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Case Report

ANTISYNTHETASE SYNDROME-A CASE REPORT

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ABSTRACT

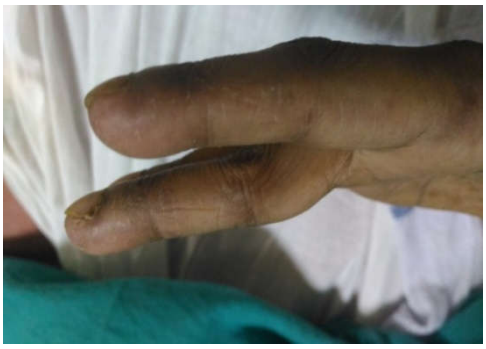
Dermatomyositis is a subtype of idiopathic inflammatory myopathies, characterized by cutaneous involvement and proximal muscle weakness. It can also have other systemic manifestations like arthritis, pulmonary and cardiac involvement. Antisynthetase syndrome is a rare variant with myositis and interstitial lung involvement characterized by autoantibodies against aminoacyl-tRNA synthetases. Here we present a case of dermatomyositis with predominant interstitial lung disease (ILD) anti-aminoacyl-tRNA synthetase antibodies (antisynthetase syndrome).

INTRODUCTION

Case Report

21 year old female from Malappuram presented to us with fever, cough and worsening of dyspnoea of 1 week duration. She had dry cough for the past 2 years followed by dyspnoea on exertion and polyarthralgia of 1 year duration. Fever was of high grade associated with chills and cough with purulent expectoration. Arthralgia was present involving wrists, elbows, shoulders and knees and was additive in nature, but not associated with any swelling or morning stiffness. There was no history of weakness, myalgia, orthopnea, or paroxysmal nocturnal dyspnoea. There was no history of oral or genital ulcerations or dysphagia. She gave history of bluish discoloration of extremities on exposure to cold. There was no history of allergy to dust or asthma. She had taken ayurvedic medications for joint pains but had no relief. She has a 3 year old son and there was no history of abortions.

On examination, she was conscious, oriented and had clubbing and cyanosis. Her pulse rate was 140/min, regular and normal in volume and character. Blood pressure was 110/80 mmHg. Saturation was 70 % with 2 L of Oxygen, and she had a respiratory rate of 46/min and was febrile with a temperature of 100^0F. General examination showed rough, cracked skin at the tips and lateral aspects of the fingers forming irregular lines which is known as mechanic's hand and erythematous plaques over knuckles suggestive of Gottrons papules (Figure-1).



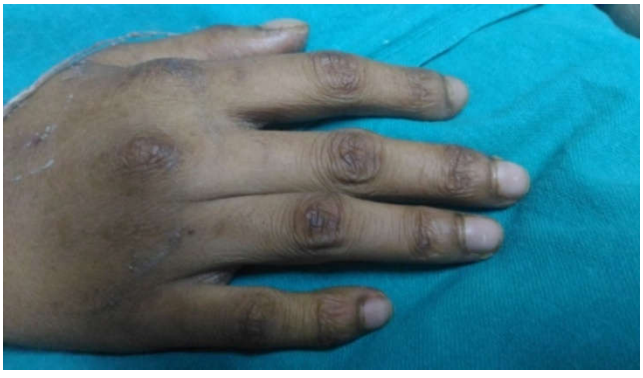


Fig 1 Mechanics hands and Gottron's Papules

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Respiratory system examination showed that Trachea was central, with Apex beat in 5th intercostal space in midclavicular line, vocal fremitus and vocal resonance was increased with coarse crepitations present in left mammary, infraaxillary, inter & infrascapular regions. Cardiovascular system examination showed a normal apex, palpable P2 and dull note in 2nd intercostal space. Auscultation showed a loud P2, abdominal examination showed no hepatosplenomegaly and there were no focal neurological defects or weakness in neurological examination.

Investigations done showed total count 28,600cells/µL with a differential count N88% L9%, hemoglobin 11.4g/dL, platelet count $6.84x10^3$/micro litre, ESR 80mm/1st hr, Blood urea 27mg/dl, Serum Creatinine 0.7mg/dl, Sodium 141meq/L , Potassium 4.6meq/L, total/conjugated bilirubin 0.7/0.4mg/dl respectively, total protein/albumin 5.9/3gm/dl, ALT 89U/L, ALP 102U/L, Serum Calcium 9mg/dL, serum phosphate 2.9mg/dL, Uric acid 3.9mg/dL, Urine routine examination normal and HIV, HBsAg, anti HCV was negative. Sputum C&S, AFB was Negative and blood C&S was sterile. Her CPK was elevated 690U/L, ANA was positive - 4 IU/L (>1.2), anti ds DNA negative, Ro52+++, Jo-1+++. CRP was 57mg/dL (normal<6), Procalcitonin- 2.60 (>2) Serum ACE 32micrograms /L (normal), TSH-1.15U/ml and RA factor – negative. Echo showed mildly dilated RA/RV, Moderate PAH, Good LV/RV function.

