ISSN Print: 2664-8881
ISSN Online: 2664-889X
Impact Factor: RJIF 8
IJMS 2023; 5(2): 12-17
www.medicinejournal.in
Received: 05-06-2023
Accepted: 11-07-2023

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Atypical presentation of GBS: Pharyngio-cervical brachial variant progressing to descending paralysis

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DOI: https://doi.org/10.33545/26648881.2023.v5.i2a.38

Abstract
Guillian-Barre Syndrome (GBS) presents typically as ascending symmetrical flaccid muscle weakness, with or without sensory symptoms and areflexia. In some patients it may present atypically as one of the multiple variants of GBS which are known. Pharyngeal-cervical-brachial variant is a rare variants and it is characterized by involvement of weakness of muscle of oropharyngeal, neck and proximal upper extremities, which may further progress to rare pattern of descending paralysis. Pharyngeal - cervical-brachial and descending paralysis variant is an unfamiliar entity, very often misdiagnosed as botulism, myasthenia gravis and brainstem stroke, causing undue delay in treatment leading to bad outcomes. This is a case report of the rare pharyngeal-cervical-brachial variant of GBS progressing to descending paralysis. A 32-year-old Indian male presented with weakness of dysphagia, bilateral upper limb and nasal intonation of voice which later involved his trunk and lower limbs. A diagnosis of pharyngeal-cervical-brachial variant of GBS with descending paralysis pattern was made after exclusion of other differentials based on clinical history, examination, cerebrospinal fluid analysis and nerve conduction study. The patient improved significantly with immunoglobulin therapy with no neurological deficit. Pharyngeal-cervical-brachial variant of GBS should be considered a differential in any patient presenting with bulbar palsy and symmetrical upper limb weakness. This is to ensure early diagnosis, treatment, and follow-up of the potential complications.

Keywords: GBS, gullian barre syndrome, PCB (pharyngeal cervical brachial), descending paralysis, acute demyelinating syndrome, immunoglobulin G

Introduction
Guillian-Barre Syndrome (GBS) is an immune-mediated, demyelinating, inflammatory polyradiculopathy, characterized by acute symmetrical flaccid muscle weakness with absent or decreased deep tendon reflexes [1]. Patients present usually with ascending paralysis that is noticed as rubbery feeling in legs associated with a tingling sensation [2]. The lower cranial nerves are also frequently involved [3]. Pharyngeal-cervical-brachial (PCB) variant of GBS very rare and is characterized by muscle weakness involving oropharyngeal, neck, and upper extremity muscles, and was reported for the first time by Ropper in 1986 [4-5].
PCB variant of GBS is unfamiliar, which is often misdiagnosed as brainstem stroke, myasthenia gravis or botulism. The additional features of ophthalmoplegia and ataxia point towards overlap with Miller-Fisher syndrome [6]. Herein, we would like to report a case of PCB variant of GBS which progressed rapidly as descending paralysis, but with early dignosis and initiaion of treatment with immunoglobulin g made a full recovery.

Case Report
Patient 32 years old male presented to Jaipur golden hospital on 28.12.2021 with chief complaints of progressive shoulder girdle weakness heaviness which started in the left side with inability to lift left arm overhead 3 days ago. The patient visited a general practioner and orthopedic surgeon for the same where he was prescribed anti-inflammatory, analgesics and physiotherapy but did not improve his condition. The weakness further progressed to involve the right shoulder and upper arm with inability to lift both arms above head and difficulty combing his hair and getting dressed. At this moment his wrist and hand muscles were intact and he could perform fine activities like buttoning unbuttoning using mobile

laptop. Thereafter he started feeling difficulty in chewing and swallowing food solids > liquids, slurring of speech and changes in his voice a nasal twang noticed by family member. Along with his shoulder weakness had progressed further to involve his elbows neck back and abdominal muscles wherein the patient was unable to get up from supine to siting position. He also started feeling weakness in both his hands with inability to hold glass of water and dropping things. He then came to the emergency department on 28.12.2021. His lower limbs was not involved and he was able to walk and get up from sitting to standing position. His bowel and bladder were intact. He has no history of any chronic illness like DM II/thyroid /HTN/CVA/heart disease. Family history was not significant. He was an occasional drinker of Alcohol, with no other addictions. He is a non-vegetarian with normal bowel bladder habits. He had history of diarrhea with low grade fever for 3-4 days during 22.12.2021 to 25.12.2021 for which patient had taken only symptomatic treatment.

