

# Acute multiple cranial neuropathy: An oculopharyngeal variant of Guillain-Barré Syndrome

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## CASE STUDY

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## ABSTRACT

We report the case of a 20-year-old male who presented to us with acute bilateral multiple cranial neuropathy in the form of bilateral total ophthalmoplegia and bulbar dysfunction. The patient had normal haematological and biochemical investigations, however, cerebrospinal fluid (CSF) analysis showed raised protein (96mg/dl) in the second week of illness. Peripheral nerve conduction studies and an MRI of the brain were normal. The patient showed gradual improvement after three weeks of supportive treatment. Considering the course of illness and the clinical and investigational profile, a diagnosis of an oculopharyngeal variant of Guillain-Barré syndrome (GBS) was made.

### Key Words

Multiple cranial nerve palsy, Guillain-Barré syndrome, Oculopharyngeal GBS

### Implications for Practice:

#### 1. What is known about this subject?

Guillain-Barré syndrome covers the heterogeneous group of acute immune-mediated polyradiculoneuropathies, usually

presenting as post-infectious ascending paralysis. Cranial nerves can be affected in a minority of cases.

#### 2. What is the key finding in this case report?

We report a rare presentation of GBS with affection of multiple cranial nerves with the exception of facial nerve and without any peripheral nerve involvement.

#### 3. What are the implications for future practice?

GBS may present rarely as multiple cranial nerve palsies without any peripheral nerve involvement, so it needs to be ruled out as a cause of pure cranial nerve palsies.

## Background

Multiple cranial neuropathies are rare and may relate to infectious, inflammatory, or neoplastic lesions of the skull base or brainstem. They may also be associated with systemic diseases, or may occur as an accompaniment of a generalised polyneuropathy. Widespread cranial nerve palsies have been noted in large series of patients with Guillain-Barré syndrome (GBS), however, GBS presenting with only multiple cranial nerve dysfunction occurs very rarely.<sup>1</sup> We present a case with sole affection of the cranial nerves, but sparing the facial nerve and other peripheral nerves. To our knowledge this presentation of GBS has not been reported from India to date.

## Case details

A 20-year-old male presented with complaints of acute onset of double vision followed by drooping of both eyelids, difficulty in swallowing, nasal twang in his voice, and nasal regurgitation of liquids. These symptoms progressed slowly over the subsequent four days. He denied any other complaints, including history of snake bite.

On examination he had bilateral ptosis (Figure 1), total ophthalmoplegia with absent pupillary reflexes and mydriasis, suggestive of bilateral third, fourth, and sixth nerve palsies. He had bulbar type dysarthria and an absent gag reflex, which were indicative of bilateral ninth and tenth cranial nerve palsy. Corneal reflexes (blink testing) were

intact bilaterally and examination of the facial nerve was normal. Other physical and neurological findings were unremarkable.

Laboratory studies, including complete blood count, ESR, electrolytes, biochemical investigation, and chest x-ray were normal. A cerebrospinal fluid (CSF) analysis performed on the sixth day of illness was normal but repeat CSF analysis on day 14 of illness showed a markedly elevated total protein concentration (96mg/dl). Peripheral nerve conduction study and an MRI of the brain were normal on day eight of illness. GQ1b antibody testing (Miller Fisher test) was negative. Electrophysiological testing of the seventh cranial nerve could not be performed for logistic reasons. A modified Tensilon (using neostigmine) challenge test did not show any change in severity of symptoms or signs.

**Figure 1: Bilateral ptosis**



The acute onset of multiple cranial nerve palsy over five days duration without other associated symptoms such as seizures, headache, vomiting, confusion, weakness of any limb, constitutional symptoms, and absence of history of snake bite with a normal MRI of the brain and normal CSF (except raised proteins in the second week of illness) made diagnosis of infections, ischaemia, vasculitis, and neoplasms unlikely. The patient did not have fluctuating ophthalmoplegia, moreover a negative modified Tensilon challenge test ruled out myasthenia gravis. The patient's presentation did not favour diagnosis of botulism as an etiology. Normal chest x-ray and haematological, serological investigations ruled out a systemic disease manifesting as bilateral cranial nerve palsy (diabetes, sarcoidosis, connective tissue disease, endocrinopathy). A normal contrast MRI ruled out hypertrophic meningitis and cytology was performed to exclude carcinomatous meningitis. These rare entities can present with multiple cranial neuropathies as well as symptoms caused by cord or root compression.

