

Case Report

Pharyngeal-cervical-brachial variant of Landry Guillain Barre syndrome after dengue infection: a case report

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ABSTRACT

Dengue is a common mosquito-borne infection in India. We reported a rare Pharyngeal cervical brachial (PCB) variant of Landry Guillain Barre syndrome (LGBS) associated with the dengue virus infection. The pathogenesis seems to be molecular mimicry between gangliosides and microbial lipo-oligosaccharides. PCB usually presents with oropharyngeal or cervicobrachial weakness. Therefore, it must be recognised early and distinguished from conditions presenting with cephalocaudal progressing weakness, such as Myasthenia Gravis, Miller-Fisher syndrome, botulism, diphtheria, porphyria or brain stem stroke. The aim of the study was to add to the limited literature on the PCB variant of LGBS after dengue infection and shed some light on presentation and management options for this rare entity.

Keywords: Pharyngeal cervical brachial variant, Landry Guillain Barre syndrome, Dengue, Cephalocaudal weakness, Oropharyngeal weakness

INTRODUCTION

Dengue is the second most common mosquito-borne disease affecting humans after malaria.¹ Dengue can lead to many neurologic complications, including dengue encephalopathy, encephalitis, ADEM, transverse myelitis, stroke, neuro-ocular (maculopathy, papilledema, oculomotor paresis) and neuromuscular disorders (different variants of Landry Guillain Barre syndrome (LGBS), myositis).²

Different variants of LGBS, including Miller Fisher, Acute motor axonal neuropathy (AMAN), Acute inflammatory demyelinating polyradiculoneuropathy (AIDP), have been reported during or after dengue infection.³ However, only a very few cases of the Pharyngeal-cervical-brachial (PCB) variant of LGBS have been reported after dengue. Here, we discuss the symptoms, examination and

management of a patient who suffered from a PCB variant of LGBS.

CASE REPORT

A 50-year-old male presented to the emergency department with complaints of rapidly progressive difficulty in swallowing, nasal intonation of speech, difficulty in closing eyes and manipulating food inside the mouth from the last four days.

Over the previous two days, he also had difficulty raising hands above his head and performing hand activities like gripping and breaking chappatis. For the last 24 hours, he had noticed difficulty getting up from a sitting position. The patient had a fever with thrombocytopenia and was diagnosed and treated as a case of dengue infection (ELISA-NS1 positive) two weeks back.

On examination- GCS was 15/15, EOM was normal.

He had bilateral facial muscle weakness, bilateral 9th, 10th, 11th and 12th cranial nerve palsy. The rest of the cranial nerves are within normal limits. His breath-holding time was 12 sec. He had proximal (grade 2) more than distal (grade 4) weakness in upper limbs with a bilateral handgrip of approximately 30%. In lower limbs, power in proximal muscle was grade 4 and distal was grade 5. Sensory examination and cerebellar tests were within normal limits. There was generalised areflexia, and plantars were downgoing bilaterally.

Possibilities of PCB variant of LGBS, myasthenia gravis, porphyria and brain stem lesion were considered. Routine investigations were within normal limits except that his ELISA IgM dengue came out to be positive. Nerve conduction studies showed demyelinating polyradiculoneuropathy involving both upper and lower limbs with impersistent F waves. Repetitive nerve stimulation studies were within normal limits.

MRI brain and MRI C-spine were within normal limits. CSF examination showed raised proteins (120 mg/dl) with normal sugar levels and cells. The rest of the CSF examination was unremarkable. The patient was diagnosed as a PCB variant of LGBS and treated with 2 g/kg of IVIG over five days. The patient responded well to the therapy.

DISCUSSION

The PCB variant of GBS is an infrequent entity. As the name suggests, it involves the oropharynx manifesting as oropharyngeal weakness and cervicobrachial weakness and loss of reflexes in upper limbs.⁴ Similar to classic LGBS, PCB has been linked with antecedent upper respiratory tract infections and diarrhoea. These include *Campylobacter jejuni*, herpes virus, cytomegalovirus, dengue virus, Epstein Barr virus, etc.⁵ A case of PCB has also been reported after a co-infection with dengue and chikungunya virus.⁶ PCB can also present para- or post-COVID-19 infection.⁷

Recent history of dengue infection, positive dengue IgM ELISA test, and endemicity of dengue virus with a seasonal spread of dengue virus during this time favours the diagnosis of dengue-associated PCB variant of LGBS in our case. The mechanism by which the dengue virus induces the development of the PCB variant of GBS has not been studied much, as only a few such cases have been reported. Still, most likely, pathogenesis seems to be molecular mimicry between gangliosides and microbial lipo-oligosaccharides.⁸

PCB variant causes weakness in the cephalocaudal (downward) direction, unlike other LGBS variants where we see ascending weakness. This makes it essential to differentiate it from similar conditions, including myasthenia gravis, Miller Fisher syndrome, botulism,

diphtheria or brain stem stroke. Clinical history, nerve conduction studies, brain imaging, and findings of albuminocytologic dissociation on CSF analysis, along with neurological examination, helps in differentiating PCB from other causes of downward progressing weakness.⁵

Some patients with PCB variant may develop mild lower limb weakness, but it is not prominent than orofacial and upper limb weakness. Patients with PCB variant may develop ataxia and ophthalmoplegia, thus leading to considerable overlap between PCB variant and Miller Fisher syndrome, supporting the view that both the syndromes form a continuous spectrum. PCB variant may be associated with the presence of IgG anti-GT1a antibodies.⁹

Recognising the disease on time and starting prompt treatment would result in a better prognosis similar to our patient's. Treatment options include IVIG and plasma exchange.¹⁰ Our patient was treated with 2 g/kg of IVIG over five days. As a result, the patient's symptoms gradually improved, and the patient had complete functional recovery at the end of 6 weeks. This suggests that treatment of PCB after dengue works similar to the treatment of PCB following other infections.

CONCLUSION

PCB variant of LGBS is an uncommon neurological complication seen during and after Dengue infection. Physicians should always suspect this entity after encountering patients presenting with oropharyngeal and cervicobrachial weakness following dengue infection. A delay in the diagnosis and treatment might affect the outcome, and early treatment with IVIG may give gratifying results. Knowledge about the presentation, differential diagnosis, and management of the PCB variant of GBS is this report's most important take-home message.

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