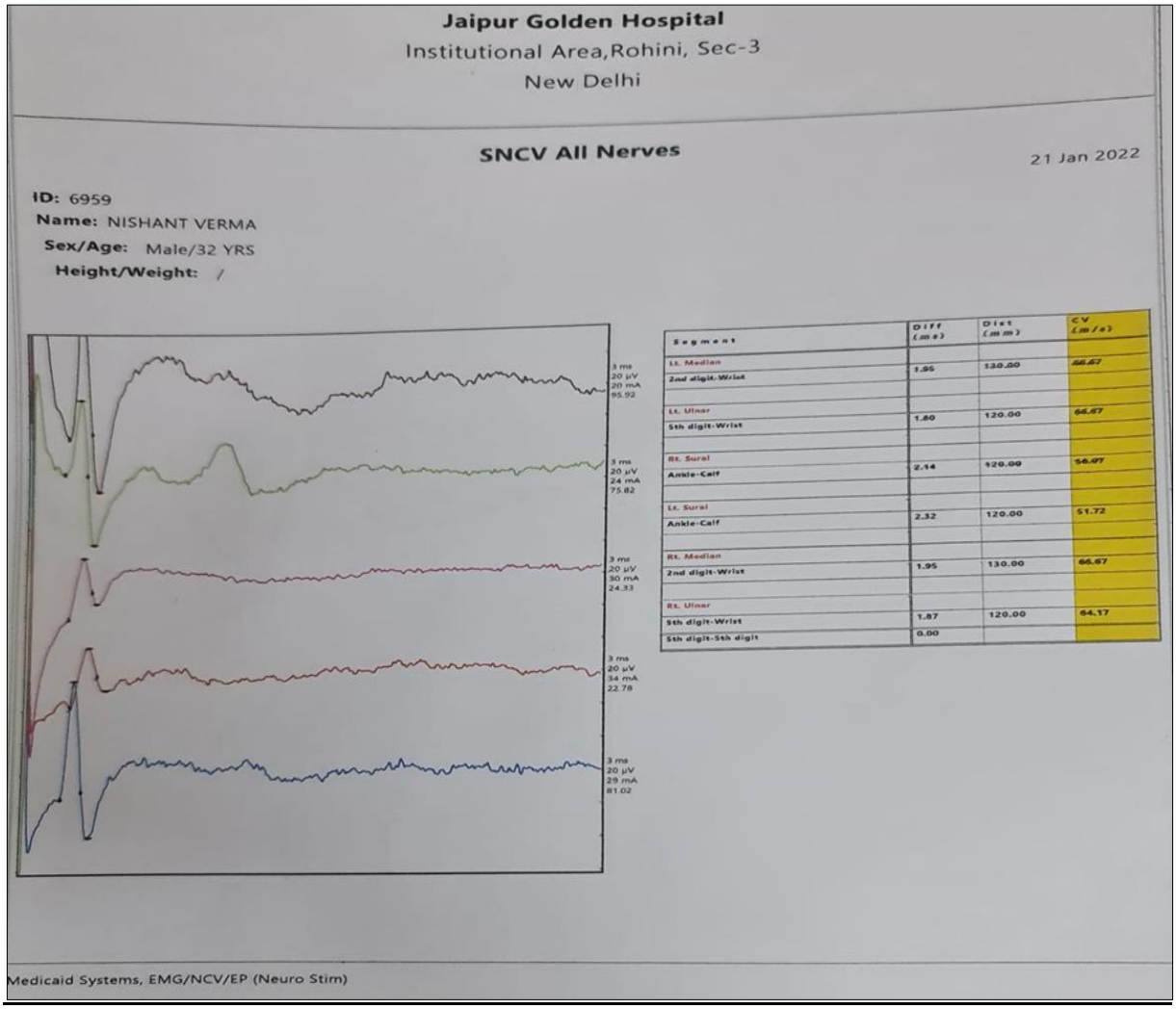
There was no history of seizures, diplopia, vertigo, numbness, parasthesia, headache, difficulty walking, backache, neck stiffness, altered sensorium, arthralgia, oral ulcers, rash, photosensitivity no history of drugs or heavy metal exposure no immunization in past 6 months.

