CASE REPORT

A typical presentation of Pharyngeal Cervical Brachial variant of Guillain Barre Syndrome (rare variant):–A case report

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Abstract:

Guillainbarre Syndrome is a clinical manifestation of acute demyelinating, resultant weakness and ascending paralysis. Guillainbarre Syndrome (GBS) consists of many variants, including acute motor axonal neuropathy, acute motor-sensory axonal neuropathy, pure sensory, pan autonomic, Miller fisher syndrome and pharyngeal-cervical-brachial subtypes. The pharyngeal-cervical-brachial subtype is the rare variant of Guillainbarre Syndrome (GBS) with typically spared lower limb weakness.

Keywords: Guillain Barre Syndrome, subtype, rare variant, bulbar palsy

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Introduction:
Guillain-Barré Syndrome (GBS) is an acute immune-mediated polyneuropathy characterized by rapidly progressive weakness andareflexia. It represents a spectrum of disorders that affect the peripheral nervous system and are often triggered by preceding infections. While the classic form of Guillain-Barré Syndrome manifests as an ascending symmetrical paralysis, several distinct clinical variants have been recognized. Among these, the pharyngeal-cervical-brachial (PCB) variant is rare and uniquely characterized by acute weakness in the oropharyngeal, neck, and upper limb muscles, typically sparing the lower limbs. This variant poses diagnostic challenges due to its overlapping features with other neuromuscular disorders, requiring a high degree of clinical suspicion.

The pathogenesis of Guillain-Barré Syndrome, including its pharyngeal-cervical-brachial subtype, is thought to involve molecular mimicry between microbial antigens and nerve gangliosides, leading to an autoimmune attack on peripheral nerves. Antibodies against gangliosides such as GT1a and GD1a are frequently implicated in the pharyngeal-cervical-brachial variant. Diagnostic confirmation is typically achieved through electrophysiological studies, including electromyography (EMG) and nerve conduction studies (NCS), which help distinguish between axonal and demyelinating processes. Treatment strategies primarily involve immunotherapy, with intravenous immunoglobulin (IVIG) and plasmapheresis being the mainstay therapies.

This case report describes a patient presenting with the pharyngeal-cervical-brachial variant of Guillain-Barré Syndrome, highlighting the clinical presentation, diagnostic workup, and favorable response to plasmapheresis. The discussion aims to enhance understanding of this uncommon Guillain-Barré Syndrome subtype, emphasizing the importance of early recognition and timely intervention to optimize patient outcomes.

Case presentation:
A 36-year-old male presented to the emergency room (ER) with complaints of difficulty in speaking, difficulty in swallowing, and weakness in both upper limbs. Notably, there was no associated weakness in the lower limbs. His symp-

toms had been preceded by an upper respiratory tract infection and a history of diarrhea lasting for 3 to 4 days. The patient reported consuming Chinese rice from a local restaurant four days prior to the onset of symptoms. The following day, he experienced nausea and profuse diarrhea, passing approximately 8 to 10 stools per day.

On arrival at the ER, arterial blood gas (ABG) analysis revealed type II respiratory failure. A chest examination showed globally decreased breath sounds, prompting immediate intubation for respiratory support. A neurologist was consulted, and the patient was diagnosed with Guillain-Barré Syndrome (GBS). Given the clinical presentation involving bulbar and upper limb muscle weakness with spared lower limbs, the pharyngeal-cervical-brachial (PCB) variant of Guillain-Barré Syndrome was suspected. The neurophysician recommended plasmapheresis as the primary therapeutic intervention.

The patient demonstrated significant clinical improvement after two sessions of plasmapheresis and was successfully extubated. Electromyography (EMG) and nerve conduction studies (NCS) were performed, confirming the diagnosis of the pharyngeal-cervical-brachial variant of Guillain-Barré Syndrome. These diagnostic modalities are critical in differentiating variants of Guillain-Barré Syndrome, with studies highlighting their utility in identifying specific patterns of demyelination and axonal involvement associated with the disease (Ropper et al.[4], Willison et al.[5]). A total of five sessions of plasmapheresis were completed, leading to a marked resolution of symptoms. His upper limb strength returned to normal, and he regained full motor control and the ability to swallow without difficulty.

