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Case Report

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Multiple cranial nerves involvement as initial presentation of Guillain-Barré syndrome

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ABSTRACT

Guillain-Barré syndrome (GBS) is an immune-mediated peripheral nerve disease with classical symptoms of progressive ascending bilateral upper and lower limb weakness. Cranial nerves involvement can be part of manifestation of GBS. Case reports on early involvement of multiple cranial nerves in this disease are limited. We hereby describe a 49 year old man who was diagnosed as GBS presenting unusually with facial diplegia and bulbar palsy that preceded lower limbs weakness and paresthesia. The diagnosis was supported with rare neuroimaging findings of bilateral facial nerves enhancement. He recovered well with supportive management.

Keywords: Guillain-Barré syndrome, facial diplegia, bulbar palsy, cranial nerves

INTRODUCTION

Guillain-Barré syndrome (GBS) is an acute inflammatory peripheral nerve disease which classically presented with rapidly progressive ascending flaccid paralysis. Multiple cranial nerves involvement are rare and usually occurs after the weakness of limbs. The importance of early recognition is to anticipate the progression of disease. Moreover, cranial nerve enhancement by magnetic resonance imaging (MRI) in GBS is rare. High index of suspicion by an attending doctor is required when encountering these unusual presentations in order to get proper diagnosis.

CASE

A 49 year old man with underlying hypertension, ischemic heart disease, heart failure and diabetes mellitus experienced sudden loss of facial expression and dribbling from his mouth while drinking. His speech also became slurred. His gait became unsteady subsequently. Otherwise, there was no history of recent infection and trauma.

His vital signs were within normal limits. Examination showed lower motor neuron type of facial diplegia (Figure A and B) and nasal speech. Gag reflex was absent. He was ataxic on his feet although power of bilateral lower limbs was Medical Research Council (MRC) Scale of 5. The power of bilateral lower limbs was reduced from MRC scale of 5 to 4 on day 4 of illness with paresthesia. All deep tendon reflexes were absent. Pain sensation and proprioception were intact.

Patient refused lumbar puncture. Nerve conduction studies showed prolonged left median nerve sensory peak latency while reduced sensory nerve action potential of bilateral ulnar and median nerves (Table 1); prolonged distal motor latencies of right medial, left ulnar and bilateral peroneal nerves (Table 2). F wave latencies were within normal range (Table 3). There were absence of response from bilateral trigeminal and facial nerves. Anti-ganglioside antibody panel was negative. MRI of the brain and spine showed enhancement of the distal intracanalicular, geniculate ganglion (Figure C) and tympanic (Figure D) segments of facial nerves bilaterally as well as the cauda equina on post-contrast T1 weighted sequences.

His GBS disability scale was 4. IVIg was not prescribed in view of thrombotic risk because of his background of coronary artery disease. He received supportive care such as physiotherapy and speech therapy. He was discharged in the second week of illness with the ability to swallow safely and no worsening of other symptoms. Upon follow up at the fourth month of illness, his symptoms much improved with being able to ambulate without any aids.

