Case Report

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Pharyngeal-Cervical-Brachial Variant of Guillain Barre Syndrome

Yashif N A[1], Kishalay Datta[2], Priya Govil[3], Shebin Althaf[4]

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Author's Affiliation: [1]Post Graduate trainee, [2]Director and HOD, [3]Associative Consultant, [4]Resident, Department of Emergency Medicine, Max Superspeciality Hospital, Shalimar Bagh, Delhi 110088, India.

Corresponding Author: Yashif N A, Post Graduate trainee, Department of Emergency Medicine, Max Superspeciality Hospital, Shalimar Bagh, Delhi 110088, India.

E-mail: yashifbava2010@gmail.com

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Abstract

Guillain-Barre Syndrome (GBS) usually present as[1] symmetrical ascending muscle weakness associated with areflexia and flaccidity with or without sensory symptoms. However,[2] in some patients it may present atypically and one of the rare variant is Pharyngeal-cervical-brachial variant, usually presented as muscle weakness extending from cervical area to the upper extremities often affected with proximal muscle groups.

A 20-year-old male presented in ER with complaints of weakness over bilateral upper limb, dysphagia and difficulty in speaking followed by quadriparesis and breathing difficulty. Diagnosis of PCB variant was made by CSF and nerve conduction study and all other possible differentials were excluded. The patient improved by administration of iv immunoglobulin and other supportive measures. Physician should think about PCB variant of GBS, whenever a patient presenting with symmetrical upper limb weakness and bulbar palsy.

Keywords: Guillain barre syndrome; Pharyngeal-cervical-brachial variant; Atypical presentation.

Introduction

Guillain-Barre Syndrome (GBS) is an immune -mediated demyelinating inflammatory polyneuropathy, typically characterized as acute onset of symmetrical muscle weakness with decreased tendon reflexes or areflexia.

Patients usually presented as an ascending paralysis having rubbery legs associated with paresthesia/numbness over bilateral lower limbs. Pharyngeal-cervical-brachial (PCB) is a rare entity of GBS is usually involving muscle groups extending from cervical area to proximal upper extremities and was first reported by Sir Ropper.[3] It is often falsely diagnosed as myasthenia gravis, brainstem stroke, diphtheritic polyneuropathy or botulism. Miller-Fisher syndrome is another entity having additional features of[4] ophthalmoplegia, ataxia and cns symptoms.

Case

A 20-year-old male presented to ER with one week history of weakness over bilateral upper limbs, difficulty in swallowing and difficulty in speaking with h/o fever and cough prior one week back. Patient was apparently normal two weeks back with h/o fever and URTI symptoms for one week followed by upper limb weakness, which was insidious in onset, gradually progressing (from proximal to distal); Patient father noticed that he had difficulty in holding objects and with overhead movements, associated with dysphagia and difficulty in speaking followed by clumsiness in lower limbs (note able to kick his motorcycle).

Patient was then admitted in outside hospital, symptoms got worsened followed by breathing difficulty and referred to our hospital. There was no history of syncope, seizure and bowel/bladder incontinence at the onset of the disease. The patient's medical history was unremarkable, did not receive any immunizations recently. Also provides no history of drugs or heavy metal exposure and no similar illness in the family.

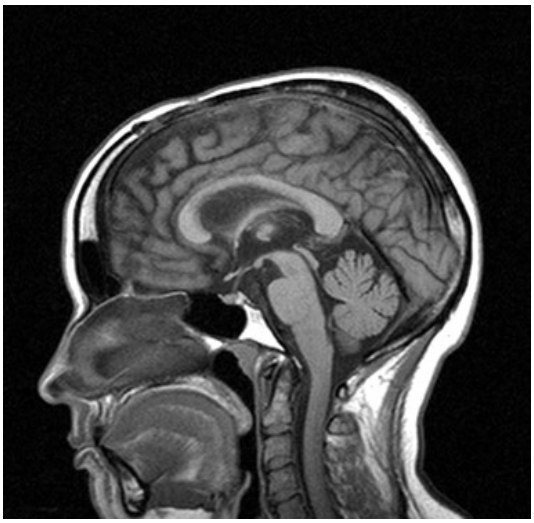
General physical examination was drowsy, with saturation around 94% on full flow NRBM, BP – 140/100 and GCS – E1V1M1, pupil-miosis with sluggishly reactive. Cranial nerve and sensory nerve examination could not be assessed due to low GCS. Power was of grade 0/5 in the upper limbs and 0/5 in the lower limbs in (MRC) grading. Deep tendon reflexes were absent in bilateral upper and lower limbs and plantars were mute. Examinations of other systems were unremarkable. Patient was intubated in view of low GCS and airway was threatened. Differential diagnosis of brainstem stroke, myasthenia gravis, botulism, Diphtheric polyneuropathy, intracranial mass occupying lesion and PCB variant of GBS were made.

Lab values

White cell count	5700
Differential count	N65 L32 E2
HB	13
Calcium	2.3
Blood urea/Creatinine	38/1
Sodium/Potassium	137/4.3

CSF analysis

White cell	4
Glucose	3
Protein	70
RBC	Nil



MRI brain and cervical spine were normal, increased latencies were observed in NCV study of the right and left median nerves (CPN) (9.44 milliseconds and 12.5 milliseconds); diagnosis pointed towards acute inflammatory demyelinating polyneuropathy. Antibody testing was not feasible due to financial issues of the patient.

Diagnosis was based on clinical scenario, CSF findings, and NCS report. He was managed conservatively with support of immunoglobulin and tracheostomized after 4 days. Gag reflex and power was patent 5/5 in bilateral upper and lower limbs at the time of discharge. On further visits the deep tendon reflexes of lower limb also recovered.

Discussion

The important symptoms in our patient were weakness of upper limbs, dysphagia, and difficulty in speaking followed by quadriparesis and breathing difficulty.

First, we rule out intracranial mass occupying lesion and brain stroke by brain imaging study which was normal. Diphtheria infection was omitted by negative history of tonsillar pain, pharyngeal bleeding and bulls' neck and no history

intake of preservatives/ honey or autonomic dysfunction, so botulism was excluded. Since tone and reflex were abnormal, myasthenia gravis ruled out. By evaluating clinically, monitoring lab parameters and electrophysiological findings, a diagnosis of GBS was made. Hence, our patient fulfilled the criteria for PCB variant.

The pathogenesis is mainly due to molecular mimicry between gangliosides and microbial lipo-oligosaccharides.[5] with prior history of upper respiratory tract infection or diarrhea. Serological studies show Campylobacter jejuni is the main culprit for this disease while patient presented to us had h/o history of URTI.[6] In patients with PCB variant of GBS, the reflexes may be absent in the arms, or may be generalized areflexia and the lower extremities are spared or mildly affected, however in our patient power in bilateral lower limbs were not preserved (i.e. 0/5) with generalized areflexia. Patients are treated usually with intravenous immunoglobulins (IVIg) or plasmapheresis.

Our patient recovered rapidly with the help of immunoglobulin, Power regained and respiratory compliance improved and patient was extubated. This can be regarded as severe manifestation PCB variant of GBS.

Conclusion

GBS may present atypically as PCB variant and likely to get easily misdiagnosed. So, we should give suspicion for PCB variant in any patient presenting with bulbar palsy and symmetrical upper limbs weakness.

Before confirming diagnosis, we should rule out all other differentials like: brainstem lesion, neuromuscular disorder, diphtheritic polyneuropathy, and botulism.

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