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A CASE REPORT OF MULTIFOCAL MOTOR NEUROPATHY WITH CONDUCTION BLOCKS (MMNCB) ON LONG TERM IMMUNOTHERAPY (WITH REVIEW OF LITERATURE)

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ABSTRACT

Multifocal motor neuropathy (MMN) with conduction block is an acquired immune-mediated demyelinating neuropathy with slowly progressive weakness, fasciculations, and cramping, without significant sensory involvement. Clinically, it may resemble ALS with predominant lower motor neuron involvement, but muscle atrophy and more rapid progression are lacking. Duration of disease prior to diagnosis ranges from several months to more than 15 years. Unlike ALS, MMN usually responds to treatment with intravenous immunoglobulin (IVIG) or cyclophosphamide, even after many years of duration. The patient discussed here is a 42- year old male who presented to our OPD as a case of slowly progressive weakness which started in his right foot 9 years back that was followed by weakness of his right hand without any sensory involvement. Patient was evaluated thoroughly and the diagnosis of MULTIFOCAL MOTOR NEUROPATHY WITH CONDUCTION BLOCKS (MMNCB) was made. Patient was treated with IVIG and showed dramatic improvement. Patient has been on IVIGs from last 5 years and is doing well.

KEYWORDS: MMNCB (Multifocal motor neuropathy with conduction block, ALS (Amyotropic lateral sclerosis), IVIG (intravenous immunoglobulin).

INTRODUCTION

Multifocal motor neuropathy (MMN) with conduction block is an acquired immune-mediated demyelinating neuropathy with slowly progressive weakness, fasciculations, and cramping, without significant sensory involvement. MMN is a rare disorder, and its lifetime prevalence is estimated to be 1 case in 100,000 population in the United States.[1] Most patients maintain productive lives despite ongoing symptoms, and up to 94% remain employed. However, gradual progression of symptoms may also lead to significant disability. Fatal outcomes have been reported only rarely, and at least some case reports describe patients with other entities, including motor neuron disease. Rarely, multifocal motor neuropathy may be associated with a B-cell lymphoma producing monoclonal antibodies against GM1 and GD1b myelin glycolipids. MMN is more common in males with a male-to-female ratio of about 3:1[2]. The mean age of onset is 40 years. Eighty percent of patients are aged 20-50 years at presentation. Rarely, children as young as 6 years may be affected. Clinically, it may resemble ALS with predominant lower motor neuron involvement, but muscle atrophy and more rapid progression are lacking. Duration of disease prior to diagnosis ranges from several months to more than 15 years. Unlike ALS, MMN usually responds to treatment with intravenous immunoglobulin (IVIG) or cyclophosphamide, even after many years of duration.

CASE PRESENTATION

A 42 year old male, non-hypertensive, non-diabetic with a history of working as an amalgam filler presented with a history of slowly progressive weakness in right foot in the form of slipping of slippers while walking without any sensory loss or paraesthesias approximately 9 years back. This remained in more or less the same condition until he noticed difficulty in putting right hand in pocket with clawing of medial 2 fingers of right hand almost 1 year after the first complaint. Over the next 2 months he noted difficulty in nail cutting with right hand with occasional difficulty in holding spoon or pen. Over the next one year there was difficulty in gripping objects with left hand. No history of any sensory, bladder/bowel or cranial nerve involvements.

History of occupational mercury exposure present for 15 years. H/o burn injury in both hands present in early childhood which did not cause any weakness but with associated contractures in both hands' 3rd and 4th fingers. No h/o diabetes mellitus or hypertension. No h/o hypothyroidism or any history of malignancies. No h/o any alcohol intake or smoking. No h/o similar illness in the family members.

On examination patient was conscious, well-oriented, pulse rate 74/min, regular and all peripheral pulses were felt, BP of 124/80mmHg, with no postural drop, normal chest, CVS and abdominal examination.

CNS Examination- HMF –normal. Cranial nerves normal. Neck flexors, extensors 5/5.

Motor system: Bulk: B/L clawing of medial 2 fingers of both hands present with some amount of contractures in both hands. Tone: normal.

Power – B/L shoulder: 5/5, elbow: 5/5, b/l wrist: 5/5. B/l adductor pollicis, extensor pollicis brevis weak. In left hand, abductor pollicis brevis were weak. B/l card test positive. In left hand, abductor pollicis brevis and EIP also weak. B/L hip: flexion: 5/5, extension- 5/5, adduction- 5/5, abduction: 5/5. B/L knee; flexion, extension: 5/5. Ankle dorsiflexion: right side: 4/5, left side: 4/5, plantar flexors- right 5/5, left 4/5. Toes: dorsiflexion: right side: 4/5, left side 3/5. Plantar flexion b/l 5/5.

