORIGIN - PL12 POSITIVE ANTISYNTHETASE SYNDROME

BACKGROUND:

Antisynthetase syndrome (ASyS) is a rheumatic disease that was initially described as part of autoimmune myositis spectrum disorder characterized by presence of anti Jo1 antibodies. It's now known to be associated with a variety of antibodies to aminoacyl-tRNA synthetases.

Clinical presentation of ASyS is diverse including inflammatory myopathy, interstitial lung disease (ILD), arthritis, cutaneous hyperkeratosis *mechanic's hands*, fever and Raynaud's phenomenon.



Antisynthetase Syndrome and Autoantibodies: A Literature Review and Report of 4 Cases. Marin FL, Sampaio HP. Am J Case Rep. 2019 Jul 25

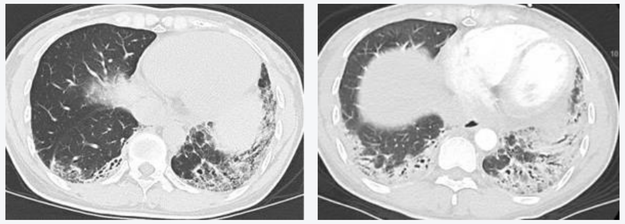
We present a rare case of ASyS manifesting with longstanding ILD, Shjogren syndrome and diagnosed in a setting of fever of unknown origin.

CASE PRESENTATION:

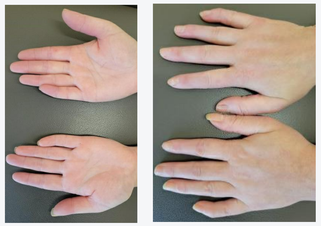
A 41-year-old woman, with history of ILD, Raynaud's phenomenon and Sjögren syndrome was hospitalized with progressive exertional, fevers, chills, and body aches for the last 3 weeks. She had received treatment with antibiotics and a short course of steroids as outpatient without effect.

On physical exam, she had low grade fever, tachycardia, tachypnea. Dry crackles were heard in lower lung lobes bilaterally. Hyperkeratotic rash was noted on palmar aspect of both hands and around nail beds.

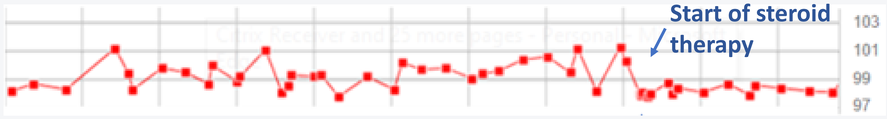
Lab work was remarkable for WBC of 31000/mcL, Hb of 8.9 g/dL, CRP of 22, RF of 40. CT chest showed worsening pulmonary fibrosis and enlarged pulmonary artery.



3 months before presentation *on presentation*



Patient was treated with broad spectrum antibiotics. However, she continued to have fevers after a week of hospitalization. Blood, sputum cultures and viral panel were negative. Broncho-alveolar lavage showed normal cells. Rheumatologic work up revealed high titer of anti-PL12 and anti-SSA-52KD. Patient was diagnosed with antisynthetase syndrome and stared on steroids. This was followed by resolution of the fever.



Patient now follows with rheumatologist and pulmonologist as outpatient, who tailor mycophenolate and prednisone regimens. PFT shows a restrictive pattern with FVC 0.99L. She requires 3L of oxygen supplementation on exertion. Patient is currently undergoing evaluation for a lung transplant.

CONCLUSION:

1. ASyS can cause long-lasting fevers meeting criteria for fever of unknown etiology.

2. Early broad rheumatologic workup is beneficial for patients with ILD.

3. ASyS can be associated with Sjögren like picture