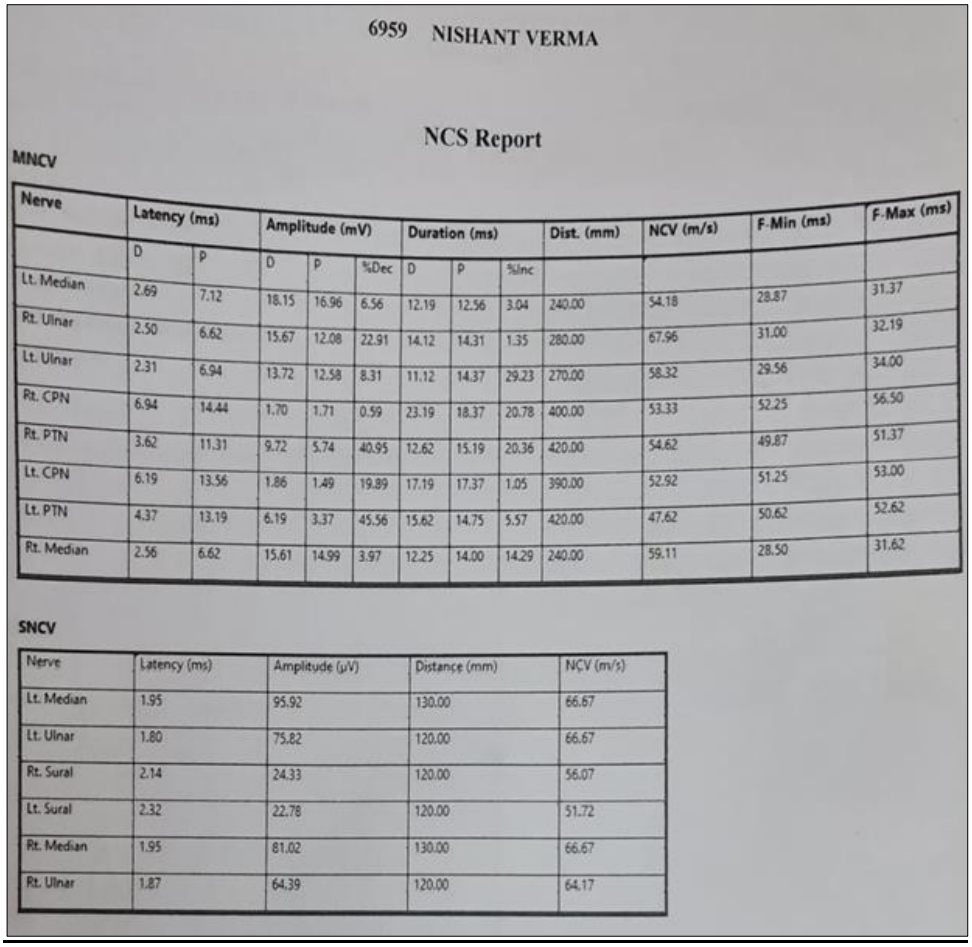
On examination patient was anxious vitals were stable CVS RS per abdomen examination were within normal limits. CNS examination was done in detail. Higher mental status were intact with MMSE score was 29/30. He had dysarthria, weak gag reflex, nasal intonation of voice and weak cough reflex diagnostic of bulbar palsy, no facial deviation or facial nerve involvement. All other normal bulk and tone in all limbs of power of 1/5 in all muscles of bilateral upper limbs with significantly depressed deep tender reflexes no dermatosomal sensory loss. Lower limbs had power of 4/5 depressed DTR, plantars bilateral acute with no sensory loss. Cerebellar signs like finger nose and dysdiadochokinesia could not be done due to bilateral upper limb motor weakness. Knee test was normal for both limbs. On admission patient was normal but with absent bilateral arm swing (? GUNSUNGER'S GAIT). There was no ataxia and Rhomberg's sign was negative with the history & physical examination differential diagnosis of intracranial mass occupying lesions, myasthenia gravis, botulism, diphtheria polyneuropathy, brainstem stoke & PCB variant of GBS were considered. Pts routine investigation were normal and mentioned in Tablet 1. Further NCCT head was done to rule out stoke or intracranial mass which was done. MRI Brain with whole spine screening with contrast done to r/o spinal lesions and brain lesions and was reported Normal in view of narrowing diagnosis to GBS variant. Nerve conduction test of all 4 limbs and facial nerve done which was suggestive of significant. Acute inflammatory demylinating polyneuropathy of all 4 limbs nut normal NCV of facial nerve with normal blink reflex. Electromyography of all 4 limbs was suggestive of neurogenic pattern with incomplete recruitment. Further a lumbar puncture consent was taken and performed CSF studies showed clear albumino cytological dissociation with CSF protein of (> 400mg/dl) and