***Case report***

# Acute bulbar palsy plus syndrome-A rare variant of Guillain–Barre syndrome : a case report.


**ABSTRACT**

**Background** : Acute Bulbar Palsy Plus – GBS is a rare presentation of GBS that requires early recognition and treatment.

**Clinical Description** : We present a case of 11year old girl presenting with diplopia, change in voice and nasal regurgitation. NCV study was done which showed decreased compound muscle action potential in bilateral common peroneal nerves and decreased sensory nerve action potential in bilateral ulnar nerves suggesting subclinical axonal involvement of both upper limbs and lower limbs. A diagnosis of acute bulbar palsy plus – Guillan Barre Syndrome was kept after ruling out Polio, diptheria, toxins and metabolic causes.

**Management and Outcome**: Child was started on Iv Immunoglobulins. The nasal intonation and bilateral LR palsy resolved within 3days of starting treatment. **Conclusion** : This case report is aimed to raise awareness about this rare variant of GBS. Its early recognition and initiation of treatment could allow full recovery.



*Keywords: Guillain Barre syndrome , Acute Bulbar palsy plus syndrome , Stroke mimic.*

## 1. INTRODUCTION

Guillain barre syndrome is an acute paralytic polyneuropathy that typically presents as symmetrical ascending paralysis with areflexia due to an autoimmune reaction, mostly post infection. Isolated multiple cranial nerve involvement is rare in GBS[1] specially in pediatric population. Acute bulbar palsy GBS can present either alone or in combination with opthalmoplegia, ataxia or both. Here we present a case of Acute bulbar palsy plus variant of GBS (with opthalmoplegia) in a pediatric patient.

## 2. CASE PRESENTATION

An 11 year old female presented to us with complaints of sore throat ten days back followed by acute onset double vision, change in voice and nasal regurgitation of fluids. There was no history of headache, fever, abnormal behavior, illicit drug intake, recent vaccination, insect or animal bite.

At presentation, the child was conscious & oriented. Motor examination revealed normal tone, power, plantar response but universal areflexia. There was bilateral VI, VII, IX and Xth cranial nerve palsy in the form of bilateral lateral rectus palsy, bilateral absent gag reflex, and uvula and bifacial weakness with no other cranial nerves involved. Clinically there were no signs of meningeal irritation, sensory, cerebellar, autonomic or bowel bladder involvement. On evaluation, routine blood investigations were normal. There were no signs of infarct or bleed on MRI brain. Fundus examination was normal. Nerve conduction studies were done which showed decreased compound muscle action potential in bilateral common peroneal nerves and decreased sensory nerve action potential in bilateral ulnar nerves suggesting subclinical axonal involvement of both upper limbs and lower limbs.

A diagnosis of acute bulbar palsy plus – Guillan Barre Syndrome was kept and child was started on IvIg at 2gm/kg total dose. The nasal intonation and bilateral LR palsy resolved within 3days of starting treatment.

## DISCUSSION

Guillain Barre Syndrome is an acute post infectious polyneuropathy occurring generally after an infective illness. Typical presentation of GBS pose no diagnostic dilemma , but some atypical variants may be missed if not considered. One such variant is Acute Bulbar Palsy plus which can present with opthalmoplegia, ataxia or both[2]. Acute bulbar palsy can be a presenting feature in various conditions like stroke, botulism and myasthenia gravis but GBS presenting with this feature is a rare entity and requires early recognition to halt disease progression. GQ1b gangliosides are expressed in the nodal region of oculomotor nerves, muscle spindle afferents, peripheral nerves and possibly in the brainstem reticular formation[3]. Cases with anti GQ1B antibodies positive suggest that weakness is due to a sustained, antibody-mediated, attack at the nodal region inducing a non-demyelinating conduction failure, as seen on NCV.

A case of acute bulbar palsy with unilateral LMN facial nerve palsy was reported in a 13yr old from Delhi[3] which had demyelinating motor neuropathy. Another case series on adult patients was reported by Kim et al[2] from Seoul which reported 11 cases of acute bulbar palsy including 2 cases with acute bulbar palsy and opthalmoplegia without limb involvement. A case of isolated bulbar involvement was reported by Hamidon et al[4] from Malaysia. This patient presented with isolated bulbar palsy with areflexia and nerve conduction studies were diagnostic of GBS. None of these cases progressed to involve the respiratory muscles and all showed good recovery. Historically, the term polyneuritis cranialis has been used to describe some patients with GBS presenting with multiple cranial neuropathies in the absence of limb weakness[5]. In a study conducted in Singapore, 15 such case reports were examined, where mean age of presentation was 40 years. In all cases, disease course was monophasic with clinical improvement within weeks or months. Initial symptoms were ocular (73%) and/or bulbar (33%). In half of cases tested for, anti-ganglioside antibodies were present and most frequently against GQ1b.

## 4. CONCLUSION

Our case is among the very few cases of acute bulbar palsy plus syndrome (with opthalmoplegia) which presented with atypical features of GBS. This case study is aimed to raise awareness on atypical variants of GBS which could lead to early recognition and initiation of treatment to allow full recovery. Children with clinical variants of Guillain-Barré syndrome are more likely to manifest rapid progression from disease onset to nadir, increasing the severity of disability, cranial nerve involvement, urine incontinence, respiratory impairment, and need for ventilator support than in typical Guillain-Barré syndrome[7].

The limitation of our study is the inability to detect the confirmatory anti antiganglioside antibodies in the CSF due to limited resources.

## CONSENT (WHEREEVER APPLICABLE)

All authors declare that ‘written informed consent was obtained from the attendant and assent was taken from the patientfor publication of this case report.

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