Steroid-responsive multifocal motor neuropathy with cranial manifestations – a case report

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Abstract

The typical presentation of multifocal motor neuropathy (MMN) is progressive asymmetric limb weakness. Cranial neuropathy is rare. We report a 28-year-old woman with cranial and bulbar palsies but with typical electrophysiological features of MMN by multifocal motor conduction blocks and serological markers of anti-ganglioside GM1 antibodies. The previous consensus on the treatment of MMN is intravenous immunoglobulins, but our patient responded to oral steroids and had clinical and electrophysiological improvement under continuous low-dose prednisolone treatment.

In summary, MMN is a treatable chronic inflammatory disease of peripheral nerves. Cranial neuropathies can be its initial presentations. Electromyography studies are crucial for MMN diagnosis and helpful in monitoring disease activity and treatment responses. Although the previous guideline did not suggest using steroids for MMN, with careful patient selection, low-dose oral steroids can be an effective treatment in patients with relatively minor symptoms.

Keywords: **Multifocal motor neuropathy, conduction block, bulbar palsy, cranial nerve, cranial neuropathy.**

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BACKGROUND

Multifocal motor neuropathy (MMN) is an immune-mediated neuropathy associated with anti-ganglioside GM1 IgM antibodies [(1)]. Its clinical manifestations are a slow progressive asymmetric weakness with distal predominant involvement, absence of sensory loss, and lack of upper motor neuron signs. The electrophysiological hallmarks of MMN are persistent, multifocal, partial motor conduction blocks (PMCBs), temporal dispersion, decreased motor nerve conduction velocity (MNCV), delayed distal motor latency, and prolonged F-wave latency of the nerves other than the common sites of nerve compressions, together with normal sensory nerve conduction studies [(2-4)]. In addition, the improvement or disappearance of PMCBs can parallel clinical improvement [(2,5-7)]. Therefore, nerve conduction studies and electromyography (NCS/EMG) is the crucial diagnostic tool for MMN. Meanwhile, serological studies of anti-ganglioside antibodies help identify MMN.

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As early as 1988, Pestronk reported two patients with asymmetric weakness, conduction blocks, and antibodies to GM1 ganglioside [8]. Later, studies of MMN showed the prevalence of 30 to 60% of anti-GM1 IgM antibodies [4, 9]. Identification of anti-GM1 IgM in blood help confirm MMN. Here, we present a case of MMN with atypical cranial manifestations but with typical NCS/EMG features and positive serum anti-GM1 IgM antibodies and responded to oral steroid treatment.

CASE PRESENTATION

Clinical scenario

A 28-year-old female had a history of acute left oculomotor palsy four years ago. This time, she came to our hospital for subacute onset dysphagia and dysarthria without limb weakness or numbness. Her deep tendon reflexes were normal for all limbs. There were no fasciculations or cramps. NCS/EMG studies showed multiple focal demyelinating motor neuropathy with conduction blocks and prolonged F-waves (Table 1). The cerebrospinal fluid (CSF) study was essentially normal, with white blood cells of 0 cell/μl and a total protein of 39.7 mg/dl. Magnetic resonance imaging (MRI) of the brain did not find any lesions. Protein electrophoresis of blood showed no paraprotein. Next, tumor surveillance by image and serum tumor markers did not find a tumor but slightly elevated CA-125 (111.4 U/mL).

She was initially diagnosed as acute inflammatory demyelinating polyradiculoneuropathy (AIDP) and treated with one cycle of double filtration plasmapheresis (DFPP). However, clinical response was limited. Later, anti-ganglioside testing showed positive results of anti-GM1 IgM antibodies in her serum. In addition, conduction blocks were exacerbated in the three-month follow-up of NCS/EMG studies (Table 1). These findings led to the final diagnosis of MMN. Because the bulbar symptoms showed no clinical deterioration from the initial encounter to the three-month follow-up, she was treated with oral prednisolone, and her dysphagia and dysarthria improved. Maintenance with low-dose prednisolone therapy (15mg every other day) continued after symptom improvement. The follow-up NCS/EMG studies in the sixth month, one year, and two years from the initial treatment showed reversal of conduction block and recovery of conduction velocity (Table 1 and Figure 1).