normal cell count. Other CSF parameters were within normal limits. Patient was given one dose of Inj. Solumedrol (Methyl prednisolone 1gm IV OD) after consultation with neurologist. During the hospital Stay the patient weakness progressed rapidly over 26-36 hours to involve both his lower limbs with a descending paralysis pattern. A diagnosis of PCB variant with descending paralysis based on BRIGHTON'S criteria table was made and patient initiated on IV immunoglobulin after wiling out any patient infections at a dose of 2 gms / kg body weight patient is 30 kgs thus total of 160 gms of I.V Ig given over 5 days by continuous IV infusion @ 25 ml /hrs (from vial of 5 gm in 100ml) i.e 30 gm in one day. The weakness did not progress any further over first 2 days with increasing power of shoulders. Unfortunately patient developed spike of high grade fever (102* f) on 3rd day of I.V Ig therapy was with hold. His antibiotics were stepped up from IV ceftriaxone to I.V piperacillin + tazobactam (4.5 gm I.V, QID) and injection linezolid (600 mg, I.V. BD) along with full fever profile which came out normal. Also his fever subsided and there were no repeat episode. Thus I.V Ig Therapy restarted on day and complete dose given without any further complications. Patient responded very well to the treatment. Patient was discharged on Day 12 of admission with normal DTR and power of 4/5 in bilateral upper limbs and 5/5 in lower limbs. His gait also normalized planters were flexors voice become normal with no difficulty in speaking or swallowing. Rest of the neurological examination was also normal one follow up focus weeks later the patient routine test were normal, his nervous system was intact he regained power of 5/5 with residual neurological weakness and was able to perform his daily routine without any problems. He was advised to continue physiotherapy and exercise. Informed written consent taken from patient to report the case and share related images.

