CASE REPORT

PHARYNGEAL-CERVICAL-BRACHIAL VARIANT OF GUILLAIN BARRE SYNDROME

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Abstract:

The Pharyngeal- Cervical-Brachial (PCB) variant of Guillain- Barre Syndrome (GBS) is a rare presentation, characterized by marked weakness of the oro- pharynx, neck and proximal upper limbs sparing power and reflexes of the lower limbs. It presents as rapid progressive pharyngeal weakness and often needs intubation and ventilation due to excessive secretions. Diagnosis is supported by high CSF protein and absent F waves in upper limbs in electro-physiological studies. Diagnosis can be confirmed by doing ant DT1a antibodies. We present the case report of a 6-year-old patient who presented with progressive difficulty in swallowing due to oro-pharyngeal muscle weakness.

Keywords: Guillain- Barre Syndrome, Pharyngeal- Cervical-Brachial variant

Introduction

Guillain Barre Syndrome (GBS) is a post infectious polyneuropathy mainly involving motor nerves with sensory and autonomic nerve involvement occurring less frequently. It is predominantly caused a demyelinating polyneuropathy, though axonal degeneration is also reported[1]. It is usually preceded by a nonspecific gastro intestinal infection or a respiratory infection. In the classic form it follows a Landry ascending paralysis where weakness begins usually in the lower extremities and progresses upwards. Onset is gradual and progresses over days or weeks[12]. Bulbar involvement occurs in about half of the cases resulting in facial weakness, dysphagia and respiratory paralysis.

There are a few variants of GBS which do not fallow the above pattern. These are namely the Miller Fisher (MF) and the Pharyngeal-Cervical-Brachial (PCB) variants. In these variants bulbar muscles are affected early in the course. Miller Fisher syndrome is characterized by opthalmoplegia, ataxia and areflexia. If cognition is impaired Bicker- staff brain stem Encephalitis (BBE) should be considered. The pharyngeal-cervical-brachial variant is rare and is characterized by acute progression of oro-pharyngeal, neck and shoulder weakness.

Received:19/09/2019 **Accepted revised version:05/12/2019** **Published: 31/12/2019**

Case report

A 6-year-old boy was transferred from a peripheral hospital for ICU care due to progressive difficulty in swallowing caused by oro-pharyngeal muscle weakness. He was first admitted to a peripheral hospital with acute onset difficulty in swallowing and drooling of saliva associated with mild fever. There had been no stridor or respiratory distress and he was not drowsy on admission to the peripheral hospital. Limb or facial weakness had not been present on examination. He subsequently needed intubation and ventilation for 2 days due to risk of aspiration caused by excessive secretions and salivation at the local hospital. He was transferred to our unit which is a tertiary care hospital for further evaluation and management as there was no improvement of symptoms after 48 hours of extubation. He was also noted to have weakness of the neck and upper limbs. He was able to walk and there was no ataxia. His cognition was not affected and there were no bladder or bowel incontinence. There was no clear history of recent infection.

On Examination, at our unit revealed bilateral lower motor neuron type facial nerve palsy with associated pharyngeal weakness. Eye movements were normal and other cranial nerves were not affected. Neck muscles were weak and upper limbs showed grade 3-4/5 weakness with hypo-reflexia. Lower limb examination was normal. Cardiovascular, respiratory and abdominal examinations were normal.

MRI brain was normal and nerve conduction studies showed absent F waves in upper and lower limbs. Lumbar puncture showed a high protein content (80 mg/dl) with normal white cell count. All other blood tests including renal and liver function test were normal. The presence of anti ganglioside anti bodies could not be assessed as this test is not available in Sri Lanka.

He was fed with a naso-gastric tube and was self-ventilating throughout hospital stay. He was treated with intravenous immunoglobulin and also received supportive therapy. Swallowing was assessed by a speech and language therapist at regular intervals. He improved gradually and was sent home after 3 weeks once he was able to eat and drink without aspiration.

Discussion

In 1986, Ropper described 3 patients with Pharyngeal-Cervical-Brachial variation and since then a few cases have been described[3]. There are a few cases reported in adults from Sri Lanka but we could not find any records on paediatric cases[4].

PCB is a rare variant of GBS accounting for 3 % of cases[1]. It typically presents as rapidly progressive oropharyngeal and cervicobrachial weakness, with associated areflexia of upper limbs. The muscle power of the lower limbs is not affected or only mildly affected indicating that PCB is a localized subtype of GBS. Affected patients display bilateral ptosis and facial weakness without involvement of the extra ocular or intra ocular muscles. If eye muscles are affected overlap with MF or BBE should be considered. The cerebro spinal fluid (CSF) protein levels in patients with PCB is high and electro physiology shows absent F waves in upper limbs and occasionally in the lower limbs as well. Differential diagnosis includes brain stem ischemia, myasthenia gravis, diphtheria and botulism[5].

The immune mechanism and neurophysiological features of GBS variants have been studied deeply and provide useful insight into the classification[3]. Auto anti bodies against

specific neuronal gangliosides have been implicated in the pathogenesis of different GBS variants.IgG anti-GM1 and anti-GD1a antibodies are associated with acute motor axonal neuropathy (AMAN), but not acute inflammatory demyelinating polyneuropathy (AIDP). In contrast, IgG anti-GQ1b antibodies are associated with MFS and BBE, both of which are characterised by ophthalmoplegia and ataxia. The strongest association for PCB is the presence of IgG anti-GT1a antibodies which is required for the confirmation of diagnosis. Unfortunately this test is not available in Sri Lanka and hence it could not be done. However we feel that the clinical picture of this patient is highly suggestive of PCB[6].

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