International Journal of Pharmaceutical Research and Applications
Volume 9, Issue 3 May-June 2024, pp: 1901-1904 www.ijprajournal.com ISSN: 2456-4494

Anti Synthetase syndrome-A Case Report

Arya Arun[1], Anfi.D.P[2], Shaiju S Dharan[3], Bincy Babu[*]

1. *Pharm D Intern (Ezhuthachan college of Pharmaceutical Sciences, Marayamuttom, Trivandrum, Kerala, India)*
2. *Pharm D Intern(Ezhuthachan college of Pharmaceutical Sciences, Marayamuttom, Trivandrum, Kerala, India)*
3. *Prof.(Dr). Shaiju S Dharan, Pricipal/HOD (Department of Pharmacy Practice, Ezhuthachan college of Pharmaceutical Sciences, Marayamuttom, Trivandrum, Kerala, India)*
4. *Dr. Bincy Babu *Assistant professor (Department of Pharmacy of Pharmacy Practice, Ezhuthachan college of Pharmaceutical Scienecs, Marayamuttom, Trivandrum, Kerala, India*

Date of Submission: 05-06-2024 Date of Acceptance: 15-06-2024

ABSTRACT
Anti-synthetase syndrome(ASS) is a rare type autoimmune disorder[1,2]. The symptoms of ASS can be quite variable, they mainly affect muscles, joints, lungs etc. The most common aminoacyl-tRNA synthetase antibody is anti-Jo 1. The autoantibodies like Jo-1, PL7 or PL12, these antibodies are strongly associated with lung diseases.The presence of myositis, non- erosive arthritis, ILD, Raynaud phenomenon, mechanic hands and fever associated with antibodies against aminoacyl-tRNA synthetase, a family of intracytoplasmic enzymes which play a vital role in protein synthesis constitute the ASS. Antibodies against aminoacyl-tRNA synthetases are the most common myositis specific-antibodies(MSA), present in patient with myositis.In ILD, lungs are involved dyspnea on exertion leads to shortness of breath. In myositis, distal muscle involvement is mostly affected by this syndrome, experiencing pain, swollen, red and warm. Prednisolone usually associated with steroid sparing agents such as azathioprine or methotrexate is used for the treatment of myositis.

KEYWORDS: Anti-synthetase Syndrome, autoimmune disorder, antibodies syndrome, ILD(Interstitial lung disease)

I. INTRODUCTION

Anti synthetase syndrome is an auto immune disorder, distinguished by the production of auto immune antibodies against aminoacyl transferase RNA synthetase (tRNA). In this the multiple systems of the body gets deeply affected and is found to be a rare medicated syndrome and a chronic disorder. This disorder is immune mediated and thereby results in unusual functioning of immune system. The remarkable symptoms that are correlated with this disorder are myositis which is a condition defining inflammation of muscles, interstitial lung disease, thickening and fissuring of the skin of hands(mechanic hands), Raynaud's phenomenon, numbing of fingers and toes and nettlesome sensation to cold condition[5].