Table 1: PTS routine investigation

Investigations	28/12/21 (D1)	30/12/21 (D3)	2/1/22 (D6)	5/1/22 (D9)
Haemoglobin (g/dl)	14.6		15	14
Tlc (10^9/l)	9.1		10	9.8
DLC	30/20/5/2/0		32/20/4/1/0	
Platelet (10^9/l)	200		246	212
S. Creatinine (mg/dl)	0.7			0.8
Bun	7			10
Serum Calcium (mg/dl)	10.1			
Ionic Calcium (mg/dl)	5			
Sodium (mmol/l)	142			140
Potassium (mmol/l)	4.8			3.9
Magnesium (mg/dl)	2			
T4/T3 (mcg/dl /ng/dl)	6/100			
S.TSH miu/L	3			
Lipid profile (triglycerides)	350			
Hba1c	5.5			

S. Vitamin B12 (pg/ml)	800			
S. Vitamin D	50			
Urine culture	Sent	Sterile		
Urine routine	Wnl			
Blood culture	Sent	Sterile		
Ana profile	Negative			
ESR mm/h	20			14
CRP (mg/dl)	6			2
CPK	4			
Phosporus(mg/dl)	4.6			
SGOT/SGPT (U/L)	54/92			44/36
S. Protein (g/dl)	8.8			
S. Albumin (g/dl)	4			
A:G RATIO	1.5			
S. Bilirubin (TOTAL) (mg/dl)	0.89			1.00
LDH (U/L)	200			
Ultrasound	Normal			
Stool routine and hanging drop	Normal			
Stool culture	Sent	Sterile		
HIV	------	Non-reactive		
HBsAG	----	Non-reactive		
HCV	------	Non-reactive		
Chest x ray	Normal			
Procalcitonin			0.12	
Ns1 antigen /typhidot/ps for mp			Negative	
Repeat cultures (blood and urine)			Sent -sterile	





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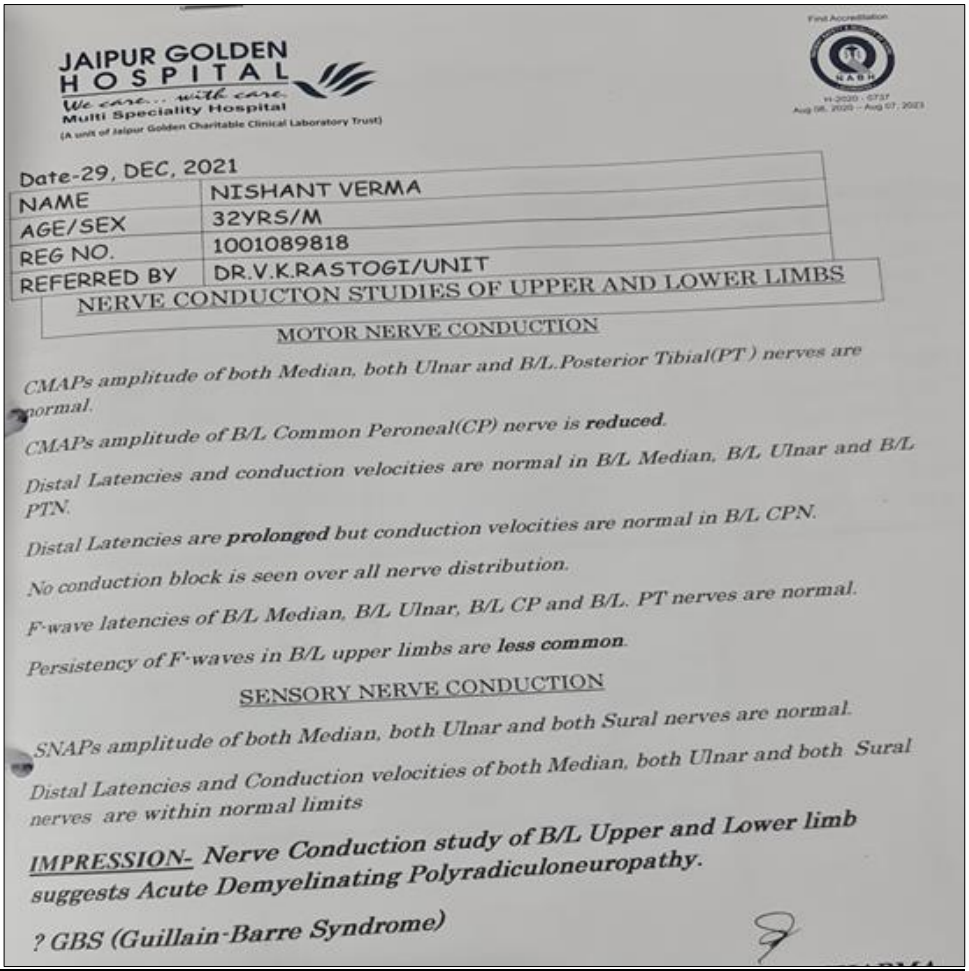
NCS Report

