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Case Report

A wolf in sheep's clothing: myositis mimicking motor neuron disease – a case report

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ABSTRACT

Idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of immune-mediated muscle disorders typically presenting with proximal muscle weakness and elevated creatine kinase levels. Atypical presentations, particularly in elderly individuals, may mimic motor neuron disease (MND), creating significant diagnostic challenges. We report a 72-year-old man who presented with a six-month history of progressive weakness beginning in the proximal lower limbs and later involving distal limbs and bulbar muscles. Examination revealed muscle wasting, spasticity, exaggerated reflexes, polyminimyoclonus, and neck flexor weakness, suggesting a mixed upper and lower motor neuron syndrome. Nerve conduction studies were normal, while electromyography demonstrated an irritable myopathic pattern without diffuse neurogenic denervation. Serum creatine kinase levels were normal. Autoimmune evaluation showed anti-thyroid peroxidase positivity, SSA/Ro52 positivity, rheumatoid factor positivity, hypocomplementemia, inflammatory polyarthritis, renal involvement, and interstitial lung disease. Whole-body muscle magnetic resonance imaging demonstrated diffuse symmetrical inflammatory changes, while FDG-PET CT excluded occult malignancy. The patient was treated with intravenous methylprednisolone followed by oral prednisolone and mycophenolate mofetil, resulting in marked clinical improvement. Idiopathic inflammatory myositis may rarely present with upper and lower motor neuron-like signs and mimic MND. Recognition of systemic autoimmune features, appropriate electrophysiological evaluation, and muscle imaging is essential to avoid misdiagnosis of a potentially treatable condition.

Keywords: Idiopathic inflammatory myositis, Motor neuron disease mimic, Amyotrophic lateral sclerosis, Inflammatory myopathy, Overlap myositis

INTRODUCTION

Idiopathic inflammatory myopathies (IIMs) are immune-mediated skeletal muscle disorders characterized by muscle inflammation and varying degrees of systemic involvement.[1–3] Subtypes include dermatomyositis, polymyositis, immune-mediated necrotizing myopathy, inclusion body myositis, and overlap myositis.[2,4]

Classically, IIM presents with subacute symmetrical proximal muscle weakness and elevated serum creatine kinase (CK) levels.[1,5] However, atypical presentations, particularly in elderly individuals, may include distal weakness, chronic progression, bulbar symptoms, and even normal CK levels.[6,7] These features may closely resemble motor neuron disease (MND), especially amyotrophic lateral sclerosis (ALS), a neurodegenerative disorder characterized by progressive upper and lower motor neuron degeneration.[8]

Misdiagnosing inflammatory myopathy as ALS has significant clinical implications because inflammatory myopathies are potentially treatable, whereas ALS carries a poor prognosis and limited therapeutic options.[9,10] We report a patient with idiopathic inflammatory myositis

presenting with combined upper and lower motor neuron-like features, clinically mimicking MND.

CASE REPORT

A 72-year-old male presented with progressive weakness for six months. The weakness initially involved the proximal lower limbs, resulting in difficulty rising from a squatting position and climbing stairs. Over time, distal lower limb weakness developed, followed by involvement of both proximal and distal upper limbs. He reported difficulty turning in bed and performing fine motor tasks.

Subsequently, he developed dysphagia predominantly to solids and hoarseness of voice. Neck flexor weakness was also noted. There was no history of sensory symptoms, bowel or bladder dysfunction, cognitive impairment, or behavioral changes.

In addition to neuromuscular symptoms, he reported inflammatory polyarthralgia, bilateral pedal edema, frothy urine, and increased urinary frequency, suggesting systemic involvement.