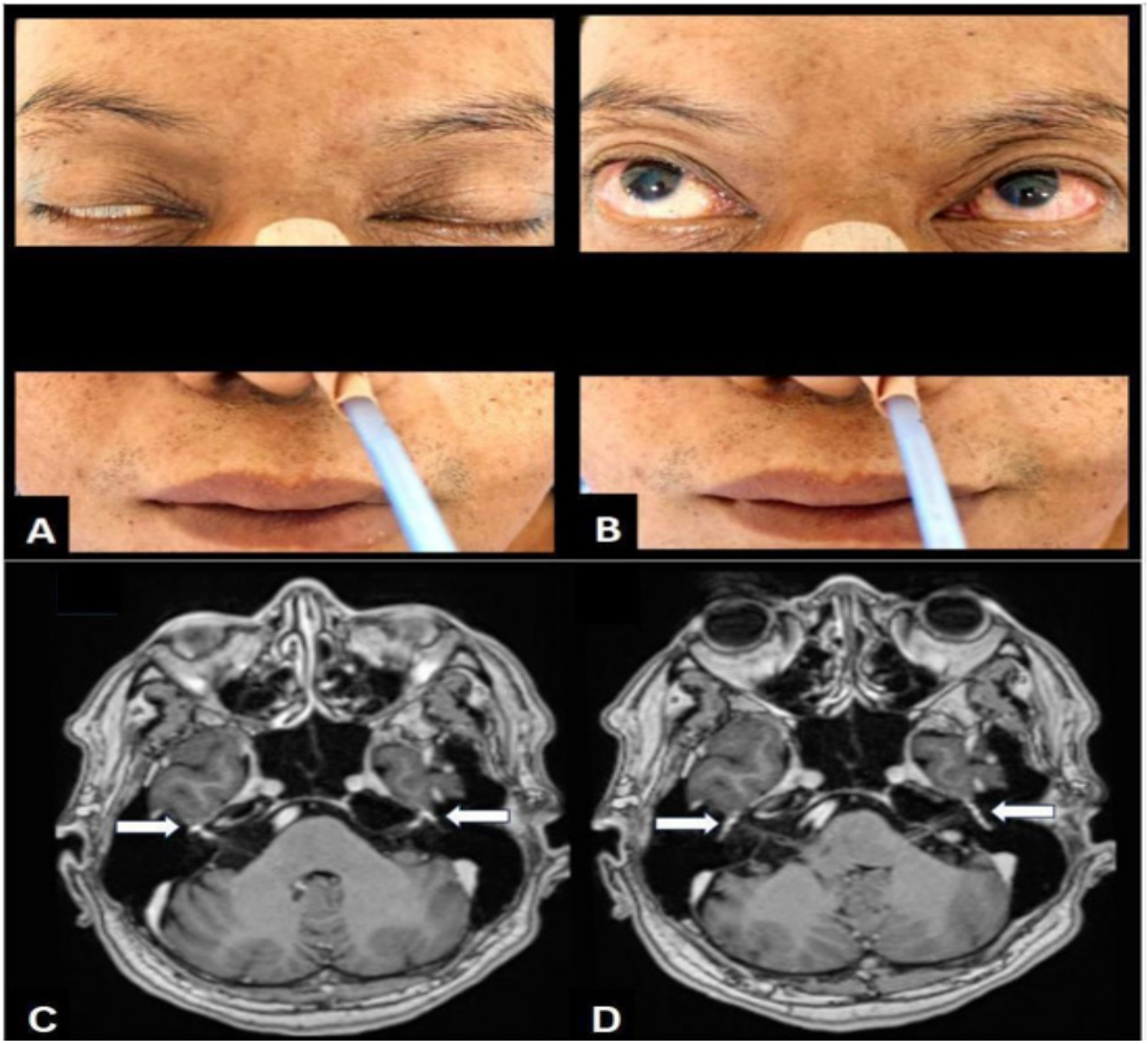

Figure. Bilateral lagophthalmos with loss of nasolabial folds and ryle tube was inserted for bulbar palsy (**A**); Weakness of bilateral frontalis muscles (**B**). Post-contrast T1MPRAGE images of the brain on axial show enhancement (arrows) of the distal intracanalicular and geniculate ganglion (**C**) and tympanic (**D**) segment of bilateral facial nerves

Table 1. Sensory nerve conduction studies

Nerve/sites	Receptor site	Onset latency (ms)	Peak latency (ms)	NP Amp (μV)	PP Amp (μV)	Segments	Distance (cm)	Velocity (m/s)
Right median-digit II (antidromic)								
Wrist	Index finger	2.71	3.60	13.8	13.3	Wrist-index	15	55
Left median-digit II (antidromic)								
Wrist	Index finger	3.08	4.29	9.8	10.8	Wrist-index	15	49
Right ulnar-digit V (antidromic)								
Wrist	Little finger	2.38	3.29	6.2	10.4	Wrist-little finger	13	55
Left ulnar-digit V (antidromic)								
Wrist	Little finger	2.29	3.25	10.4	10.5	Wrist-little finger	13	57
Left sural (antidromic)								
Calf	Ankle	2.60	3.54	16.1	21.0	Calf-ankle	14	54
Right sural (antidromic)								
Calf	Ankle	2.71	3.65	19.0	21.8	Calf-ankle	14	52
Left superficial peroneal - ankle								
Lateral leg	Ankle	1.94	2.75	11.2	15.0	Lateral leg-ankle	11	57
Right superficial peroneal - ankle								
Lateral leg	Ankle	2.19	2.98	8.3	8.7	Lateral leg-ankle	13	59