MNCV

Nerve	Latency (ms)		Amplitude (mV)			Duration (ms)			Dist. (mm)	NCV (m/s)	F-Min (ms)	F-Max (ms)
	D	P	D	P	%Dec	D	P	%Inc				
Lt. Median	2.69	7.12	18.15	16.96	6.56	12.19	12.56	3.04	240.00	54.18	28.87	31.37
Rt. Ulnar	2.50	6.62	15.67	12.08	22.91	14.12	14.31	1.35	280.00	67.96	31.00	32.19
Lt. Ulnar	2.31	6.94	13.72	12.58	8.31	11.12	14.37	29.23	270.00	58.32	29.56	34.00
Rt. CPN	6.94	14.44	1.70	1.71	0.59	23.19	18.37	20.78	400.00	53.33	52.25	56.50
Rt. PTN	3.62	11.31	9.72	5.74	40.95	12.62	15.19	20.36	420.00	54.62	49.87	51.37
Lt. CPN	6.19	13.56	1.86	1.49	19.89	17.19	17.37	1.05	390.00	52.92	51.25	53.00
Lt. PTN	4.37	13.19	6.19	3.37	45.56	15.62	14.75	5.57	420.00	47.62	50.62	52.62
Rt. Median	2.56	6.62	15.61	14.99	3.97	12.25	14.00	14.29	240.00	59.11	28.50	31.62

SNCV

Nerve	Latency (ms)	Amplitude (μV)	Distance (mm)	NCV (m/s)
Lt. Median	1.95	95.92	130.00	66.67
Lt. Ulnar	1.80	75.82	120.00	66.67
Rt. Sural	2.14	24.33	120.00	56.07
Lt. Sural	2.32	22.78	120.00	51.72
Rt. Median	1.95	81.02	130.00	66.67
Rt. Ulnar	1.87	64.39	120.00	64.17



JAIPUR GOLDEN HOSPITAL
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(A unit of Jaipur Golden Charitable Clinical Laboratory Trust)

Date-29, DEC, 2021

NAME	NISHANT VERMA
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REG NO.	1001089818
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NERVE CONDUCTON STUDIES OF UPPER AND LOWER LIMBS

MOTOR NERVE CONDUCTION

CMAPs amplitude of both Median, both Ulnar and B/L Posterior Tibial(PT) nerves are normal.

CMAPs amplitude of B/L Common Peroneal(CP) nerve is ***reduced****.*

Distal Latencies and conduction velocities are normal in B/L Median, B/L Ulnar and B/L PTN.

Distal Latencies are ***prolonged*** *but conduction velocities are normal in B/L CPN.*

No conduction block is seen over all nerve distribution.

F-wave latencies of B/L Median, B/L Ulnar, B/L CP and B/L. PT nerves are normal.

Persistency of F-waves in B/L upper limbs are ***less common****.*

SENSORY NERVE CONDUCTION

SNAPs amplitude of both Median, both Ulnar and both Sural nerves are normal.

Distal Latencies and Conduction velocities of both Median, both Ulnar and both Sural nerves are within normal limits

IMPRESSION- ***Nerve Conduction study of B/L Upper and Lower limb suggests Acute Demyelinating Polyradiculoneuropathy.***

? GBS (Guillain-Barre Syndrome)

Fig 1: NCV of all four limbs showing acute demyelinating polyradiculopathy

Discussion

The cardinal symptoms in our patient were bilateral upper limbs weakness, dysphagia,
and nasal twang of voice. First, differential of intracranial mass occupying
lesion or brain stroke was considered but there were no signs of raised intracranial pressure, and confirmed with extensive brain and spine imaging which were all normal. Differential of diphtheritic polyneuropathy was ruled out as on basis of absence of history of sore throat, bleeding pharyngeal membrane and bull's neck. There was no history of intake of honey, iridoplegia or autonomic dysfunction which ruled out botulinisim. Based on clinical, laboratory, CSF and electrophysiological findings of our patient with progressive weakness of upper limb, pharyngeal muscle and areflexia, which progressed as descending paralysis a diagnosis of GBS was formulated. Besides, our patient fulfilled the criteria for PCB variant of GBS in initial period.

