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An atypical presentation of Guillain-Barré Syndrome with isolated third crania nerve involvement and descending paralysis

MAKT Marasinghe[1], PA Jayasinghe[2]

[1]Postgraduate Institute of Medicine, Colombo, Sri Lanka. [2]National Hospital Galle, Sri Lanka.

Correspondence: Dr. Tharushi Marasinghe
e-mail*: tharushi1992824@gmail.com*
https://orcid.org/0009-0001-0038-1152
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Introduction

Guillain-Barré syndrome (GBS) is the most common and severe acute paralytic neuropathy, affecting about 100,000 people worldwide each year (1). It is an antibody-mediated post-infectious autoimmune disorder (2). Classical GBS typically presents with bilateral, symmetrical lower-limb weakness and areflexia, progressing in an ascending pattern over days to weeks (1). However, different variants of GBS such as Miller Fisher syndrome (MFS), which is characterised by the triad of ophthalmoplegia, ataxia, and areflexia, and pharyngeal-cervical-brachial (PCB) variant presenting predominantly with bulbar and facial muscle weakness are well-known (1-3). This report describes a rare case of descending GBS with isolated 3rd cranial nerve dysfunction, emphasizing the importance of early diagnosis, specific treatment, and supportive care.

Case presentation

A 46-year-old previously healthy female presented to the National Hospital Galle with an acute onset of unsteadiness and asymmetrical bilateral ptosis that progressed over one day. On the morning of the admission day, she felt difficulty in maintaining her posture while standing and walking, describing a sensation of imbalance and feeling as though she might fall. There was no preceding trauma or loss of consciousness. A few hours later, she noticed mild drooping of the left upper eyelid, described as heaviness of the left eyelid and a reduced visual field. Her symptoms progressively worsened, and by the evening, she noticed mild drooping of her right upper eyelid as well.

She denied double vision, facial weakness, dysarthria, limb weakness, or sensory deficits at onset. No prior history of neurological or autoimmune disease was reported.

Notably, she had a watery diarrhoea episode with low-grade fever one week prior, which resolved spontaneously after a few days. She had no recent history of respiratory tract infection, vaccination, or toxin exposure.

On admission, the patient was an average-built female with a BMI of approximately 25 kg/m^2. She was hemodynamically stable and afebrile. On neurological examination, she had asymmetrical bilateral ptosis, left more prominent than the right side, with normal ocular movements and preserved pupillary reflexes. Motor power of all four limbs was 5/5 with no focal weakness and preserved sensation. however, deep tendon reflexes were absent globally. Her single breath count was 28, with preserved neck flexor and extensor strength, and no bulbar or cerebellar signs were noted other than ataxia. Other systemic examinations, including respiratory, cardiovascular, and abdominal systems, were unremarkable. Importantly, no oropharyngeal pseudomembrane was noted on examination.

Initial investigations, including full blood count, renal and liver function tests, inflammatory markers,

and a non-contrast CT (NCCT) of the brain, were normal. due to her atypical presentation, we faced a diagnostic dilemma. A multidisciplinary team approach was taken for further management and referred to the neurology team for expert input. Laboratory investigations are tabulated in Table 1.

The patient complained of a sudden onset of heaviness in her bilateral upper limbs and difficulty holding her arms against gravity on the morning of the second day of admission. A few hours later, she noticed progressive weakness in both of her lower limbs. On examination, bilateral upper limb power was reduced to 3/5, and lower limb power was 4/5. By evening, her neck muscle power declined, and her single breath count deteriorated to 8, raising the concern for respiratory failure.

Patient was intubated and arranged ICU care and started her on IVIG 0.4 mg/kg following neurology opinion. MRI Brain was reported as normal, and Nerve Conduction Studies (NCS) and Electromyography (EMG) revealed demyelinating polyneuropathy, including prolonged distal latencies, reduced conduction velocities, and conduction block in tibial and common peroneal nerve in the lower limbs, as well as the median and ulnar nerves in the upper limb. Electromyography (EMG) additionally suggested demyelinating involvement of the superior division of the oculomotor nerve.

IVIG was continued for a total of 5 days, and the patient showed gradual clinical improvement. She was successfully extubated after 48 hours of mechanical ventilation. At the time of discharge, her ptosis had partially recovered, and motor power in all four extremities had improved to 4/5. Neck flexor and extensor strength were fully regained, and she remained haemodynamically stable throughout recovery.

On discharge, she was referred for outpatient physiotherapy and neurology for continued rehabilitation and monitoring.

CSF analysis performed on the tenth day of presentation showed albuminocytologic dissociation with a protein level of 90.8 mg/dL and a white blood cell (WBC) count of 4/μL (lymphocytes) with normal sugar levels. In order to exclude other causes for descending paralysis, we performed a serum assay for botulinum toxin and diphtheria antibody, which were normal.

The results of the nerve conduction examination, the investigation of the cerebrospinal fluid, and the improvement in clinical symptoms with intravenous immunoglobulin (IVIG) led to the diagnosis of an variant of Guillain-Barré Syndrome (GBS).

