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A CASE OF GUILLAIN-BARRÉ SYNDROME PRESENTED WITH ABNORMAL PUPIL WITHOUT OPHTHALMOPLÉGIA

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ABSTRACT:

A 50-year-old man, resident of Balochistan, Pakistan presented with acute progressive tetraparesis associated with a unilateral dilated pupil. He had normal ocular movement, but the right pupil was dilated and unresponsive to light or accommodation reflexes. This is the first case from Pakistan.

Keywords: Guillain-Barré syndrome; ophthalmoplegia, pupillary light reflex

INTRODUCTION

Guillain-Barré syndrome (GBS) is a common paralytic neuropathy with well recognized clinical and pathological variants¹. An anomalous autoimmune response is triggered due to an infection which damages the peripheral nerves and their spinal roots. Within days of immune stimulation triggered by a preceded infection, the patient suffers from acute limb paresis in conjunction with sensory or cranial nerve involvement². In 20-30 percent of the cases, the disease results in respiratory failure. The only effective therapy is the administration of intravenous immunoglobulin or plasma exchange with continuous symptomatic treatment³. It is of crucial importance that the diagnosis of GBS is made as soon as possible since it is a rapidly progressing disease which may lead to respiratory failure and death⁴. The diagnosis which is largely based on clinical features and electrodiagnostic studies, NCS/EMG, and CSF analysis is sometimes hindered by the non-availability of biomarkers for certain variants of the disease. Identification of different clinical variants and prognostic biomarkers are some of the challenges researchers face. Here we reported a rare variant of GBS with the involvement of the pupils characterized by anisocoria, muscle weakness, failure of pupil accommodation in the left eye, and absence of tendon reflexes in a 50-year-old patient.

CASE PRESENTATION

A 50-year-old male, married, a farmer by occupation, resident of Turbat, Balochistan presented to the outpatient department with complaint of weakness in all four limbs for the past one week. He had no known comorbidities. According to the patient, he initially experienced mild weakness in his lower limbs which progressed rapidly to involve the upper limbs almost within a few hours of onset of his initial symptoms. The patient was unable to stand without support at the time of his admission to the neuromedicine ward, Jinnah Postgraduate Medical Center. The patient claimed that his symptoms were preceded by a mild upper respiratory infection with associated fever a month back. Patient suffered from constipation. No history of difficulty in swallowing or shortness of breath was documented. Patient did not complain of any vision disturbance. On examination, the patient was found to be of average height and built with normal vitals and a Glasgow coma scale score of 15. Higher mental functions were intact. Speech was intact. Gait could not be assessed as the patient was unable to walk by himself. On a neurological exam, it was found that pupils were not bilaterally equally reactive to light (BERL). The pupil of the right eye was dilated (measuring 6 mm in diameter) and unresponsive to light reflex or accommodation. The left pupil was 3 mm in diameter and reacted normally. Extraocular muscle

movements were normal. Fundoscopic examination did not reveal any abnormality. There was no facial weakness reported. Pharyngeal and palatal functions were normal. The uvula was centrally placed. Upon examining the motor system, bulk was normal with decreased tone in all four limbs. The power was reported to be 3/5 in all four limbs with moderately weaker hand grip, and absent deep tendon reflexes. Plantar reflex caused a downward response of the hallux (big toe) which was normal for a healthy adult.

Other examinations including cerebellar functions, deep tendon reflexes (DTRs), superficial reflexes, and sensations were all normal. White blood cell count was 22,000 cells per mm³, but other laboratory tests including the red blood cell count, hematocrit, hemoglobin, urinalysis, serum glucose, urea, creatinine, and other electrolytes were within normal range. Electroencephalogram was also normal. The differential diagnoses included Guillain-Barré syndrome, neuromuscular junction disease such as botulism or Lambert-Eaton myasthenic syndrome and electrolyte disturbance. The nerve conduction studies (NCS) showed that right median and right ulnar nerves had low amplitude, prolonged distal latencies, and low velocities. There was no response detected from tibial and peroneal nerves. The sensory nerve conduction studies showed that the Sural, Median, and Ulnar nerve had prolonged latencies, low amplitudes, and low velocities. The findings of the nerve conduction studies were indicative of acute sensory-motor axonal neuropathy (AMSAN a variant of GBS). The diagnosis of GBS was confirmed by the combination of nerve conduction studies and cerebrospinal fluid analysis. The elevated protein levels of 74 mg/dL (normal range = 15 to 40 mg/dL) with no increase in white blood cells confirmed the diagnosis of acute sensory-motor axonal neuropathy – a variant of Guillain Barre Syndrome.

