Guillain–barré syndrome without limb weakness: A rare variant with acute bulbar palsy

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ABSTRACT

A 76-year-old male was brought to the emergency room with an acute onset of breathlessness and difficulty swallowing. Examination revealed bilateral ptosis, bilateral vocal cord abductor palsy with diaphragmatic paralysis. He did not have any limb weakness. A diagnosis of acute bulbar palsy was made. Cerebrospinal fluid showed albumino-cytological dissociation. Magnetic resonance imaging of the brain (MRI) was normal, and a nerve conduction study (NCS) showed Acute Motor and Sensory Axonal Neuropathy (AMSAN). Guillain–Barré syndrome with acute bulbar palsy was considered. Here, we report a case of suspected Acute Bulbar Palsy plus (ABPp) syndrome. ABPp may be considered as a variant of GBS between the Miller–fisher and Pharyngeal-cervical-brachial variant and does not have any definite limb weakness. This patient also had ABPp with diaphragmatic palsy. However, whether this syndrome is an isolated variant of GBS or a continuum between the Miller-fisher syndrome (MFS) and Pharyngo-cervical brachial (PCB) variants remains to be elucidated. This case is relevant to primary care physicians as the disability with GBS remains high and may render a large burden to carers. The initial symptom of acute dysphagia must lead on the primary care physician to keep this disease in mind to prevent an unwarranted delay in diagnosis.

Keywords: ABPp, acute bulbar palsy, Guillain–Barré syndrome, variant

Case Presentation

A 76-year-old male was brought to the emergency room with an acute onset of breathlessness and difficulty swallowing. He was normal 2 days back when he developed difficulty swallowing and speaking, associated with drooping of eyelids for 2 days. Finally, he presented with breathlessness lasting for 2 h.

On examination, he had stable vitals. Neurological examination revealed the presence of bilateral ptosis. He had no evidence of facial palsy. He had no evidence of palatal palsy and his gag reflex was normal. On attempting swallowing, he developed sudden stridor, indicating vocal cord palsy. His neck and tongue movements were normal. Motor examination showed normal power, tone, and reflexes. Sensory examination was normal. There were no cerebellar or meningeal signs.

An emergency otolaryngology consult was sought, and a video laryngoscopy showed bilateral vocal cord abductor palsy. An ultrasound of the diaphragm was done, denoting diaphragmatic weakness. He was intubated on an emergency basis for airway protection.

Due to the sudden presentation of multiple cranial nerves, disseminated in 48 h, a diagnosis of acute bulbar paralysis secondary to demyelination was made. MRI brain ruled out a mass lesion.

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out structural causes, and contrast-enhanced Computed Tomography of the neck and thorax ruled out malignancies. A lumbar puncture was done in view of demyelinating disorders, which showed albumino-cytological dissociation. CSF analysis showed a high protein value of 75 mg/dL and no cells on the CSF.

A nerve conduction study (NCS) was done. NCS showed prolonged distal latencies with reduced amplitudes in all four limbs, and hence, acute motor and sensory axonal and demyelinating radiculo-neuropathy was confirmed. Phrenic nerve conduction was also done, which showed prolonged amplitudes [Table 1]. Therefore, the Acute Motor and Sensory Axonal Neuropathy (AMSAN) variant of GBS was considered. Antibody workup was done. He was negative for GM1, GM2, GM3, GD1a, GD1b, and GQ1b antibodies [Table 2]. Bickerstaff encephalitis was not considered part of the differential as there was no ophthalmoplegia, hyperreflexia, or encephalopathy [Table 1].

**Course**

He was initiated on intra-venous immunoglobulin (IVIG) at 0.4 g/kg × 5 days. His deep tendon reflexes disappeared on Day 3, but he did not develop any limb weakness. He was tracheostomized on day 10 and was continued on the ventilator. He had fluctuating blood pressure levels indicative of Dysautonomia, controlled with oral Nifedipine tablets. His diaphragmatic weakness improved by day 30. He was weaned off the ventilator, with Bi-level positive airway pressure (BIPAP) therapy, over the next 28 days. A repeat NCS showed resolution of AMSAN and showed normal distal latencies of the diaphragm—phrenic nerve. He was successfully decannulated 58 days after admission into the hospital. He was discharged on follow-up 60 days after discharge.