Neurological examination revealed normal higher mental function, cranial nerve examination with proximal-predominant muscle wasting, increased tone in all four limbs, diffuse exaggerated deep tendon reflexes including trapezius reflex ansent jaw jerk, mute plantar responses, polyminimyoclonus of the outstretched hands, neck and truncal muscle weakness with proximal more than distal weakness in bilateral upper limbs and lower limbs. The combination of progressive weakness, muscle wasting, hyperreflexia, spasticity, and bulbar symptoms initially raised strong suspicion for motor neuron disease, particularly ALS.[8]

Laboratory investigations demonstrated a normal CBC, RFT, Serum electrolytes, liver function test with hypoalbuminemia, normal serum CK level, hypothyroidism-TSH-41, elevated anti-thyroid peroxidase antibodies-78 (ref-30), anti-thyroglobulin-773 (ref 100), positive SSA/Ro52 antibodies, positive rheumatoid factor, low complement C3 levels, and proteinuria.

Nerve conduction studies were normal. Electromyography demonstrated short-duration, low-amplitude polyphasic motor unit potentials with early recruitment, consistent with an inflammatory myopathy. There was no evidence of widespread chronic neurogenic denervation typically seen in ALS.[11]

MRI of the brain and cervical spine showed no corticospinal tract pathology. Whole-body muscle MRI demonstrated diffuse symmetrical STIR hyperintensities involving multiple muscle groups, suggestive of inflammatory myopathy.[12] CT chest revealed interstitial lung disease. FDG-PET CT excluded occult malignancy.The constellation of systemic autoimmune manifestations, characteristic MRI findings, and myopathic electromyographic abnormalities supported the diagnosis of idiopathic inflammatory myositis with overlap autoimmune features.

Based on the clinical, electrophysiological, serological, and radiological findings, a diagnosis of idiopathic inflammatory myositis with systemic autoimmune involvement was established. The patient was treated with intravenous methylprednisolone 1 g daily for five consecutive days. This was followed by oral prednisolone 40 mg daily. Mycophenolate mofetil was initiated at a dose of 500 mg twice daily as a steroid-sparing immunosuppressive agent. The patient demonstrated significant clinical improvement following treatment. Muscle strength improved substantially, with marked recovery of proximal limb and neck flexor weakness. Dysphagia and hoarseness also improved considerably.

At follow-up, he was able to walk independently without support, rise from a chair unassisted, and perform overhead activities that had previously been difficult. The favorable response to immunosuppressive therapy strongly supported an inflammatory autoimmune etiology rather than a neurodegenerative motor neuron disorder. The patient continues to undergo regular follow-up with assessment of muscle strength, functional status, pulmonary involvement, renal parameters, and treatment-related adverse effects.

DISCUSSION

This case highlights an unusual presentation of inflammatory myopathy mimicking motor neuron disease. The coexistence of progressive weakness, muscle wasting, hyperreflexia, spasticity, polyminimyoclonus and bulbar symptoms initially suggested ALS.[8] However, several clinical and investigative findings argued against a diagnosis of MND.

First, the presence of systemic autoimmune manifestations, including inflammatory polyarthritis, hypocomplementemia, proteinuria, interstitial lung disease, SSA/Ro52 positivity, rheumatoid factor positivity and anti-TPO positivity, strongly suggested a multisystem immune-mediated disorder rather than isolated motor neuron degeneration.[2,4,13] Anti-TPO positivity indicated autoimmune thyroiditis and supported an underlying autoimmune diathesis. Autoimmune clustering is well recognized in overlap myositis syndromes.[2,4] Second, electrophysiological findings were inconsistent with ALS. Classical ALS demonstrates widespread chronic neurogenic motor unit potentials with evidence of active and chronic denervation across multiple body regions.[11] In contrast, our patient demonstrated a myopathic pattern with early recruitment and absence of diffuse neurogenic abnormalities. Third, muscle MRI provided important diagnostic information. Diffuse symmetrical STIR hyperintensities are characteristic of active inflammatory myopathy and are not typical of ALS.[12] MRI has emerged as a valuable tool in differentiating inflammatory

myopathies from neurodegenerative disorders and in identifying muscles suitable for biopsy.