The studies involving human participants were reviewed and approved by the institutional review board of Chang Gung Medical Foundation, with approval number 201700701A3. Written informed consent to participate in this study was provided by the participant.

Table 1. Serial electromyographic studies

(A) Motor conduction studies

	Initial	3 month	6 month	1 year	2 year
CMAP amplitude (distal/proximal, mV)					
Left median	9.1/3.8	10.4/1.7	10.5/4	9.1/6.2	10.6/8.2
Right median	8.3/4	9.4/2.2	10.3/4.2	10.8/8.1	10.5/8
Left ulnar	3.6/1.8	3.9/1.2	5.4/3.3	7.2/4.2	8.2/5.2
Right ulnar	10.5/5.8	9.6/4.6	12.3/9	12/10.2	12.7/11.9
Left peroneal	3.7/2.9	5.8/3.7	4.9/3.6	5.3/3.9	5.8/4.2
Right peroneal	5.8/4.9	8.2/5.6	7.5/6.6	7.3/6.3	6.1/5.7
Left tibial	8.7/2.4	8.8/2.2	10/3.8	10.5/4.8	13.1/4.5
Right tibial	5.7/1.1	4.6/0.3	7.5/1.1	10.9/2	11.2/5.2
Motor conduction velocity(m/s)					
Left median	30	22	26	37	41
Right median	44	46	44	46	48
Left ulnar	39	30	35	41	47
Right ulnar	45	44	42	47	49

Table 1. Serial electromyographic studies (continue)

	Initial	3 month	6 month	1 year	2 year
Left peroneal	38	38	39	41	43
Right peroneal	43	40	41	42	47
Left tibial	40	32	35	39	40
Right tibial	33	30	37	36	40
Distal motor latency (ms)					
Left median	2.6	2.8	2.9	2.5	2.5
Right median	2.5	3	3	2.5	2.5
Left ulnar	2.6	2.4	2.6	2	2
Right ulnar	2.1	2.4	2.6	2.1	2.1
Left peroneal	4.3	4.5	4.9	3.7	3.7
Right peroneal	3.3	3.6	3.8	2.8	2.8
Left tibial	4.4	4.1	4.2	3	3
Right tibial	4.9	4.6	5.7	3.1	3.1
F-wave latency (ms)					
Left median	44.4	46.3	46	28.5	30.2
Right median	N/A	N/A	N/A	N/A	27.1
Left ulnar	34.8	40.8	36.9	27.2	27.8
Right ulnar	N/A	N/A	N/A	N/A	25.7
Left peroneal	47.4	46.8	48.4	45.6	55.8
Right peroneal	N/A	N/A	N/A	N/A	40.9
Left tibial	48	52.3	48.8	46.3	45.1
Right tibial	N/A	N/A	N/A	46.3	36.5
H reflex					
Left tibial	30.6	NR	30.7	27.8	27.4
Right tibial	29.2	NR	30.9	28	27.1

(B) Sensory conduction studies

	Initial	3 month	6 month	1 year	2 year
Distal SNAP amplitude (mV)					
Left median	79	75	89	96.7	135.8
Right median	75	N/A	61	85.6	101.5
Left ulnar	47	72	77	72	103
Right ulnar	55	N/A	74	89.7	86.7
Left sural	N/A	18	13	22.1	22.2
Right sural	14	17	13	23.2	26.2
Sensory conduction velocity (m/s)					
Left median	62	70	70	65	63
Right median	64	N/A	61	67	66
Left ulnar	64	60	62	61	62
Right ulnar	56	N/A	62	63	65

No spontaneous activity was recorded in the electromyography at initial encounter, 3-month, 6-month, 1-year, and 2-year studies. Abbreviations: CMAP, compound muscle action potential. SNAP, sensory nerve action potential. N/A, not available. NR, no response.