Table 1: Laboratory investigation results

Test	Result	Test	Result
WBC	8.81 x 10^9/L	Erythrocyte Sedimentation Rate (ESR)	5 mm/hr
Haemoglobin (Hb)	12.3 g/dL	Serum Creatinine	52.3 μmol/L
Platelets (Plt)	250 x10^9/L	Bilirubin	18.5 μmol/L
Sodium (Na^+)	136 mmol/L	Aspartate Aminotransferase (AST)	28 U/L
Potassium (K^+)	4 mmol/L	Alanine Aminotransferase (ALT)	20 U/L
Calcium (Ca^{+2})	2.4 mmol/L	Alkaline Phosphatase (ALP)	59 U/L
Magnesium (Mg^{+2})	0.8 mmol/L	Albumin	40 U/L
C-Reactive Protein (CRP)	21 mg/L		

Figure 1: The photo taken at the time of admission demonstrates bilateral ptosis, more pronounced on the left side

Discussion

Guillain-Barré Syndrome (GBS) was first described in 1916 by Charles Guillain, Jean Alexandre Barré, and André Strohl in France. They identified it as a type of areflexic paralysis in soldiers during World War I, notable for its normal cerebrospinal fluid (CSF) cell count but elevated protein levels (1, 3, 4). It is the most common cause of acute acquired paralysis worldwide. In 60-70% of cases, it occurs 1 - 3 weeks after a preceding respiratory or gastrointestinal infection, such as *Campylobacter jejuni*, cytomegalovirus, or Epstein-Barr virus (2-4).

The exact pathogenesis of Guillain-Barré Syndrome (GBS) remains incompletely understood, but the current hypothesis is molecular mimicry**,** where immune responses generated after an infection cross-react with epitopes on peripheral nerves. This leads to the formation of autoantibodies that damage Schwann cells and myelin, resulting in Acute Inflammatory Demyelinating Polyneuropathy (AIDP)**,** or target axons, causing Acute Motor Axonal Neuropathy (AMAN) or Acute Motor and Sensory Axonal Neuropathy (AMSAN) (3)**.** This immune-mediated injury disrupts nerve signal conduction and causes muscle weakness. Recovery involves remyelination and axonal regeneration, which may take weeks to months. The clinical features, subtype, and prognosis depend on the specific autoantibodies involved and the extent of nerve damage. Common subtypes include classical GBS with AIDP (85-90% of cases), AMAN**,** AMSAN**,** Miller Fisher Syndrome (MFS)**,** the Pharyngeal-Cervical-Brachial (PCB) variant, and Bickerstaff Brainstem Encephalitis (1-3, 6).

The case in the current report demonstrates an atypical presentation of GBS, which doesn't align with previously suggested variants.

As seen in our patient, descending paralysis with isolated cranial nerve involvement is extremely rare and mimics conditions like myasthenia gravis, botulism, diphtheria, or brainstem stroke (6-9). Myasthenia gravis is ruled out due to the absence of fluctuating weakness and NCS findings. Botulism was excluded by negative serum toxin assays, absence of autonomic dysfunction, and no exposure history (9). Diphtheritic neuropathy was ruled out due to negative serology and absence of pseudomembranes (7).

Due to isolated ptosis without ophthalmoplegia or pupillary involvement, initially suggested to be a cranial mononeuropathy. However, rapid progression to quadriparesis, global areflexia, respiratory compromise, and classic CSF and NCS/EMG findings supported a diagnosis of GBS (6). Involvement of the superior division of the oculomotor nerve is rare but has been reported in variants like Miller Fisher syndrome; unfortunately, our patient did not fulfill its diagnostic triad (8).

In the literature, only a few cases of GBS presenting with descending paralysis have been reported. A similar case was reported by Ralapanawa, *et al.,* which involved unilateral ptosis without ophthalmoplegia progressing to severe quadriparesis and multiple cranial nerve palsies (11). In 2020, a case of descending paralysis was reported in the United States (1). Another case reported in the literature in 2013 documented a GBS case initially presenting with bilateral claw hands (10). Most recently, there was a case report published in Sri Lanka with descending paralysis presenting with right upper limb weakness (3). These rare presentations emphasize the clinical variability of GBS and the importance of considering atypical forms in diagnosis.

Guillain-Barré Syndrome (GBS) is primarily a clinical diagnosis, which can be confirmed by nerve conduction studies and the presence of cytoprotein dissociation in CSF analysis (3, 5). Unfortunately, antibody testing was not available in Sri Lanka due to limited availability. Therefore, it is challenging to diagnose atypical variants of GBS. Publishing of more atypical cases helps to improve early detection of GBS variants, which has a strong clinical impact, because it raises the

clinician's awareness of those variants and leads to the specific treatment as early as possible (10).

Treatment with immunotherapy with either IV immunoglobulin (IVIG) or plasma exchange (PLEX) is equally effective for GBS (2, 3, 5). The combination of plasma exchange followed by IVIG is not significantly more effective than using either treatment alone. Additionally, there is no strong evidence to support the use of a second course of IVIG in patients with GBS who continue to deteriorate after the initial treatment (5). There is no proven benefit with corticosteroids (3).

In our case, with the contribution of a multidisciplinary team, our patient was given IVIG, which was continued for 5 days, and was initiated on limb, facial, and chest physiotherapy, which aided her recovery. Our case emphasizes the importance of multidisciplinary involvement and early recognition and treatment of the atypical variant of GBS.

Conclusions

This case highlights a rare and atypical variant of Guillain-Barré Syndrome presenting with descending paralysis and isolated cranial nerve involvement. Early diagnosis of the disease with the help of expert opinion and early initiation of IVIG therapy and a multidisciplinary approach contributed to a favorable outcome. Guillain-Barré Syndrome should be considered in patients presenting with isolated cranial nerve symptoms, particularly when accompanied by areflexia or rapid progression of the symptoms to the extremities.

Informed written consent has been obtained from the patient to publish this case report with photographs.

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