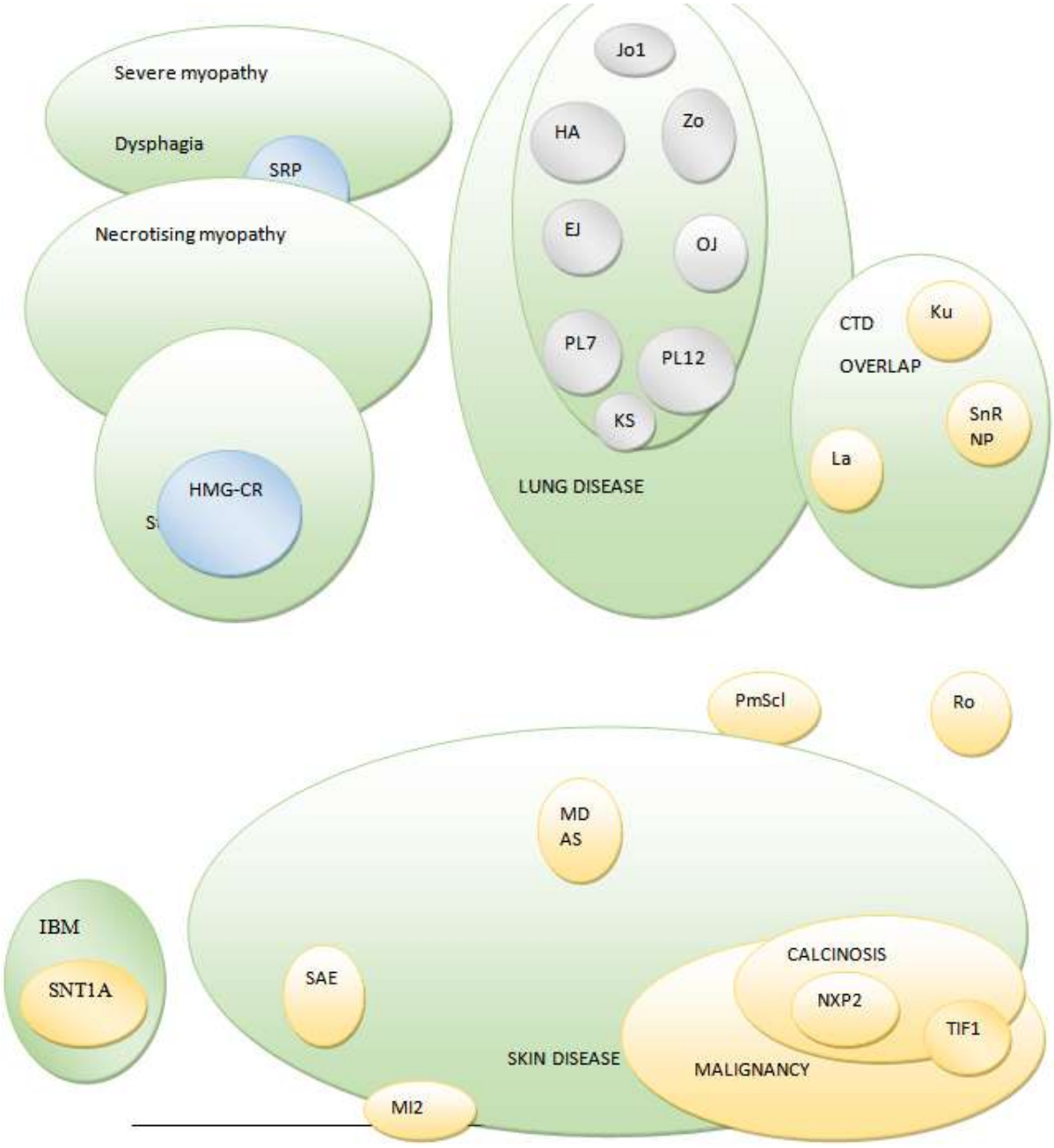
EPIDEMOLOGY

Anti synthetase syndrome first discovered in 1990 among 29 patients with polymyositis/dermo-myositis and ILD. Overall incidence of IIM (idiopathic inflammatory myopathy) is about 6 to 10 per million, incidence of jo-1 is 1.2 to 2.5 per million and prevalence is 5 per 100,000. Estimated ratio is about 2.1 in some series.[11].

PATHOPHYSIOLOGY

Aminoacyl-tRNA synthetases are enzymes involved in binding of specific amino acids to matching tRNA during the translation phase of protein synthesis. 20 different synthetases, each one is specific to one amino acid. These synthetase stimulate macrophages to produce TNF-alpha, unclear. Also a T-cell in tissue damage[4].

International Journal of Pharmaceutical Research and Applications
Volume 9, Issue 3 May-June 2024, pp: 1901-1904 www.ijprajournal.com ISSN: 2456-4494



TREATMENT

Predniosolone -1mg/kg/day remains the drug of choice for ASS. When ILD is prominent, cyclophosphamide administered in monthly intravenous pulses(1gr/body m^2). Among the steroid sparing agents, azathioprine -2.5mg/kg/day, methotrexate (0.3mg/kg/week)[2]

II. CASE PRESENTATION

A 36yrs old male patient, working as an Indian army officer (Havildar) has been symptomatic in 2018 with Raynaud's phenomenon. There was no suggestive history of digital ulcerations/pitting on him. The digital ulceration is a condition marked by poor blood flow to the finger tips due to narrowing of blood vessels.

International Journal of Pharmaceutical Research and Applications
Volume 9, Issue 3 May-June 2024, pp: 1901-1904 www.ijprajournal.com ISSN: 2456-4494

In February 2020, he developed joint pain and swelling involving bilateral PIP (Proximal inter phalangeal) joints. Few days later he developed myalgia and proximal muscle weakness of upper and lower limbs. There was no sensory/bowel bladder/ cerebellar symptoms.

He was evaluated at CH[EC] and upon MRI scanning of calf, he was detected with myositis. Immunological workup revealed negative ANA (antinuclear antibodies) and anti jol positivity in LIA (local infiltration analgesia). At the time of arrival in the department of Rheumatology his vital were as follows Pulse: 70/min, BP: 120/80mmHg, RAS: No pallor, edemaSkin: no skin thickening/no Gottron's sign(flat red rash over the back of the fingers, elbows or knees).

Inmusculoskeletal system, the gait was normal, no abnormality detected in upperlimb, lower limb, spine

MMT 8:80/80, Chest: NAD,CNS: NAD, CVS: NAD.

He was managed as a case of anti-synthetase syndrome with oral steroids, methotrexate with good response to treatment. Steroids were tapered and stopped 6 months back.

Laboratory parameters include Hb 15.6 g/dl, CRP 1.0 mg/l, platelet 1.84 L/cumm. Urine RE: NAD, ANA by IIF (August 2021): Negative, Creatinine 0.97 md/dl, C3,C4: 1.0/0.17 g/L, ALT/AST-19/38 IU/L, CPK-113/U/L, LDH-220U/L, Ig profile normal, PFT:FVC 105% predicted, The ECG found to be normal.

