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A CASE REPORT AND ROLE OF INTRAVENOUS IMMUNOGLOBULIN IN THE TREATMENT OF IMMUNE - MEDIATED NECROTIZING MYOSITIS

Nguyen Manh Linh[1]
Vu Thu Trang[1]*, Vu Thi Mai Huyen[1],
Tran Le Linh Trang[1], Nguyen Thi Phuong Nhung[1]
Tran Thi Mai[1], Nguyen Lan Anh[1]

ABSTRACT

Objective: To report a rare case of idiopathic autoimmune myopathy that was successfully managed with intravenous immunoglobulin (IVIG) therapy.

Methods: We described the clinical course, diagnostic workup, and therapeutic response of a patient definitively diagnosed with IMNM at the 108 Military Central Hospital.

Results: A 62-year-old male presented with symmetrical proximal muscle weakness and markedly elevated creatine kinase (CK) levels (> 9,000 U/L). Serological testing was positive for anti-signal recognition particle (anti-SRP) antibodies, and muscle biopsy confirmed myofibre necrosis consistent with IMNM. Initially, the patient was treated with high-dose corticosteroids combined with mycophenolate mofetil, which resulted in poor response and progressive worsening of muscle weakness. Subsequent adjunctive therapy with IVIG (2 g/kg body weight was administered over 5 days, repeated after one month), along with optimized immunosuppression resulted in significant clinical improvement in muscle strength, restored mobility, CK levels and no significant adverse effects were observed during the treatment period.

Conclusion: Intravenous immunoglobulin is an effective and well-tolerated therapeutic intervention for immune-mediated necrotizing myopathy, particularly in cases refractory to standard treatment.

Keywords: Immune mediated necrotizing myopathy, intravenous immunoglobulin, myositis.

Corresponding author: Vu Thu Trang, Email: bsvtrang0807@gmail.com

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[1]*Military Central Hospital 108*

1. INTRODUCTION

Immune-mediated necrotizing myositis (IMNM) is a distinct subset of idiopathic inflammatory myopathies (IIMs), characterized by severe, symmetrical proximal muscle weakness and significantly elevated serum creatine kinase (CK) levels [1]. IMNM is categorized into three main serological subtypes based on specific autoantibodies: anti-signal recognition particle (anti-SRP) positive, anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (anti-HMGCR) positive, and seronegative IMNM [2]. Among these, anti-SRP-positive IMNM is often associated with a more aggressive clinical course, severe muscle necrosis on biopsy, and a higher resistance to standard immunosuppressive therapies [1, 2]. In addition to muscle damage, extramuscular involvements may also occur, increasing morbidity and mortality rates [1].

In clinical practice, managing IMNM remains challenging. While high-dose corticosteroids combined with steroid-sparing agents are the first-line treatment, a substantial proportion of patients fail to achieve remission or experience relapses during steroid tapering. Intravenous immunoglobulin (IVIG) has emerged as a highly effective adjunctive therapy for refractory IMNM, particularly in the anti-SRP subgroup. Current evidence, however, is largely limited to case series and individual reports. In Vietnam, reports on anti-SRP-positive IMNM remain scarce, potentially due to underdiagnosis or limited access to specialized antibody testing. This hinders early diagnosis and the selection of optimal treatment protocols.

We reported a case of anti-SRP and anti-Ro52 positive IMNM associated with interstitial lung disease, which was refractory to initial immunosuppression but showed a remarkable response to IVIG.

2. CASE PRESENTATION

- Patient profile: A 62-year-old male was admitted to the Military Central Hospital 108 with a diagnosis of IMNM (anti-SRP and anti-Ro52 positive) and concomitant interstitial lung disease (ILD).

- Medical history: Chronic gastritis.

- Presenting illness: Nine months prior to admission, the patient experienced fatigue and weight loss. One week before admission, he developed mild muscle pain in the proximal limbs, followed by progressive weakness.

- Physical examination: On admission, the patient was conscious with stable vital signs. No fever, dyspnea, or dysphagia was noted. Neuromuscular examination revealed normal muscle bulk but reduced proximal muscle strength (Upper limbs: 3-4/5; Lower limbs: 3/5). Distal muscle strength and deep tendon reflexes were preserved.

- Diagnostic workup:

+ Laboratory Investigations: Significantly elevated muscle enzymes: CK = 9,272 U/L, CK-MB = 368.8 U/L, AST = 375 U/L, and ALT = 240 U/L.

