



INSIGHTS ON MULTIFOCAL MOTOR NEUROPATHY : A CASE SERIES OF 3 PATIENTS

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ABSTRACT Multifocal motor neuropathy (MMN) is a rare immune-mediated motor neuropathy characterized by asymmetric weakness that preferentially affects distal upper limb muscles. The diagnosis of MMN is based on clinical, laboratory and electrophysiological characteristics demonstrating conduction block . Steroids and therapeutic plasma exchange are ineffective in MMN and may lead to worsening of the disease. However, more than 80% of patients with MMN experience improvement after IVIG. This article emphasizes the importance of early recognition and that appropriate treatment can have positive impact on quality of life.

KEYWORDS :

INTRODUCTION:

Multifocal motor neuropathy (MMN) is an acquired , immune mediated demyelinating motor neuropathy . It was first described in 1982 by Parry and Clark but the term MMN, its association with anti-GM1 IgM and its response to immunotherapy was described in 1988 by Pestronk and associates. The estimated prevalence is less than 1 per 100000 population .^[1]

The ratio of prevalence in male:female is about 3:1 with onset in fourth to fifth decade of life^[2]. It presents clinically as progressive asymmetric pure motor weakness involving the distal upper limb more commonly. It is characterised by partial motor conduction block . Here we report a case series of 3 patients who were diagnosed as MMN and treated promptly with immunotherapy .

CASE PRESENTATION :

Case 1 is a 42 years old male who presented with pure motor distal followed by proximal left lower limb weakness since 6 months . It was followed by right distal upper limb weakness since 2 months . Examination revealed power of : 1/5 in right distal lower limb , 3/5 in right proximal lower limb with wrist drop and weak hand grip . NCS-EMG was suggestive of conduction block in the right radial nerve. Serum IgM Ganglioside panel was negative . He was treated with I.V immunoglobulin (IVIg) 2gm/kg over 5 days and showed significant improvement from mRS 3 to mRS 1 .

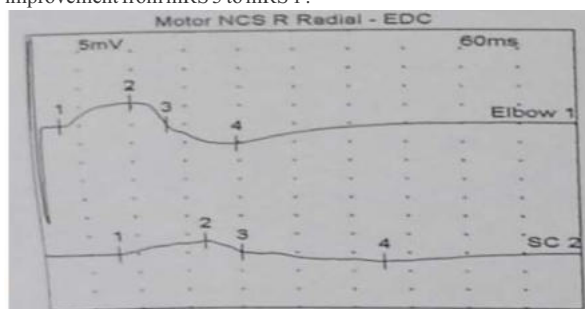


Figure 1: NCS demonstrating conduction block in right radial nerve in case 1 .



Figure 2 : Right sided foot drop in case 1 .

Case 2 is a 34 years old male who presented with pure motor distal right lower limb weakness with wasting since 1 year followed by left upper limb distal weakness since 2 months . Examination showed power : right distal lower limb - 3/5 and weakness in the left upper limb in the form of wrist drop . NCS EMG revealed conduction block in the left radial and right common peroneal nerves. Serum IgM Ganglioside panel was positive. Patient was treated with Injection IVIg 2gm/kg over 5 days. There was significant improvement in mRS from 3 at admission to mRS of 1 at discharge .

Ganglioside IgM		
	Observed Value	Reference Range
GM1	Positive,20	Negative
GM2	Positive,37	Negative
GM3	Positive,16	Negative
GD1a	Negative, 7	Negative
GD1b	Positive,30	Negative
GT1b	Negative,0	Negative
GQ1b	Positive,20	Negative

Figure 3 : Serum IgM Ganglioside Panel of case 2.

Case 3 is a 59 years old female who presented with bilateral sequential distal upper limb weakness in the form of wrist drops since 4 months . NCS - EMG revealed conduction blocks in both radial nerves . Serum IgM Ganglioside panel was positive . She received IVIG 2gm/kg over 5 days with clinical stabilisation into the course of her weakness.



Figure 4 : Bilateral wrist drop in case 3 .

DISCUSSION

Multifocal motor neuropathy (MMN) most commonly affects the distal upper limbs and is characterized by asymmetric weakness along the distribution of the individual nerves, unaccompanied by pain or sensory loss. Two of the above three patients had onset of symptoms in the lower limbs , a finding that can be present in about 20%^[3] of patients . There can be aggravation of the weakness in about 83 %^[4] of patients on cold exposure . The key distinguishing electrophysiologic feature of MMN is conduction block at noncompressible sites are present in 2 or more nerve distributions, then a diagnosis of MMN can be strongly considered^[5] . As MMN is a rare disease, it mimics other diseases, such as amyotrophic lateral sclerosis, chronic inflammatory demyelinating polyradiculoneuropathy, hereditary neuropathy with liability to pressure palsy, inclusion body myositis, focal neuropathies, and radiculopathies, which may be suspected before the identification of MMN. Approximately 50% of MMN patients test positive on a ganglioside panel, with the majority of positive results showing antibodies against the GM1 ganglioside, specifically as IgM antibodies as is noticed in two of our three patients^[6] .

The pattern of weakness, absence of sensory findings or pain, and conduction block at noncompressible sites are key features that can raise suspicion of MMN. Early Intravenous immunoglobulin is the only therapy with proven efficiency in MMN patients by providing improvement of muscle strength as noticed in two of the three patients .

Declaration Of Patient Consent :

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial Support And Sponsorship: Nil

Conflicts Of Interest

There are no conflicts of interest

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