Case report

Multi focal motor neuropathy presenting as acute quadriparesis – acute multifocal motor neuropathy (AMMN)

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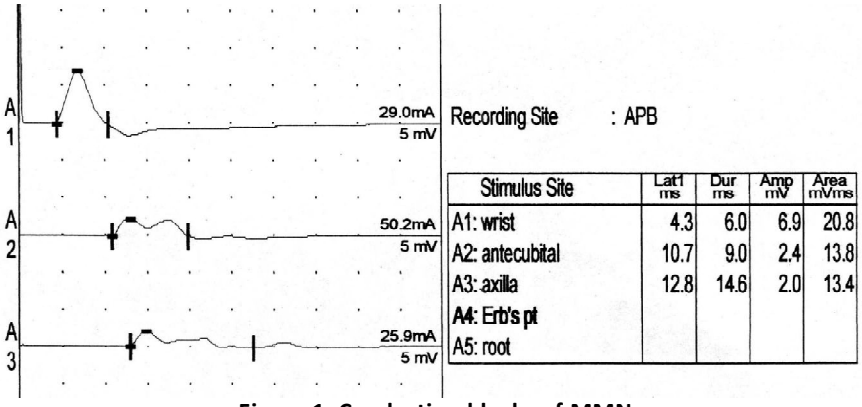
Introduction

Multifocal motor neuropathy (MMN) is an acquired immune-mediated condition[1]. It usually progresses insidiously in a stepwise fashion with lower motor neuron type weakness, cramps, fasciculations, myokimia and later atrophy. Sensory symptoms are generally not present. Sometimes MMN is mistakenly diagnosed as motor neuron disease (MND). In most cases it is asymmetric and is strikingly multifocal and begins in one or both arms[2]. Rarely MMN can present as acute quadriparesis. Only a few cases of this rare atypical presentation are reported in world literature[3,4]. This variant known as acute-onset MMN (AMMN) should be differentiated from other immune-mediated neuropathies such as acute inflammatory demyelinating (AIDP) or axonal (AMAN) polyneuropathy, acute motor conduction block neuropathy (AMCBN), acute-onset chronic inflammatory demyelinating polyneuropathy (CIDP). Persistent motor conduction blocks (CBs) at sites not exposed to compression or entrapment, absence of sensory symptoms, normal CSF protein and IgM reactivity against ganglioside GM1 favors AMMN over the other conditions. We report such a case of AMMN in a Sri Lankan patient and this is probably the first such report from Asia.

Case report

A 39-year old female presented with difficulty in walking of one day's duration. She was unable to stand, walk or sit within the next few days. Weakness was present in all four limbs but was more apparent in the upper limbs. Reflexes were diminished and there were no fasciculations. On direct questioning she reported some weakness in her hands over the preceding few months which progressed slowly to involve proximal upper limb muscles. Examination revealed predominantly distal bilateral asymmetric weakness in all 4 limbs. Reflexes were markedly diminished. There was no wasting or fasciculations. She did not have any sensory symptoms or signs. Rest of the examination was unremarkable. Serial EMGs done showed features of predominant motor neuropathy with persistent conduction blocks (CBs). Sensory involvement was demonstrable only in median nerve (probably a co existing CTS). CBs continued to appear in the subsequent studies making conditions such as AMAN / AIDP rather unlikely (Usually CBs disappear within weeks in these conditions). CSF analysis (done after day 10) was normal. CSF protein level was only 20 mg/dl and there was no increase in cells. This was further evidence against AIDP, AMAN or acutely presenting CIDP. Renal function tests, electrolytes, liver function tests, full blood count and inflammatory markers were normal. A diagnosis of acute onset multi focal motor neuropathy (AMMN) presenting with quadriparesis and persistent CBs was made. She was treated with a course of intravenous immunoglobulin (IVIg) for 5 days (0.4g/kg/day × 5 days) to which she responded dramatically. She was treated with a second cycle within a few weeks after which she regained normal power.



Stimulus Site	Lat1 ms	Dur ms	Amp mV	Area mVms
A1: wrist	4.3	6.0	6.9	20.8
A2: antecubital	10.7	9.0	2.4	13.8
A3: axilla	12.8	14.6	2.0	13.4
A4: Erb's pt				
A5: root				

Figure 1. Conduction blocks of MMN.

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Practice Points

- MMN is a rare but treatable motor neuropathy with an excellent prognosis
- Early accurate diagnosis demands a high degree of clinical suspicion
- MMN can be misdiagnosed as MND, hence important to exclude in all cases of MND
- In suspected but unconfirmed cases of MMN masquerading as MND a good response to a course of IVIG helps to differentiate the two
- Very rarely MMN can present as acute quadriparesis (AMMN) as described here
- Persistent CBs, normal CSF, asymmetrical weakness, ant GM1 antibodies helps differentiating AMMN from AIDP, AMAN, CIDP
- Magnetic resonance imaging shows gadolinium enhancement and/or hypertrophy of the brachial plexuses
- Unlike in CIDP treatment with steroids or plasma exchange can be harmful in MMN

Discussion

Multifocal motor neuropathy (MMN) usually is an indolent chronic distal multifocal disease which progresses slowly with lower motor neuron-type weakness, without sensory symptoms[1]. Diagnostic criteria include motor conduction block (CBs) at sites not exposed to compression or entrapment[2]. CBs may persist or reverse irrespective of clinical outcome. However CBs can persist for longer periods in MMN as opposed to AMAN and AIDP. It is a treatable condition with a good prognosis which is sometimes mistakenly diagnosed as Motor Neurone Disease (MND) which has a very poor prognosis. MMN is a very rare condition, affecting only about 1 per 100,000 people in the population. Men are about three times as likely to be affected as women. Most patients are between the ages of 30 and 50 years when symptoms are noted, with the average age of onset being 40 years. The diagnosis rests on identifying the typical clinical and electro-physiological features. Electro diagnostic studies reveal conduction block outside of common sites of entrapment in motor but not sensory nerves[1].

Acute-onset MMN (AMMN) as in this case should be differentiated from other immune-mediated neuropathies such as AMAN, AIDP, AMCBN and CIDP. Asymmetry, persistent CBs, normal CSF and predominate upper limb weakness is in favor of AMMN. CIDP is less likely in this case due to asymmetrical, predominantly distal weakness and normal CSF protein level. MMN show an excellent treatment response to repeated cycles of IVIg. We found only nine previously reported cases of AMMN in world literature[3,4]. We believe that ours is the first such case from Asia. The cases described so far are very similar to ours with the key features being the acute quadriparesis with areflexia, persistent CBs on EMG, normal CSF protein and an excellent response to treatment with IVIg.

In MMN cerebrospinal fluid examination may be normal, but a mild elevation of protein is not uncommon[5]. Elevated anti ganglioside antibodies, including GM1 IgM antibodies though likely to be helpful in confirming the diagnosis of MMN their absence does not exclude it[4]. Furthermore, elevated GM1 IgM can be found in patients with other neuropathies, motor neuron disease, or even normal individuals[5]. IVIg is the mainstay of treatment. Many other immunosuppressant medications have been tried in MMN. Of these, only cyclophosphamide has been shown to be of any significant, reproducible benefit. MMN does not usually respond to steroids or plasma exchange, and these treatments may worsen it[6,7,8].

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