+ Immunology: Myositis-specific antibody panel was strongly positive for anti-SRP and anti-Ro52, which was pivotal for the diagnosis.

+ Electromyography (EMG): Findings were consistent with an autoimmune-mediated myogenic process.

+ Muscle biopsy: Histopathology showed predominant myofiber necrosis and degeneration with sparse inflammatory cell infiltration (Figure 1), confirming the diagnosis of IMNM.

+ Imaging & pulmonary function: HRCT of the chest revealed non-homogeneous interstitial thickening in both lungs. Pulmonary function testing showed mild restrictive ventilatory defects, consistent with ILD.

- Diagnosis: IMNM (anti-SRP and anti-Ro52 positive) with associated Interstitial Lung Disease.

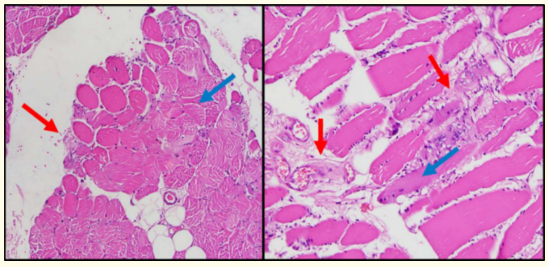

Figure 1. Muscle histopathology. The specimen reveals degenerating muscle fibers (blue arrows), extensive myofiber necrosis and atrophy, accompanied by scattered lymphocytic infiltration (red arrows).

- Therapeutic Intervention and Clinical Course:

+ Phase 1 (Initial): High-dose corticosteroids (Methylprednisolone 500 mg/day for 3 days) followed by Mycophenolate sodium. During this period, the patient's condition deteriorated; proximal muscle strength decreased to 1-2/5, and he became unable to perform basic activities of daily living. After one month, clinical response was minimal.

+ Phase 2 (Escalation): The patient received intensified therapy with IVIG at a total dose of 2 g/kg (1 g/kg/day over 5 days), repeated after one month. Concurrently, the immunosuppressive regimen was optimized with a combination of MMF (2 g/day) and Tacrolimus (4 mg/day).

- Outcome: Following two cycles of IVIG and optimized immunosuppression, the patient showed significant improvement in muscle strength and resumed normal daily activities. Laboratory markers (CK, LDH, and liver enzymes) nearly normalized (CK = 200 UI/L, LDH = 188 UI/L, AST = 23.9 UI/L, ALT = 20.8 UI/L) (Figure 2). The patient was discharged with a long-term outpatient maintenance plan. No common side effects (headache, fever, rash) or serious adverse events (anaphylaxis, acute renal failure, thromboembolism) were reported during IVIG administration.

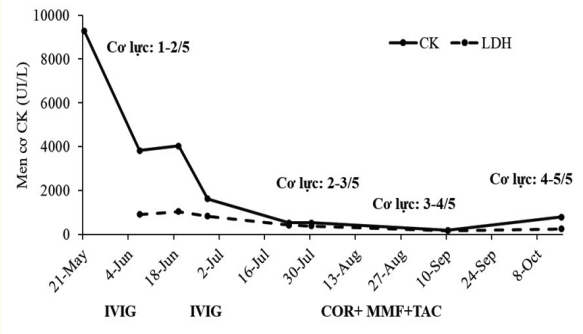

Figure 2. Integrated therapeutic protocol and longitudinal clinical/laboratory monitoring. The chart illustrates the clinical progression of muscle strength in correlation with the reduction of serum muscle enzymes (CK and LDH) following IVIG administration and optimized immunosuppressive therapy.

3. DISCUSSION

This case represents a classic presentation of anti-SRP-positive IMNM, characterized by a severe, rapidly progressive course and resistance to conventional therapy. Currently, standardized diagnostic criteria for IMNM are still evolving, but the diagnosis in this case was firmly established based

on proximal muscle weakness, markedly elevated CK, and the presence of anti-SRP and anti-Ro52 antibodies. The presence of anti-SRP is not only a diagnostic hallmark but also a poor prognostic indicator, often associated with steroid resistance [1, 2]. While the independent prognostic role of anti-Ro52 in IMNM remains to be fully elucidated, it is frequently linked to extramuscular manifestations, particularly ILD, as observed in our patient [3].

Initial therapy with corticosteroids and MMF is appropriate for IMNM with ILD; however, it was insufficient in this case. The progression of muscle weakness and the development of dysphagia-a critical "red flag" in myositis-necessitated early treatment escalation [4, 5]. Dysphagia significantly increases the risk of aspiration pneumonia, a life-threatening complication. In refractory cases, IVIG is a highly effective second-line therapy capable of inducing rapid clinical response.