Table 2. Motor nerve conduction studies

Nerve/sites	Muscle	Latency (ms)	Amplitude (mV)	Area (mVms)	Duration (ms)	Relative amptitude (%)	Segments	Distance (cm)	Latency difference (ms)	Velocity (m/s)	Relative velocity (%)
Right median- abductor pollicis brevis (APB)											
Wrist	APB	5.1	6.9	28.7	10.33		Wrist-APB	8			
Elbow	APB	9.31	5.7	27.0	10.46	83	Elbow-wrist	25	4.21	59	100
Right ulnar-adductor digiti minimi (ADM)											
Wrist	ADM	2.88	4.4	14.6	7.77	100	Wrist-ADM	8			
Below elbow	ADM	6.81	3.5	9.9	6.81	80.8	Below elbow-wrist	26	3.94	66	100
Above elbow	ADM	11.23	2.6	7.7	5.69	75.1	Albow-below elbow	13	4.42	29	44.6
Left ulnar- adductor digiti minimi (ADM)											
Wrist	ADM	4.19	6.1	19.5	6.52	100	Wrist-ADM	8			
Below elbow	ADM	8.69	5.0	16.7	6.83	82.6	Below elbow-wrist	26	4.50	58	100
Above elbow	ADM	11.98	3.4	13.6	7.17	67.1	Albow-below elbow	15	3.29	46	
Left peroneal- extensor digitorum brevis (EDB)											
Ankle	EDB	6.79	2.0	9.2	7.6	100	Ankle-EDB	8			
Below fibula head	EDB	14.29	1.7	8.1	7.6	85.8	Below fibula head-ankle	32	7.50	43	100
Above fibula head	EDB	15.67	1.8	8.9	8.0	105	Above-below fibula head	10	1.37	73	
Right peroneal-extensor digitorum brevis (EDB)											
Ankle	EDB	4.81	4.3	18.7	8.19	100	Ankle-EDB	8			
Below fibula head	EDB	15.65	3.2	16.3	9.27	75.7	Below Fibula head-ankle	36	8.83	41	100
Above fibula head	EDB	15.27	4.2	20.9	9.19	130	Above-below fibula head	10	1.62	62	
Left tibial-anterior tibialis (AH)											
Ankle	AH	5.33	13.9	50.4	8.29	100	Ankle-AH	8			
Knee	AH	15.58	11.1	46.9	9.06	80	Knee-ankle	40	10.25	39	100
Right tibial-anterior tibialis (AH)											
Ankle	AH	4.73	8.9	37.3	9.21	100	Ankle-AH	8			
Knee	AH	15.85	7.1	34.5	8.96	79.9	Knee-ankle	40	11.13	36	100

Table 3. F wave

F wave	Minimum F latency (ms)	Maximum F latency (ms)	Mean F latency (ms)
Right ulnar-ADM	25.5	38.0	30.1
Right median-APB	32.6	34.2	33.2
Left ulnar-ADM	29.4	30.2	29.8
Left peroneal-EDB	42.2	44.3	43.4
Left tibial-AH	30.9	39.6	35.2
Right peroneal-EDB	41.8	66.8	47.9
Right tibial-AH	40.0	45.9	42.9

ADM: Adductor digiti minimi, APB: Abductor pollicis brevis, EDB: Extensor digitorum brevis, AH: Anterior tibialis

DISCUSSION

GBS typically manifests as ascending limb weakness. Cranial nerve involvement is seen in 50% of patients with GBS and it usually follows limb involvement.[1] However, facial diplegia and bulbar palsy as initial manifestation of GBS followed by lower limb weakness is a rare occurrence.[1-3] Recognition of early cranial nerve involvement as part of the GBS spectrum is important to anticipate the typical disease progression.[4-6] These prominent symptoms could be features of GBS variants such as sensory ataxic variant, facial diplegia with paresthesia and acute bulbar palsy plus.

GBS is diagnosed clinically and supported with various investigations such as cerebrospinal fluid analysis, nerve conduction studies, neuroimaging or serum anti-ganglioside

antibodies. MRI could be a supplementary diagnostic modality to exclude infectious, vascular or neoplastic causes. Rare findings of bilateral facial nerves enhancement in this patient provide supportive evidence for nerve inflammation in GBS.[7,8]

Decision of prescribing IVIg was weighed between risk of thrombosis and progression of symptoms. There is lack of clinical evidence to support the safe use of IVIg in patients with severe cardiovascular disease. There are cases of myocardial infarction after use of IVIg.[9] Thus, this patient did not receive IVIg in view of his symptoms which did not further progress with risk consideration of his comorbidities. There is also limited evidence for IVIg in milder forms and variants of GBS.[10]

CONCLUSION

Initial cranial nerve involvement is the unusual presentation of GBS which should not be missed in the clinical practice. Future study is required to explore the safety of IVIg use in patients with ischemic heart disease and its benefits in milder or variant forms of GBS.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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