Landry Guillain-Barré Strohl syndrome Atypical Variant of Guillain-Barre Syndrome: Case Report and Review

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Abstract: Guillain-Barre syndrome (GBS) is the most common cause of acute or sub-acute generalized paralysis in practice. It is an autoimmune polyradiculopathy, which manifests as areflexic flaccid paralysis with variable sensory and autonomic dysfunction. Atypical variety of GBS are being reported from various parts of the world. There are reports of normo-reflexic or hyper-reflexic varieties of GBS, here we present a case of GBS with preserved reflexes

Keywords: Guillain-Barre syndrome with preserved reflex, atypical variant

1. Introduction
Guillain-Barre syndrome (GBS) is the most common cause of acute or sub-acute generalized paralysis in practice. It is an autoimmune polyradiculopathy, which manifests as areflexic flaccid paralysis with variable sensory and autonomic dysfunction. In a typical case, there is symmetrical weakness that is noticed first in lower limbs with variable sensory and autonomic involvement and absent reflexes. Typical cerebrospinal fluid (CSF) picture shows albumino-cytologic dissociation. Pathologically GBS has been divided into two subtypes: Demyelinating and axonal. Increasingly, atypical varieties of GBS are being reported from various parts of the world. There are reports of normo-reflexic or hyper-reflexic varieties of GBS from Chinese, Japanese and European populations. These varieties are not very common in Indian Subcontinent, and only a few such atypical cases have been reported till now.

2. Case Report
A 17 year young boy complained of sore throat (1st day), on next day he experienced change in voice with regurgitation of water, enroute to hospital patient developed lower limb and upper limb weakness and had to be mobilized with assistance of two person. O/e limbs were flaccid, asymmetrical weakness of both upper and lower limbs, there was multiple cranial nerve involvement (VII, IX, X, XI), with truncal muscle weakness, power of proximal and distal muscle of lower limb was 2/5 & 3/5 respectively(MRC grading). The power in upper limb proximal and distal muscle was 3/5 & 3/5. Deep tendon reflexes were preserved, there was no sensory deficit or autonomic dysfunction, B/L flexor plantar, normal abdominal reflex, no bladder bowel involvement, After one day of admission patient developed respiratory distress and was intubated.

3. Investigations
All hematological work up including creatine kinase & CSF study was normal. MRI BRAIN study was also normal: based on strong clinical suspicion and NCV study was done which revealed evidence of a predominantly axonal motor polyradiculoneuropathy, in addition conduction block was seen in both ulnar nerve across the elbow.
4. Management

Patient was managed on routine supportive therapy and standard therapy for GBS IVIg (400mg/kg/day) for 5 days. Tracheostomy was also done and after 10 days patient was discharged in stable condition with mild difficulty in closing eye.

5. Discussion and Review

Acute ascending weakness was first described by Landry in 1859, but the full extent of the disease and its characteristics were described by Guillain, Barré and Strohl in 1916.9 The disease gained international notoriety under the name that remains today, Guillain Barré Syndrome.10 GBS is now the world’s most common cause of acute neuromuscular paralysis.2 Our understanding of GBS has evolved many folds with many atypical variants being reported across the world with cases reported of normo-reflexia and even hyper-reflexia. No longer is areflexia an essential criteria for making a diagnosis of GBS. The typical CSF picture can take 48 h to a week to evolve making albumino-cytologic dissociation an unreliable indicator for early diagnosis and treatment.

Our patient's clinical presentation and disease course was typical of GBS except for preservation of reflexes. Preserved reflexes and even hyperreflexia may occur in patients with pure motor GBS. 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. 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. As tendon jerks are dependent on synchronized volley of impulses, a purely axonal lesion would preserve tendon jerks better than a demyelinating lesion. In our patient, no electrodiagnostic correlation of peripheral nerve demyelination was found.

References


