Pakistan Journal of Neurological Sciences (PJNS)

Volume 12 | Issue 4 Article 9

12-2017

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Date of submission: July15, 2017 ***Date of revision:*** August 25, 2017 ***Date of acceptance:*** September 20, 2017

ABSTRACT

Guilain-Barre Syndrome (GBS) is a common cause of Neuro-muscular paralysis with a spectrum of clinical and electrophysiology variants. We present a case of thePharyneo-cervico-brachial type. A 25 yr old male came to the Medical OPD with the complains of one day history of upper limb weakness and decreased flexion of the neck , which was progressively worsening, along-with associated shortness of breath. His systemic inquiry and past medical/surgical histories were insignificant. Physical exam correlated withthe diagnostic criteria of PCB variant of GBS. Diagnosis was confirmed on EMG/NCS,which showed axonal neuropathy involving axillay and musculocutaneousnerve, along with absent F –waves latencies inleft median nerve. He showed significant improvement in his weakness over a course of 12 days. Such a case has not been reported to the best of our knowledgefrom our part of world, as of yet.

KEYWORDS

Guillain-Barre Syndrome, Pharyngeo-cervico-brachial variant

INTRODUCTION

Guillain -Barre syndrome(GBS) is a set of clinical syndromes that present as acute inflammatory polyradiculopathy with resulting weakness and diminished reflexes and has several variants[1].Pharyngeal-cervico-brachial (PCB) variant is among the few rare variants of GBS which manifest as rapidly progressive oropharygeal and cervico-brachial weakness with areflexia in the upper limb and characterized by axonal rather than demylinating neuropathy.[2] Because of the rarity of this variant it is often misdiagnosed as a brainstem stroke, myasthenia gravis or botulism.[2]Noteworthy overlap in clinical, immunological and neuro-physiological profiles have revealed that PCB forms a continuous spectrum with Fischer syndrome and represent a localized form of GBS. [3,4]. In view of the infrequency of this disease presentation, we present a case of PCB variant of GBS in our area.

CASE REPORT

25 yr old male, right handed, no known co morbid, came to Medical OPD with complain of one day history of upper limb weakness which was progressive along with shortness of breath. The patient was sitting at his work desk, writing when he started feeling limited ability to move his arms and hands bilaterally. The weakness started progressing over a course of 2 days limiting his daily activities due to which he required help in dressing and feeding himself. He did not any history of swallowing difficulty or change of voice or any change in his smile or drooping of eye lids. He does not report any prior history of infection, vaccination, urinary or gastrointestinal symptoms. He has no previous drug history and his past medical and surgical histories were insignificant.

On physical examination his heart rate and blood pressure were normal at 80beats per min and 130/80mmhg, respectively. Heart sounds were normal with s1 and s2. There were no added sounds. JVP was normal. Muscle bulk and tone were normal but there was decreased neck flexion. Powers in the upper limbs were reduced as follows:

Table 1: Powers In Upper Limbs

Powers in the lower limbs, however, remained relatively unaffected.

DTRS were as follows:

Powers	Right Upper Limb	Left Upper Limb
Shoulder abduction	2/5	2/5
Elbow flexion	2/5	2/5
Elbow extension	3/5	3/5
Wrist flexion	3/5	3/5
Wrist Extension	3/5	3/5

Table 2: Deep Tendon Reflex

Reflexes	Right	Left
Biceps Jerk	Mute	Mute
Triceps Jerk	+	+
Supinator jerk	Mute	Mute
Knee jerk	++	++
Ankle jerk	+	+
Plantars	Down-going	Down-going

There was no evidence for swallowing difficulty(gag reflex was normal) or ptosis. His extra ocular movement wasfull, .his palatal movements were normal. Rest of Cranial nerve, sensory and cerebellar exams wasnormal.

MRI Brain plain was unremarkable. Laboratory investigations including CBC, routine chemistry, renal, liver and thyroid profiles were normal. EMG/NCS(table 3 &4) was suggestive of axonal neuropathy in bilateral Axillary and Musculocutaneous nerves, along-with conduction block, at proximal (erb-Axila) level, in bilateral median and ulnar nerves (An unusual finding for PCB since it is an axonal motor neuropathy not demyelinating type). F wave of left Median nerve was absent. The findings were suggestive of cervico-brachial variant of Guillain-Barre Syndrome.

