

## Pharyngeal–Cervical–Brachial Variant: A Rare Form of Guillain–Barre Syndrome in a Paediatric Patient

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This case report describes a successfully managed rare variant of Guillain-Barre´ Syndrome (GBS), the pharyngeal-cervical-brachial (PCB) variant, in a 5-year-old child which is characterized by the involvement of oropharyngeal, neck and upper limb muscles, without disturbed consciousness or ataxia. Since early treatment and supportive care yield successful outcome, it is important for the treating physician to be familiar with other diseases that mimic the disease and variants for prompt and accurate diagnosis.

**Key words:** Pharyngeal-Cervical-Brachial variant of Guillain-Barre´, bulbar palsy, PCB variant of GBS

### Introduction

Guillain-Barre Syndrome is recognized as one of the commonest causes of acute flaccid paralysis in children.<sup>1</sup> Pharyngeal-cervical-brachial variant is a rare form of GBS, which can be easily misdiagnosed if not properly evaluated.<sup>1</sup> Prompt recognition of variants of GBS enables anticipatory monitoring for disease complications and early initiation of definitive treatment.<sup>6</sup>

### Case History

A 5-year-old previously well female child was admitted to paediatric unit, National Hospital Kandy, with a complain of refusal of food, abnormal unsteady gait, change in voice, which was attributed by the parents to a psychiatric illness following an argument with her class teacher. When further inquired on admission, she was found to have dysphagia for liquids, drooling of saliva, nasal regurgitation of

liquids, nasal intonation of voice, cough and unsteady gait for the last 5-6 days. Symptoms were acute in onset and progressive. She had a recent history of mild upper respiratory tract infection which resolved spontaneously. There was no bladder or bowel incontinence, no history of animal bites, stings or any injections. Vaccination status of the child was up to date.

On examination, she was afebrile, GCS 15/15, but anxious. Body weight was 20kg (BMI-16.4kg/m<sup>2</sup>). Her face was deviated to left side with drooling of saliva. Pulse rate was 74 beats/min, regular in rhythm. Blood pressure was 92/60 mmHg. Respiratory rate was 28/min, with equal bilateral chest movements. Few coarse crepitations were audible on right lower lung zone, measured vital capacity was about 20-25 ml/kg spontaneous tidal volume was 6 ml/kg. Peripheral oxygen saturation (SpO<sub>2</sub>) was 97% on air. Abdominal examination revealed normal findings.

On nervous system examination, right-sided lower motor neuron type of facial nerve palsy, with 9<sup>th</sup> and 10<sup>th</sup> cranial nerve (CN) palsy were noted. Cough and gag reflexes were significantly impaired. All other CNs were normal. Neck muscle power was graded as 3-4. Proximal upper limb muscle power was grade 3-4 and lower limb power was 4. All the deep tendon reflexes in upper limbs and lower limbs were absent. Plantar response in both sides were down going. Sensory system and cerebellar examination revealed no abnormality. There

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were no signs of meningeal irritation. Pupils were equal in size (3mm) and reacted to light. Fundal examination revealed normal findings.

Nerve conduction study (NCS) and the CT brain with brain stem done on day 5 of the illness revealed normal findings. Study of sensory nerves were normal. All her baseline investigation results for full blood count, renal and liver function tests were within the normal range. White cell count  $12.8 \times 10^9/L$ , platelet count  $230 \times 10^9/L$ , serum potassium 4.4 mmol/L, serum sodium 140 mmol/L, fasting blood glucose 112 mg/dl. Chest X-ray revealed a right lower zone haziness. Cultures for Salmonella, Shigella and serological examination for mycoplasma pneumonia were negative.

PCB variant of GBS, acute demyelinating encephalomyelitis and brainstem stroke were considered as main differential diagnoses. She was started on IV immunoglobulin (IG) 2mg/kg for 5 days in the ICU following a clinical diagnosis of a Guillain Barre variant.