An additional diagnostic challenge in this case was the normal CK level. Although elevated CK is considered a hallmark of inflammatory myopathies, normal values do not exclude the diagnosis. Chronic disease, overlap syndromes, and certain inflammatory myopathies may present with normal or minimally elevated CK levels.[11] The dramatic clinical response to corticosteroids and mycophenolate mofetil further supported an inflammatory etiology. Such improvement would be highly unusual in ALS and underscores the importance of identifying treatable MND mimics.[9,14,10] This case emphasizes the need for a comprehensive diagnostic approach when evaluating patients presenting with apparent upper and lower motor neuron syndromes. Careful assessment of systemic features, autoantibody profiles, electrophysiological findings, and muscle imaging can prevent misdiagnosis and facilitate appropriate therapy.

CONCLUSION

Idiopathic inflammatory myositis can rarely present with combined upper and lower motor neuron-like features and closely mimic motor neuron disease. The presence of systemic autoimmune manifestations, characteristic muscle MRI abnormalities, and myopathic electromyographic findings should prompt consideration of inflammatory myopathy even when serum CK levels are normal. Early recognition is essential because timely immunosuppressive treatment can result in substantial clinical improvement and prevent unnecessary diagnostic labeling as a neurodegenerative motor neuron disorder.

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REFERENCES

1. Dalakas MC. Inflammatory muscle diseases. N Engl J Med. 2015;372(18):1734-47.
2. Lundberg IE, de Visser M, Werth VP. Classification of myositis. Nat Rev Rheumatol. 2018;14(5):269-78.
3. Dimachkie MM, Barohn RJ. Idiopathic inflammatory myopathies. Semin Neurol. 2012;32(3):227-36.
4. Allenbach Y, Benveniste O, Stenzel W, Boyer O. Immune-mediated necrotizing myopathy: clinical features and pathogenesis. Nat Rev Rheumatol. 2020;16(12):689-701.
5. Greenberg SA. Inflammatory myopathies: evaluation and management. Semin Neurol. 2008;28(2):241-9.
6. Needham M, Mastaglia FL. Inclusion body myositis: current pathogenetic concepts and diagnostic and therapeutic approaches. Lancet Neurol. 2007;6(7):620-31.
7. Lundberg IE, Miller FW, Tjärnlund A, Bottai M. Diagnosis and classification of idiopathic inflammatory myopathies. J Intern Med. 2016;280(1):39-51.
8. Brooks BR, Miller RG, Swash M, Munsat TL. El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. Amyotroph Lateral Scler Other Motor Neuron Disord. 2000;1(5):293-9.
9. Turner MR, Talbot K. Mimics and chameleons in motor neurone disease. Pract Neurol. 2013;13(3):153-64.
10. Aitchison C, Romanowski C, Barker A, Jenkins T. Motor neuron disease mimics in practice: a case series. BMJ Case Rep. 2022;15(7).
11. de Carvalho M. Electrodiagnosis of amyotrophic lateral sclerosis: a review of existing guidelines. J Clin Neurophysiol. 2020;37(4):294-8.
12. Malartre S, Bachasson D, Mercy G, Sarkis E, Anquetil C, Benveniste O, et al. MRI and muscle imaging for idiopathic inflammatory myopathies. Brain Pathol. 2021;31(3):e12954.
13. Marie I, Hatron PY, Dominique S, Cherin P, Mouthon L, Menard JF. Short-term and long-term outcomes of interstitial lung disease in polymyositis and dermatomyositis: a series of 107 patients. Arthritis Rheum. 2011;63(11):3439-47.
14. Je Y, Park YE, Shin YB. Differentiating inclusion body myositis from amyotrophic lateral sclerosis based on the features of dysphagia: insights from a patient with rapidly progressive dysphagia. J Clin Neurol. 2025;21(1):83-5.

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