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Anti-Synthetase Syndrome (PL-7 Positive) Presenting with Necrotizing Inflammatory Myositis and Right-Sided Pneumonia: A Diagnostic Challenge

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Abstract: Anti-Synthetase Syndrome (ASS) is an autoimmune disorder characterized by autoantibodies against aminoacyl-tRNA synthetases, including the rarer anti-PL-7 antibody. This variant is often associated with severe multi-system involvement, including necrotizing myopathy, interstitial lung disease (ILD), and non-erosive arthritis. The clinical manifestations can overlap with other systemic conditions, posing challenges in diagnosis and management. The case report describes about a 57-year-old woman with diabetes, hypertension, and morbid obesity was bedridden due to growing limb weakening and edema for a month. Her diagnosis included necrotizing inflammatory myositis, pneumonia, quadriparesis, hypokalemia, and anemia, along with Anti-Synthetase Syndrome (PL-7 positive). Imaging showed thoracic compressive myelopathy and myositis. Clinical improvement was achieved by the use of IVIG, corticosteroids, antibiotics, and supportive care.

Keywords: *Amino Acyl-tRNA Synthetases, Interstitial Lung Disease, Myositis, Anti PL-7.*

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I. INTRODUCTION

Anti-Synthetase Syndrome (ASS) is a rare and complex autoimmune disorder characterized by the presence of autoantibodies against aminoacyl-tRNA synthetases, most notably anti-Jo-1, but also including less common variants such as anti-PL-7. Clinically, it presents with a constellation of symptoms, including inflammatory myopathy, interstitial lung disease (ILD), non-erosive arthritis, Raynaud's phenomenon, and characteristic skin manifestations like "mechanic's hands." Among these, the anti-PL-7 antibody subset often presents with a more aggressive pulmonary involvement and may have atypical or overlapping features compared to classic anti-Jo-1 positive cases.[1]

Necrotizing autoimmune myopathy (NAM) is a severe form of inflammatory myopathy marked by muscle fiber necrosis with minimal inflammatory infiltrate. It is frequently associated with significant muscle weakness and can lead to debilitating complications such as quadriparesis. The association of NAM with Anti-Synthetase Syndrome, especially in the setting of anti-PL-7 positivity, is rare and carries a high risk of morbidity, especially when complicated by pulmonary infections.[2]

This case report describes a patient diagnosed with PL-7 positive Anti-Synthetase Syndrome who presented with necrotizing myopathy complicated by quadriparesis, severe anemia, hypokalemia, and candiduria. The patient also exhibited pulmonary involvement suggestive of bronchogenic spread of infection, raising differentials of pulmonary tuberculosis or atypical pneumonia, in addition to gastrointestinal complications such as gastric ulcers and erosive duodenitis.[1][2]

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The complexity of the clinical presentation required a multidisciplinary approach to management, involving antimicrobial therapy, immunomodulation with intravenous immunoglobulin (IVIG) and corticosteroids, mucolytic support with N-acetylcysteine, gastric protection with proton-pump inhibitors, and supportive care for hematological and electrolyte disturbances.[4][5]

This case highlights the importance of early recognition and aggressive management of Anti-Synthetase Syndrome with multi-organ involvement and underscores the critical role of targeted immunotherapy in improving outcomes in patients presenting with severe myositis and respiratory complications.[3]

II. CASE REPORT

A 57 yr old female patient came to the hospital with a complaint of pain in the right leg for 1.5 months, swelling in B/L legs for 15 days, bedridden for 1 month presented with paraparesis at private hospital with B/L lower limbs and truncal weakness where the patient was diagnosed as necrotizing inflammatory myositis with hepatopathy, coagulopathy & Rt sided pneumonia.

➢ *Past Medical History:*
K/C/O DM, HTN, morbidly Obese.

