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Respiratory Medicine

ANTI-PL 12 POSITIVE ANTISYNTHETASE SYNDROME PRESENTING AS SEVERE ILD- A CASE REPORT

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ABSTRACT Anti-synthetase syndrome (ASS) is a rare systemic autoimmune disease characterized by a combination of ILD , myopathy, fever and polyarthralgia and Raynaud's phenomenon and mechanic's hand . Hallmark of ASS is the presence of antibodies against aminoacyl tRNA synthetase enzyme.

The most commonly encountered autoantibody is anti-JO 1 (around 80%), less commonly encountered are anti pl 7, anti-PL 12 anti E J. (1)

Here we report the case of a 45 year old lady presenting with features of ASS where anti PL 12 was positive but with a negative anti JO 1.

KEYWORDS : Anti-synthetase syndrome, ILD, Anti-pl 12

BACKGROUND:

- Interstitial lung diseases (ILDs) comprise of disorders with variable etiology but with similar clinical and radiological presentation. Predominant site of involvement is the interstitium , but may also involve alveoli and pulmonary vasculature (6). ILD seen in Anti-synthetase syndrome usually progress rapidly and does not respond to glucocorticoids. (5)

CASE REPORT:

- A 45-year- old lady, home maker, with no comorbid illness presented with progressive shortness of breath, dry cough, muscle weakness, multiple joint pain, features of Raynaud's phenomenon and unexplained fever since 2 years.

No relevant past history, occupational history, No h/o any pets in home. No H/o any longterm drug intake. Non- smoker, Non-alcoholic

O/E:

- on examination patient was obese, with low saturation 70 % at room air, pulse rate 90/minute, BP 130/90mmHg, Afebrile, on auscultation bilateral crepitations were present, Other system examination were within normal limits. Gottron 's papules and mechanic hand were present on general inspection.

Investigations:

CT Chest revealed patchy ground glass opacities in both lungs with interstitial septal thickening with honeycombing in both lungs predominantly involving bilateral lower lobes- features suggestive of NSIP.RA factor was negative, anti jo 1 was negative but anti pl 12 was +, anti-Ro 52 positive, ANA positive with cytoplasmic pattern, CRP was elevated, Serum creatinine kinase –upper limit, S.LDH negative. ESR and CRP levels were elevated. 2D Echocardiogram showed normal study with no evidence of PAH.

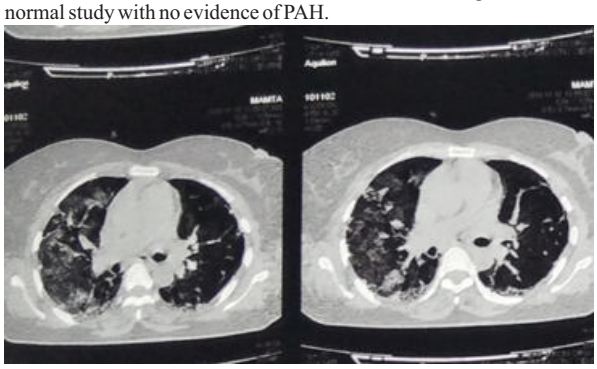

Image 1: CT showing features of ILD

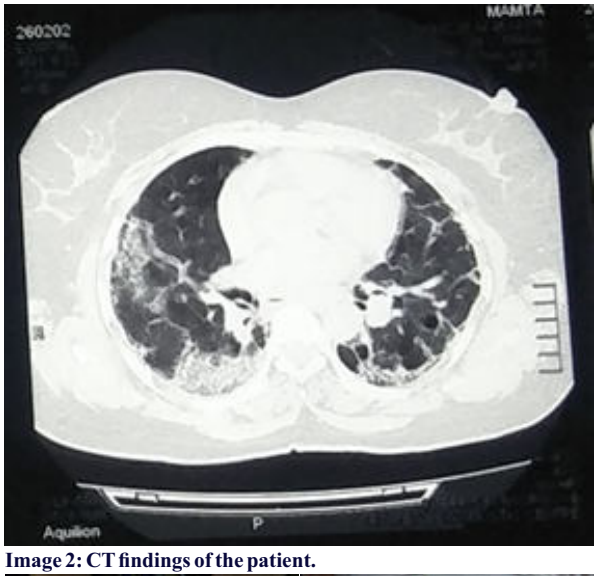

Image 2: CT findings of the patient.

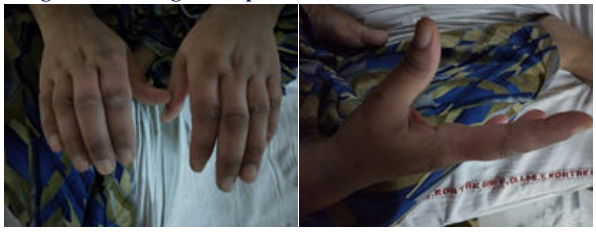

Image 3: Gottron's papules and mechanic 's hand

Treatment:

- Patient was initially treated with glucocorticoids and symptomatic therapy but there was no much improvement in symptoms, then Immunosuppressive drug, Azathioprine was a started and there was much improvement in saturation.

DISCUSSION:

- ASS firstly described by Marguerie et al in 1990 forms a major subgroup of inflammatory myopathies.(1, 2)
- ASS is 2 to 3 times more common in women than men with a mean age of onset varying from 43 to 60 years. (3)
- ILD is the most frequently occurring manifestation of ASS being

reported in 80-90 % cases, lung involvement is the most important prognostic indicator. (4)

- NSIP is the most frequently reported histological pattern in ASS and presence of fibrosis is an adverse predictor for response to therapy. (4)
- Patients with ASS have corticosteroid resistant myositis or ILD and they require additional immunosuppressive medications. (1)

CONCLUSION:

- Anti-synthetase syndrome is a rare, autoimmune disorder and must be considered while evaluating a patient with ILD.
- Usually the antibody seen in anti-synthetase syndrome is anti Jo 1, but may also present as anti-PL 12 positive with anti Jo 1 negative so an extensive panel of Anti-aminoacyl-tRNA synthetase antibodies should be tested.
- Anti-synthetase syndrome responds poorly to systemic glucocorticoids and requires additional immunosuppressive agents.

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