Case Report

Slowly Progressive Asymmetrical Flaccid Quadriparesis: Is it Multifocal Motor Neuropathy with Conduction Block or Progressive Muscular Atrophy?

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Abstract

Introduction: Multifocal motor neuropathy (MMN) is a slowly progressive, asymmetrical neuropathy with autoimmune etiology, exclusively involving motor nerves, electrophysiologically characterized by multiple conduction blocks, and is treatable. Therefore differential diagnosis is of critical importance.

Case presentation: We report a case of 60-year-old lady with chronic progressive asymmetrical flaccid quadriparesis in conjunction with phrenic nerve involvement and brisk deep tendon reflexes. Differential diagnosis between multifocal motor neuropathy and motor neuron disease (MND) is important.

Conclusion: In case of slowly progressive asymmetrical flaccid quadriparesis MMN should always be taken in account. Electrophysiological and other supportive diagnostic tests should be performed. Also in case of respiratory insufficiency in a patient with MMN, the rare possibility of involvement of the phrenic nerve should be remembered.

INTRODUCTION

Multifocal motor neuropathy is a slowly progressive, asymmetrical neuropathy with autoimmune etiology [1], exclusively involving motor nerves, electrophysiologically characterized by multiple conduction blocks [2,3]. Clinical features are weakness, atrophy and cramps in the muscles innervated by the involved nerves. Fasciculations and absence of deep tendon reflexes (DTR) are frequently encountered. The presence of conduction blocks at least in two, or more nerves and a normal sensory conduction are two basic diagnostic criteria. Guideline of clinical and electrophysiological diagnostic criteria of MMN has been recommended [4].

CASE PRESENTATION

A 60 year old lady presented with difficulty in walking. History revealed weakness and numbness starting in lower extremities and then progressing up to the upper extremities for last three years.

Examination showed flaccid quadriparesis more pronounced in the distal muscles of lower extremities and on the left side. Muscle atrophy was prominent on the left side and distal muscles of upper extremities. Deep tendon reflexes were brisk in all extremities. Despite the presence of sensorial complaints, all the modalities of sensory examinations were normal.

These data suggested a generalized, chronic, progressive, asymmetrical motor neuron involvement. The differential diagnosis includes motor neuron disease (MND)’s, cervical and lumbar spinal canal stenosis, asymmetric polymyopathies.

Nerve conduction study (NCS) revealed an asymmetrical motor polyneuropathy with multiple, segmental demyelination including conduction blocks. Needle electromyography (EMG) showed subacute and chronic neurogenic involvement. No giant motor unit action potentials (MUAPs) were present.

Biochemical work-up, and markers of vasculitic, tumors and infections were normal. Anti-GM1 antibodies were negative.
Whole spinal Magnetic Resonance Imaging (MRI); abdomen and thorax computed tomography (CT) examinations were normal.

These data were consistent with multifocal motor neuropathy (MMN). Intravenous immune globulin (IVIG) administered at a doses of 2mg/kg. A modest improvement in strength but not in NCSs was observed one month after treatment.

Differential diagnosis between MND and MMN is of crucial importance, because MMN is a treatable condition [1,5]. However, differentiation in some cases may be remarkably difficult. Parry and Clarke presented five cases initially diagnosed as MND because of distal prominent muscle atrophy, fasciculations, and brisk DTR, but the condition eventually was diagnosed as MMN after electrophysiological evaluation. The diagnoses were verified by nerve biopsy [6]. Pestronk et al. [5] also reported two cases previously diagnosed as MND because of hyperactive tendon reflexes, but turned out to be MMN after the determination of a high titer of anti-GM1 antibodies [7]. In our case, anti-GM1 ganglioside was negative. However, the absence of antiganglioside antibodies is not required for the exclusion of the diagnosis of MMN [1,7].

Three years later the patient readmitted with slight dyspnea and respiratory hypersecretion Examination revealed progressed weakness and muscle atrophy. DTR were brisk but no other upper motor neuron involvement signs were present.

NCS revealed a more severe asymmetrical motor polyneuropathy with multiple, segmental demyelination including conduction blocks. Needle EMG showed inactive chronic neurogenic involvement without giant MUAPs.

Thorax CT showed infiltration consistent with pneumonia. Respiratory function tests showed an obstructive and restrictive type of respiratory insufficiency. Phrenic nerve conduction studies revealed a marked prolongation of latencies of both phrenic nerves and reduced muscle (diaphragmatic) APs. She died abruptly on the first day of hospital admission. Consent for an autopsy was rejected by her family.

The involvement of cranial nerve was rarely reported at early or late stage in the course of MMN [4,8,9]. Beydoun presented a case with bilateral phrenic neuropathy of acute onset [4]. Magistris also reported a case diagnosed as MMN 13 years after the onset with symptoms, such as weakness of neck, tongue muscles and the presence of respiratory distress [10]. A phrenic NCS was performed because of the presence of a mild dyspnea, which has shown the bilateral involvement of phrenic nerves.

CONCLUSIONS

In case of slowly progressive asymmetrical flaccid quadriaparesis MMN should always be taken in account. The electrophysiological and other supportive diagnostic tests should be performed. Also in case of respiratory insufficiency in a patient with MMN, the rare possibility of involvement of the phrenic nerve should be remembered.

REFERENCES