➢ *On Examination:*
The patient was conscious, coherent, and cooperative. The pulse rate was 78 bpm, blood pressure was 130/70mmHg, spo2 was 92%, and the respiratory system (RS) examination indicated regular bilateral air entry (BAE+) with right basal crept+. Cardiovascular examination: Both heart sounds were heard with no murmur. There was no evidence of pallor, cyanosis, icterus, clubbing, or lymphadenopathy. In Neurological examination -No neurological deficits, Per-abdomen examination: Abdomen is soft and nontender. Anasarca +, muscle tenderness+, and CNS examination suggest that Motor power was reduced bilaterally, with strength graded as 3/5 in upper limbs and 2/5 in lower limbs. Bilateral extensor plantar responses were noted. These findings suggest bilateral upper motor neuron involvement, likely affecting the corticospinal tracts.

➢ *Laboratory Findings:*
HRCT chest revealed segmental consolidation in the right lower lobe suggestive of infective etiology, along with centrilobular nodules and patchy consolidations in the right upper, middle, and left lung lobes—findings consistent with a bronchogenic spread of infection, with differentials including atypical pneumonia. COOMBS test – Negative indicates no immune-mediated haemolysis is occurring. Muscle Biopsy provisional –Necrotising myositis. MRI spine – degenerative disc, thecal sac indentation from D6-D7 to D9-D10 (compressive myelopathy). MRI of bilateral lower limbs showed features of myofascitis, indicating an underlying inflammatory process involving both muscle and fascia. USG abdomen shows Grade 2 fatty liver. Urine examination revealed plenty of budding yeast forms. UGIE impression - gastric ulcers are described, and erosive duodenitis. Urine for culture indicates no bacterial growth. Chest Xray – increased broncho vascular markings.

- **Renal function test & Electrolytes levels:** Blood urea 33 mg/dL, serum creatinine 0.33 mg/dL, sodium 135 mEq/L, potassium 2.6 mEq/L (hypokalemia), chloride 94 mEq/L.
- **Liver function tests:** Total bilirubin 1.43 mg/dL, direct bilirubin 0.34 mg/dL, SGOT 28 U/L, SGPT 41 U/L, ALP 121 U/L.
- **Proteins:** Total protein 5.53 g/dL, albumin 2.6 g/dL, globulin 3 g/dL.
- **Uric acid:** 7.99 mg/dL.
- **C3 and C4 levels:** C3 - 104 mg/dL, C4 - 35 mg/dL.

- *Complete Blood Picture:*

✓ Haemoglobin: 9.5 g/dL (anemia)
✓ Total WBC count: 8.3 x10^3/µL
✓ Platelets: 168 x10^3/µL
✓ Neutrophils: 88.7%, Lymphocytes: 5.1%, Eosinophils: 0.4%, Monocytes: 5.8%, Basophils: 0.0%
✓ MCV: 96.5 fl, MCH: 30.5 pg, MCHC: 31.7 g/dL, RDW-CV: 24.9%, RBC count: 3.11 million/µL, HCT: 30.0%

Table 1 Illustrates the Findings and Clinical Interpretation of Important Muscle Injury and Antibody Indicators used to Identify and Distinguish between Inflammatory and Autoimmune Muscle Disorders.

Antibodies	Result
FDP 8	NEGATIVE
SPR	NEGATIVE
JO-1	NEGATIVE
HMG COA	NEGATIVE
CPK	ELEVATED
PL 7	POSITIVE
Antinuclear antibody	NEGATIVE
Antineutrophil cytoplasmic antibody	NEGATIVE

(FDP 8 - Fibrin Degradation Product 8, SPR – Scleroderma Profile/Panel Report ,JO-1 – Histidyl-tRNA Synthetase Antibody, HMG-CoA – 3-Hydroxy-3-Methylglutaryl-Coenzyme A Reductase Antibody,CPK – Creatine Phosphokinase ,PL-7 – Threonyl-tRNA Synthetase Antibody)

Finally, the patient is diagnosed with Anti-Synthetase Syndrome (PL-7 positive) with necrotizing inflammatory

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myositis with right-sided pneumonia, complicated by quadriparesis, hypokalaemia, and severe anemia.

Initially, the patient was managed with broad-spectrum antibiotics to address the suspected pulmonary infection. Nebulizer Budecort was administered to control airway inflammation and improve respiratory symptoms. Injection N-acetylcysteine (NAC) at a dose of 1.2 g IV twice daily was given as a mucolytic agent to facilitate mucus clearance. Injection Pantoprazole 40 mg IV was used to treat existing gastric ulcers and prevent further gastrointestinal complications.