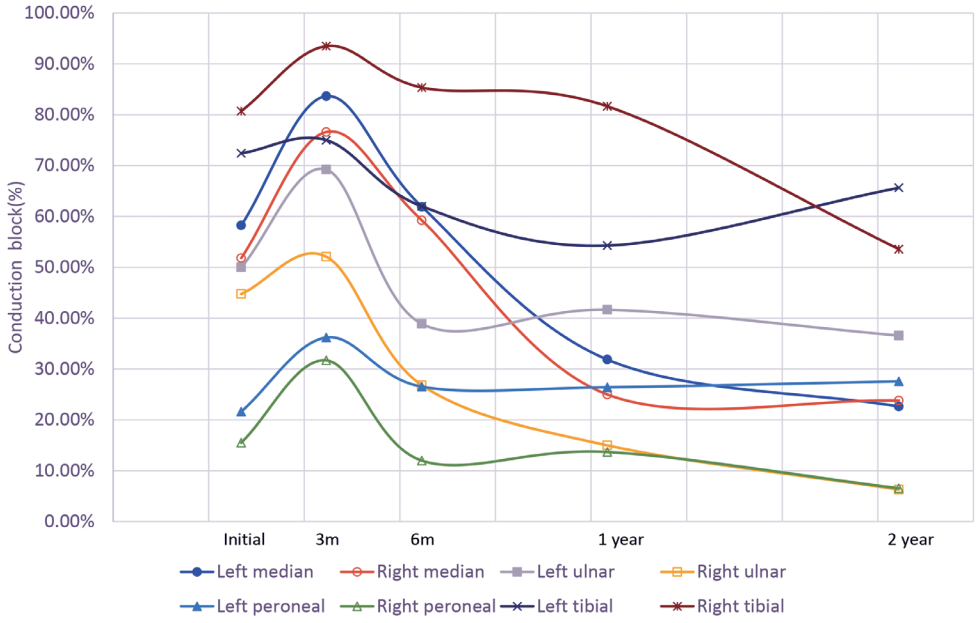

Fig. 1. Degree of conduction blocks in serial electromyographic studies
The severity of the conduction blocks of each nerve was quantified in the degree of conduction block (%), which was the percentage of loss of amplitude of the proximal CMAP relative to the distal CMAP. That was, conduction block (%) = (1 - proximal CMAP amplitude/distal CMAP amplitude) x 100%.
At the patient's initial admission, the nerve conduction studies showed multiple conduction blocks. Therefore, the patient received one cycle of plasmapheresis under the tentative impression of AIDP. However, conduction locks aggravated in all tested nerves and reached peaks in the third-month follow-up studies. Therefore, we started oral prednisolone therapy for the patient. In the sixth-month follow-up, all tests showed improvement in conduction blocks, and the conduction returned to normal at the right peroneal nerve. At one year, nerve conductions kept improving in bilateral median nerves and right ulnar nerve. Later in the second-year follow-up, the degree of conduction blocks was kept stationary in all tested nerves.

DISCUSSION AND CONCLUSIONS

Clinical manifestation of MMN

Cranial, bulbar, and respiratory muscle involvement are rare but possible in MMN. Four years ago, our patient experienced an episode of oculomotor palsy, but the diagnosis remained idiopathic because no extensive NCS/EMG study had been done. Four years later, dysphagia and dysarthria developed without typical limb weakness, but comprehensive NCS/EMG and serological studies led to the diagnosis of MMN.

