Case report

Rapidly progressive multifocal motor neuropathy with a dramatic response to therapy

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Abstract

We report a young female with multifocal motor neuropathy (MMN) who had unusually rapid progression involving both distal and proximal weakness, which responded dramatically to treatment, without any relapses up to now.

Introduction

Multifocal motor neuropathy with conduction blocks is an acquired immune-mediated demyelinating neuropathy with slowly progressive weakness, fasciculations, and cramping, without significant sensory involvement. MMN is generally a slowly progressive illness with predominant distal paresis.

Clinically, it may resemble chronic inflammatory demyelinating polyneuropathy (CIDP) and in advanced cases, motor neuron disease (MND) with predominant lower motor neuron involvement.

The course and the management of MMN are different from those of CIDP and MND. Therefore, early and correct diagnosis is important to prevent disability.

A typical case of MMN was first documented in Sri Lanka in 1998[1].

Case report

A 25-year old female, diagnosed to have diabetes mellitus eight months back was admitted to Teaching Hospital Karapitiya, with progressive weakness and paresthesia of hands for six months. At the onset, she had weakness in fingers of the left hand followed by weakness and numbness in both hands over five months. Her day-to-day activities were severely compromised due to the illness. There was no proximal muscle weakness in the upper limbs, muscle cramps, or muscle twitching.

Two months after the onset of the disease, she had noticed difficulty in getting up from squatting position and climbing stairs, with more marked weakness on left lower limb. There was no history suggestive of cranial nerves or bulbar involvement. She denied any history of preceding febrile illness, diarrhoea, or exposure to toxic substances including heavy metals. She had no family history of similar disease.

On examination, she was not pale and had an average built. Cranial nerves and optic fundi were normal. Examination of the upper limbs revealed bilateral wrist drop (Figure 1) with no evidence of muscle wasting or fasciculations. There was reduced muscle power in all the small muscles of the hands. She had muscle power of grade 2 on the left hand and grade 3 on the right. Proximal muscle power of the upper limbs was normal. Deep tendon reflexes in upper limbs were diminished with no objective sensory loss.

Figure 1. Bilateral wrist drop.

In the lower limbs, proximal muscle power was grade 2 on the left side and grade 3 on the right. Power of the distal muscles of the lower limbs was normal. She had diminished deep tendon reflexes in lower limbs with flexor plantar response and normal sensation. The rest of the neurological examination was normal. Cardiovascular, respiratory, and abdominal examination were unremarkable.

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Table 1. Nerve conduction study

Motor nerve conduction study

Site	*Latency (ms)*	*Amplitude*	*Area*	*Segment*	*Distance (ms)*	*Interval (ms)*	*NCV (m/s)*	*NCV N.D.*
Median, R								
Wrist	3.96ms	6.00mV	15.11mVms	Wrist		3.96ms		
Elbow	10.05ms	342.00uV	956.40uVms	Wrist – Elbow	190mm	6.09ms	31.2m/s	
Ulnar, R								
Wrist	2.76ms	8.74mV	18.88mVms	Wrist – Midforearm		2.76ms		
Elbow	5.58ms	3.29mV	7.55mVms	Midforearm – below elbow	90mm	2.82ms	31.9m/s	
Axilla	8.7ms	1.80mV	5.42mVms	Below elbow – Above elbow	115mm	3.12ms	36.9m/s	
Site 4	9.72ms	1.70mV	5.43Vms	Axilla	60mm	1.02ms	58.8m/s	

F–wave study

Nerve	*Stim.site*	*F-Lat*	*F-Lat. N.D.*	*M.Lat.*	*F-M Lat*	*F-occur.*	*Distance*	*FWCV*	*N.D.*
Median R	Wrist					0/16.0%			
Ulnar R	Wrist	2.75ms				0/16.0%			

Sensory nerve conduction study

Site	*Latency*	*Amplitude*	*Area*	*Segment*	*Distance (mm)*	*Interval (ms)*	*NCV (m/s)*	*NCV N.D*
Ulnar, R								
Wrist	3.16ms	18.40uV	0.00uVms	Wrist	125mm	3.16ms	39.6m/s	
Median, L								
Wrist	2.76ms	25.20uV	0.00uVms	Wrist	135mm	2.76ms	48.9m/s	
Median, R								
Wrist	0ms							