Table 2: Diagnostic criteria for the pharyngeal–cervical–brachial variant of Guillain–Barre syndrome

Features required for diagnosis	Features strongly supportive of the diagnosis
1. Oropharyngeal weakness with neck and arm weakness which is relatively symmetrical and arm areflexia/ hyporeflexia	1. Antecedent infectious symptoms
2. Disturbed consciousness, prominent leg weakness and absence of ataxia	2. Cerebrospinal fluid albumin cytological dissociation
3. Monophasic pattern of illness with interval between onset and nadir of oropharyngeal or arm weakness between 12 hours and 28 days followed by subsequent clinical plateau	3. Evidence of neuropathy on neurophysiological studies
4. All identified alternative diagnosis are absent	4. Presence of IgG anti-GT1a or anti-GQ1b antibodies

Differential diagnosis of Guillain–Barré syndrome

The differential diagnosis of Guillain–Barré syndrome are multiple and very dependent on the clinical features of the individual patient. Following is an overview of the most important differential diagnoses depending on location in the nervous system

CNS
Infection or inflammation of the brainstem (eg. Neuromyelitis optica, sarcoidosis, sjogrens syndrome or myelin oligodendrocyte glycoprotein antibody-associated disorder)
Brainstem stroke
Infection or inflammation of the spinal cord (eg, Sjögren syndrome, sarcoidosis or acute transverse myelitis)
Malignancy (eg. Neurolymphomatosis, leptomeningeal metastases)
Vitamin deficiency (eg. Wernicke encephalopathy, vitamin B1 deficiency, or (SACD) subacute combined degeneration of the spinal cord, due to vitamin B12 deficiency)
Compression of spinal cord or brainstem
Anterior horn cells
Acute flaccid myelitis (eg polio, rabies, west nile virus, enterovirus D68 or A71, Japanese encephalitis virus)
Nerve roots
Infection (eg. HIV, Lyme disease, cytomegalovirus, varicella zoster virus, ebstein barr virus)
Leptomeningeal malignancy
Compression
Peripheral nerves
Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
Metabolic or electrolyte disorders (hypoglycaemia, porphyria, copper deficiency, hypothyroidism,)
Vitamin deficiency (beri-beri due to vitamins B1 deficiency, vitamin B12 or vitamin E deficiency)
Toxins (drugs, alcohol, thallium, lead, arsenic, organophosphate, ethylene glycol, methanol, diethylene glycol)
Vasculitis
Critical illness polyneuropathy
Neuralgic amyotrophy
Infection (eg diphtheria or HIV)
Neuromuscular junction
Myasthenia gravis
Lambert–Eaton myasthenic syndrome
Neurotoxins (eg. botulism, tick paralysis, tetanus, snakebite envenomation)
Organophosphate intoxication
Muscles
Electrolyte or metabolic disorders (eg. hypokalaemia, thyrotoxichypokalaemic periodic paralysis, hypophosphatasemia, hypomagnesaemia)
Inflammatory myositis
Acute rhabdomyolysis
Drug-induced toxic myopathy (eg. induced by emetine, colchicine, chloroquine, statin)
Mitochondrial disease

Conclusion

GBS may present atypically as PCB variant and likely to get misdiagnosed. So, a high index of suspicion for PCB variant of GBS should always be considered in every patient with presenting complaints of symmetrical upper limbs weakness and bulbar palsy. But at the same time before confirming a diagnosis of PBC variant, all other possible differentials like brainstem lesion, neuromuscular disorder, diphtheritic polyneuropathy, and botulism should be ruled out. The diagnosis was confirmed with CSF studies and electrophysiological studies at n early stage and timely

initiation of treatment resulted in good prognosis decreasing morbidity and mortality.

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