



ANTI-SRP ANTIBODY–ASSOCIATED IMMUNE-MEDIATED NECROTIZING MYOPATHY PRESENTING WITH SEVERE QUADRI-PARESIS AND DYSPHAGIA: A CASE REPORT

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ABSTRACT

Background: Immune-mediated necrotizing myopathy (IMNM) is an uncommon autoimmune muscle disorder characterized by progressive muscle weakness and marked elevation of serum creatine kinase. Anti-signal recognition particle (SRP) antibodies identify an important subtype that may present with severe proximal weakness and substantial functional disability. **Case Presentation:** A 65-year-old woman presented with approximately two months of progressively worsening weakness involving all four limbs, associated with dysphagia. Her weakness progressively impaired mobility and resulted in wheelchair dependence. Neurological examination demonstrated severe symmetrical, predominantly proximal weakness, with power approximately 3/5 in the upper limbs and 2/5 in the lower limbs. Deep tendon reflexes were preserved and no significant sensory deficit was documented. CK/CPK was approximately 6887 U/L, with elevated ESR and CRP. Whole-body MRI for myositis screening demonstrated STIR hyperintense signal/edema involving bilateral shoulder-girdle muscles, posterior paraspinal muscles, bilateral gluteal, piriformis and obturator muscles, and bilateral thigh muscles, supporting active inflammatory muscle involvement. Myositis immunoblot showed strong anti-SRP positivity (+++), with additional anti-Ku (++), anti-Ro52 (++), and weak anti-Mi-2 α/β positivity; antisynthetase antibodies including Jo-1, PL-7, PL-12, EJ and OJ were negative. The overall clinical, biochemical, radiological and serological findings were consistent with anti-SRP antibody-associated IMNM. **Treatment and Outcome:** The patient received intravenous methylprednisolone 1 g as an initial dose, followed by 500 mg intravenously once daily for five days. After five days of therapy, limb power improved to approximately 4/5. She was discharged on oral prednisolone (Omnacortil) 40 mg once daily for five days followed by 20 mg once daily for ten days, with advice for regular follow-up. **Conclusion:** Anti-SRP-associated IMNM should be considered in patients with rapidly progressive proximal weakness, dysphagia and markedly elevated CK. Muscle MRI and myositis-specific antibody testing provide important diagnostic support. Early recognition and treatment may result in meaningful improvement in muscle strength and functional status.

KEYWORDS

Anti-SRP Antibody; Immune-Mediated Necrotizing Myopathy; Inflammatory Myositis; Proximal Weakness; Dysphagia; Quadripareisis.

1. INTRODUCTION

Idiopathic inflammatory myopathies comprise a heterogeneous group of autoimmune disorders characterized primarily by skeletal muscle weakness, elevated muscle enzymes and variable extramuscular manifestations. Immune-mediated necrotizing myopathy (IMNM) is a distinct subgroup characterized by prominent myofiber necrosis and regeneration, usually with relatively little inflammatory-cell infiltration on muscle biopsy. Clinically, it is notable for acute, subacute or rapidly progressive symmetrical proximal muscle weakness and often marked elevation of serum creatine kinase.

Anti-signal recognition particle (anti-SRP) and anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (anti-HMGCR) antibodies define important IMNM phenotypes. Anti-SRP-positive myopathy is particularly associated with severe muscle involvement and substantial functional limitation. Dysphagia may occur and is clinically important because it can contribute to aspiration and nutritional compromise.

Diagnosis requires integration of clinical findings, muscle enzymes, electromyography and/or muscle MRI, myositis-specific serology, and muscle biopsy when indicated. MRI commonly demonstrates muscle edema and helps identify active muscle involvement. Early recognition is important because untreated IMNM may result in persistent weakness and disability.

We report a 65-year-old woman with severe progressive quadripareisis and dysphagia, marked CK elevation, characteristic multifocal muscle edema on MRI and strong anti-SRP antibody positivity, with early improvement following high-dose intravenous corticosteroid therapy.

2. Case Presentation

A 65-year-old female presented with approximately two months of progressive weakness involving all four limbs. The weakness was predominantly proximal and progressively interfered with mobility and activities of daily living. Her functional status deteriorated to wheelchair dependence. She also developed difficulty in swallowing.

Neurological examination revealed severe symmetrical, predominantly proximal limb weakness. Motor power was approximately 3/5 in the upper limbs and 2/5 in the lower limbs at presentation. Deep tendon reflexes were preserved at approximately 2+. No significant sensory deficit was documented. The pattern of weakness, preserved reflexes and absence of a prominent sensory component favored a primary myopathic process.

No focal central nervous system deficit was documented in the available records. The clinical presentation prompted evaluation for inflammatory myopathy.

3. Investigations

Laboratory evaluation revealed a markedly elevated CK/CPK level of approximately 6887 U/L, with elevated ESR and CRP. Exact ESR and CRP values were not available in the supplied records.

Whole-body MRI for myositis screening demonstrated subtle STIR hyperintense signal involving muscles of both shoulder girdles; posterior paraspinal muscles in the dorsolumbar region, more prominent in the lumbar region; and ill-defined STIR hyperintense signal involving bilateral gluteal, piriformis and obturator muscles and bilateral thigh muscles. The radiological impression was multifocal muscle edema suggestive of inflammatory myositis.