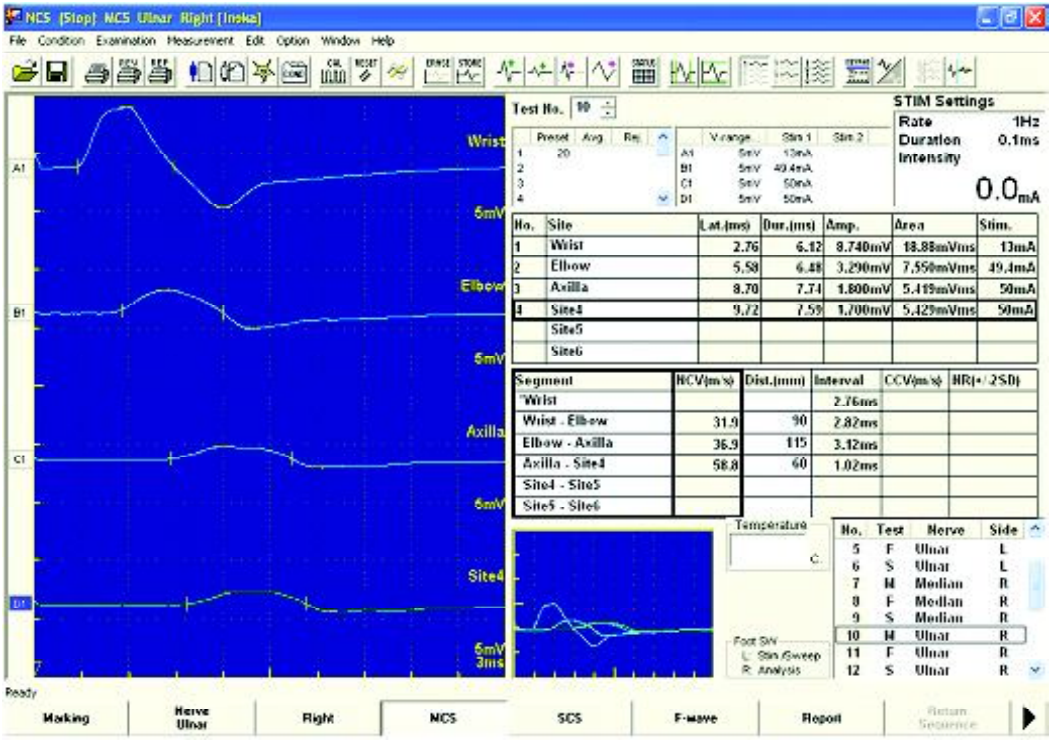

Figure 2. Ulnar motor studies.

Nerve conduction study showed evidence of segmental demyelination at multiple sites with delayed distal motor latencies, reduced motor nerve conduction velocities and multiple motor conduction blocks, i.e. the amplitude of the compound muscle action potential is significantly lower on stimulation of the proximal segments than that from the distal segment (Figure 2 and Table 1). Conduction blocks were noted at the sites other than the entrapment sites. F waves were absent. Sensory nerve conduction studies were normal.

Cerebrospinal fluid analysis was normal. Her FBS was 79mg/dl and HbA1c was 6.2%. Rest of the biochemical and haematological tests were normal. The diagnosis of MMN was made on clinical and neurophysiological grounds. She was given high doses (2g/kg) of intravenous immunoglobulin for three days and physiotherapy was continued. She made a rapid recovery with almost normal functional state within two weeks. Repeat nerve conduction study revealed improved conduction blocks.

Discussion

MMN is a rare disease with an estimated prevalence of 1-2/100,000 individuals[3]. It is more frequent in men than women with an approximate ratio of 3:1[2]. The mean age at disease onset is 40 years. Clinically MMN is characterized by slowly progressive or stepwise progressive, asymmetrical, and more distal paresis. The upper limbs are usually affected earlier and more severe than the lower limbs[2,3,5]. The most common initial symptom is wrist drop or finger drop and impaired grip strength. Cranial nerve involvement is uncommon. Most patients develop a slowly progressive disease course over several years. Beside, relapsing forms of MMN showing acute deterioration, stepwise progression, as well as spontaneous remission have occasionally been described[3,5]. After extensive literature search, we were unable to find a case of MMN with fairly rapid disease progression over few months leading to severe disability. The most prominent electrophysiological feature in MMN is multifocal persistent partial conduction blocks, i.e. the failure of a nerve impulse to propagate through a structurally intact axon present in motor but not sensory nerve fibers and located outside the common entrapment sites[3]. Approximately 40-50% of patients have IgM serum antibodies directed against GM1 ganglioside[3].

By contrast, with CIDP, treatment with plasma exchange and prednisolone is generally not effective in MMN and even associated with worsening of the disability in some patients[2,5]. Many clinical trials have shown that treatment with high dose intravenous immunoglobulin leads to improvement of muscle power in patients with MMN[3]. It is important to recognize this condition early considering the differences in the management and outcome from other demyelinating neuropathies.

References

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