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A case of acute motor sensory axonal neuropathy presenting reversible conduction block

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Reversible conduction block (RCB) was rare in patients with acute motor sensory axonal neuropathy (AMSAN). A-46-year-old man presented with paresthesia, weakness, diplopia, and dysarthria. Nerve conduction study (NCS) exhibited axonal changes with conduction block in motor and sensory nerves. His symptoms were rapidly progressed and recovered. Conduction block was disappeared in the follow-up NCS performed after 2 weeks. The AMSAN case with RCB showed rapid progress and rapid recovery of clinical symptoms as acute motor axonal neuropathy patients with RCB.

Key words: Guillain-Barre Syndrome; Nerve conduction; Muscle weakness

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Acute motor axonal neuropathy (AMAN) is a subtype of Guillain-Barre syndrome (GBS), which is relatively common in East Asia including Japan, China, and Korea.[1,2] According to some reports, AMAN is more prevalent than typical GBS known as acute inflammatory demyelinating polyradiculoneuropathy (AIDP).[3-5] On the other hand, acute motor sensory axonal neuropathy (AMSAN) is a rare disease compared with AMAN, but the pathophysiology and clinical course of AMSAN are reported to be similar to those of AMAN except sensory nerve involvement.[2]

Reversible conduction block (RCB) which was known to be associated with a sodium-channel dysfunction in node of Ranvier has been reported in some AMAN patients. AMAN with RCB is known to show relative rapid recovery and good prognosis.[1,2] However, RCB was rarely reported in patients with AMSAN and the clinical course and outcome in patients with AMSAN with RCB were not well-known. Recently, we experienced a case of AMSAN with RCB in serial electrophysiologic studies.

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CASE

A 46-year-old man presented with tingling sensations of 4 limbs, double vision, and speech disturbance developed the day before of admission. He was healthy and had no history of any particular disease and medication. He was diagnosed and treated for acute enterocolitis by diarrhea a week ago. Bilateral horizontal diplopia, dysarthria, dysphagia, bifacial palsies, quadriparesis (MRC grade 4), and gait ataxia were observed on neurologic examinations. Deep tendon reflexes on 4 limbs were absent. Nerve conduction study (NCS) which was performed on the second day of the admission exhibited the decreased sensory nerve action potential (SNAP) amplitude and compound motor action potential (CMAP) potential of both upper extremities with sparing the both lower limbs. Conduction block was observed in both ulnar nerves. F-waves of both median, ulnar, and common peroneal nerves and H-reflexes in both calf muscles were not formed (Fig. 1A). Cerebrospinal fluid test was normal. Enzyme-linked immunosorbent assay for IgG and IgM antibodies against GM1, GM2, GD1a, GD1b, GT1a, GT1b, GQ1b, and GD3 were all negative. No specific abnormal finding was observed in CBC, blood chemistry, urine analysis, and serology and virology tests. Stool culture for Campylobacter jejuni was negative.

He was treated with IV immunoglobulin (0.4g/kg) for 5

Sensory Nerve Conduction Studies

Nerve / Sites	Latency ms	Amplitude μV	Distance cm	Vel. Pk m/s
Left MEDIAN				
1. Fingr-Wrist	3.25	3.5	15	46.2
2. Palm-Wrist	2.25	4.6	9.5	42.2
3. Wrist-Elbow	4.20	12.0	23	54.8
4. Elbow-Axilla	2.60	5.3	16.8	64.6
Right MEDIAN				
1. Fingr-Wrist	3.35	2.7	15	44.8
2. Palm-Wrist	2.45	7.0	9.5	38.8
3. Wrist-Elbow	4.35	17.9	25	57.5
Left ULNAR				
1. Fingr-Wrist	2.90	1.8	13	44.8
2. Wrist-Elbow				
3. Elbow-Axilla	3.10	1.5	16	51.6
Right ULNAR				
1. Fingr-Wrist	3.00	1.2	13	43.3
2. Wrist-Elbow	4.90	7.2	27	55.1
Left SURAL				
1.	4.05	12.6	14	34.6
2.				
Right SURAL				
1.	4.20	8.2	14	33.3
2.				
Left SUPERFICIAL PERONEAL				
1.	3.65	5.1	12	32.9
2.				
Right SUPERFICIAL PERONEAL				
1.	3.50	13.2	12	34.3
2.				

