A Unique Presentation of Autonomic Dysautonomia Requiring a Pacemaker in a Teenager with Acute Motor and Sensory Axonal Neuropathy: A Case Report

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Introduction

Acute motor and sensory axonal neuropathy (AMSAN) is a rare, severe axonal variant of Guillain-Barré syndrome (GBS). Autonomic neuropathy is an important complication that may present in these individuals. These autonomic abnormalities include a range of cardiac arrhythmias including sinus tachycardia, bradyarrhythmias and even asystole [1]. This is likely secondary to either an excess activation or suppression of the parasympathetic or sympathetic pathways.

These abnormalities in GBS are usually transient and reversible, but in severe cases, they may persist due to permanent damage to the cardiac conduction system. These individuals with autonomic dysautonomia are candidates for implantation of permanent pacemakers.

Case Description

Setting: Academic Acute Inpatient Rehabilitation Hospital

Patient: A 19-year-old male with Acute Motor and Sensory Axonal Neuropathy (AMSAN) complicated with autonomic dysautonomia requiring a pacemaker

Case Presentation: The patient is a 19-year-old male with no past medical history who presented to the hospital with a rapidly progressive ascending tetraplegia. He developed acute respiratory failure and required intubation. Eventually, a G-tube and trach were placed. A lumbar puncture was performed and revealed cytoalbuminologic dissociation in the cerebrospinal fluid. He was treated with IVIG and completed 5 sessions of plasma exchange. An EMG nerve-conduction study demonstrated severe non-length-dependent sensorimotor polyneuropathy with axonal features consistent with AMSAN (table 1). His hospital course was complicated by spontaneous episodes of symptomatic bradyarrhythmia leading to sinus arrest likely associated with autonomic dysfunction. As a result, he was treated with implantation of a leadless pacemaker. He also had orthostatic hypotension requiring an alpha-agonist. He had impaired mobility and activities of daily living and was sent to acute inpatient rehabilitation.

Assessment / Results

With comprehensive interdisciplinary therapies, he was able to improve in strength, endurance, and trunk control. He progressed from maximum assistance with ambulation to ambulating >150 feet with platform walker with minimum assistance. On discharge, he continued to be limited by decreased strength but continued therapies. On outpatient follow-up, he demonstrated significant improvement, ambulating modified independent with a standard walker.

Sensory Nerve Conduction Study

Nerve	Onset Latency (ms)	Peak Latency (ms)	Peak-Peak Amp. (µV)
Left Ulnar	NR	NR	NR
Left Radial	NR	NR	NR
Left Sural	NR	NR	NR

Motor Nerve Conduction Study

Nerve (Recording site)	Onset Latency (ms)	Peak-Peak Amp. (mV)	Base-Peak Amp. (mV)	Duration (ms)
Left Median (APB)	NR	NR	NR	NR
Left Ulnar (ADM)	NR	NR	NR	NR
Left Common Peroneal (EDB)	NR	NR	NR	NR
Left Tibial (AH)	NR	NR	NR	NR
Left Common Peroneal (TA)	NR	NR	NR	NR
Right Axillary (Deltoid)	NR	NR	NR	NR

Needle Electromyography Study

Muscle (Left side)	IA	Fib	PSW	Fasc
Deltoid	Increased	None	1+	None
Flexor Carpi Radialis	Increased	None	3+	None
First Dorsal Interosseous	Increased	None	3+	None
Tibialis Anterior	Increased	1+	2+	None
Vastus Medialis	Increased	None	1+	None

Table 1: Nerve conduction study and needle EMG showing non-length-dependent sensorimotor polyneuropathy with axonal features. Amp, amplitude; APB, abductor pollicis brevis; ADM, abductor digiti minimi; EDB, extensor digitorum brevis; AH, abductor hallucis; TA, tibialis anterior; IA, insertional activity; Fib, fibrillation; PSW, positive sharp waves; Fasc, fasciculations.

Discussion

Autonomic dysfunction can be observed in patients with GBS including the severe AMSAN variant. The risk of autonomic dysautonomia is higher in patients with tetraplegia, respiratory failure or bulbar involvement [2]. Labile hypertension, sinus tachycardia and orthostatic hypotension are the commonly seen autonomic abnormalities; however, there are cases of tachyarrhythmia, bradyarrhythmia and even asystole [1]. Bradyarrhythmia occurs in up to 50% of patients with severe GBS and is secondary to parasympathetic overactivity [3]. These patients can experience a range from bradycardia to periods of sinus arrest, which occurs spontaneously. More severe bradyarrhythmias are seen in severely affected patients, including those that require mechanical ventilation [4]. Current literature does not show any standardized treatment for symptomatic bradyarrhythmia secondary to autonomic dysautonomia. Patients with GBS who experience multiple episodes of bradycardia and asystole should be evaluated and considered as potential candidates for pacemaker implantation.

Conclusions

Patients with AMSAN usually experience a prolonged and incomplete recovery. Complicated cases may require a pacemaker for symptomatic bradyarrhythmia secondary to autonomic dysautonomia. Early identification and aggressive goal-specific rehabilitation in the inpatient and outpatient setting is vital to maximize independence and lead to better outcomes in this patient population.

References

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