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Case Report

Pharyngo-Cervico-Brachial Variant of Guillain-Barré Syndrome: A Case Report

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Abstract

Objective: To report a case of a patient with Pharyngo-Cervico-Brachial syndrome, as an unsual variant of Guillain-Barré Syndrome.

Methods: Patient data were obtained from medical records from "Clínica Universidad de Navarra", Pamplona, Spain.

Results: We report the case of a 41-year-old male with no relevant medical history or recent vaccinations, who presented to the emergency department with a 24-hour history of progressive dysphagia, dysarthria, choking episodes, and ascending paresthesias involving the limbs and tongue. Neurological examination revealed moderate dysarthria, nasal voice, weakness of soft palate elevation, and generalized areflexia, without motor or sensory deficits in the limbs or ocular motility impairment. Persistent tachycardia was observed during hospitalization. Brain MRI was unremarkable. Cerebrospinal fluid analysis revealed albumin-cytological dissociation. Anti-ganglioside antibodies (anti-GT1a, anti-GT1b, and anti-GQ1b) were detected. Electrophysiological studies showed a mild reduction in motor and sensory nerve conduction amplitudes in the upper limbs, with normal findings in the lower limbs. The patient received intravenous immunoglobulin (2 g/kg over 5 days) with good tolerance and progressive clinical improvement.

Keywords: *Guillain-Barré syndrome, anti-ganglioside antibodies, Pharyngo-Cervico-Brachial, anti-GT1a, anti-GQ1b.*

Introduction

Guillain-Barré syndrome is a rapidly progressive, monophasic, presumably post-infectious disease characterized by weakness of the limbs and areflexia or hyporeflexia, and associated with peripheral nerve damage, either demyelinating or axonal, with an immune basis. It has been linked to prior exposure to pathogens, most commonly Campylobacter jejuni, and even to previous vaccination history. The diagnosis typically relies on clinical and electrophysiological characteristics, supported by the demonstration of antibodies against peripheral nerve gangliosides. Its treatment is still based on the use of immunoglobulins or plasmapheresis, with typically favorable response rates.[1]

There are various reported variants with different degrees of involvement: sensory, ataxic, autonomic, ophthalmoplegic, as well as different electrophysiological patterns: axonal, demyelinating, or mixed. Some varieties present predominantly a focal involvement, which constitutes the minority of cases, such as Miller Fisher syndrome, the pharyngo-cervico-brachial variant, or Bickerstaff encephalitis.[1]

The pharyngo-cervico-brachial variant is a rare entity, constituting approximately 3% of all Guillain-Barré syndrome cases. It is characterized by facial, pharyngeal, cervical, and upper limb involvement, with sparing of the lower limbs. The electrophysiological pattern most commonly associated is axonal damage.[2] The most frequently associated antibodies against gangliosides are anti-GT1a, which are often related to oropharyngeal involvement, likely due to the predominant presence of these gangliosides in this anatomical area.[3]

Case Presentation

We describe the case of a 41-year-old male, with no significant medical history except for an upper respiratory tract infection 7 days prior, which had resolved by the time of consultation, and no recent vaccination history. He presented to the emergency department of our hospital with a 24-hour history of progressive dysphagia, which caused choking and dysarthria, along with ascending paresthesias in the upper and lower limbs and the tongue. The patient did not report weakness in other locations or visual symptoms. On physical examination, weakness in the elevation of the soft palate was noted, along with moderate dysarthria and a nasal voice, accompanied by generalized reduction in osteotendinous reflexes. No limitation of extraocular movements, weakness, or sensory disturbances in the extremities were observed. During his stay, he had a constant tendency toward tachycardia. Complementary imaging studies were performed, with normal cerebral MRI findings. A lumbar puncture was done for cerebrospinal fluid (CSF) analysis, which revealed albuminocytological dissociation. Anti-ganglioside antibodies were detected: anti-GT1a, anti-GT1b, and anti-GQ1b. Electrophysiological studies showed a slight decrease in the amplitude of motor and sensory conduction velocities in the upper limbs, with normal findings in the lower limbs. Immunoglobulins were started at a dose of 2g/kg, administered over 5 days, with good tolerance and progressive improvement in symptoms overall.

Discussion

The pharyngo-cervico-brachial variant is characteristically very uncommon as a presentation of Guillain-Barré syndrome. Diagnosis requires relatively symmetric oropharyngeal, cervical, and upper limb weakness, along with decreased osteotendinous reflexes, a monophasic course, and a time interval between the onset and clinical nadir of between 12 hours and 28 days. Additionally, a prior infectious history, the presence of albuminocytological dissociation in CSF, a compatible electrophysiological pattern, and the presence of anti-GT1a antibodies are strong diagnostic indicators.[2] Although our case did not show clinically evident weakness in the neck or upper limbs, the characteristic pharyngeal pattern, along with the changes in CSF, detection of anti-GT1a antibodies, and the electrophysiological pattern, suggest a milder but relatively characteristic presentation. The progressive dysphagia and its potential complications justified the early initiation of immunoglobulins, resulting in satisfactory recovery. Although tachycardia was persistent, no other signs of autonomic involvement were observed. Compared to other reported cases, the common feature is oropharyngeal involvement, though without facial[2], cervical, or brachial[4,5] involvement. While paresthesias were initially noted in the lower limbs, they did not correspond to lower limb involvement as seen in some atypical pediatric cases[6]. The study and consideration of these rare variants allow for early identification and anticipation of potential complications that could compromise the patient's vital status, enabling timely initiation of targeted therapies to contain it.

Conclusion

We present a clinical case of a male patient with bulbar involvement in the context of a rare variant of Guillain-Barré syndrome, which, although he did not develop clear clinical signs of upper limb muscle weakness, constitutes an early presentation that will aid in recognizing future cases, enabling both diagnosis and timely management.

Conflicts of Interest

The authors declare no conflict of interest.

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