In the literature review, cranial nerve manifestations were rare but possible in MMN. For example, Kaji presented two cases of MMN with cranial involvement of the hypoglossal nerve [10]. Galassi also reported two MMN cases with unilateral hypoglossal and abducens nerve palsies [11]. In addition, Pringle introduced a 45-year-old man who presented with bilateral abducens nerve palsies, facial diplegia, left trochlear nerve palsy, impaired taste, dysarthria, bilateral sternocleidomastoid and trapezius muscles weakness, and areflexia of limbs. Comprehensive electrophysiological studies revealed characteristic features of MMN in this patient [12]. Although clinical diagnostic criteria for MMN in the guideline of the European Federation of Neurological Societies and the Peripheral Nerve Society listed the absence of cranial nerve involvement in the supportive criteria, it footnoted the exceptional cases [3, 13]. Therefore, the literature review and our case study highlight the possibilities of cranial nerve involvement in MMN. In our case, oculomotor,

glossopharyngeal, and vagus nerves were involved.

Anti-ganglioside antibodies in MMN

Gangliosides are a family of sialic acid-containing glycosphingolipids with polymorphic structures and functions and are widely distributed in the nervous system. The functions of gangliosides include protecting nerves from immune attacks [14], being the composition of the node of Ranvier [15], and modulating the function of ion channels [16, 17]. However, due to molecular mimicry, gangliosides can become targets of autoimmune reactions and lead to demyelinating diseases [18].

The ganglioside GM1 is abundantly located in the white matter of the brain [16], spinal cord, and peripheral motor and sensory nerves [19]. In addition, GM1 is present in all 12 cranial nerves in humans [20]. Its wide distribution explains the various clinical disorders related to anti-GM1 antibodies, including Guillain-Barres syndrome [21] and Alzheimer's disease [22, 23]. Anti-GM1 antibodies in MMN [24] showed positive correlations with more severe weakness, more disability, and more axon loss, and negative correlations with muscle powers by Medical Research Council scores [9]. In addition, the distribution of GM1 gangliosides included the cranial nerves. The immunocomplex formation of anti-GM1 antibodies and GM1 gangliosides on the cranial nerves can result in cranial nerve injury and present as cranial nerve palsies, as reported in the previous case studies and this case report [10].

Besides anti-GM1 antibodies, several other autoantibodies have been associated with MMN. For example, anti-GM2 IgM and anti-GD1b IgM are estimated to be present in 6 and 9% of MMN patients [9]. Antibodies against myelin-associated-protein (MAG) [25] and antibodies targeting neurofascin-186 and glimodin, a cell adhesion molecule at node of Ranvier of axon and its ligand at Schwann cell, are linked to MMN [26]. The antibodies bind on the neuronal surface, induce complement activation via the classical pathway, disrupt sodium channel clusters, and result in the characteristic conduction blocks [21, 27, 28]. In addition, the higher the complement activity, the more severe the muscle weakness and axonal loss in the patients with MMN [28].

Electrophysiologic features of MMN

The electrophysiologic features of MMN are motor conduction blocks, temporal dispersion, slow MNCV, delayed distal motor latencies, and prolonged F responses, but sparing of sensory nerves. However, other demyelinating neuropathies also have similar characteristics and need careful differentiation. For example, AIDP has conduction blocks and slow MNCV. However, the conduction block in AIDP is a single-peaked event and usually improves or disappears after treatment. Besides, prolonged demyelinating also characterizes chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), but it usually affects some sensory nerves and is rarely restricted to motor involvement. In addition, albumin-cytological dissociation with elevated protein levels and normal total nucleated cell counts are common in AIDP and CIDP but are not characteristic of MMN. Another differential diagnosis of asymmetric pure motor weakness is motor neuron disease. However, conduction blocks of motor nerves are not for the case of motor neuron disease. Accordingly, unremarkable CSF biochemistry and persistent pure motor demyelinating conduction blocks of selected nerves in our patient weighted the diagnosis of MMN from other differential diagnoses.

Conduction blocks are crucial for MMN diagnosis and also a disease course. The reversal of conduction blocks is associated with clinical improvement [5-7]. However, the changes in conduction blocks during treatment are complex regarding asymmetricity of proximal and distal sites, different nerves, and individual responses to immunotherapy. Paradoxical relapse of conduction blocks may be observed during treatment, and Cappellari et al. proposed eight models representing various situations of conduction blocks and their changes during treatment [6]. According to the model theory, an in-depth understanding of the pathophysiology of MMN and overall observations of the dynamic electrophysiological changes is essential for adequately evaluating conduction blocks.

