Case Report

Guillain-Barre syndrome with preserved reflexes and normal cerebrospinal fluid: a case study

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ABSTRACT

Guillain-Barre syndrome (GBS) is an acute fulminant polyradiculopathy, which typically manifests as areflexic paralysis with variable sensory and autonomic involvement. Typical cerebrospinal fluid (CSF) picture consists of an elevated CSF protein without pleocytosis (albumin-cytologic dissociation). There have been many case reports of atypical presentations of GBS; with normo-reflexia or even hyper-reflexia from Chinese/Japanese and European population but only a few from Indian Subcontinent. Also the typical CSF picture if not found, makes the diagnosis of GBS even more difficult. A 24-year-old man presented with weakness of all 4 limbs of 4 days duration with the antecedent history of loose stools and fever. On examination, there was flaccid paralysis involving all the 4 limbs (lower limb weakness more than the upper limb) with preserved reflexes, no sensory or cranial nerve deficit, no bladder-bowel involvement; and a normal CSF study at presentation, which 1 week later showed albumin-cytologic dissociation. On electro-diagnostic studies, it was proven as a case of acute motor axonal neuropathy. Patient was managed with routine empirical antibiotics and intravenous methyl prednisone; after 3 weeks, patient was discharged in a stable condition without any residual deficit. Our understanding about the GBS has changed manifolds over the last few decades with many atypical variants being reported across the world. This case study is to lay stress on the fact that even in the absence of typical clinical features and a normal CSF study the diagnostic possibility of GBS should be kept if there is strong clinical suspicion.

Keywords: Guillain-Barre syndrome, Guillain-Barre Strohl syndrome, Landry-Guillain-Barre-Strohl syndrome, Acute motor axonal neuropathy, Normo-reflexia, Hyper-reflexia, Areflexic motor paralysis

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.1 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.1 Pathologically GBS has been divided into two subtypes: Demyelinating and axonal.2 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.3-6 These varieties are not very common in Indian Subcontinent, and only a few such atypical cases have been reported till now.7,8

CASE REPORT

A 24-year-old unmarried male, laborer by occupation presented with weakness of all four limbs for 4 days, which began as an inability to button his shirt. Progressively, all the muscle groups were involved and on the 4th day he...

required assistance to get up from the bed. There was no history of bladder, bowel complaints, no history of fever, no history of facial deviation, no history of blurring of vision, difficulty in speaking or swallowing, and no history of trauma to neck or back. Preceding this weakness (around 10-12 days back) there was a history of episodes of loose stools (3-4/day) associated with low-grade fever, which persisted for 2 days.

On examination

Patient was moderately built, well nourished, conscious, well oriented to time, place and person. General physical examination was unremarkable, vitals were stable.

There was no atrophy in any of the muscle groups in upper or lower limb, no fasciculation. Tone was normal in muscles of upper limb, but slightly decreased in all muscle groups of the lower limb. Power was 3/5 in upper limb flexors/extensors both; 1/5 in lower limb flexors/extensors both. Deep tendon reflexes were normally present in all the four limbs. Plantar response was flexor. No cranial nerve deficit, no sensory deficit, no heart rate variability with posture, and no other evidence of vasomotor involvement. Rest of the systemic examination was unremarkable.

On CSF examination on day 5 of illness: Cells - 4, proteins - 26 mg/dl. On magnetic resonance imaging study of the spine, no evidence of compressive myelopathy was evident. Based on strong clinical suspicion a nerve conduction study was done, which revealed acute motor axonal neuropathy (AMAN) (Figure 1-10). A repeat CSF study was done on Day 13: Cells - 4, proteins - 130 mg/dl. Rest all the investigations were within normal limits. Due to local unavailability, Campylobacter jejuni serology and anti-GM1 antibodies could not be done.

Management and outcome

Due to unaffordability, standard therapy of GBS could not be followed, and the patient was started on injection methyl prednisolone from 4th day of onset of symptoms. On 3rd day of starting steroid injections and 7th day of illness, patient was able to lift his legs on his own; though still he was unable to bear his weight on his legs (lower limb power 4/5), and was able to feed and dress himself (upper limb power 4/5). On 8th day of starting steroids, he was able to stand with the help of a person and with support. On 10th day, he could walk with crutches. On 15th day, he could walk with slight assistance. On 17th day, he could walk unaided. Starting from the 18th day steroids were tapered and then stopped.

At the time of discharge patient had no weakness, power was 5/5 in all muscle groups of upper and lower limbs.
DISCUSSION

About a century ago, three French neurologists Guillain, Barre and Strohl described two soldiers, who developed acute paralysis with areflexia that spontaneously recovered. They reported the combination of increased protein concentration with a normal cell count in the CSF, or albumino-cytological dissociation, which differentiated the condition from polio-myelitis.

Since then, 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.\(^9\) 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\(^10\) making albumino-cytologic dissociation an unreliable indicator for early diagnosis and treatment.

In our case the patient presented with quadriparesis without areflexia with no sensory level, no history suggestive of diphtheria and botulism. CSF study was normal, but there was antecedent history of loose stools that made us think of GBS and so electrodiagnostic study was performed, which then showed albumino-cytologic dissociation.

Also, the standard treatment modality for GBS includes intravenous immunoglobulin and plasmapheresis, which in our setup might not be a feasible option always due to the cost factor. There is a lot of debate weather steroids can have a role in GBS or not and most of the studies are against it.\(^11\) In our case, standard treatment could not be given, patient was managed with steroids, which fortunately could be started early in the disease course as the patient presented...
very early and he showed remarkable recovery and thus steroids can have a role in GBS, however it requires further studies to be proven.

This case study is to alert fellow clinicians of such atypical varieties of GBS that can cause diagnostic dilemma and if not identified at the earliest can be rapidly fatal.

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