**Discussion**

Guillain–Barré Syndrome has an overall incidence of 1 to 2 cases per 100,000 every year.[1]

This illness is relevant to the practice of primary care physicians, as Guillain–Barré Syndrome is associated with a lot of disability, which in turn destabilizes the support of the family. A study done by Wachira et al.[2] showed that the years lived in disability due to GBS were 57.52.

About 90% of cases presented with limb weakness of the arms and legs. Only 1% presented with no weakness. The absence of deep tendon reflexes is usual, but 2% of cases would have normal reflexes.[3] It was atypical that our patient manifested without any limb weakness or areflexia. Areflexia had only developed later in the disease course; however, his limb power remained normal. It was seen that 85% of cases have CSF cell counts of less than five cells.[4] The NCS showed AMSAN. Our patient also had no history of antecedent infection.

**Variants of GBS**

**Miller–fisher syndrome**

There are variations within GBS [Table 3]. Miller–Fisher syndrome is a GBS variant that presents with ataxia and ophthalmoplegia.[5] About 25% of these cases present with no limb weakness.[6] Antibodies are directed to GQ1b in 85% of cases.[7] However, cross-reactivity is expressed with GT1a antibodies.[8]

**Pharyngeal-cervical-brachial variant**

This variant manifests with weakness of the neck muscles and shoulders and may have swallowing difficulty.[9] Antibodies are directed to GT1a in 51.0% of patients and GQ1b in 39.0% of patients.[10]

**Acute bulbar palsy plus (ABPP) syndrome**

This may be a subtype between Miller–Fisher Syndrome and PCB variant.[11] Here, acute bulbar palsy without limb weakness may develop. Over the course of illness, this may transition into a differentiated Guillain–Barré syndrome. This cannot be classified into PCB or MFS variants [Figure 1].

Kim et al.[12] attempted to identify acute bulbar palsy without limb weakness. The most frequent positivity was IgG anti-GT1a antibodies (100%) followed by IgG anti-GQ1b antibodies (55%).

According to Cao et al.[13] the question arises as to whether ABPP syndrome can be considered along a spectrum between subtypes of PCB and MFS, or should be considered a separate GBS subtype. The study attempted to classify ABPP into six categories:

1. ABPP with ophthalmoparesis
2. ABPP with ophthalmoparesis and ataxia
3. ABPP with ataxia
4. ABPP with facial palsy
5. ABPP with masticatory and tongue weakness
6. ABPP with ophthalmoparesis, facial palsy, masticatory, and tongue weakness.

The study concluded that ABPP syndrome falls between GBS and MFS and is more likely to be a separate subtype of GBS rather than an overlap of distinct subtypes. However, in this most extensive case series of ABPP, no cases had reported diaphragmatic palsy, such as seen in our case.

### Table 1: Phrenic nerve conduction study

| Region       | Latency (ms) | Reference range | Amplitude (µV) | Reference range |
|--------------|-------------|-----------------|---------------|----------------|
| Left phrenic | 6.2         | 6.3±0.8         | 55.9          | 600±140        |
| Right phrenic| 12.88       | 6.3±0.8         | 121           | 600±140        |

### Table 2: Antibody profiling

| Antibody tested | Test result |
|-----------------|-------------|
| GM1, GM2, GM3   | Negative    |
| GD1a, GD1b, GT1b, GQ1b | Negative     |
| GT1a            | ?           |
The limitation in our case is that we could not test for GT1a antibodies. In the context of our case, their role may require further evaluation. Furthermore, given the variable predictability of these antibodies, one must go by clinical diagnosis, in this case. Acute bulbar palsy without limb weakness may be considered a distinct sub-entity of GBS.

**Summary**

In conclusion, an isolated presentation of bulbar palsy must warrant consideration of GBS. As a part of GBS, acute bulbar palsy usually occurs as a transitional syndrome and shares some features with either the MFS or PCB variant. However, whether this syndrome is an isolated variant of GBS or a continuum between the MFS and PCB variants remains to be elucidated. This case is relevant to primary care physicians as the disability with GBS remains high and may render a large burden to carers. The initial symptom of acute dysphagia must lead on the primary care physician to keep this disease in mind, to prevent an unwarranted delay in diagnosis.

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**Conflicts of interest**

There are no conflicts of interest.

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