Table 3 : Motor Nerve Conduction Studies

Nerve-Muscles	Stimulus site	Latency (ms)	Distance (cm)	Amplitude (mv)	NCV (ms)	F.LAT. (ms)	Duration (ms)
Rt. Median APB	W	2.8	7.0	4.4	-	24.0	-
`` `` `` `` ``	E	6.3	21.0	4.0	60.0	-	-
`` `` `` `` ``	Axila	8.7	-	3.9	-	-	-
`` `` `` `` ``	...	13.1	-	160uv	-	-	-
Rt. Ulnar ADQ	W	2.6	7.0	3.3	-	-	-
`` `` `` `` ``	DE	5.9	19.0	2.8	56.0	-	-
`` `` `` `` ``	PE	7.6	10.0	2.0	55.0	-	-
`` `` `` `` ``	Axila	9.6	-	605uv	-	-	-
`` `` `` `` ``	Erb's	12.1	-	560uv	-	-	-
Rt. Post. Tibial AH	A	4.5	10.0	7.4	-	54.6	-
`` `` `` `` ``	K	14.0	40.0	7.0	42.0	-	-
Rt. Peroneal EDB	A	4.7	8.0	3.7	-	55.0	-
`` `` `` `` ``	DK	11.2	31.0	3.5	47.7	-	-
`` `` `` `` ``	PK	13.4	10.0	3.5	45.0	-	-
Lt. Post. Tibial AH	A	4.1	10.0	2.9	-	55.0	-
`` `` `` `` ``	K	13.9	40.0	2.7	41.0	-	-
Lt. Peroneal EDB	A	4.7	8.0	2.8	-	54.0	-
`` `` `` `` ``	DK	11.2	30.0	2.4	47.5	-	-
`` `` `` `` ``	PK	13.5	10.0	2.4	44.6	-	-
Lt. Peroneal TA	DK	2.8	10.0	3.4	-	-	-
`` `` `` `` ``	PK	4.9	10.0	3.2	47.6	-	-
Rt. Facial	Mastoid	2.4	-	2.4	-	-	
Lt. Facial	Mastoid	2.2	-	2.3	-	-	

Table 4: Electromyography

Muscles	fibs	Psw*	Others	Amp	Duration	Polys	Recurit	Interfrence
Lft biceps	nil	nil	none	normal	normal	no	normal	normal
Lft FDI	nil	nil	none	normal	normal	no	RFR*	reduced
Lft TA	nil	nil	none	normal	normal	no	RFR*	reduced
Lft Triceps	+	+	none	No effort	No effort	No effort	No effort	No effort
Lft Deltoid	nil	nil	none	normal	normal	no	normal	normal

TABLE 4: ELECTROMYOGRAPHY DETAILS.
Psw = positive sharp waves., RFR= rapid firing units

DISCUSSION

PCB is a rare variant of Guillain-Barre Syndrome. It usually affects the bulbar muscles and causes ophthalmoplegia and facial palsy, eventually involving the neck flexors and proximal muscles of the upper limbs.[3,5,6] It is one of the rare variants that may recover after a short disease course.[2,3] The diagnostic criteria includes: Symmetrical neck and arm muscle weakness and arm areflexia with relative sparing of lower limb muscles .It has a monophasic pattern of illness . There is relative sparing of the lower limbs, with or without decreased deep tendon reflexes, and lack of sensory deficits.

We present a case of cervico-brachial variant of Guillain-Barre syndrome due its rarity and the fact that it has not been reported from our area of the world, according to best of our literature search. In our patient, the weakness descended from the neck flexors to the proximal muscles of the upper limbs, bilaterally, along-with shortness of breath, which may be secondary to diaphragmatic weakness. In our case, as compared to other reported cases, there was sparing of the bulbar and pharyngeal muscles as he did not have swallowing problem. He did, however, report some degree of dysarthria, which could not be documented on examination. Our patient fulfilled the diagnostic criteriaPCB variant of GBS described by Wakerley BR et al. [2]. As per diagnostic criterion, there was symmetrical neck and arm muscle weakness and arm areflexia with relative sparing of lower limb muscles.The pattern of illness was monophasic. Sensory level was not present. MRI of brain and cervical spine were normal, which ruled out alternative diagnoses. EMG/NCS was suggestive of axonal neuropathy due to low amplitude of CMAP in in bilateral Axillary and Musculocutaneous, ulnar and left post tibial nerves,F wave of left Median nerve was also absent. However we also found conduction block, at proximal (erb-Axila) level, in bilateral median and ulnar nerves. This finding is unusual as PCB is reported to be an axonal type and conduction blocks is indicative of demylinating type. Since rest of features fulfilled the criteria we believe this needs further investigation. The patient recovered after a twelve-day course of the disease, with conservative management. Unfortunately, CSF studies could not be performed at the time of disease presentation. Anti-ganglioside antibodies aid the diagnosis of PCB variant- a test, which remained unperformed due to unavailability of test in our country.

CONCLUSION

It is important to recognize therare variants of GBS, which present infrequently. It is high clinical suspicion that leads to diagnosis of rare cases.

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Conflict of interest: Author declares no conflict of interest.
Funding disclosure: Nil

Author's contribution:
Hazim Brohi; concept, data collection, data analysis, manuscript writing, manuscript review
Marium Mustaqeem; data collection, data analysis, manuscript writing, manuscript review
Sundus Khan; data analysis, manuscript writing, manuscript review