Upon discharge, the patient was fully alert and oriented, with a Glasgow Coma Scale (GCS) score of 15/15. He was able to move all four limbs without restriction and had no residual dysphagia. This case highlights the importance of early recognition and aggressive management of the pharyngeal-cervical-brachial variant of Guillain-Barré Syndrome, emphasizing the efficacy of plasmapheresis in achieving rapid clinical recovery. Studies have demonstrated that timely plasmapheresis significantly reduces the duration of mechanical ventilation and improves functional outcomes in patients with severe Guillain-Barré Syndrome, including rare variants like pharyngeal-cervical-brachial.

Discussion:

The pharyngeal-cervical-brachial (PCB) variant of Guillain-Barré Syndrome (GBS) represents a rare and distinct subtype of this immune-mediated neuropathy. Unlike the classic form of Guillain-Barré Syndrome, which typically presents with ascending paralysis, the pharyngeal-cervical-brachial variant is characterized by weakness predominantly affecting the oropharyngeal, cervical, and upper limb muscles, with lower limb strength generally preserved. The diagnosis of this variant requires a high index of suspicion, particularly in patients presenting with acute bulbar symptoms and upper limb weakness without significant lower limb involvement (Nagashima et al[3]).

The pathophysiology of Guillain-Barré Syndrome, including its pharyngeal-cervical-brachial variant, involves an aberrant autoimmune response triggered by molecular mimicry between microbial antigens and peripheral nerve components. The production of antibodies against gangliosides, specifically GT1a and GD1a, is associated with this variant and contributes to the dysfunction of motor and sensory neurons (Willison et al.[5]). The clinical history of a preceding upper respiratory or gastrointestinal infection, as noted in this case, is a common antecedent.

Electromyography (EMG) and nerve conduction studies (NCS) are pivotal in confirming the diagnosis of Guillain-Barré Syndrome and its variants. These diagnostic modalities help differentiate between axonal and demyelinating forms of neuropathy and provide insights into the specific distribution of nerve involvement. In the pharyngeal-cervical-brachial variant, findings typically show reduced compound muscle

action potentials (CMAPs) with relatively preserved conduction velocities in lower limbs, aiding in distinguishing it from other Guillain-Barré Syndrome subtypes (Ropper et al.[4]).

Therapeutically, the pharyngeal-cervical-brachial variant of Guillain-Barré Syndrome responds well to immunomodulatory treatments, including plasmapheresis and intravenous immunoglobulin (IVIG). Plasmapheresis, which was employed in this case, has been shown to accelerate recovery by removing circulating autoantibodies and immune complexes (Khan et al.[2]). Clinical trials and systematic reviews have consistently demonstrated its efficacy in reducing morbidity and shortening the duration of ventilatory support in severe Guillain-Barré Syndrome cases (Dimachkie & Barohn,[1]).

Multidisciplinary management is critical in optimizing patient outcomes. This includes neurologists for diagnostic evaluation and treatment planning, pulmonologists for respiratory management, and rehabilitation specialists to ensure functional recovery. Close monitoring of respiratory function is particularly important, as bulbar involvement increases the risk of respiratory failure, necessitating timely intervention, as seen in this case.

In conclusion, the pharyngeal-cervical-brachial variant of Guillain-Barré Syndrome, while rare, should be considered in patients presenting with bulbar and upper limb weakness following an infectious trigger. Early diagnosis and prompt initiation of plasmapheresis or intravenous immunoglobulin can significantly improve prognosis. Further research into the immunopathogenesis of this variant may lead to more targeted therapeutic strategies.

Conclusion:

The pharyngeal-cervical-brachial variant of Guillain-Barré Syndrome, while rare, should be considered in patients presenting with bulbar and upper limb weakness following an infectious trigger. The specific triggers, detailed history, and early examination play crucial roles in the timely identification of Guillain-Barré Syndrome. Early diagnosis and prompt initiation of plasmapheresis or intravenous immunoglobulin can significantly improve prognosis. Multiple comparative trials evaluating intravenous immunoglobulin versus plasmapheresis have shown that plasmapheresis offers promising results, particularly in severe cases. A multidisciplinary approach involving neurology, critical care, and rehabilitation ensures comprehensive care and better outcomes. Continued research into the immunopathogenesis and tailored therapeutic strategies for Guillain-Barré Syndrome variants is essential for advancing treatment outcomes.

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Role and contribution of authors:

Shazaf Masood Sidhu, collected the data, references and did the initial writeup.

Muhammad Irfan Malik, collected the data and helped in introduction writing.

Dawood Sohail, collected the data, references and helped in discussion writing.

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