Table 1. Findings of Nerve Conduction Studies

Nerve Muscles	Stimuli Site	Distance (cm)	Latency (ms)	Amplitude (mV)	NCV (m/s)
Median (APB)	W	7.0	3.5	0.8	
	E	20.0	8.4	0.6	40.9
Ulnar (ADQ)	W	7.0	3.0	0.5	34.9
	D.E	20.0	8.7	0.3	23.5
	P.E	10.0	13.0	0.2	-
Post Tibial	A	Non-responsive	Non-responsive	Non-responsive	Non-responsive
	K	Non-responsive	Non-responsive	Non-responsive	Non-responsive
Peroneal (EDB)	A	Non-responsive	Non-responsive	Non-responsive	Non-responsive
	D.K	Non-responsive	Non-responsive	Non-responsive	Non-responsive
	P.K	Non-responsive	Non-responsive	Non-responsive	Non-responsive
	D.F.H	Non-responsive	Non-responsive	Non-responsive	Non-responsive
Peroneal (TA)	P.F.H	Non-responsive	Non-responsive	Non-responsive	Non-responsive
	P.F.H	Non-responsive	Non-responsive	Non-responsive	Non-responsive
H. Reflex	P.Fossa	Non-responsive	Non-responsive	Non-responsive	Non-responsive

The patient was administered the intravenous immunoglobulin to which he responded positively as improvement was detected upon NCS. Patient was discharged on the 7th day of admission. The patient was lost to follow-up.

DISCUSSION

The patient in the present study, fulfilled the diagnostic criteria for GBS 5. He presented with progressive loss of sensory and motor function, with decreased muscle tone and power in all four limbs. Absence of fever, increased protein in cerebrospinal fluid, and supporting nerve conduction studies confirmed the diagnosis of GBS. The unilateral pupillary involvement with dilatation and unresponsiveness to light reflex or accommodation without the involvement of extraocular muscles made this case peculiar. GBS is a result of an immune-mediated attack against the peripheral nerve which may be preceded by a viral infection among other trigger factors⁶. There are several remarkably overlapping variants of GBS that share a common characteristic of acute symmetrical paralysis with or without sensory loss⁶⁻⁷. An important variant of GBS, is known as, "Miller Fisher Syndrome" which may present with ocular involvement however, in contrast to the present case, Miller Fisher Syndrome is associated with multiple extraocular muscles paralysis⁷. Unlike MFS, the extraocular muscles were not involved in the present study case. As stated earlier, GBS is usually antecedent by a viral-like disease that may manifest up to six weeks prior to the onset of neurological deterioration. The patient in this case reported contracting a mild upper respiratory infection, four

weeks before he started developing neurological deficits. Other known triggers include *Campylobacter jejuni*-induced gastroenteritis, certain parasitic infections including malaria, malignancy, and bone marrow transplantation.⁸ The symptoms of GBS develop abruptly with distal limb weakness with little or no sensory deficit usually accompanied by unbearable radiculopathy⁹. The symptoms are rapidly progressive and may lead to death within hours of admission. Up to thirty percent of patients progress to respiratory failure. Other complications include sepsis, deep vein thrombosis (DVT), and aspiration.⁹ Therefore, the patients should be started on immunotherapy as early as possible which is the mainstay of treatment¹⁰.

CONCLUSION

GBS is a medical emergency which can lead to respiratory failure and death. Therefore, prompt diagnosis and treatment are crucial to patient outcome. We reported a rare variant of GBS with the involvement of the pupils characterized by anisocoria and absence of deep tendon reflexes in a 50-year-old patient. Plasmapheresis - a first line of immunotherapy is the mainstay of treatment for all major variants of GBS.

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Suneel Kumar; data analysis, manuscript writing, manuscript review