ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 03 | March - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A CASE OF ANTISYNTHETASE SYNDROME

Pharmacy

Dr.Mohan Varughese* Associate Professor And Consultant Department Of .medicine Belivers Church Medical College, Thiruvalla, India* Corresponding Author

Dr. Blessy Rachal boban clinical pharmacist Belivers church medical college, Thiruvalla, India.

Anjali.R Nazareth College of Pharmacy Othera, Thiruvalla, India.

ABSTRACT

Anti-synthetase syndrome is a rare medical condition seen across the globe. Here we have discussed the presentation of this uncommon medical illness

KEYWORDS

INTRODUCTION

The inflammatory myopathies constitute a heterogeneous group of chronic autoimmune disorders of unknown etiology. Polymyositis (PM) and dermatomyositis (DM) are the two types of disease in the group. As with other systemic autoimmune disorders, PM and DM are associated with serum autoantibodies, some of which are detected almost exclusively in these diseases.

A chief subgroup in inflammatory muscle disease is the antisynthetase syndrome considered by the presence of anti-synthetase antibodies and systemic clinical features are in muscles (myositis), lungs (interstitial lung disease), and joints (chronic polyarthritis) [1]

Table 1: Antisynthetase antibodies

Aminoacyl-tRNA synthetase (antigen)	Antisynthetase autoantibody
Histidyl-tRNA synthetase	Jo-1
Alanyl-tRNA	PL-12
Isoleucyl-tRNA	OJ
Threonyl-tRNA	PL-7
Glycyl-tRNA	EJ

Clinical features

Antisynthetase syndrome patients present with similar clinical features which include one or more among three involvement: myositis, interstitial lung disease, and joint involvement[2]. Raynaud's phenomenon and fever are also frequently observed.

Clinical manifestations	Prevalence (%)
Myositis	>90
Interstitial lung disease	60
Arthritis	50
Raynaud's phenomenon	40
Mechanic's hands	30
Fever	20

In several retrospective studies, the annual incidence of idiopathic inflammatory myopathies has been reported to be 2 to 10 new cases per million adults per year.[3] Antisynthetase antibodies are detected in 20% to 40% of such cases.[4] The disease is two to three times more common in women than in men.[5]

Interstitial lung disease may be the only manifestation in the absence of clinical myositis,[6] and patients can be falsely diagnosed as idiopathic pulmonary fibrosis when antisynthetase syndrome is not suspected. The confirmation of the diagnosis can be with thoracic high-resolution computed tomography.[7]

Lung transplantation is only currently available treatment for idiopathic pulmonary fibrosis.

Antisynthetase antibody is protective against an underlying malignancy.[8] But several recently published case studies showed various malignancies occurring within 6 to 12 months of the diagnosis of antisynthetase syndrome.[5,9]

Anti-Jo-1 antibody test is high specificity for correct clinical diagnosis [10]

Treatment

Glucocorticoids are considered the mainstay of treatment. Prednisone and azathioprine resulted in better functional outcomes.[11] The combination of methotrexate and azathioprine may be beneficial in patients who previously had inadequate responses to either of these agents alone.[12]

Concomitant therapy with prednisone and azathioprine in a randomized clinical trial resulted in better outcomes and a remarkably lower prednisone dose for maintenance therapy at 3 years than with prednisone alone.[13] For severe pulmonary involvement associated with antisynthetase syndrome, monthly intravenous infusion of cyclophosphamide is effective.[14] Cyclosporine has also been effectively used in a case of interstitial lung disease associated with anti-Jo-1 syndrome[15]. Rituximab, a monoclonal antibody to B lymphocyte antigen CD20, can also be used efficaciously in refractory disease[16]

CASE REPORT

76-year-old male patient. 17yrs is a known case diabetic, hypertensive presented with on OHA and calcium channel blocker presented with

1. Cough since 2013
2. Joint pain since 2016
3. Breathlessness since 2013

Cough since 7 years the mostly dry, during exacerbation presented with sputum, mucoid, not associated with haemoptysis. Exertional breathlessness present for 7 years. H/o muscle weakness and intermittent myalgia. No H/o Raynaud's phenomenon. No H/o cyanosis and no H/o palpitation.

Medical History: The patient was getting treatment for Diabetic and Hypertension with OHA, Sulfonylurea, Metformin, and Glimepiride. Vaccinated against Pneumococci and Influenza. The patient was recently admitted to a hospital for breathlessness and cough.

Occupational History: Employed in coal mine. Chronic exposure to cement, coal, construction material, silica, and asbestos dust.

Vitals: Pulse: 72/min, BP: 120/80mmHg, RR: 21/min, Temp: 98.4C, SPO2: 98%, No clubbing, there is significant desaturation present during 6-minute walk test.

CVS: S1S2 Normal, RS: Few scattered rales, CNS: Vibrational sense impaired big toe.

