Guillain Barre syndrome with brisk reflexes-another variant

Sir,

Guillain Barre syndrome (GBS) is a common cause of acute peripheral neuropathy and is characterized by hyporeflexia or areflexia. Hyperreflexia has been reported with acute motor axonal neuropathy. Recently there have been reports of hyperreflexia with two variants of Guillain Barre Syndrome-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. Even though hyporeflexia or areflexia is necessary for diagnosis of Guillain Barre Syndrome, hyperreflexia doesn’t exclude a GBS variant.

A 14-year-old boy was admitted with progressive weakness of limbs of 10 days duration. He had difficulty in getting up from squatting position, climbing stairs, difficulty in lifting upper limb above shoulder and difficulty in holding objects. There was no cranial nerve symptoms, sensory symptoms or bladder symptoms. There was no muscle pain. There was no preceding fever, diarrhoea, vaccination or drug intake. On examination he was afebrile, had bilateral lower motor neuron type facial weakness, weakness of proximal muscles of lower limb (3/5 MRC) and distal muscles (4/5). Upper limb muscles showed weakness in proximal (4-/5) and distal muscles (4/5). There was marked trunk and neck flexion weakness. Limbs were facecied. Deep tendon reflexes were brisk. Plantars were flexor bilaterally. Sensory system was normal. There were no cerebellar signs. He needed one-person support to walk. Investigations showed normal total and differential leukocyte counts, sedimentation rate (ESR 8 mm/1 h), creatine kinase and electrolytes (Na, K, Mg, Phosphate). CSF study showed normal protein with no cells. Nerve conduction study showed definite conduction blocks (outside usual entrapment sites) or probable conduction block/temporal dispersion in all nerves tested with normal sensory conduction [Table 1]. F responses were normal or minimally prolonged. Conduction velocities were normal and distal latencies were normal or slightly prolonged He was treated with IVIG (400 mg/kg/day for 5 days) and he improved. We could not do immunological testing due to local unavailability.

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 indicators of rapid clinical recovery.\(^1\) In Europe, patients with pure motor GBS had preserved tendon reflexes up to MRC grade 3 paresis\(^2\) and more recently an AMAN patient with hyperreflexia has been reported.\(^3\) Acute facial diplegia with hyperreflexia has been described as a Guillain Barre syndrome variant and nerve conduction studies in limbs were normal in these reports.\(^4\) Another variant has been described recently with retained reflexes-acute motor conduction block neuropathy.\(^5\) Two patients developed symmetric proximal and distal weakness without sensory abnormalities after enteritis. Tender reflexes were normal in one patient and brisk in the other.

Electro physiology showed early partial motor conduction block in intermediate and distal nerve segments, normal sensory conduction even across the sites of conduction block and normal somatosensory evoked potential. Because some clinical, electrophysiologic and laboratory findings matched the features of multifocal motor neuropathy (MMN), the term “acute multifocal motor neuropathy” has been proposed.\(^6\) In conclusion, 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 CBs and avoiding the pathophysiologic implication that all CBs are demyelinating in nature. They propose that conduction block in their case were due to axonal block by the antibody at node of Ranvier with out demyelination. Bereano et al suggested that it could be still conduction block due to demyelination.\(^7\)

In our case there was conduction block/temporal dispersion in several nerves suggesting demyelination as the pathological mechanism underlying conduction block.

GBS should be considered in differential diagnosis in patients with acute quadriparesis even when there are brisk reflexes and nerve conduction studies are invaluable in this regard.

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Distal latency (Normal upper limit)</th>
<th>Amplitude distal/proximal duration</th>
<th>Velocity m/s (Normal lower limit)</th>
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<tr>
<td>Rt median</td>
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<td>13.7/3</td>
<td>10.7/16.5</td>
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<td>2.3 (3)</td>
<td>3.9/1.1</td>
<td>15.6/17.1</td>
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<td>Left median</td>
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<td>13.7/1.9</td>
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<tr>
<td>Left ulnar</td>
<td>2.8 (3)</td>
<td>4.4/1.7</td>
<td>13.8/21.8</td>
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<td>Rt CPN</td>
<td>4.2 (5)</td>
<td>4.1/0.6</td>
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<td>1/1.4</td>
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<td>Lt PTN</td>
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</tbody>
</table>

References

5. Accepted on 14-01-2006

Table 1: Motor conduction study

Letter to Editor
Bullet injury to the atlanto-axial region

Sir,

Gunshot wounds (GSWs) of the atlanto-axial region are uncommon. These injuries to the upper cervical spine are often devastating with loss of spinal cord function and respiratory compromise, sometimes leading to death.

A bullet penetrating the atlas without producing neurological deficits is exceptional.

A 21-year-old male was admitted with history of gunshot wound to the left side of neck 5 days before. Other than a left lower motor neuron facial palsy, there was no neurological deficit. An entry wound was present on left side of neck just below the mastoid. There was no exit wound. X-ray of the cervical spine showed a bullet in the region of C1 vertebrae, posterior to the anterior arch [Figure 1]. CT of the spine demonstrated that the bullet was lying obliquely behind the anterior arch of the atlas, about 3 mm from the odontoid [Figures 2 and 3]. Flexion and extension films of the cervical spine and craniovertebral junction did not demonstrate any mobility.

Since the patient was neurologically intact with no evidence of any sensorimotor deficit, he was managed conservatively. However, about a week after admission, he started having pus discharge from the wound, which persisted despite antibiotics. A decision was therefore made to remove the bullet. A vertebral angiogram done prior to surgery showed that the vertebral artery was free from the bullet.

The patient was operated through the far lateral approach. The mastoid bone and the sub-occipital triangle were exposed surgically [Figure 4]. The bullet was hidden behind the mastoid process which was partially drilled off to visualize the bullet. The patient was managed in the Neurology Department, Senior Lecturer in Neurology, Medical College, Thiruvananthapuram, Kerala, India.

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References


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