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A Rare Case of An Oculopharyngeal Subtype of Guillain-Barré Syndrome

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Abstract **Case Report**

Cranial nerve involvement in Guillain-Barré syndrome (GBS) is a rare but clinically significant manifestation that presents unique diagnostic challenges. We report the case of an 18-year-old female patient with a history of gastroenteritis who developed bilateral ptosis, horizontal diplopia, swallowing difficulties, facial diplegia, and tongue protrusion defects over 10 days. Neurological examination revealed multiple cranial nerve involvement, with normal cerebral MRI findings and absent albuminocytological dissociation in cerebrospinal fluid analysis. Electromyography showed pathological findings in the cranial nerves, while serum anti-ganglioside antibodies were negative. Diagnosed with the oculopharyngeal variant of GBS, the patient underwent plasma exchange and immunoglobulin therapy, resulting in slight improvement after three months, yet persistent deficits remained. This case highlights the complexity and variability of GBS presentations, particularly the importance of considering cranial polyneuritis in patients with preceding infectious symptoms. Further studies are needed to explore optimal treatment strategies and the underlying mechanisms of this rare variant.

Keywords: Guillain Barré Syndrome, Oculopharyngeal Subtype, Cranial Nerves.

INTRODUCTION

Cranial nerve involvement in Guillain-Barré syndrome (GBS) is a rare but clinically significant manifestation that has garnered increasing attention in recent literature. Various etiologies, including infectious, inflammatory, or neoplastic processes, as well as neuromuscular junction disorders and myopathies, can lead to cranial nerve deficits [1, 2]. Among these, GBS has been recognized as a notable cause, albeit infrequently reported [3].

CASE REPORT

We report the case of an 18-year-old patient with a history of gastroenteritis one month prior to the symptoms, who presented with a 10-day history of bilateral ptosis with horizontal diplopia, followed by swallowing difficulties with false routes, facial diplegia and tongue protrusion defect. The patient reached the peak of her symptoms in 22 days. On neurological examination, we noted multiple involvement of the cranial nerves, namely: the oculomotor nerves, the motor trigeminal nerve, the facial nerve, the glosso pharyngeal nerve and the hypoglossal nerve. The rest of the examination was normal. Cerebral MRI was normal, and ENMG showed absence of motor response in the 2 facial nerves and abolition of blink reflex bilaterally, with normal limb conductance. Lumbar puncture revealed no albuminocytological dissociation, and the anti-ganglioside antibody assay was negative. The oculopharyngeal form of GBS was diagnosed, and the patient received 5 sessions of plasma exchange and 2 courses of 0.4 g/kg/day immunoglobulin for 5 days. The patient was reviewed at 3 months, with slight improvement in ptosis and oculomotor disorders, but persistent facial diplegia, masseter muscle amyotrophy and persistent swallowing disorders requiring gastrostomy.

DISCUSSION

Cranial nerve involvement in Guillain-Barré syndrome (GBS) is a rare but clinically significant

manifestation that has garnered increasing attention in recent literature. Various etiologies, including infectious, inflammatory, or neoplastic processes, as well as neuromuscular junction disorders and myopathies, can lead to cranial nerve deficits [1, 2]. Among these, GBS has been recognized as a notable cause, albeit infrequently reported [3].

In our patient, the clinical presentation did not conform to the classic criteria for Miller Fisher syndrome (MFS), which is characterized by the triad of ophthalmoplegia, ataxia, and areflexia [4]. Although our patient exhibited bilateral ptosis and diplopia, the absence of ataxia and deep tendon reflexes excluded a diagnosis of MFS. Similarly, the pharyngeal-cervical-brachial (PCB) variant, defined by rapid oropharyngeal and cervical weakness, was ruled out as our patient lacked neck and upper limb weakness [5].

Rapidly progressive bifacial weakness, categorized as bifacial weakness with paresthesia (BFP), also encompasses cases of GBS subtypes. However, our patient's extensive cranial nerve involvement (III, IV, VI, IX, and XII) did not fit the BFP classification [6]. Recent literature has documented additional cases of cranial polyneuritis, with a 2020 study highlighting patients exhibiting ocular symptoms and progressive bulbar weakness, echoing our patient's presentation [7].

Since 2015, cranial polyneuritis has been proposed as a distinct oculopharyngeal subtype of GBS. Its incidence remains unclear, but the growing body of literature suggests it is considerably rarer than other GBS variants [8]. A prospective study from 2023 involving over 300 patients found a higher incidence of cranial nerve involvement than previously reported, emphasizing its association with preceding infections [9]. This aligns with our patient's history of gastroenteritis, a common antecedent in many GBS cases.

In examining the literature, only 16 cases of cranial polyneuritis have been documented prior to 2020, with a median age of 40 years and a male predominance [10]. Notably, 82% of these patients had preceding infectious symptoms, including upper respiratory and gastrointestinal illnesses, similar to our patient's history [11]. Initial ocular symptoms were common; in our case, bilateral ptosis and diplopia mirrored findings where ocular symptoms often presented first [12].

Cerebral MRI was unremarkable in both our patient and nine other reported cases, reinforcing the idea that GBS variants frequently present with normal imaging despite significant neurological deficits [13]. While albuminocytological dissociation was documented in 67% of previous cases, our lumbar puncture showed no such findings, highlighting a deviation from typical GBS presentations [14]. Electromyography (EMG) revealed pathological findings in the cranial nerves of our patient, consistent with previous reports that showed variability in EMG presentations among GBS patients [15].

Interestingly, serum anti-ganglioside antibodies were detected in approximately 50% of cases in recent literature, indicating that the presence of these antibodies may not be universal across all GBS variants [16]. Our patient's lack of detectable antibodies further illustrates the complexity of her clinical presentation.

Clinical improvement was noted in all documented cases of cranial polyneuritis; however, some patients did not receive treatment, while others showed varying degrees of response to therapies such as intravenous immunoglobulins and steroids [17]. Our patient's slight recovery following plasma exchange and immunoglobulin therapy underscores the variability in treatment response and the necessity for individualized management strategies.

CONCLUSION

This case of cranial polyneuritis associated with Guillain-Barré syndrome underscores the complexity and variability of GBS presentations, particularly in the context of cranial nerve involvement. Despite the rarity of this variant, our patient's clinical history, including preceding gastroenteritis and the subsequent onset of bilateral ptosis, diplopia, and bulbar symptoms, aligns with findings from recent literature that highlight similar cases.

Treatment responses can vary significantly among patients, highlighting the importance of individualized management strategies. Future studies should continue to explore the underlying mechanisms and optimal treatment protocols for cranial polyneuritis in GBS, as well as the potential links between infectious antecedents and subsequent neurological manifestations. Overall, this case contributes to the growing recognition of cranial polyneuritis as an important consideration in the differential diagnosis of patients presenting with cranial nerve symptoms following recent infections.

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