Sensory system: B/L touch, pain and vibration, temperature and proprioception normal.

DTR	BJ	SJ	TJ	KJ	AJ
R	2+	2+	2+	2+	2+
L	2+	2+	2+	2+	2+

Bilateral pectoralis: 2+, Hand flexors absent. Jaw jerk absent
Cerebellar signs: normal
Gait normal

Investigations

Hemogram CBC - normal Coagulation profile- normal Lipid profile- normal **CPK** : 250 **CRP** : 0.6 **HbA1C** : 5.1%	**Biochemistry :** **RBS**: 120mg/Dl **LFT**: normal **KFT**: normal **Ca/PO4** -normal **24 hour urinary mercury level:** <6ug/L (normal) **S. Uric acid** : 8.1mg%	**ECG**- Normal **CXR**- Normal **Vitamin B12 levels** -1032 pg/mL(211-911) **MRI spine:** degenerative disc changes in C4, C5, C6,C7 with reduction in disc space.	**Autoimmune profile** **Anti GM1 IgM -Positive** GM 2- neg GM 3- neg GD 1a -neg Gd 1b – neg GT 1b – neg GQ 1b - neg dsDNA-neg Nucleosome- neg Histone – neg SmDl – negative PCNA- neg PO- neg SS-A/Ro 60- neg SS-A/Ro 52- negative SS-B/La- equivocal CENP-B- neg SCL70 – neg Ul-snRNP- negative AMA M2- negative Jo1 – negative PM-Scl – neg Mi-2 – neg Ku - negative

Nerve conduction studies
Motor Nerve Conduction

Nerve and Site	Latency	Amplitude	Segment	Latency difference	Distance	Conduction velocity
Median R						
Wrist	2.9ms	11.8mV	Abductor pollicis brevis wrist	2.9ms	mm	m/s
Elbow	6.8m/s	11.8mV	Wrist-elbow	3.9ms	200mm	51m/s
Ulnar R						
Wrist	2.5 ms	8.9mV	Wrist below elbow	10.3 ms	mm	m/s
Below elbow	12.8ms	2.9mV	Wrist below elbow	10.3ms	230	22m/s
Median L						
Wrist	2.7ms	8.0mV	Abductor pollicis brevis wrist	2.7ms	mm	m/s
Elbow	6.4ms	7.7mV	Wrist -elbow	3.7ms	220mm	59m/s
Ulnar L						
Wrist	2.5 ms	11.0mV	Wrist -Below elbow	6.1ms	mm	m/s
Below elbow	8.6ms	1.4mV	Wrist -Below elbow	6.1ms	230mm	38m/s
Peroneal L						
Ankle	4.2ms	7.6mV	Extensor digitorum brevis ankle	4.2ms	mm	m/s
Fibula (head)	10.4ms	6.7mV	Ankle – fibula (head)	6.2ms	320mm	52m/s
Tibial L						
Ankle	3.8 ms	8.5mV	Abductor hallucis-ankle	3.8ms	mm	m/s
Popliteal fossa	11.1ms	0.7mV	Ankle popliteal fossa	7.3ms	380mm	52m/s
Peroneal R						
Ankle	4.4ms	6.1mV	Extensor digitorum brevis- ankle	4.4ms	mm	m/s
Fibula (head)	10.4ms	6.1mV	Ankle fibula (head)	6.1ms	310mm	51m/s
Tibial R						
Ankle	3.4ms	12.0mV	Abductor hallucis- ankle	3.4ms	mm	m/s
Popliteal fossa	12.4ms	30.0mV	Ankle – Popliteal fossa	9.0ms	400mm	44m/s
Ulnar. R (inching method)						
Wrist	2.5ms	10.7mV	Wrist-6cm wrist	ms	mm	m/s
6cm Wrist	3.7ms	7.4mV	Wrist- 6cm wrist	ms	mm	m/s
10cm Wrist	6.8ms	6.4mV	6cm Wrist-10cm wrist	ms	mm	m/s
14cm Wrist	8.2ms	5.8mV	Wrist- 14cm wrist	ms	mm	m/s
18cm Wrist	8.6ms	5.2mV	Wrist -18cm wrist	ms	mm	m/s
20cm Wrist	9.5ms	3.5mV		ms	mm	m/s
22cm Wrist	10.1ms	4.9mV		ms	mm	m/s
24cm Wrist	11.0ms	4.8mV		ms	mm	m/s

EMG Summary Table

	Spontaneous					MUAP			Recruitment
	IA	**FIB**	**PSW**	**FASC**	**H.F**	**AMP**	**DUR**	**PPP**	**Pattern**
R. First D Inteross	N	None	None	R	None				
L. Deltoid	N	None	None	NONE	None				
L. Gastroc N(MED)	N	None	None	R	None				

