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(CASE REPORT)

'When nature breaks the rules': Guillain-barré syndrome presenting with descending paralysis

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Abstract

Guillain-Barré Syndrome (GBS) typically presents with ascending paralysis, but we report an atypical case involving descending paralysis in a 36-year-old male. The patient experienced upper limb weakness and respiratory muscle involvement, progressing to respiratory distress. Initial cerebrospinal fluid (CSF) analysis and nerve conduction studies (NCS) were normal, delaying the diagnosis. Treatment with intravenous immunoglobulin (IVIG) led to partial recovery. This case highlights the diagnostic challenges of atypical GBS presentations and the importance of early intervention.

Keywords: Autonomic Nervous System Diseases; Guillain-Barre Syndrome; Miller Fisher Syndrome; Paresis; Muscle Weakness

1. Introduction

Guillain-Barré Syndrome (GBS) has always been looked upon as an important differential in a case of ascending paralysis [1][2][3][4]. Although this description is classical and frequently encountered, there are still many possible varied presentations of this disease [6][8]. Recent literature has highlighted sporadic cases of atypical GBS presenting as descending paralysis making it necessary to consider GBS as a rare but possible differential for descending paralysis also [7].

2. Case Report

A 36-year-old male, accountant by occupation, presented to the general emergency department with a short 3 day history of gradually progressive bilateral upper limb weakness. His chief complaints were difficulty in everyday tasks like combing his hair and lifting things. He experienced an episode of gastroenteritis 4 weeks back which was treated with antibiotics on an out-patient basis. There was no history of recent vaccination, or trauma. Neurological examination revealed symmetrical weakness in the upper limbs with diminished reflexes. Cranial nerve function was intact, and no sensory deficits were noted. The lower limbs were spared at presentation. On general examination, the patient's vital signs were within normal limits, with a temperature of 98.6°F, a respiratory rate of 16 breaths per minute, a blood pressure of 120/80 mmHg and a pulse rate of 82 beats per minute.

2.1. Investigations

A comprehensive diagnostic assessment was conducted to evaluate the patient's condition, starting with laboratory investigations. A complete blood count revealed a white blood cell count of 6.8 x 10^9/L, hemoglobin of 14.1 g/dL, and platelet count of 245 x 10^9/L. Electrolytes were unremarkable, with sodium at 138 mEq/L, potassium at 4.0 mEq/L, and

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chloride at 102 mEq/L. Renal function tests revealed a serum creatinine of 0.9 mg/dL, and liver function tests were within normal limits (ALT 32 U/L, AST 28 U/L).

A cerebrospinal fluid (CSF) analysis was performed on day 1. The results indicated that all parameters are within the normal reference ranges (Table 1).

Table 1 Summary of Cerebrospinal Fluid Analysis results as of Day 1

CSF Analysis Parameter	Measurement	Normal Reference Range
Opening Pressure	120 mm H_2O	70 to 180 mm H_2O
Protein Level	32 mg/dL	15 to 60 mg/dL
Glucose Level	65 mg/dL	50 to 80 mg/dL
White Blood Cells (WBCs)	0 WBCs	0 to 5 WBCs (all mononuclear)

A nerve conduction study (NCS) performed on day 3 revealed normal conduction velocities and latencies, with no evidence of demyelination or axonal damage. Magnetic resonance imaging (MRI) of the brain and spine was normal, with no signs of central nervous system lesions.

By day 7, the patient developed respiratory distress, necessitating intubation and mechanical ventilation. A repeat NCS at this time revealed prolonged F-wave latency (upper limbs: 40 ms, reference < 32 ms) and conduction block, suggestive of GBS.

The patient's motor examination revealed normal tone in all limbs, but strength was decreased with Medical Research Council (MRC) grades of 3/5 in the upper limbs and 2/5 in the lower limbs. Reflexes were absent in all limbs. Sensory examination revealed diminished sensation to light touch, pain, and temperature in a glove-and-stocking distribution. Vibration and proprioception were intact. Cranial nerve examination was normal, with no signs of facial weakness or ophthalmoplegia. Autonomic testing showed normal results, with no orthostatic hypotension or arrhythmias. Laboratory investigations, including complete blood count, electrolytes, liver and kidney function tests, were within normal limits. Cerebrospinal fluid analysis showed an elevated protein level (75 mg/dL) with no pleocytosis, consistent with albuminocytologic dissociation. Nerve conduction studies demonstrated reduced conduction velocities and prolonged distal latencies, suggesting a demyelinating process.

2.2. Management

A final diagnosis of atypical Guillain-Barré Syndrome (GBS) was made. Thus, the patient was started on intravenous immunoglobulin infusion at the dose of 0.4 gram/kg/day for a period of 5 days. To relieve the respiratory distress due to muscle weakness, ventilatory support was provided. Alongside, our patient received supportive care including physical therapy and muscle training to reduce complications of immobility.

2.3. Outcome

Gradual improvement was noted in upper limb strength over two weeks, with a Medical Research Council (MRC) grade of 3/5 improving to 4/5. The patient remained ventilator-dependent for three weeks, after which he was successfully weaned off mechanical ventilation. By week 4, he had recovered most of his forequarter strength and was referred to a physiotherapist with the advice to continue physical therapy and rehabilitation. At a 6-month follow-up, the patient showed full recovery of upper limb strength (MRC 5/5) and very mild weakness in respiratory function.

3. Discussion

Guillain-Barré Syndrome (GBS) is an acute polyradiculoneuropathy that presents as a diagnostic dilemma to the clinician. The pathogenesis lies in an immune-mediated demyelination of peripheral nerves. As per existing literature, the reported incidence of GBS ranges from 1 to 4 cases per 100,000 individuals annually. In epidemiological terms, it affects people of all ages with a bimodal distribution [1]. The textbook presentation is of a patient who develops rapidly progressing symmetrical limb weakness, sensory disturbances and areflexia, typically following a harmless infection [5][9]. The paralysis is described as an ascending type of paralysis, which appears first in the lower limbs and then gradually progresses cranially [10].

Descending pattern of paralysis in GBS is rare, accounting for only about 15% of cases in the acute inflammatory demyelinating polyneuropathy (AIDP) variant of GBS [1]. This form of paralysis typically begins in the upper limbs and progresses downward, which can lead to significant diagnostic challenges. For instance, a case study described a patient with descending paralysis starting in the upper limbs, eventually leading to respiratory complications requiring ventilator support [1]. This highlights the need to look at GBS as an entity with extremely varied manifestations.

4. Conclusion

This case report described an extremely atypical presentation of Guillain-Barré Syndrome. Contrary to the classical complaints of ascending paralysis, our patient presented with descending paralysis which caused a delay in the diagnosis. Thus, physicians should consider GBS as an improbable yet possible differential even in the cases of descending paralysis. Nerve conduction studies on peripheral nerves, if positive for demyelination, could greatly aid in confident diagnosis.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

No ethical approval required for publishing this case report.

Statement of informed consent

Informed consent was obtained from the patient included in the study.

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