The course of the illness, pattern of cranial nerve involvement with raised proteins in the second week of illness favoured a diagnosis of cranial variant of GBS after excluding other causes of multiple cranial nerve palsy.

The patient was treated with supportive treatment and intravenous immunoglobulin (IVIG). His ptosis, diplopia, dysphagia, and nasal regurgitation started to improve gradually after 21 days. He has received regular follow-up for more than two years and has shown consistent improvement over the time with achievement of full recovery after five months and no recurrence of any symptoms.

## Discussion

Multiple cranial nerve dysfunctions<sup>1,2</sup> can occur with primary intracranial pathology (neoplasms, demyelinating lesions, ischaemia, aneurysms, congenital defects, dural vein thrombosis, infection, cranial nerves neuropathy, vasculitis), neuromuscular junction disorder (myasthenia gravis, botulism), myopathy, and as a manifestation of a systemic disorder (sarcoidosis, diabetes mellitus, endocrinopathy, connective tissue disorder, heavy metal poisoning, diphtheria, porphyria). However, when the clinical syndrome is acute in onset, self-limiting, and without any other significant complaints except cranial nerves dysfunction, most of the above possibilities are less likely to be the causative factors.

GBS is an acute, frequently severe and fulminant polyradiculopathy that is autoimmune in nature. GBS manifests as rapidly evolving areflexia, motor paralysis with or without sensory disturbances. Tingling, prickling, or pins and needles sensation are usually followed within hours or days by symmetric leg weakness and trouble walking. Weakness of the upper limbs and ocular, oropharyngeal, and facial muscles develops with variable frequency and severity.

In the early 1920s, Georges Guillain<sup>3</sup> proposed a clinical classification of the GBS based on topographic consideration and divided GBS in four categories:

1. Patient with involvement of extremities only.
2. Patient with involvement of both extremities and cranial nerves.
3. Patient with syndrome confined to cranial nerves only.
4. Patients with polyradiculopathy and mentation changes.

A number of variant or restricted forms are included under the same umbrella term of GBS because they share many of the same clinical, electrophysiological, and CSF findings, or there is a transition to the typical disease after an infectious illness. Cranial nerve dysfunctions may be preceded or followed by extremities involvement. Out of all cranial nerves, facial nerve and oculomotor nerve involvement is present in 50 per cent, and 10–20 per cent of patients, respectively.<sup>6</sup> GBS presenting with only multiple cranial nerve dysfunction occurs rarely.<sup>1, 3, 7–10</sup>

Pure cranial nerve involvement was described by Guillain and Kreis<sup>3, 7</sup> as well as by Van Bogart.<sup>8</sup> Van Bogart proposed that there might also exist a syndrome of bilateral cranial neuropathy without clinical involvement of the limbs. Munsat and Barnes<sup>1</sup> reported five patients aged 14 months to 40 years, with acute multiple cranial neuropathy. Because their clinical features and course were similar to those of patients with typical GBS, the investigators suggested their patients had a regional variant (i.e., a cranial form) of GBS.

In most of the previously reported cases authors described ophthalmoplegia with sparing of pupillary function and supranuclear gaze palsies when ophthalmoplegia recovers, suggesting interruption of gaze pathways in brain stem and lower motor neuron type of involvement of other cranial nerves because of peripheral mechanism of disease process. Only small proportions were having internal ophthalmoplegia as seen in our patient. Facial diplegia was also frequently observed in previously reported cases of cranial variants of GBS. The pattern of involvement of cranial nerves in the form of complete ophthalmoplegia with bulbar dysfunction with sparing of facial nerves is atypical in our case in comparison to previously reported cases.

### Conclusion

In summary, pure cranial variant of GBS should be considered when clinical presentation is of acute multiple cranial nerve palsies without signs and symptoms suggestive of other etiologies of cranial nerve dysfunction. An oculopharyngeal variant is likely if the facial nerve is spared. GBS has various ramifications presenting with atypical variants but the course of illness, pattern of involvement of the nervous system, laboratory and electro-diagnostic studies are the clues for diagnosis. A high index of suspicion and systematic workup is required to rule out other causes, which can then lead to the correct diagnosis and early institution of treatment.

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### PEER REVIEW

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### CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

### PATIENT CONSENT

The authors, Dosi R, Ambaliya A, Patel N, Patell R, and Shah M, declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.