27. A NEW PRESENTATION OF INTERSTITIAL LUNG DISEASE

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Introduction: Patients presenting with new onset interstitial lung disease (ILD) should be assessed for secondary, potentially treatable, causes. Some patients with rheumatic disease may present with ILD as the first manifestation of their condition. Early diagnosis and treatment of an underlying rheumatic disease can improve patient outcome. As autoimmune screening has evolved to include extended myositis and scleroderma panels, increasing numbers of patients with ILD are being referred for review by a rheumatologist. This case highlights one such patient diagnosed with new onset ILD during an acute hospital admission but subsequently found to have an underlying connective tissue disease (CTD).

Case description: A 54-year-old previously fit and well man presented to the Emergency Department with a three-month history of progressively worsening shortness of breath and dry cough. He denied any other symptoms, including those in keeping with a connective tissue disease (CTD). His past medical history was unremarkable. His father had a possible diagnosis of Sjögren's syndrome and his sister had Sjögren's syndrome with ILD. He had a 20 pack year smoking history.

On admission his could only walk ten meters before having to stop due to shortness of breath. On auscultation of his chest he had fine inspiratory crepitations in the mid and lower zones. Blood tests revealed an elevated c-reactive protein (CRP) of 96 and erythrocyte sedimentation rate (ESR) of 120. He failed to improve with intravenous antibiotics and his CRP rose to 122. Computer tomography (HRCT) of the chest, abdomen and pelvis revealed bilateral reticulation peripherally in the mid and lower zones and paraseptal/centrilobular emphysematous changes in the upper zones. Pulmonary function tests (PFTs) demonstrated normal spirometry but reduced gas transfer. Anti-cyclic citrullinated peptide (anti-CCP) antibodies, rheumatoid factor, anti-neutrophil cytoplasmic antibodies (ANCA) and HIV screen were negative. Creatine kinase was normal. ANA was positive, with positive anti-Ro-52 antibodies and positive anti-PL12 on an extended myositis panel.

A diagnosis of anti-synthetase syndrome was made. He was treated with three 1 gram doses of intravenous methylprednisolone on consecutive days then switched to 40mg of oral prednisolone daily. His inflammatory markers improved and he was discharged home. Monthly cyclophosphamide infusions were commenced and he has received two doses thus far. Although subjectively the patient does not report much improvement in his breathing as yet, he attends his appointments independently and is able to walk over 50 meters without stopping. Repeat PFTs and HRCT chest are scheduled.

Discussion: Patients presenting with ILD with no identifiable cause should be assessed and screened for CTDs. Evaluation should include a thorough history and examination, looking for associated conditions. ILD may be associated with rheumatoid arthritis, systemic sclerosis, polymyositis, dermatomyositis, anti-synthetase syndrome, sarcoidosis, Sjogren's syndrome, mixed connective tissue disease and systemic lupus erythematous (SLE).

Pulmonary function tests may demonstrate a restrictive pattern on spirometry, although this can be normal. Gas transfer is often reduced and a carbon monoxide transfer factor of $< 40\%$ is indicative of advanced disease. Imaging, usually in the form of high-resolution CT (HRCT), can allow assessment of the pattern of ILD and the potential for reversibility. It can often help avoid the need for lung biopsy.

If no clear alternative cause of ILD (eg. drugs, occupational exposure, inhaled substances, infection, radiation) then physicians should consider sending bloods to help excludes CTDs, including creatine kinase, rheumatoid factor, anti-CCP antibodies, anti-nuclear antibodies (with extended myositis and scleroderma panels) and ANCA, even in asymptomatic patients. Case series have demonstrated that ILD may be the only presenting feature in a proportion of those with anti-synthetase syndrome, particularly in patients with anti-PL7 or PL-12. The classic triad of clinical features for anti-synthetase syndrome consists of ILD, myositis and arthritis (mechanic's hands). Our patient had no CTD symptoms at presentation but has gone on to develop Raynaud's and stiffness in his fingers over the last 6 months. He has not at any stage had evidence of myositis, either clinically or serologically, which is in keeping with case series of patients with ILD and PL-12 positivity reporting a proportion as being amyopathic. Particular factors that should prompt screening for anti-synthetase syndrome include female gender, middle age, clinical signs suggestive of a CTD and an NSIP pattern on HRCT.

Key learning points: Rheumatic conditions which can present with ILD include rheumatoid arthritis, systemic sclerosis, polymyositis, dermatomyositis, anti-synthetase syndrome, sarcoidosis, Sjögren's syndrome, mixed connective tissue disease and SLE. Newly presenting ILD patients with no identifiable cause should be checked for rheumatoid factor, anti-CCP antibodies, anti-nuclear antibodies (with extended myositis and scleroderma panel) and ANCA.

ILD associated with anti-synthetase syndrome, polymyositis or dermatomyositis often warrants early treatment with steroid and cyclophosphamide or another immunosuppressive agent such as rituximab.

Patient characteristics which should prompt screening for anti-synthetase syndrome with extended myositis panel testing include clinical suspicion of CTD, female gender, middle age, CTD symptoms or signs and NSIP pattern on HRCT.

All patients presenting with ILD should have pulmonary function tests and imaging to assess severity of disease.

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