This patient admitted with history of slowly progressive pure motor weakness in right foot followed by both hands with wasting without proximal weakness, pain or bowel/bladder or cranial nerve involvement with NCV s/o conduction blocks and anti GM1 IgM positive, was diagnosed as a case of multifocal motor neuropathy with conduction blocks (MMNCB). Patient was treated with intravenous immunoglobulins, starting with a loading dose of 2g/kg followed by a maintenance dose of 1g/kg. Patient has been doing well on immunotherapy from last 5 years and is on regular follow up. The maintenance dose is administered according to patient's symptoms on in patient basis. Patient improves significantly after receiving IVIGs. He is able to carry out his daily chores and is still employed.

DISCUSSION

MMN is an autoimmune peripheral neuropathy without a known cause. Rarely, MMN may develop following treatment with tumor necrosis factor (TNF) – α antagonists. Rarely, multifocal motor neuropathy may be associated with a B-cell lymphoma producing monoclonal antibodies against GM1 and GD1b myelin glycolipids.

Electrodiagnostic evaluation may document the presence of asymptomatic conduction blocks in other clinically unaffected nerves. Positive serology for anti-GM1 antibodies is supportive of the diagnosis of MMN, particularly higher titers.

Neuroimaging studies are not routinely performed in patients with suspected MMN. Magnetic resonance imaging (MRI) of the brachial plexus may show an increased signal intensity on the T2-weighted images, usually without contrast enhancement. Neuromuscular ultrasound frequently shows enlargement of multiple nerves in patients with MMN. Nerve conduction study (NCV) with needle electromyography (EMG) is essential in demonstrating the presence of multifocal involvement without significant sensory component. When MMN is defined clinically, some patients may not have demonstrable conduction block on conventional NCS. Other signs of demyelination may be present, including decreased velocities, prolonged terminal latencies, temporal dispersion, and delayed (or absent) F waves. Sensory NCS findings are normal, even across the same segments with demonstrated motor conduction block. Additionally, electrodiagnostic evidence of axonal degeneration has been demonstrated in at least one nerve from as many as 50% of patients.

Nerve biopsy is not routinely performed in the evaluation of patients with suspected MMN. Sural nerve biopsy findings may be normal, but findings may also show mild demyelination and poor remyelination in the absence of significant inflammation.

Infusion-related adverse effects are less common with SCIG than with IVIG, and SCIG may be given at home by the patient and his or her family. Long-term IVIG treatment improves muscle strength and functional disability, but the responsiveness may decrease over time. If IVIG is not (sufficiently) effective, then alternative treatments (eg, cyclophosphamide, rituximab, cyclosporine) should be considered. Intravenous immunoglobulin neutralizes circulating myelin antibodies through anti-idiotypic antibodies. It down-regulates proinflammatory cytokines, including INF-gamma.

Most complications are related to treatment. IVIG can lead to aseptic meningitis, thromboembolic events, and kidney failure; cyclophosphamide can lead to myelosuppression, hemorrhagic cystitis, and bladder carcinoma.

Prognosis is usually good, and 70-80% of patients respond to treatment. Even in patients who do not respond to therapy, weakness is only slowly progressive and up to 94% of patients remain employed.

More than 20 years ago Roth et al.[1] reported a patient with chronic asymmetric, distal motor neuropathy without sensory loss. Electrophysiological examination revealed proximal multifocal persistent conduction blocks (CBs) outside the common entrapment sites. Soon afterwards, others described individuals with similar characteristics.[2,3] The term 'multifocal motor neuropathy' (MMN) was coined in 1988 by Pestronk et al.[4] who first recognized the association of MMN with anti-GM1-IgM antibodies and the responsiveness to immune-modulating therapies. Since then, systematic clinical and electrophysiological evaluation of larger patient cohorts increased our pathophysiological understanding of MMN and paved the way for more effective treatments.[5,6,7,8,9,10] Especially the successful application of intravenous immunoglobulins (IVIgs) marked a cornerstone in MMN therapy and is nowadays regarded as the gold standard.[10,11,12,13,14,15,16] More recently, diagnostic criteria for this rare neuropathy have been proposed by various European and American neurological associations[17,18] which help to delineate MMN from other neuropathies such as chronic inflammatory demyelinating polyneuropathy (CIDP) or multifocal acquired demyelinating sensory and motor (MADSAM) neuropathy (Lewis-Sumner syndrome) and motor neuron disease (MND).

