CASE REPORT

Acute Motor Sensory Axonal Neuropathy: A Variant of Guillain–Barré Syndrome—A Rare Case Report

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Abstract

Background: Guillain–Barré syndrome (GBS) is an immune-mediated disorder of the nervous system that shows acute or subacute onset. It is also known as Landry's paralysis. It is characterized by muscle weakness of legs and arms, limb paresthesias, and total or relative areflexia. Acute motor sensory axonal neuropathy (AMSAN) is a distinct subtype of GBS. It is not only a rare but severe variant that involves axonal degeneration in motor and sensory nerve fibers and has a prolonged recovery course.

Case description: A 60-year-old male presented to the emergency department having complaints of weakness, numbness, and tingling sensation in feet for the last fortnight, which ascended gradually towards the calves. He was observed to have a sensory disorder in hands, but not flaccid paralysis. The patient history and nerve conduction studies were indicative of AMSAN variant of GBS.

Discussion: In patients of AMSAN, the reduction of sural nerve amplitude is more pronounced as compared to acute inflammatory demyelinating polyneuropathy (AIDP) patients. In case of our patient, the electrophysiological feature indicated a more than 50% decrease in SNAP. A marked reduction in sensory nerve action potential and compound muscle action potential with only slightly decreased conduction velocities is a requirement for the diagnosis of axonal neuropathies, which is the trait seen in the reported case.

Keywords: Acute motor sensory axonal neuropathy, Compound muscle action potential, Guillain–Barré syndrome, Nerve conduction studies, Sensory nerve action potential.

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Introduction

Guillain-Barré syndrome (GBS) is an immune-mediated disorder of the nervous system that shows acute or subacute onset. It is also known as Landry's paralysis.[1] It is characterized by muscle weakness of legs and arms, limb paresthesias, and total or relative areflexia[2] GBS or postinfective polyneuritis may not be a single disease but a cluster of acute neuropathies with several related immune-mediated pathogenic mechanisms. The peripheral nerves in GBS are infiltered by lymphocytes and macrophages and causes endoneural inflammation of spinal nerve roots and entrapment sites.[3] GBS is considered to have several variants like acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor sensory axonal neuropathy (AMSAN), and Miller-Fisher syndrome.[4]

Acute motor sensory axonal neuropathy (AMSAN) is a distinct subtype of GBS. It is not only a rare but severe variant that involves axonal degeneration in motor and sensory nerve fibers and has a prolonged recovery course.[5] AMSAN occurs in approximately 1–15% of GBS patients. In it, a sensory disturbance is an essential feature in addition to motor symptoms and signs affecting limbs. The course of the disease is more severe and prolonged with frequent ventilator dependency, slow recovery, and significant residues.[6–11]

In addition to clinical features, nerve conduction study and neurophysiological variables confirm the diagnosis and allow for further categorization of a variant of GBS.

Case description

A 60-year-old male presented to the emergency department having complaints of weakness, numbness, and tingling sensation in feet for the last fortnight, which ascended gradually towards the calves. He was observed to have a sensory disorder in hands, but not flaccid paralysis. The patient also reported that ten days ago, he suffered from mild fever and four days of self-limiting diarrhea. He did not have any respiratory disease symptoms. Moreover, there was not any history of vaccination. MRI of the brain and spine did not show any acute changes. During the second day of admission, nerve conduction studies were conducted on the patient whereby tibial, peroneal, and sural nerves were studied. These studies showed acute sensory and motor neuropathy affecting the lower limb (Table 1).

The patient history and nerve conduction studies were indicative of the AMSAN variant of GBS. Electro-diagnosed test performed revealed that the right peroneal nerve was non-stimulable while the left personal nerve showed CMAP amplitude with less than 90% of a lower limit of motor amplitude. Motor nerve

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Table 1: Nerve conduction study report of the patient

Motor nerve conduction study	*Latency (ms)*	*Amplitude (mv)*	*Velocity (m/s)*
Right tibial nerve	3.85	2.5*	38.89
Left tibial nerve	3.65	2.9*	38.82
Right peroneal nerve	Non-stimulable		
Left peroneal nerve	3.85	1.5*	39.08
Sensory nerve conduction study	*Latency (ms)*	*Amplitude (μv)*	*Nerve conduction velocity (NCV) (m/s)*
Right sural nerve	2.04	7.2*	43.92
Left sural nerve	1.96	9.3*	46.12

*Abnormally reduced value

conduction velocity was reduced mildly in both tibial and peroneal nerve bilaterally but no conduction block was visualized.

In the case of the sural nerve, SNAP amplitude was reduced bilaterally. It was less than 50% of the lower limit of normal sensory amplitude. According to NCS, neurophysiologically, there was no evidence of demyelination, i.e., latencies were unaffected. Cerebral spinal fluid (CSF) analysis was also done within two days of initial presentation. It revealed an albumin cytological disassociation with nil white cells present and an elevated protein level of 103 mg/dL against the normal value (i.e., from 45 to 60 mg/dL).

On the third day of hospitalization, the patient was put on intravenous immunoglobulin (IVIG) therapy for the treatment of suspected AMSAN. The patient showed gradual improvement with IVIG therapy. After a hospital stay of 10 days, the patient was discharged with 4/5 power in lower limbs, 5/5 power in upper limbs, and normal sensory examination.

Discussion

This case report describes a patient of AMSAN. The patient did not have any history of diabetes or neoplastic or hematological disorder. The patient had symptoms of paresthesia along with little objective sensory loss which is considered as the most common initial symptom of GBS. This case is regarded as different because of its characteristic sensory clinical presentation, a significant decrease in amplitude of motor and sensory action potential, and mildly reduced nerve conduction velocities. Latency is unaffected indicating sparing of demyelination injury. Elevated CSF proteins as seen in this case finding are a characteristic feature in 50% of cases of GBS as also seen in other studies.[6–12]

Acute axonal neuropathies, such as AMAN and AMSAN are observed to have a fast progression as well as a prolonged recovery time as compared to acute demyelinating neuropathies.[10,11] The clinical difference can be attributed to fundamental differences in the mechanism, as the axolemma and nodes of Ranvier are affected in AMAN and AMSAN compared to Schwan cells being affected in AIDP.

In patients of AMSAN, the reduction of sural nerve amplitude is more pronounced as compared to AIDP patients.[12] In case of our patient, the electrophysiological feature indicated a more than 50% decrease in SNAP. A marked reduction in sensory nerve action potential and compound muscle action potential with only slightly decreased conduction velocities is a requirement for the diagnosis of axonal neuropathies, which is the trait seen in the reported case.

Conclusion

AMSAN is a rare variant of GBS and it can often be missed due to variations in clinical presentations. Diligent nerve conduction analysis helps in diagnosing the above variant as it presented the affected neurophysiological variables in CMAP, SNAP in unambiguous terms. Knowledge of potential clinical manifestations and clear-cut diagnosis of AMSAN is very important as it can help manage these patients and for providing counseling to their family members.

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