Case Report

An Acute Motor Axonal Neuropathy (AMAN) Case With Motor Conduction Blocks in Childhood

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Abstract

Objective

Acute motor axonal neuropathy (AMAN), characterized with decreased compound muscle action potentials (CMAP) and absence of demyelinating findings in electrophysiological studies, is a subtype of Guillain-Barre Syndrome (GBS). A 4 yr-old male patient presented with ascending weakness, dysarthria and dysphagia to İstanbul Dr. Lütfi Kırdar Kartal Training and Research Hospital Neurology outpatient for three days to in 2012. Dysphonia, restricted eye movements, flaccid tetraplegia and areflexia were found in neurological examination. There were motor conduction blocks in all peripheral nerves in electrophysiological studies.According to these findings the patient was diagnosed as Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP). Reduction of CMAP amplitudes in posterior tibial nerve, absence of CMAPs in median, ulnar and peroneal nerves and loss of motor conduction blocks were found in following electrophysiological studies. According to these findings, patient was diagnosed as AMAN. Motor conduction blocks may appear in early stage of AMAN and they disappear in later examinations. That's why electrophysiological studies must be repeated in patients with GBS.

Keywords: Acute motor axonal neuropathy;Motor conduction blocks; Guillain-Barre Syndrome; Acute motor axonal neuropathy

Introduction

There are three major types of Guillain-Barre Syndrome (GBS): acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor axonal neuropathy (AMAN) and acute motor sensory axonal neuropathy (AMSAN). It is difficut to distinguishthses 3 from according to clinical findings. Electrophysiological findings play determinant role in diagnosis and classification of GBS (1). AMAN diagnose is based on decreased compound muscle action potentials (CMAP) and absence of demyelinating findings (2).

AMAN with conduction blocks is rare, and usually recovers completely(3). That's why AMAN patients may be diagnosed as AIDP in early stages. AMAN may appear after Campylobacter jejuni infection. It is less often in children then adults and more often in boys then girls. AMAN usually appears after third year of life(4). Clinical findings in childhood are similar to adults, but meningeal irritation findings, cranial nerve involvement, flask quadriplegia and dysautonomia can appear more

frequent (5).

Here we report a 4 yr-old patient having AMAN with motor conduction blocks in early period of disease.

Case Report

A 4yr -old male patient presented with ascending weakness, dysarthria and disphagia to İstanbul Dr. Lütfi Kırdar Kartal Training and Research Hospital Neurology outpatient for 3 days in 2012. The patient had stomacache, nausea, vomitting and diarrhea 1 week earlier. Dysphonia, restristed vertical and horizontal eye movements, bilaterale peripheral facial paralysis, absence of velum-pharyngeus reflex, flaccid tetraplegia, areflexia and unresponsive plantar reflexes were found in neurological examination. He had urinary encontinance and constipation. Hemograme, serume glucose, liver functions, kidney functions, electrolites, vitamine B12, folic acid, thyroid functions, urine analysis and cerebrospinale fluid analysis were normal. Sensory nerve conduction velocities and sensory action pottentials (SAP) were normal in electrophysiological studies. Partial motor conduction blocks in elbow segment of median nerves and popliteal segment of posterior tibial nerves, complete motor conduction blocks in elbow segment of unlar nerves, prolonged distal motor latency and decreased CMAP amplitudes in peroneal nerves were found (Table I). F-waves were absent in median and unlar nerves. There was no denervation potential and motor unit pottentials in needle electromyography (EMG). Serological tests for C. jejuni were negative. The patient was diagnosed as AIDP and treated with 0.4 gr/kg/d intravenous immunoglobuline (IVIG) for 5 d.

At fifteenth day, patient's clinical findings did not recover. In electrophysiological studies, sensory nerve conduction studies were normal. Median, unlar and peroneal nerve CMAP amplitudes were absent. Posterior tibial nerve CMAP amplitudes were decreased, distal motor latency and motor conduction velocities were normal. Our diagnose was changed to AMAN according to these findins.

At 40th day, velum-phryngeal reflex was positive. Muscle strenghts were 3/5 in humerus abductors and adductors, 2/5 in elbow flexors and extensors and 0/5 in other muscles. Median and ulnar nerve CMAPs were absent, posterior tibial and peroneal nerve CMAPs were decreased and motor conduction velocities were normal. Denervation pottentials (Possitive sharp waves and fibrillation) were found in all proximale and distal muscles in upper and lower limbs, and MUPs were absent.

At the end of second month, dysphonia and dysphagia recovered. Muscle strenghts were 4/5 in humerus abductors and adductors, 2/5 in elbow flexors and extensors, 3/5 in thigh flexors and extensors and 0/5 in other muscles. CMAP amplitudes were decraesed in all peripharal nerves in upper and lower limbs and motor conduction velocities were normal.

Discussion

Conduction blocks appear when saltatory conduction stops but the axon remains intact. In practice, this situation is seen as abnormal amplitude/area CMAP reduction on proximal stimulation compared with distal CMAP. Conduction block is usually associated with segmental demyelination. Electrophysiological studies in early stages of diseases can not distinguish the cause of reduction in CMAP amplitude, which may appear due to reversible conduction block or length-dependent conduction failure (6).