Due to the presence of severe anemia, the patient underwent transfusion with one unit of packed red blood cells (PRBCs) and was started on oral iron-folic acid (IFA) supplementation along with vitamin C to enhance hematinic absorption. For immunomodulation, intravenous immunoglobulin (IVIG) was initiated at a dose of 0.4 mg/kg per day (five vials daily), along with high-dose oral prednisolone 60 mg/day, aiming to control the autoimmune-mediated necrotizing myopathy associated with PL-7 positive Anti-Synthetase Syndrome.

III. DISCUSSION

Anti-synthetase syndrome (ASS) is a rare autoimmune condition characterized by the presence of autoantibodies directed against aminoacyl-tRNA synthetases, most commonly anti-Jo-1. However, patients with non-Jo-1 antibodies, such as anti-PL-7, often present with more severe pulmonary involvement and relatively milder or variable muscle symptoms. Our patient, with PL-7 positivity and Jo-1 negativity, exhibited a complex and aggressive disease course.[1]

The hallmark features of ASS include myositis, interstitial lung disease (ILD), arthritis, Raynaud phenomenon, and mechanic's hands. In this case, the patient presented with progressive quadriparesis, hypokalemia, anemia, and signs of respiratory infection, complicating the clinical picture.[2] The HRCT chest findings of segmental consolidation, centrilobular nodules, and patchy opacities suggest bronchogenic spread of infection, with differentials including pulmonary tuberculosis and atypical pneumonia, both of which are known to coexist or mimic autoimmune lung involvement.

The muscle biopsy findings of necrotizing myositis confirm a severe inflammatory myopathy, consistent with the muscle involvement seen in ASS. MRI of the lower limbs revealed myofascitis, which further supports an active inflammatory process involving muscle and fascia. Compressive myelopathy due to degenerative spinal changes noted on MRI spine could have contributed partially to the patient's weakness but would not account for the widespread muscle inflammation seen on biopsy and imaging.

Significant laboratory findings included hypokalemia (2.6 mEq/L), anemia (Hb 9.5 g/dL), hypoalbuminemia (2.6 g/dL), elevated RDW (24.9%), and candiduria (budding yeast forms in urine), likely secondary to immunosuppression or underlying systemic inflammation. The normal Coombs test ruled out immune-mediated hemolysis. Although ANA and ANCA were negative, the strong PL-7 positivity confirmed the autoimmune nature of the disease.

Interestingly, the patient also had gastrointestinal complications (gastric ulcers and duodenitis) on upper GI endoscopy, possibly related to stress, chronic steroid use, or systemic inflammation. USG abdomen showing grade 2 fatty liver suggested metabolic comorbidities which could further impact the disease course.

Overall, this case highlights the challenges in diagnosing and managing PL-7 positive ASS, especially when complicated by opportunistic infections, metabolic derangements, and multi-organ involvement. Early recognition and a multidisciplinary approach involving rheumatology, pulmonology, neurology, and infectious diseases are essential for optimal outcomes.

IV. CONCLUSION

This case underscores the importance of considering Anti-Synthetase Syndrome (particularly non-Jo-1 variants like PL-7) in patients presenting with inflammatory myopathy, interstitial lung involvement, and systemic features such as Raynaud phenomenon and mechanic's hands. Necrotizing myopathy with superimposed suspected pulmonary infection, candiduria, and electrolyte imbalances significantly worsened the patient's prognosis.

The combination of HRCT findings, positive PL-7 antibodies, muscle biopsy features, and clinical presentation confirmed the diagnosis. A prompt multidisciplinary approach, careful infection screening, judicious use of immunosuppressive therapy, and supportive care (especially for hypokalemia and nutritional deficiencies) are vital. Prognosis largely depends on the severity of lung involvement, the patient's response to therapy, and early management of infections and systemic complications.

Early diagnosis and aggressive management can improve functional outcomes, reduce morbidity, and potentially prevent life-threatening complications in PL-7 positive Anti-Synthetase Syndrome.[4]

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