Lab Investigations : Total Leucocyte Count 9600 /µL 4800 – 10800, Poly H 75 % 43 - 72 , Eosin 02 % 0 – 4, Lymph 20 % 20 – 50, Hb 12.50 g/dL 12.00 - 17.00, RBC Count L 4.16 Million 4.40 - 6.00, Packed Cell Volume L 37.40 % 42.00 - 52.00, MCV 90.0 fL 80.0 - 100.0, MCH 30.0 pg 27.0 - 34.0, MCHC 33.3 g/dL 31.5 - 36.0 Mentzer Index** H 21.63

15 – 19, RDW 13.80 % 12.10 - 14.00, Platelets Count 2.08 Lakh/µL 1.50 - 4.50, PDW** 16.30 %, Mean Platelet Volume (MPV) 9.1 fL 7.5 - 11.2, Reticulocyte Count 2.91 % Adult: 0.2 - 2.0 %. ESR- 25 mm/hr

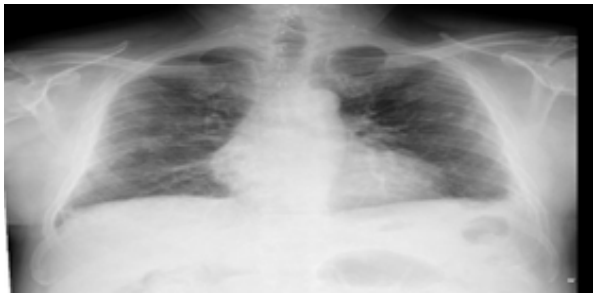

Figure 1:x-ray chest (PA)

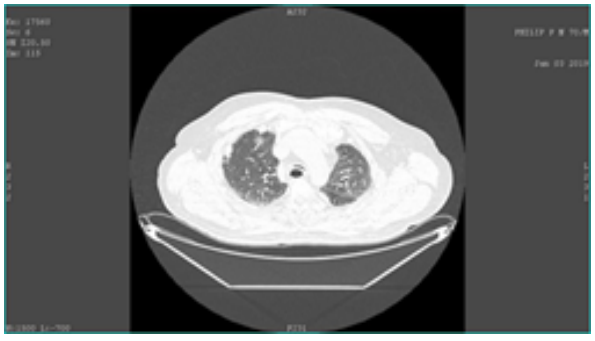

Figure 2:HR CT CHEST

LIVER FUNCTION TESTS

Bilirubin, Total, Serum 0.92 mg/dL 0.20 - 1.00, Bilirubin, Direct, Serum H 0.21 mg/dL 0.00 - 0.20, Bilirubin, Indirect, Serum 0.71 mg/dL 0.20 - 1.00, Calculated Protein, Total, Serum 6.60 g/dL 6.00 - 8.00, Albumin, Serum 4.22 g/dL 3.50 - 4.80, Globulin, Serum L 2.4 g/dL 2.5 - 3.5, A:G Ratio 1.77 1 – 2, AST, (SGOT),Serum H 47 U/L 10 – 40, ALT (SGPT),Serum H 80 U/L 10 – 40, Alkaline Phosphatase, Serum 67 U/L 30 - 120 UV

Hemoglobin A1c (HbA1c) - - 9.2%

Urea, Serum 41.6 mg/dL 17.0 - 49.0, BUN 19.44 mg/dL 7.00 - 23.00, Creatinine, Blood** H 1.47 mg/dL 0.60 - 1.30, BUN Creatinine Ratio** 13.22 6 – 22, e Glomerular Filtration Rate L 47 mL/min/1.7 3m2 60 – 160, Uric acid 4.64 mg/dL 4.40 - 7.60

PSA - 0.85 ng/mL

Microalbumin Creatinine Ratio H 32.74 mg/g Creat 0 – 30,C-Reactive Protein (CRP), Serum 1.1 mg/L 0.0 - 10.0 ,TSH (Thyroid Stimulating Hormone) 3.80 µIU/mL 0.40 - 8.90 ,Hepatitis C Virus (HCV) Antibody,Serum NonReactive , Hepatitis B virus Surface Antigen (HBsAg),Serum NonReactive

URINE ROUTINE EXAMINATION

URE. Albumin Nil,URE: Sugar 1+ ,Urine Microscopic Examination (UME): Micro - Pus 2-4 Nil / Hpf, Micro - Epith 1-2 Nil / Hpf, Micro - RBC 0-1 Nil / Hpf

AFB Sputum: Not Seen.

DISCUSSION AND CONCLUSION

A 75-year-old male patient presented with cough and breathlessness despite having treatment for Bronchial Asthma. He was a coal mine worker for more than 25 years. He is a known case of HTN for 20 years, T2DM for the last 17 years, Nephropathy, Carpal Tunnel Syndrome, and ILD. Medication history- sulfonylurea, metformin, linagliptin, Atorvastatin, and on bronchodilator inhalers as he has occasional complaints of wheeze. Vaccinated with influenza and pneumococcal vaccine. In 2019, the patient presented with joint pain and myalgia for which he was referred to the Rheumatology department and investigations showed a positive anti-Jo-1 and CK (147.6 IU/L). CT scan showed features of Interstitial Lung Disease and initially, he was managed with corticosteroid and subsequently with Mycophenolate, Pregabalin, and corticosteroids.

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