Reversible conduction failure in acute motor axonal neuropathy

Sir,

Guillain Barré syndrome (GBS) is a fairly common neurological disorder, which warrants immediate attention and management to prevent significant morbidity and mortality. The diagnosis depends on identification of the characteristic clinical pattern and exclusion of mimics with proper investigations. Based on electrodiagnostic criteria GBS is subdivided into axonal (acute motor axonal neuropathy [AMAN] and acute motor sensory axonal neuropathy) and demyelinating variants (acute inflammatory demyelinating polyradiculoneuropathy [AIDP]).[1] In patients with AMAN Nerve conduction studies typically show normal sensory nerve action potentials and progressive decline

in compound muscle action potential (CMAP) amplitude. AIDP is characterized by slowing of conduction in motor and sensory fibers with conduction blocks. Primary axonal variants however rarely may produce immune mediated conduction failure in nodes of Ranvier, which can only be recognized by serial nerve conduction studies. Sometimes such cases are initially categorized as acute multifocal neuropathy by treating physicians.[2] Here, we report three such cases of AMAN with reversible conduction failure closely mimicking demyelinating variants of GBS. First patient was a 56-year-old male with acute quadriparesis with progression to maximum disability over 7 days making him bedbound. No history of any antecedent illness was obtained. He had hypotonia in all limbs and symmetrical limb weakness with Medical Research Council grade 2 power proximally and grade 3 power distally. Ocular, facial and bulbar muscles were spared. Routine blood investigations and cerebrospinal fluid (CSF) study were normal. Initial nerve conduction study showed multiple motor conduction blocks in non-compression sites with normal sensory conduction. Serial nerve conduction studies were done four times during the hospitalization, which showed an evolving pattern of improvement in proximal amplitude and disappearance of conduction blocks without prolongation of distal latencies or excessive temporal dispersion of CMAPs.

Second patient was a 58-year-old female with acute weakness of all four limbs reaching the nadir in 5 days. She had a history of fever with loose stools 1 week prior to onset of symptoms. On examination, she had lower limb weakness with grade three power both proximally and distally and bilateral hand grip weakness. Sensory examination was normal. Deep tendon reflexes were ineluctable. Routine investigations and CSF study were normal. Initial electrophysiolological profile and subsequent evolution of CMAPs were similar to patient 1.

Third patient was a 65-year-old female who presented with disabling acute pure motor lower motor neuron quadriparesis with the nadir of her illness reached in 7 days. She had no history of any antecedent illness. On examination, she was areflexic without sensory or bladder involvement. Nerve conduction study results were similar to those of former two patients. Electrophysiological testing helps to distinguish between the types of GBS.

In AMAN the electrophysiologic profile expected is axonal, i.e., normal studies early in the course of illness followed by progressive loss of CMAP amplitude with relatively preserved conduction velocities and distal latencies followed by return of CMAP amplitude to normal with clinical improvement. Whereas in AIDP we expect prolonged distal latencies, decreased conduction velocities with motor conduction blocks early in the course of illness followed by a characteristic evolution. This evolution is characterized by progressive prolongation of distal latencies and temporal dispersion of CMAPs during the phase the patient shows clinical improvement.[3] In contrast the cases we have described show typical reversible conduction failure in axonopathies. It is produced by anti-ganglioside antibodies at the axolemma of the nodes of Ranvier.

Conduction block is the term used to describe the condition in which salutatory conduction is stopped, but the axon remains intact. Operationally, it is recognized by an abnormal amplitude/area CMAP reduction on proximal stimulation as compared with CMAP on distal stimulation. Conduction block is usually considered to be the electrophysiological correlate of segmental demyelination. In the early disease stage and based on only one recording, no electrodiagnostical distinction between demyelinating conduction block and other causes of abnormal amplitude reduction of proximal CMAP, is possible. The concept of reversible conduction failure is proposed to denote the rapid recoveries of prolonged distal motor latencies, reduced distal CMAP amplitudes and conduction block, none of which are explained by remyelination. It is produced by antibodies against GM1/GalNAc-GD1a attacking axolemma at nodes of Ranvier. In contrast, resolution of demyelinative conduction block in AIDP is usually associated with conduction slowing and increased CMAP duration with remyelinating slow components in serial recordings.[4]

Patients we have presented showed typical clinical presentation of GBS. Initial nerve conduction studies showed a pattern very difficult to differentiate from a demyelinating variant. However subsequent studies showed an improving pattern with progressive increase in proximal amplitude resulting in disappearance of conduction block pattern. Analysis of the sequential studies show typical reversible conduction failure described in AMAN. These cases we have reported show how axonal variants of GBS may masquerade as demyelinating pattern in initial nerve conduction studies and how do serial studies help in understanding the underlying pathophysiology.

Sreejith Peediackal, James Jose, V. Abdul Gafoor, B. Smitha

Department of Neurology, Government Medical College, Kozhikode, Kerala, India

For correspondence: Dr. Sreejith Peediackal, Department of Neurology, Government Medical College, Kozhikode, Kerala, India. E-mail: speediackal@gmail.com

References

1. Hadden RD, Cornblath DR, Hughes RA, Zielasek J, Hartung HP, Toyka KV, *et al*. Electrophysiological classification of Guillain-Barré syndrome: Clinical associations and outcome. Plasma Exchange/ Sandoglobulin Guillain-Barré Syndrome Trial Group. Ann Neurol 1998;44:780-8.
2. Verma R, Chaudhari TS. Two unusual variants of Guillain-Barre syndrome. BMJ Case Rep 2012 Nov 15;2012. pii: bcr2012007088. PubMed PMID: 23162026.
3. Hiraga A, Mori M, Ogawara K, Hattori T, Kuwabara S. Differences in patterns of progression in demyelinating and axonal Guillain-Barré syndromes. Neurology 2003;61:471-4.
4. Kokubun N, Nishibayashi M, Uncini A, Odaka M, Hirata K, Yuki N.Conduction block in acute motor axonal neuropathy. Brain 2010;133:2897-908.

Access this article online

Quick Response Code:

Website: www.annalsofian.org

DOI: 10.4103/0972-2327.128601