Possibility of aspiration was suspected, and child was transferred to paediatric ICU after inserting a nasogastric tube. IV piperacillin tazobactam and metronidazole were prescribed and continued for 7 days. Sputum and blood cultures yield negative results.

On admission to ICU, the decision was taken to intubate the child for airway protection. She was intubated with 5mm ID non-cuffed ET tube and the throat was packed with a throat swab. Practically it required to change the throat swab every 1-2 hours, as it was soaked frequently with saliva. Considering the increased risk of aspiration with a fully soaked pack the ET tube was changed to a 5.5mm ID cuffed ET tube. Child was sedated and ventilated in the SIMV mode and weaned down to CPAP (FiO<sub>2</sub> 0.5, pressure support 7, PEEP 4) over 24 hours. Tidal volume was maintained at 120-140ml with a respiratory rate of 16-18/min. Regular chest and limb physiotherapy were carried out. Nasogastric feeding was continued with high protein diet.

Throughout the ICU stay, there were no signs

of autonomic instability. IV immunoglobulin was continued for 5 days, but no improvement was observed. Her muscle weakness remained static. With neurology opinion after 2 days of stopping IV IG, she was scheduled for plasmapheresis every other day. She showed marked improvement of upper limb power and dysphagia with 2 cycles of plasmapheresis. Upper limb power was improved to 4-5/5 and neck muscle power improved to 4/5. Following the 3<sup>rd</sup> cycle of plasmapheresis, she was extubated. MRI brain was carried out after extubation, which revealed normal findings.

Phonation was difficult initially. Speech therapy and psychological support was arranged. 5 cycles of plasmapheresis were completed. She was fully mobilized, and speech improved.

Lumbar puncture performed on the 15<sup>th</sup> day of the illness revealed elevated protein level of 70 mg/dl with one lymphocyte and no red cells. She was discharged to ward from ICU on day 14 and discharged home on day 19 following complete recovery.

### Discussion

Multiple cranial neuropathies in GBS are rare that account for about 5% of patients, even rarer in children.<sup>1</sup> The PCB variant of GBS is a rare, localized subtype of GBS, which is defined by rapidly progressive oropharyngeal and cervicobrachial weakness associated with areflexia in the upper limbs. Generally the lower limb power is preserved or minimally affected.<sup>2,3,6</sup> Since the child in our case had a similar presentation, except for ataxia, with typical CSF abnormality of “albumino cytological dissociation”, with no any CT/MRI brain and brain stem abnormality, a clinical diagnosis of PCB variant of GBS with possible overlap with acute ataxic neuropathy was made according to the revised criteria for diagnosis of PCB variants of GBS,<sup>6</sup> even though the nerve conduction studies revealed normal findings.

Abnormality in nerve conduction studies in the form of demyelinating neuropathy or axonal variety are almost always seen in cases of GBS.<sup>1</sup> However, a study conducted by Luigetti *et al* in 2015, revealed that 37% of the patients

who underwent an early NCS ( $\leq 4$  days) showed normal neurophysiological results.<sup>5</sup> This might have been the reason for the normal nerve conduction study that was observed in our patient, as we performed the study on the 5<sup>th</sup> day of illness.

Additionally, a positive titre of anti-GT1a IgG antibodies would have been very helpful to support the diagnosis of PCB syndrome in this child as it is a common occurrence in PCB,<sup>6</sup> but these serological studies were not performed due to the unavailability in our setting.

As for any case of GBS, even for PBC variant, intravenous immunoglobulin and plasmapheresis/ total plasma exchange are effective modalities of treatment.<sup>4</sup> Supportive care including prevention of aspiration, prevention of chest infection, provision of respiratory support, physiotherapy, nutrition, psychological support and rehabilitation also played a crucial role for a successful outcome as in this case.

### Conclusion

Pharyngeal-cervical-brachial variant of GBS should be considered in patients presenting with acute onset bulbar and upper extremity weakness in differential diagnosis to enable early initiation of definitive treatment and supportive care.

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