Presently he hadcomplaints re-occurence of myalgia, not associated with weakness for last 2 months. He had no history of joint pain, skin rash/ shortness of breath and excertion. The treatment under taken includes T. Methotrexate 20mg once a week(Saturday), T. Folic acid 5mg twice a week(Tuesday/Thursday), T. Calcium 500mg OD, T. Amlodipine 5mg OD, Sachet Vit D3 60000IU once monthly, T. Pantoprazole 40mg OD.

III. DISCUSSION

This condition is more susceptible to females than in males. Anti-ARS antibodies are considered to be the markers of the disease and the activity of disease may tally up with the level of antibodies that are generate in the body. Anti-synthetase syndrome is an idiopathic inflammatory myopathy(IIM), with high prevalence of ILD compared to dermatomyositis(DM) and polymyositis. Here the patient had the symptom of Raynaud phenomenon, myalgia, swelling and pain in PIP. In this case the treatment provided for subsiding the symptom is Tab.Methotrexate 20 mg once a week, Tab.Folic acid 5mg twice a week, Tab.Calcium 500mg OD. Connors et al. and Solomon et al. proposed diagnostic criteria for the anti-synthetase syndrome. According to Connors et al., the presence of an anti-aminoacyl tRNA synthetase an antibody is required as well as one or more of several clinical features, namely, Raynaud's phenomenon, arthritis, interstitial lung disease, fever and mechanic hands. According to Solomon etal,the presence of an anti-aminoacyl tRNA synthetase antibody is required plus two major or one major and two minor criteria, the major criteria being interstitial lung disease and polymyositis or dermato-myositis, the minor criteria being arthritis, Raynaud phenomenon and mechanic hands. The patient described here fulfilled both the set of criteria for the diagnosis of anti- synthetase syndrome proposed either Connors et al or Solomon et al. It is a rare disease no controlled clinical trials have been performed for the assessment of different treatment options. The response to immunosuppressive medication is quite favourable.

IV. CONCLUSION

This case report presents a detailed analysis of a 36yrs old male patient, working as an Indian army officer (Havildar) and he becomes symptomatic in 2018 with Raynaud's phenomenon. In February 2020, he developed joint pain and swelling involving bilateral PIP (Proximal inter phalangeal joints). Few days later he developed myalgia and proximal muscle weakness of upper and lower limb. He was with case of anti-synthetase syndrome and managed with oral steroids, methotrexate with good response to treatment. Steroids was tapered and stopped 6 months back. The report discusses the clinical presentation including the presence of developed symptoms and managed with treatment options.

REFERENCE:

[1]. Katzap E, Barilla-LaBarca ML, Marder G. Antisynthetase syndrome. Current rheumatology reports. 2011 Jun;13:175-81.

[2]. Witt LJ, Curran JJ, Strek ME. The diagnosis and treatment of antisynthetase syndrome. Clinical pulmonary medicine. 2016 Sep 1;23(5):218-26.

[3]. Imbert-Masseau A, Hamidou M, Agard C, Grolleau JY, Chérin P. Antisynthetase

syndrome. Joint Bone Spine. 2003 Jun 1;70(3):161-8.

[4]. E Mirrakhimov A. Antisynthetase syndrome: a review of etiopathogenesis, diagnosis and management. Current medicinal chemistry. 2015 May 1;22(16):1963-75.

[5]. Cojocaru M, Cojocaru IM, Chicos B. New insights into antisynthetase syndrome. Maedica. 2016 Jun;11(2):130.

[6]. Noguchi E, Uruha A, Suzuki S, Hamanaka K, Ohnuki Y, Tsugawa J, Watanabe Y, Nakahara J, Shiina T, Suzuki N, Nishino I. Skeletal muscle involvement in antisynthetase syndrome. JAMA neurology. 2017 Aug 1;74(8):992-9.

[7]. Marder GS, Greenwald R. Antisynthetase syndrome. The inflammatory myopathies. 2009:191-206.

[8]. Brulhart L, Waldburger JM, Gabay C. Rituximab in the treatment of antisynthetase syndrome. Annals of the rheumatic diseases. 2006 Jul 1;65(7):974-5.

[9]. Chatterjee S, Prayson R, Farver C. Antisynthetase syndrome: not just an inflammatory myopathy. Cleve Clin J Med. 2013 Oct 1;80(10):655-66.

[10]. Marie I, Josse S, Decaux O, Dominique S, Diot E, Landron C, Roblot P, Jouneau S, Hatron PY, Tiev KP, Vittecoq O. Comparison of long-term outcome between anti-Jo1-and anti-PL7/PL12 positive patients with antisynthetase syndrome. Autoimmunity reviews. 2012 Aug 1;11(10):739-45.

[11]. Kane BS, Mbodji AB, Dieng M, Longo CS, Fall BC, Ndao AC, Niasse M, Berthe A, Sow M, Ly F, Ndongo S. Epidemiological, clinical and immunological aspects of antisynthetase syndrome: a multicentre study in Dakar. The Egyptian Journal of Internal Medicine. 2023 Apr 27;35(1):34.