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Diagnosis of Acute Bulbar Palsy Plus Syndrome: A Rare Variant of Guillain–barre Syndrome

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Authors' contributions

This work was carried out in collaboration between both authors. Author SS involved in the diagnosis and management of this case, wrote the first draft of this report. Author NR also involved in the diagnosis and management of this case. The author edited the draft and approved the final version of this report. Both authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Presenting case of a 11-year-old girl with sudden cranial nerve weakness, universal areflexia but without limb pain or weakness. NCV showed subclinical axonal involvement in limbs. A diagnosis of acute bulbar palsy plus–GBS was made and child was started on Iv Immunoglobulins which showed significant improvement. This report is for raising awareness about this variant of GBS.

Keywords: Guillain barre syndrome; viral infection; clinical presentation.

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. Many micro-organisms have been associated with GBS like *Campylobacter jejuni*,

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Zika virus, and in 2020, the severe acute respiratory syndrome coronavirus 2 [1]. The two common types include – AIDP (Acute Inflammatory Demyelinating Polyneuropathy) and axonal type (AMAN and AMSAN) [2]. The typical clinical presentation along with nerve conduction study forms the basis of early diagnosis and institution of immunoglobulin therapy to halt the progress to respiratory involvement. Isolated multiple cranial nerve involvement is rare in GBS [3] specially in pediatric population. Acute bulbar palsy GBS is a scarcely reported entity that 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.

1.1 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. No history of similar illness in the past or any major ailment in the past.

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.

2. MANAGEMENT AND OUTCOME

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 (Fig. 1) 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.

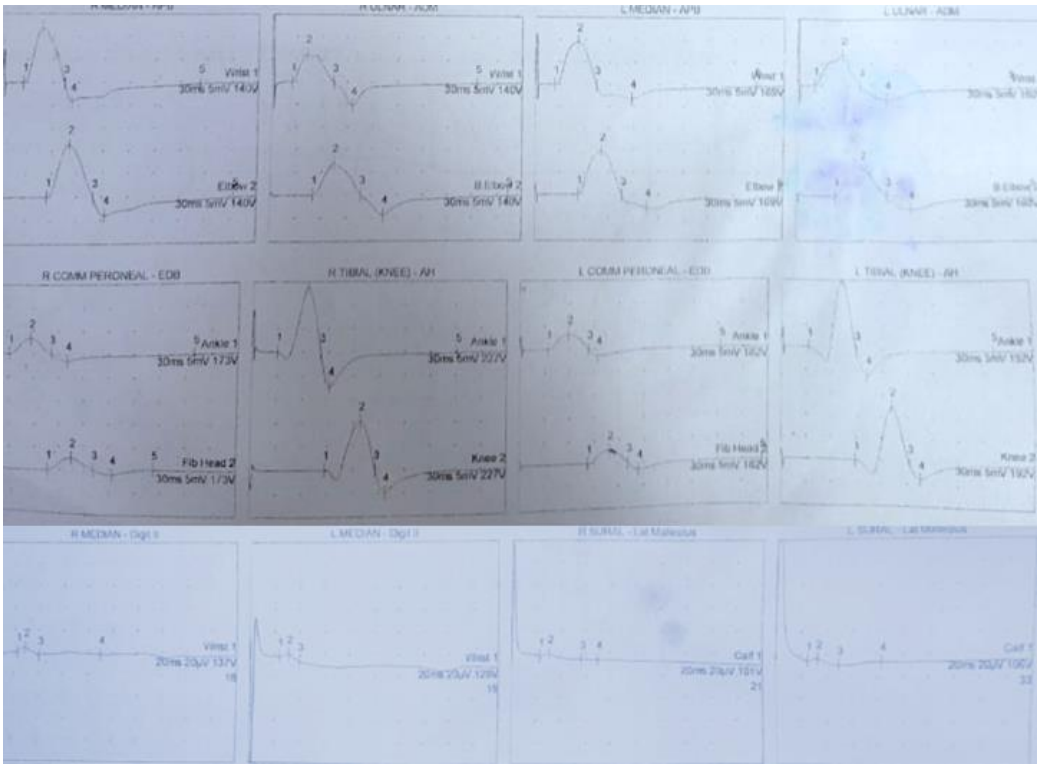

Fig. 1. NCV study of the patient

3. DISCUSSION

Guillian 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 [4]. 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 [4]. 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 similar case of acute bulbar palsy with unilateral LMN facial nerve palsy was reported in a 13yr old from Delhi [5] who presented with facial weakness following an upper respiratory tract infection. The child had no limb weakness, but universal areflexia. NCV showed demyelinating motor neuropathy. The child was followed closely for any worsening and she improved with regular physiotherapy. Another case is reported in a 3yr child with facial weakness following influenza vaccination [6]. The illness progressed to respiratory weakness in 2 days. Child was then evaluated which revealed abnormal enhancement of cranial nerves on MRI Brain and increased IgG levels in CSF. The child was started on Iv Immunoglobulins to which she responded well. The youngest case reported in literature is of a 10month baby [7] who presented with facial weakness with diminished tendon reflexes without any apparent limb weakness. The child improved with conservative management and close foloow up over a span of 4 months. As is evident in these cases and our study, management of Acute Bulbar Palsy Plus – GBS in pediatric population requires high clinical suspicion, detailed neurological examination and early intervention to prevent progression of the disease. In the 3 yr old child – NCV was not done, since MRI and CSF findings guided the diagnosis, wherein in our case, MRI was normal, but NCV showed subclinical limb involvement.

Another case series on adult patients was reported by Kim et al. [8] from Seoul which reported 11 cases of acute bulbar palsy including 2 cases with acute bulbar palsy and opthalmoplegia without limb involvement. Around 40% cases had normal NCV study, MRI Brain was done in 8 patients and none of them showed any abnormal results. All 11 cases had positive IgG anti-GT1a antibodies in serum. A case of isolated bulbar involvement was reported by Hamidon et al. [9] 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. In a study conducted in Singapore [10], 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. MRI brain was normal in 89%cases but NCV studies showed abnormality in 92% cases.

All these adult studies have shown that MRI brain can have normal study, but NCV is mostly abnormal in cases of ABP-GBS which guides to diagnosis. Detection of serum/CSF antibodies is not done in many cases due to financial reasons or non availability.

Our study is one of the few studies on GBS in pediatric population presenting with multiple cranial nerve involvement without clinical limb weakness.

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 [11].

5. STUDY LIMITATION

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

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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