Although MMN has meanwhile been identified as a distinct nosological entity and significant success has been made in elucidating important aspects of the disease, several issues remain to be clarified. For example, there are still unsettled questions concerning the etiology of MMN, the biological basis of CBs as well as the optimum long-term therapy.[8,19,20,21]

Therapy

Because MMN is supposed to be an immune-mediated disease, various immunomodulatory treatment strategies have been applied to date in MMN patients. In contrast to CIDP and Lewis-Sumner syndrome, numerous studies have demonstrated that corticosteroids and plasma exchange are ineffective in MMN. In fact, they even worsen the symptoms in up to 20% of MMN patients, underlining that different pathophysiological mechanisms must be functional.[3,4,16,85,125,126] Nowadays, IVIgs are regarded as first-line therapy and their efficacy in MMN has meanwhile been proven in 4 large double-blind, placebo-controlled trials.[17,23,63,127,128,129] In addition, 2 retrospective trials confirmed that IVIg is initially effective in 70–86% of the patients by most individuals require periodic treatment for several years.[130,131] Whether the subcutaneous route of IVIg administration is advantageous compared to regular intravenous infusions with respect to steady IVIg plasma concentrations, patients' quality of life or cost-effectiveness needs further evaluation.[132,133,134] Similar to other neurological disorders, the exact mechanism of action of IVIg in MMN is still unclear at present.[135] This also applies for the question whether patients with high titers of anti-GM1 antibodies respond better to IVIg compared to those with lower titers.[7,25,95] The clinical effect of IVIg is usually impressive and muscle strength improves substantially within the first week of treatment.

Otherwise, the diagnosis should be reconsidered, although chronic paresis and muscle atrophy do not recover after IVIg application in most of the cases. While anti-GM1 antibody titers are not affected by IVIg and thus, are not suitable as therapeutic markers, disappearance of partial CB sometimes parallels clinical improvement.[136,137,138] The common IVIg dose at the beginning of the disease is 2 g/kg body weight given on 2–5 consecutive days. However, the treatment effect usually rapidly declines after several weeks. Therefore, it is important to find an applicable maintenance regime with individualized IVIg doses (e.g. 0.4 g/kg IVIg once weekly or 1–2 g/kg IVIg in monthly intervals) in order to optimize the cost-to-benefit ratio.[62,139] Nevertheless the efficacy of IVIg decreases after several years of treatment in most of the patients, necessitating higher dosage or shortened infusion intervals to stabilize the symptoms.[94,140] The recent observation that higher doses of IVIg might be superior already at the initial stage[141] and be able to prevent secondary axonal degeneration or promote remyelination[142] needs to be confirmed in larger studies and valid data on the long-term efficacy of IVIg in MMN are missing.

Soon after the initial description of MMN, cyclophosphamide was tested for this indication in several small uncontrolled trials. Taken together, high doses of cyclophosphamide seem to have a moderate effect, especially when given intravenously while lower oral doses could not influence disease progression.[4,12,26,143] Brannagan et al.[144] recently reported a patient with refractory MMN who experienced sustained disease remission after high-dose cyclophosphamide (50 mg/kg body weight over 4 days) without stem cell rescue. In contrast, myeloablative cyclophosphamide followed by autologous stem cell transplantation worsened the symptoms in another patient.[145] Hence, further studies are clearly needed to finally judge the therapeutic potential of aggressive immunosuppressive regimens in MMN. Given its problematic risk-to-benefit ratio, cyclophosphamide is currently only recommended if IVIg is not sufficiently effective.[17]

Many other immunomodulatory or immunosuppressive agents such as azathioprine, methotrexate, cyclosporin A, mycophenolate mofetil or β-interferons have occasionally been tested in MMN but in most cases revealed conflicting results and controlled trials on these substances are missing.[3,146,147,148,149] Data concerning the efficacy of the monoclonal antibody rituximab, which targets the CD20 molecule on B cells and might be able to reduce pathological autoantibody levels in MMN, are likewise inconclusive and larger trials are needed.[150,151,152]

CONCLUSION

During the past 20 years numerous clinical and electrophysiological studies have helped to shed light on the pathophysiology of MMN and led to significant advances in its diagnosis and treatment. IVIg can restore muscle strength and delay disease progression. However, therapies with proven long-term efficacy or even strategies able to cure the disease are still lacking underlining the need to continue the search for innovative treatment approaches. Although MMN is typically characterized by CB most likely caused by focal demyelination, the pathophysiological abnormalities probably extend far beyond. Novel morphological and electrophysiological findings highlight the importance of axonal degeneration and impaired axon–myelin interactions, which probably occur already at early stages of MMN. Finally, anti-GM1 antibodies seem to represent a valid diagnostic marker rather than the true trigger of the disease and other possible targets of the immune response in MMN await identification.

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