Abstract

Anti-synthetase syndrome is an overlap syndrome affecting lung, skeletal muscle, joints and skin. It is characterized by the presence of anti-synthetase antibodies, often anti-Jo-1 antibodies. This rare syndrome is seen in 40-60 years age group with female predominance. And it can be missed if not specifically consider in patients whose initial presentation is interstitial lung disease.

We report a case of anti-synthetase syndrome in a 64-year-old male from a rural area whose initial presentation was interstitial lung disease. He presented with progressive dyspnea on exertion, symmetrical polyarthritis, proximal muscle weakness and exfoliative erythematous rash. He had “mechanic’s hands”, proximal myopathy, bilateral fine end inspiratory crepitations in the lung bases.

He had elevated CPK, normal serum creatinine, chest Xray evidence of lung fibrosis and lung function tests of restrictive lung disease. His HRCT chest revealed interstitial lung disease suggestive of Usual interstitial pneumonia (UIP) pattern. His serology for ANA, rheumatoid factor was negative. Anti-U1RNP antibody was also negative. Jo-1 antibodies and Scl 70(PM-1) antibodies were positive. Anti-smith, anti-Ro (SSA), anti-La (SSB) antibodies were negative. Muscle biopsy was suggestive of polymyositis.

Patient was diagnosed as having anti-synthetase syndrome with myositis and interstitial lung disease. Patient was started on immunosuppressive therapy with corticosteroid and Azathioprine as steroid sparing agent. He was offered multidisciplinary care consisted of a rheumatologist and a pulmonologist. He had remarkable improvement of his symptoms, lung functions and biochemical markers at follow-up.

Anti-synthetase syndrome is a rare clinical syndrome which has features that overlap with other connective tissue diseases such as Raynaud's phenomenon, non-erosive arthritis, myositis and interstitial lung disease. Therefore, special serological investigations are required for diagnosis. And treatment with immunosuppressive therapy carries good prognosis. Absence of anti-Jo-1 antibodies and absence of myositis are poor prognostic factors.