Motor Nerve Conduction Studies

Nerve / Sites	Latency ms	Amplitude mV	Duration ms	Distance cm	Velocity m/s
Left MEDIAN – Abductor Pollicis Brevis					
1. Wrist	4.10	2.7	13.00		
2. Elbow	8.10	2.7	12.55	24	60.0
Right MEDIAN – Abductor Pollicis Brevis					
1. Wrist	3.95	2.7	11.00		
2. Elbow	8.40	2.5	11.35	26	58.4
Left ULNAR – Abductor Digiti Minimi					
1. Wrist	2.35	11.3	16.05		
2. Below Elbow	6.30	10.1	15.55	25.5	64.6
3. Above Elbow	8.90	0.2	12.50	8	30.8
4. Axilla	11.45	0.1	8.15	14	54.9
Right ULNAR – Abductor Digiti Minimi					
1. Wrist	2.00	11.7	13.35		
2. Below Elbow	6.25	6.9	13.45	29	68.2
3. Erb's point	9.35	1.6	12.10	8	25.8
Left COMMON PERONEAL – Extensor Digitorum Brevis					
1. Ankle	4.30	1.6	12.65		
2. Fibular Head	12.10	1.3	13.85	32.5	41.7
3. Popliteal Fossa	14.00	1.1	16.10	8	42.1
Right COMMON PERONEAL – Extensor Digitorum Brevis					
1. Ankle	4.15	3.1	12.10		
2. Fibular Head	10.55	2.4	13.35	32.5	50.8
3. Popliteal fossa	12.40	2.2	12.10	8	43.2
Left TIBIAL (KNEE) – Abductor Hallucis					
1. Ankle	4.90	18.4	13.35		
2. Knee	13.10	15.2	14.25	38	46.3
Right TIBIAL (KNEE) – Abductor Hallucis					
1. Ankle	3.35	31.0	11.55		
2. Knee	12.40	21.3	13.60	38	42.0
R FACIAL - Orb Oculi					
1. Anterior Ear	1.05	3.0	22.05		
L FACIAL - Orb Oculi					
1. Anterior Ear	1.25	1.9	18.85		

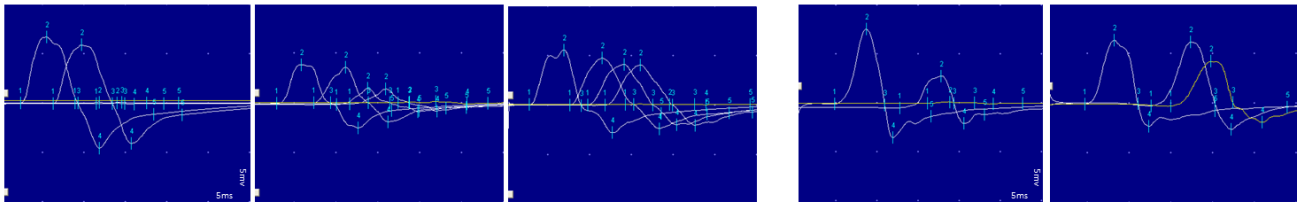
F-Wave

Nerve	Latency ms
Left MEDIAN	No
Left ULNAR	No
Left COMM PERONEAL	No
Left TIBIAL (KNEE)	48.65
Right MEDIAN	No
Right ULNAR	No
Right COMMON PERONEAL	No
Right TIBIAL (KNEE)	50.10

H-Reflex

Nerve	Latency ms
Left TIBIAL (KNEE) – Soleus	No
Right TIBIAL (KNEE) - Soleus	No

A



B 2nd day 17th day 31th day C 17th day 31th day

Fig. 1. Nerve conduction studies in the present case. (A) Initial nerve conduction study which was performed on the second day of admission exhibited the decreased sensory nerve action potential amplitude and compound motor action potential amplitude of both upper extremities. Conduction block was observed in both ulnar nerves. F-waves of both median, ulnar, and common peroneal nerves and H-reflexes in both calf muscles were not formed. (B) Conduction block in the left ulnar motor nerve was firstly appeared on the second day of admission, began to reverse on the seventeenth day of admission, and disappeared on the thirty-first day of admission. (C) Conduction block in the left common peroneal nerve was firstly appeared on the seventeenth day of admission and disappeared at the thirty-first day of admission.

