RESEARCH PAPER

Dermatomyositis with Acute Motor-Sensory Axonal Neuropathy Variant of Guillain-Barré Syndrome: A Rare Coexistence

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ABSTRACT

Dermatomyositis is a rare inflammatory disease characterized by muscle weakness and distinctive skin rashes.[1,2] It is a type of autoimmune condition where the immune system attacks skin and muscle tissue, often causing purple/red rashes on eyelids (heliotrope rash) and knuckles (Gottron papules). Guillain-Barré Syndrome (GBS) is an acute immune-mediated polyradiculoneuropathy characterized by rapidly progressive ascending weakness, areflexia, and variable sensory involvement. Guillain-Barré Syndrome Coexistence of dermatomyositis with Guillain-Barré syndrome is exceedingly rare and may pose significant diagnostic challenges due to overlapping clinical manifestations.[3,4]

A 33-year-old male presented with generalized body pains for one month, facial puffiness for 25 days, and progressive weakness for 15 days.

This case highlights the rare coexistence of dermatomyositis and the AMSAN variant of Guillain-Barré syndrome presenting as a diagnostic overlap syndrome. Atypical clinical manifestations may initially mimic primary inflammatory myopathies, resulting in diagnostic uncertainty. Early electrophysiological evaluation, detailed neurological examination, and multidisciplinary assessment are essential for accurate diagnosis and timely initiation of immunotherapy, which can substantially improve clinical outcomes.[5,6]

Keywords: Guillain-Barré Syndrome, Acute Motor-Sensory Axonal Neuropathy, Dermatomyositis, Inflammatory Myopathy.

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INTRODUCTION

Dermatomyositis is an idiopathic inflammatory myopathy characterized by proximal muscle weakness, elevated muscle enzymes, and characteristic cutaneous manifestations.[1,2]

The global incidence of Guillain-Barré syndrome is estimated to be 1–2 cases per 100,000 persons per year, with a slightly higher prevalence in adult males.[4] In many patients, the syndrome follows an infectious illness such as gastroenteritis caused by Campylobacter jejuni, or viral infections including Cytomegalovirus infection, Epstein–Barr virus infection, and COVID-19.[3,5] Clinically, patients typically present with symmetrical ascending weakness, reduced or absent deep tendon reflexes, sensory symptoms, and autonomic dysfunction. Several variants of GBS have been described, including Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN), and Miller Fisher Syndrome.[5,7,8]

Although peripheral neuropathy may occasionally coexist with inflammatory myopathies, overlap with Guillain-Barré syndrome, particularly the AMSAN variant, is exceedingly rare and sparsely reported in literature.[2]

The prevalence of dermatomyositis is approximately 1.1 per 100,000 population, whereas Guillain-Barré syndrome has an incidence of approximately 1–2 per 100,000 population annually.[1,4] The coexistence of dermatomyositis and the Acute Motor-Sensory Axonal Neuropathy (AMSAN) variant of Guillain–Barré syndrome is extremely rare. To date, no population-level prevalence data are available, and the association has been described only sporadically in case reports.[2]

CASE PRESENTATION

A 33-year-old male presented with generalized body pains for one month, facial puffiness for 25 days, and progressive weakness for 15 days. Weakness initially involved the proximal muscles of the lower limbs and subsequently progressed to involve the distal lower limbs and upper limbs. The patient also developed difficulty in squatting, climbing stairs, swallowing solids, and opening the mouth.

Initial clinical evaluation favoured inflammatory myopathy with possible angioedema. On neurological examination, higher mental functions and cranial nerve examination were normal. Motor system examination revealed symmetrical proximal predominant weakness involving both lower and upper limbs with reduced muscle power. Deep tendon reflexes were diminished in the upper limbs and absent at bilateral knee and ankle jerks, suggestive of lower motor neuron involvement. Plantar responses were bilaterally flexor. Sensory examination was largely unremarkable, with preserved superficial and deep sensations, no sensory level, and no bowel or bladder involvement, thereby favouring a predominantly motor neuropathic process.

Laboratory investigations demonstrated markedly elevated serum creatine kinase levels (4369 IU/L), suggestive of active muscle injury.[1] ANA profile extended panel was negative for anti-dsDNA, nucleosomes, histones, Ro-52, SS-A, SS-B, Sm/RNP, Mi-2α, Mi-2β, CENP-A, CENP-B, Sp-100, and PML antibodies.

Myositis antibody profile demonstrated strong positivity for transcription intermediary factor 1 gamma (TIF1γ) and PM-Scl antibodies, along with positivity for nuclear matrix protein 2 (NXP2) antibodies. The panel showed strong positivity for TIF1γ (band intensity 114), PM-Scl75 (band intensity 91), and PM-Scl100 (band intensity 69), with positive NXP2 antibodies (band intensity 11), while antibodies against Jo-1, Mi-2, SRP, Ku, PL-7, PL-12, EJ, OJ, SAE1, and MDA5 were negative. These findings were suggestive of associated autoimmune inflammatory myopathy with overlap features.[9]

Nerve conduction studies revealed findings consistent with acute motor-sensory axonal neuropathy (AMSAN) variant of Guillain-Barré syndrome, characterized by reduced compound muscle action potential amplitudes with evidence of motor axonal involvement and associated sensory nerve involvement. Generalized hyporeflexia with absent lower limb reflexes further supported peripheral neuropathic involvement.[5,7,8]

MRI pelvis with screening of bilateral thighs demonstrated T2/STIR hyperintensities involving bilateral anterior, anteromedial, and posterior compartment muscles of the thighs, including rectus femoris, vastus lateralis, intermedius and medialis, adductor group, and hamstring muscles, with mild subcutaneous oedema, suggestive of inflammatory myositis.

Magnetic resonance imaging of the whole spine and Thigh was normal, excluding compressive or structural aetiologies.

The patient was treated with intravenous immunoglobulin (IVIG) therapy followed by corticosteroids, resulting in significant clinical improvement with improvement in muscle strength and functional status. Early recognition of this rare neurological overlap syndrome and prompt

initiation of immunotherapy were crucial in achieving favourable clinical outcomes.[6,8]

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying clinical details.

Conflict of Interest

The authors declare no conflict of interest.

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