Treatment of MMN

MMN is a chronic, treatable immune-mediated demyelinating neuropathy. The previous consensus stated intravenous immunoglobulin (IVIG) for the choice of treatment for MMN [3, 13]. IVIG can bring clinical and electrophysiological stabilization and improvement after persistent treatments, although the effects decay after

some time and requires maintenance doses [5, 29]. In a double-blind, placebo-controlled study, IVIG showed its ability to control MMN disease activity [30]. More recently, subcutaneous immunoglobulin (SCIG) has become another patient-friendly treatment choice of MMN [31, 32].

Therapeutic apheresis, including plasma exchange and plasmapheresis, is considered ineffective or exacerbating for MMN because of antibody and cytokine rebound after apheresis [33, 34]. Therefore, therapeutic aphesis is not recommended in chronic immune-mediated neuropathy, like CIDP [35] and MMN [36-38]. The initial failure of DFPP treatment under the tentative impression of AIDP in our patient could be related to this situation and led to the following exacerbation of conduction blocks in the three-month follow-up.

In contrast, glucocorticoids, the steroids used in controlling immune-mediated diseases, have potent immunosuppressive and anti-inflammatory effects with complex mechanisms on multiple immune reactions. Glucocorticoids suppress almost all immune cells, innate immunity, and adapted immunity. The number of circulating monocytes and macrophages are reduced, synthesis of pro-inflammatory cytokines is decreased, circulating T cells and B cells are lowered, leukocyte differentiation is altered, and vessel permeability and adhesion molecules are suppressed [39, 40]. Intravenous and oral steroids are widely used in controlling chronic immune-mediated neurological diseases. For example, they are the treatment of CIDP and inflammatory myopathies, maintenance therapy for myasthenia gravis, first-line treatment of autoimmune encephalitis, and initial treatment for acute attacks of neuromyelitis optic and multiple sclerosis [41].

In the earliest records of MMN in 1982, Lewis treated five patients with multifocal demyelinating neuropathy, and two patients who took steroids improved [42]. However, controversial responses to steroids appeared in the following case reports. Some patients did not respond to steroids [43], and some patients' weakness appeared or deteriorated after using steroids [44]. A systematic review by Nobile-Orazio in 2001 collected over 60 patients from case reports and accounted for a steroid-response rate of 11% of the patients [4]. The guideline for MMN, which recommends against the use of steroids, was established based on these case studies [13]. Not negligible, certain MMN patients were beneficial from steroids while others required IVIG to have clinical improvement. In our patient, her symptoms of bulbar palsies persisted but did not worsen in the first three months; however, NCS/EMG deteriorated with more severe conduction blocks (Figure 1). Weighted the cost-effectiveness of treatments, the risk of allergy to IVIG, and the clinical-electrophysiological dissociation, we used oral steroids for the relatively mild symptoms. After two years of follow-up, low-dose oral prednisolone remained effective in reaching clinical and electrophysiological improvement in this patient. Therefore, we reasoned that steroids could be an alternative agent under careful patient selection.

Conclusions

Cranial neuropathies can be the initial presentations of MMN. For idiopathic isolated cranial neuropathies, an extensive electrophysiologic study seems needed for a more accurate diagnosis. To be noticed, clinical-electrophysiological dissociation with symptoms resolving but persistent conduction blocks could happen after treatment. Continuous following and maintenance of immunotherapy are mandatory for best practice. IVIG is the choice of immunotherapy for MMN, but oral steroids may be an alternative in patients with minor symptoms with persistent conduction blocks.

Availability of data and materials: The raw data supporting the conclusions of this article will be made available by the authors under reasonable request, without undue reservation.

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