Determining of proximal conduction blocks is an important finding of acute GBS, but is found in about 40% of patients (7). Diffuse demyelination replaces conduction blocks in following weeks. Conduction blocks are usually found on distal nerve ends, nerve roots and nerve entrapment sites where the blood-nerve barrier is thought to be weak (8). Conduction blocks can be determined in 67% of AMAN patients, and patients can be diagnosed as AIDP (1). Hadden et al. evaluated electrophysiological examinations of 369 patients from 11 west countries repeated with 4 week intervals. After first examination, 69% of patients were diagnosed as AIDP, and 3% AMAN. AIDP and AMAN rates were found very near after following examinations (2). We found conduction blocks in the first examination of our patient, however, we found axonal polyneuropathy findings in second electrophysiological examination. The patient was diagnosed as AMAN according to these findings.

Demyelination findings as CMAP amplitude reduction and conduction blocks may be found in patients

with anti-ganglioside antibodies in early stages (8). Conduction blocks in AMAN in early stages may appear due to anti GM-1 antibody associated with sodium channel destruction, but it is stil a matter of controversial (9). Pathological changes in Ranvier nodes were seen in immunohistochemical examinations done on rabbits made AMAN with sensitization with GM-1(10). Antibodies in AMAN patients cause binding of membrane attack complexes to nodal axolemma, destruction of sodium channels, elongation of nodal area and seperation of paranodal myeline from axons (11). The last finding is similar to paranodal demyelination, but the main problem is axonal degeneration (6). Cytoskeletal degeneration and mytocondrial dysfunction due to calcium ions enter from membrane attack complex pores is the main cause (1,12). We could not analyze the anti GM-1, anti GD1a and anti GD1b antibodies because of technical lack.

Capasso et al. reported two patients, who had acute symmetric weakness without sensory loss. These two patients had diarrhea and high levels of anti GM-1, anti GD1a and anti GD1b IgG. Campylobacter jejuni was positive in one of them. Electrophysiological studies showed motor conduction blocks in forearm segments and normal sensory coduction. Distal CMAPs were normal. Conduction blocks and weakness resolved 2-5 weeks without temporale dyspertion of distal and proximale CMAPs (3). In patients who have acute symmetric weakness that progressed monophasic and recovered rapidly, diarrhea due to C. jejuni and anti GM-1 antibodies should be accepted as a subtype of GBS(3). Patients having prolonged clinical findings, motor conduction blocks may have AMAN (1,12). Our patient had nausea, vomiting and diarrhea before acute symmetric weakness and serological testes for C. jejuni were negative.

AMAN is characterized with rapidly progressive weakness. Respiratory failure usulally occurs and recovers quickly (13). Level of residual involvement is associated with AMAN in electrophysiological examinations and clinical findings (14). Blood pressure imbalance, tashycardia, pupil and sweating abnormalities usually appears in AMAN patients as autonomic dysfunction (15). Lee et al. found urinary dysfunction in 50% of AMAN patients (16).

GBS is the most possible diagnose in a patient with acute, progressive paresis, and AIDP is the most type of GBS. Electrophysiological examinations are usually normal in early stage. Conduction blocks are diagnostic for AIDP, but these can appear in early stage of AMAN. This information should not be forgotten and electrophysiological examinations must be repeated in patients with prolonged clinical findings.

In conclusion, AMAN may present with conduction blocks, which causes confusion in making diagnoses of GBS, in early stages of disease. More studies are required to clarify how conduction block occurs in AMAN patients.

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Authors' Contribution:

Serhan Yıldırım MD: Drafting, designing of the work, analysis, and final approval of the work.

Rahşan Adviye İnan: Drafting, and final approval of the work.

Hakan Levent Gül: Designing of the work, intrpretation and final approval of the work.

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All authors agreement to be accountable for all aspects and integrity of the work.

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Table 1. Motor Nerve Conduction Studies at Onset

Nerve	Stimulation Point	Record Point	Latency (ms)	Velocity(m/s)	Amplitde (mV)
Right Median	Wrist	APB	2.0		4.5
	Elbow		8.3	23.8	1.7
Left Median	Wrist	APB	2.1		5.5
	Elbow		8.0	25.4	2.0
Right Ulnar	Wrist	ADM	1.8		6.2
	Elbow		4.5	55	5.5
	Axilla		0	0	0
Left Ulnar	Wrist	ADM	1.9		7.5
	Elbow		4.7	53.5	6.2
	Axilla		0	0	0
Right Peroneal	Ankle	EDB	3.5		3.5
	Head of fibula		7.7	42.8	3.0
	Poplitea		12.3	21.7	1.2
Left Peroneal	Ankle	EDB	3.2		3.8
	Head of fibula		7.2	45	3.5
	Poplitea		12.5	18.8	1.0
Right Tibial	Ankle	AHL	7.2		2.0
	Poplitea		17.5	24.3	1.8
Left Tibial	Ankle	AHL	7.5		1.5
	Poplitea		18.0	23.8	1.3

ADM: Abductor digiti minimi, AHL: Abductor hallucis longus, APB: Abductor pollicis brevis, EDB: Extensor digitorum brevis, ms: milisecond, m/s: metre/second, mV: milivolt

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