The mechanism of IVIG in autoimmune diseases is multifaceted [6]: (1) blockade of Fc receptors on macrophages, reducing phagocytosis of antibody-coated myofibers; (2) inhibition of the complement cascade (binding C3b/C4b), preventing the formation of the membrane attack complex; (3) modulation of the cytokine network by decreasing pro-inflammatory and increasing anti-inflammatory cytokines; (4) neutralization of pathogenic autoantibodies via anti-idiotypic antibodies; and (5) expansion of regulatory T cells (Tregs). These actions collectively attenuate muscle inflammation and necrosis.

The multicenter study conducted by Allenbach et al. [7] provided robust evidence regarding the high therapeutic efficacy of intravenous immunoglobulin (IVIG) in patients with IMNM, specifically those harboring anti-SRP and anti-HMGCR autoantibodies. Their findings demonstrated that IVIG significantly bolsters muscle strength and facilitates a substantial reduction in serum creatine kinase (CK) levels, particularly in a high percentage of patients who remained refractory to conventional corticosteroid therapy. Furthermore, a comprehensive systematic review published in 2021 [8] corroborated that the integration of IVIG into the management of idiopathic inflammatory myopathies (IIMs) yields superior disease control, more rapid symptomatic resolution, and a markedly enhanced quality of life compared to standard immunosuppressive monotherapy. A pivotal clinical advantage of IVIG is its rapid onset of action, which allows for the immediate mitigation of acute inflammatory processes and catastrophic myogenic destruction. This rapid intervention serves as a critical pharmacological bridge, providing the necessary window for slow-acting immunosuppressive agents, such as mycophenolate mofetil (MMF) or tacrolimus, to reach their full therapeutic steady-state. Research indicates that while IVIG generally maintains a favorable safety profile with high tolerability, it is associated with a spectrum of adverse events ranging from mild constitutional symptoms to rare, life-threatening complications. The most prevalent side effects, including pyrexia, cephalalgia, and rigors, have been reported in 3% to 41% of cases across various cohorts. More severe but infrequent complications, such as thromboembolic events, acute renal impairment, and aseptic meningitis (AMS), represent significant clinical risks, typically occurring in patients with pre-existing risk factors or those receiving high-dose regimens. These risks can be effectively mitigated through clinical strategies including slow infusion rates, aggressive pre-procedural hydration, and the administration of prophylactic premedication [8].

Following the initial induction of remission, a robust triple-therapy maintenance regimen consisting of corticosteroids, mycophenolate mofetil, and tacrolimus is considered an essential therapeutic strategy for severe IMNM, particularly the aggressive anti-SRP subtype. Mycophenolate mofetil, an established cornerstone of immunosuppressive therapy, exerts its effects by selectively inhibiting lymphocyte proliferation and suppressing the synthesis of pathogenic autoantibodies [9]. Complementing this, tacrolimus-a potent calcineurin inhibitor-effectively hampers T-cell activation and has been documented to achieve significant clinical responses in refractory myositis cases, especially when utilized as part of a multi-drug combinatorial approach (10). The integration of these three agents creates a potent synergistic effect which, when coupled with the immunomodulatory properties of IVIG, targets multiple discrete facets of the autoimmune pathogenesis of IMNM. Consequently, this multi-targeted approach optimizes long-term disease stabilization, facilitates a more rapid and successful tapering of maintenance corticosteroids, and minimizes the debilitating morbidity associated with prolonged steroid exposure.

In spite of not being a first-line therapy in all guidelines, IVIG is strongly recommended for refractory cases or life-threatening presentations (e.g., respiratory muscle weakness or severe dysphagia) [11]. Biological agents like Rituximab also show promise in anti-SRP IMNM [12], but

in the Vietnamese context, cost and infection risks remain significant barriers. Therefore, the timely integration of IVIG with traditional immunosuppressants represents a rational and effective strategy.

4. CONCLUSION

Intravenous immunoglobulin (IVIG) provides rapid and significant therapeutic benefits in immune-mediated necrotizing myositis, particularly in patients refractory to standard immunosuppressive regimens. Throughout the treatment of this case, IVIG was well-tolerated with no significant adverse events. This case reinforces that when indicated and administered correctly, IVIG is a safe and highly effective option that significantly improves the prognosis of IMNM.

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