Case Report

Guillain–Barre syndrome with hyperreflexia: A variant

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ABSTRACT

Guillain–Barre syndrome (GBS) is a common cause of acute peripheral neuropathy and is characterized by hyporeflexia or areflexia. Hyperreflexia has been rarely reported with acute motor axonal neuropathy. A 10-year-old boy presented with asymmetrical weakness of upper and lower limbs and change of voice. Weakness progressed in the hospital with involvement of multiple cranial nerves, preserved deep tendon jerks with extensor plantar, and normal abdominal reflexes. He was treated with IV immunoglobulin and IV methylprednisolone. He was able to walk with support with normal voice at the time of discharge. GBS should be a differential diagnosis in patients with acute quadriparesis even if there are preserved deep tendon reflexes.

Key words: Guillain–Barre syndrome, preserved reflexes, quadriparesis

Introduction

Guillain–Barre syndrome (GBS) is a common cause of acute peripheral neuropathy and is characterized by hyporeflexia or areflexia. Even though hyporeflexia or areflexia is necessary for diagnosis of GBS, hyperreflexia does not exclude a GBS variant. Recently there have been reports of hyperreflexia with two variants of GBS – acute motor conduction block neuropathy and acute facial diplegia with hyperreflexia. We describe a variant with brisk reflexes throughout the illness, acute motor conduction block neuropathy.

Case Report

A 10-year-old boy, who had an episode of upper respiratory tract infection, on the 10th day of the illness noticed pain and weakness in both upper and lower limbs more on the right side and of the distal muscles with inability to stand associated with change of voice. At admission he had asymmetrical weakness of both upper and lower limbs and in the hospital his weakness progressed with involvement of shoulder muscles and cranial nerve involvement (bilateral weakness of facial muscles) with loss of voice. On neurological examination there was bilateral 7th nerve, 10th, 11th, and 12th cranial nerve deficits, generalized hypotonia, and preserved deep tendon jerks with extensor plantar and normal abdominal reflex. There was no sensory deficit or autonomic dysfunction. Nerve conduction study was suggestive of motor axonal polyneuropathy. He was treated with intravenous immunoglobulin (IVIG) and in view of progression of disease even after 2 days of ivig therapy he was also treated with IV methylprednisolone. He recovered slowly with improvement of muscle power and was able to walk with support at the time of discharge (2 weeks).

Discussion

Areflexia is one of the two clinical features required for diagnosis of GBS. Normal reflexes or hyperreflexia throughout the course of GBS is unusual. Deep tendon reflexes may be preserved throughout the disease course in patients with acute motor axonal neuropathy (AMAN) and have been considered as indicators of rapid clinical recovery. Our patient's clinical presentation and disease course was typical of GBS except for preservation of reflexes. As for diverse disease entities within the GBS spectrum, the clinical and electrophysiological findings of the patient best fit in AMAN, in which the

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primary immunological attack is supposed to be directed against motor axons. Although hyperreflexia is a controversial symptom in patients with GBS, these findings indicate that there is functional corticospinal tract involvement in patients with a GBS variant. Moreover, 48% of Chinese and 33% of Japanese patients with AMAN showed hyperreflexia in the recovery phase.[1,2] In another study, patients with pure motor GBS had preserved tendon reflexes up to MRC grade 3 paresis.[3] Acute facial diplegia with hyperreflexia has been described as a GBS variant and nerve conduction studies in limbs were normal in this report.[2] Preserved reflexes and even hyperreflexia may occur in patients with pure motor GBS and are not inconsistent with the diagnosis. It is more appropriate to classify this neuropathy as a GBS variant, which Capasso *et al*., suggest calling "acute motor conduction block neuropathy," emphasizing the presence of conduction blocks and avoiding the pathophysiologic implication that all conduction blocks are demyelinating in nature.[4] They propose that conduction block in their case was due to axonal block by the antibody at the node of Ranvier without demyelination. Recently, electrophysiologic evidence of lower motor neuron hyperexcitability has been reported in some patients with AMAN. It has been suggested that the most common sites of nerve involvement in patients with GBS are not randomly distributed throughout the nerve. The distal motor nerves, the proximal segments, and the sites prone to compression are most vulnerable.[4-6] Early in the disease, electrophysiologic abnormalities are often mild or nonspecific. Motor conduction blocks (CB) have been documented in only 2–15% of patients with GBS within 3 weeks from disease onset, and CB in intermediate nerve segments in the first days of the disease is uncommon.[4,5,7] It is possible that mechanical impairment of the blood–nerve barrier at entrapment site may render these nerve segments more vulnerable to immunologic attack.[6] The pathologic basis of CB is thought to be acute demyelination. Experimental studies of synchronously demyelinating lesions showed that recovery from CB is characterized by dyssynchronization. It might be possible to explain the preservation of tendon reflexes in GBS by following factors. The presence of normal sensory nerve function rather than motor is required for tendon jerks.[1] As tendon jerks are dependent on synchronized volley of impulses, a purely axonal lesion would preserve tendon jerks better than a demyelinating lesion.[1] In our patient, no electrodiagnostic correlate of peripheral nerve demyelination was found.

Conclusion

GBS should be considered in differential diagnosis of patients with acute quadriparesis even when there are brisk reflexes and nerve conduction studies should be part of evaluation to confirm the diagnosis.

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