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days, however, quadriparesis (MRC 1 in upper limbs or 2 in lower limbs), dysphagia, dysarthria and facial diplegia were worsen and bilateral extraocular muscles (EOM) limitation and dyspnea were newly developed. He had to undergo artificial ventilator therapy in intensive care unit for 9 days from the third day of admission. Respiratory hold and dyspnea was rapidly recovered, and he could remove the ventilator at the eleventh day of admission.

NCS which was performed at seventeenth day of admission exhibited the decreased SNAP and CMAP amplitudes in 4 limbs with newly developed conduction block in both common peroneal nerves. Conduction blocks in both ulnar nerves were partially recovered. F-waves in both median and ulnar nerves were recovered but F-waves in both common peroneal and H-reflexes in both calf muscles were still not formed (Fig. 1B). At the same day, quadriparesis (MRC 2 in upper limbs and 3 in lower limbs), dysarthria, dysphagia, and facial diplegia were improved. He can walk with assistance and swallow and speech without obstacle. However, EOM limitation and sensory symptoms still remained.

At the thirty-first day of admission, SNAP amplitude in 4 limbs was still decreased, however, CMAP amplitude of 4 limbs returned to normal range and conduction block of both ulnar and common peroneal nerves disappeared (Fig. 1C). Quadriparesis (MRC 4 in upper limbs and MRC 4-5 in lower limbs) was much improved. He can walk without assistance. Slight EOM limitation in both horizontal directions and sensory symptoms still remained.

DISCUSSION

AMAN and AIDP are major subtypes of GBS. In general, patients with AMAN present with more rapid progression and poor recovery of neurological deficits compared with patients with AIDP. The natural course of AMAN depends on the immunopathophysiology, which are classified as axonal degeneration and RCB. AMAN patients with axonal degeneration tended to show rapid progression, late recovery and poor outcome, whereas AMAN patients with RCB tended to show fast recovery and good outcome.[1,4] RCB was reported in about 45% among patients with AMAN in two recent retrospective studies.[6,7] RCB was observed in 12 patients (70.6%) among 17 patients with AMAN in a study of Korea.[8]

RCB is known to be caused by a transient sodium-channel disruption and the detachment of paranodal myelin which was initiated by the ganglioside antibody deposition and subsequent complement activation at node of Ranvier and paranodal area.[1,2,6] The pathophysiology and clinical course in AMSAN patients with RCB are likely to be similar to AMAN patients with RCB because both diseases shared many clinical and laboratory similarities. RCB has also been reported in AMSAN, Miller-fissure syndrome, and pharyngeal-cervical-brachial GBS as well as AMAN.[2,9] The present case exhibited a rapid progression and rapid recovery with minor residual symptoms. Serial NCS exhibited RCBs in motor nerves. Conduction block was reversed after two weeks. These findings are consistent with the previous reports in AMAN patients with RCB. This report supports that AMAN and AMSAN are one clinical spectrum. However, any anti-ganglioside antibodies were not detected. The serum sample for the anti-ganglioside antibody assay was obtained on the second day of admission and third day of onset. The possibility of false-negative due to the fast sample acquisition time and method of measurement should be considered. Follow-up antibody test using more sensitive assay can increase the sensitivity of the anti-ganglioside antibody assay. Further investigation to evaluate the clinical course and immunologic characteristic in patients with AMSAN patients with RCB is needed.

Conflicts of Interest

The authors have no conflicts to disclose.

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