Myositis immunoblot demonstrated strong anti-SRP positivity (+++; intensity 133), with anti-Ku (++), anti-Ro52 (++), and weak anti-Mi-2 α/β positivity. Anti-TIF1 γ , MDA5, NXP2, SAE1, PM-Scl100, PM-Scl75, Jo-1, PL-7, PL-12, EJ and OJ were negative. Thus, the dominant serological finding was strong anti-SRP positivity and the panel did not support antisynthetase syndrome.

Abdominal ultrasonography was largely unremarkable apart from a few mildly enlarged lymph nodes in the left epigastric region; this finding was nonspecific.

4. Diagnostic Assessment

The combination of subacute progressive symmetrical proximal weakness, severe functional impairment, dysphagia, markedly elevated CK, multifocal muscle edema on MRI and strong anti-SRP antibody positivity supported a diagnosis of anti-SRP antibody-associated immune-mediated necrotizing myopathy (IMNM).

The negative antisynthetase antibodies, including Jo-1, PL-7, PL-12, EJ and OJ, argue against classifying the patient as having antisynthetase syndrome. Although weak anti-Mi-2 α/β reactivity was present, characteristic dermatomyositis skin manifestations were not documented in the supplied clinical information. A muscle biopsy report was not provided; therefore, the diagnosis is presented on the basis of the clinical, biochemical, MRI and serological phenotype.

5. Treatment

The patient was treated with high-dose intravenous corticosteroid

therapy. She received methylprednisolone (MPS) 1 g intravenously as an initial dose, followed by 500 mg intravenously once daily for five days.

After five days of therapy, a clinically meaningful improvement in muscle strength was observed, with limb power improving from approximately 3/5 in the upper limbs and 2/5 in the lower limbs to approximately 4/5.

At discharge, she was prescribed oral prednisolone (Omnacortil) 40 mg once daily for five days, followed by 20 mg once daily for ten days, and was advised regular follow-up.

6. Outcome and Follow-Up

The patient showed early neurological improvement following high-dose corticosteroid therapy. Limb power improved to approximately 4/5 after five days. She was discharged in improved condition on oral corticosteroids with advice for regular follow-up. Long-term follow-up data, including serial CK levels, functional recovery, relapse status and the need for steroid-sparing immunosuppression, were not available at the time of manuscript preparation.

7. DISCUSSION

Anti-SRP-associated IMNM is an uncommon autoimmune myopathy characterized by prominent muscle involvement. Patients commonly present with acute or subacute symmetrical proximal weakness and markedly elevated CK levels. Anti-SRP-positive disease may be particularly severe and can cause substantial disability.

In this patient, severe proximal quadriparesis, dysphagia and marked CK elevation raised strong suspicion of inflammatory myopathy. MRI demonstrated multifocal STIR hyperintensity and muscle edema involving the shoulder and pelvic girdles, thighs and paraspinal musculature, providing supportive evidence of active muscle disease.

The most significant serological finding was strong anti-SRP positivity. Anti-SRP is closely associated with IMNM and helps distinguish this phenotype from antisynthetase syndrome. In this patient, Jo-1 and other tested antisynthetase antibodies were negative. Additional anti-Ku and anti-Ro52 positivity represented accompanying antibody reactivity, while the dominant phenotype remained anti-SRP-associated IMNM.

High-dose corticosteroids are commonly used for severe autoimmune inflammatory myopathy, often followed by steroid-sparing immunosuppression depending on disease severity and response. Anti-SRP disease can require intensive and prolonged immunotherapy, and refractory cases may require additional agents such as intravenous immunoglobulin or rituximab. Close follow-up is important because recovery may be incomplete and relapse can occur.

The early improvement in limb power after intravenous methylprednisolone in this patient highlights the importance of early recognition and treatment. Serial CK measurements and objective functional assessment are important during follow-up to evaluate sustained response and guide corticosteroid tapering.

8. CONCLUSION

Anti-SRP antibody-associated immune-mediated necrotizing myopathy is an important cause of severe, rapidly progressive proximal muscle weakness. In patients with quadriparesis, dysphagia and markedly elevated CK, inflammatory myopathy should be considered even when the initial presentation resembles a neurological disorder. In this case, the combination of severe proximal weakness, preserved reflexes, high CK, characteristic MRI muscle edema and strong anti-SRP positivity supported the diagnosis. Early high-dose corticosteroid therapy was associated with significant short-term improvement in muscle power.

9. Learning Points

- Severe subacute symmetrical proximal weakness with markedly elevated CK should prompt evaluation for autoimmune inflammatory myopathy.
- Dysphagia can be an important manifestation of severe inflammatory muscle disease and should be actively assessed.
- Muscle MRI demonstrating multifocal STIR hyperintensity and edema provides supportive evidence of active myositis.
- Strong anti-SRP positivity is highly relevant to anti-SRP-

associated IMNM and should not be mislabeled as antisynthetase syndrome.

- Early immunosuppressive treatment may improve muscle strength, but long-term follow-up is required.

10. Declarations

Patient Consent: Written informed consent for publication, including clinical details and relevant investigation images, should be obtained from the patient or an authorized representative before journal submission. This should be confirmed before submission.

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