CT Thorax showed subpleural reticular opacities with macrocystic honeycombing and traction bronchiectasis, which increased from the apex to the bases of the lungs suggestive of interstitial pneumonia (Figure-2). In view of the presence of mechanic's hand, ILD, Jo 1 positivity, arthritis patient was diagnosed as a case of antisynthetase syndrome and managed with steroids and cyclophosphamide with which she improved.

Fig 2 CT Thorax showing interstitial lung disease

DISCUSSION

Inflammatory myopathies consists of a group of acquired autoimmune disorders characterized by skeletal muscle inflammation, which include dermatomyositis, polymyositis, necrotizing autoimmune myositis, inclusion-body myositis and overlap myositis.[1] Being an autoimmune disease autoantibodies are detected in the serum which could be myositis-associated and myositis-specific antibodies (MAAs and MSAs). Myositis associated autoantibodies are not specific for myositis and may be seen in other autoimmune diseases like SLE ,scleroderma also.eg: antinuclear antibodies (ANAs), anti-SSA/Ro antibodies, and anti-U1 ribonucleoprotein (anti-U1-RNP) are seen in 52%,12% and 11% cases of inflammatory myositis respectively

Myositis specific antibodies Include antibodies against components like

1. Nucleosome remodeling complex (anti-Mi-2)
2. RNA degradation and processing (anti-PM/Scl)
3. Ribonucleoproteins involved in translational transport (anti-signal recognition particle, or anti-SRP)
4. Ribonucleoproteins involved in protein synthesis (anti-aminoacyl-tRNA synthetase antibodies, also known as antisynthetase antibodies, or anti-ARS).Anti ARS antibodies include anti Jo1, Anti PL-12, Anti PL-7, Anti-OJ, Anti-KS, Anti-EJ, Anti-ZO

Antisynthetase syndrome consists of presence of anti-ARS antibodies, ILD, and clinical features like fever, arthralgias, Raynaud's phenomenon, and exanthema on the hands (also referred to as mechanic's hands). Anti ARS antibodies are directed against cytoplasmic enzymes that catalyse the formation of the aminoacyl-tRNA complex from an amino acid and its related tRNA. Anti-Jo-1 was the first anti-ARS to be discovered and the most commonly detected, and the strongest predictor of ILD.

Diagnostic criteria proposed for Antisynthetase syndrome (Table -1)[2]

Presence of an Anti ARS antibody plus 2 Major criteria or 1 Major and 2 minor criteria
Major criteria
1.ILD(not explained by environmental, occupational or drug exposure and not related to any other underlying disease)
2. Polymyositis or Dermatomyositis according to Bohan and Peter criteria
Minor criteria
1.Arthritis
2.Raynaud's phenomenon
3.Mechanic's hand.

Myositis: It is not universal and can develop later in life following the diagnosis of antisynthetase syndrome

Arthritis: is usually nondeforming, symmetrical, involving the wrists and metacarpal/phalangeal joints, whereas the proximal interphalangeal joints, shoulders, knees, and elbows are less affected.

ILD: Usually insidious in onset, nonspecific interstitial pneumonia (NSIP) is common. Anti PL-12 antibodies have a higher incidence of ILD (70–100%) and a lower incidence of biochemical myositis. Increased incidence of ILD seen among women.

Treatment

Systemic glucocorticoids are the mainstay of treatment of ILD in patients with PM or DM, Prednisolone 1mg/kg/day) with a maximum dose of 60mg daily and is tapered. Patients with impending respiratory failure due to rapidly progressive interstitial pneumonitis are treated with high-dose intravenous glucocorticoids (eg, methylprednisolone 1 gram daily for three days), followed by oral glucocorticoid therapy. A second immunosuppressive agent is often added to glucocorticoid therapy either for a glucocorticoid-sparing effect or because the ILD is more severe or progressive. Second agents that can be used are azathioprine, mycophenolate mofetil, calcineurin inhibitors, cyclophosphamide, or methotrexate. For mild to moderate ILD, azathioprine is often the first choice, Mycophenolate mofetil is an alternative if the patient does not tolerate azathioprine.[3] In patients with impending respiratory

failure due to rapidly progressive interstitial pneumonitis, cyclophosphamide(300 to 800 mg/m ^{2} every four weeks) plus high dose glucocorticoids with a transition to mycophenolate plus glucocorticoids after six months is suggested. Salvage therapy: In patients were the above treatments fail drugs like rituximab and IvIg have been tried.

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