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RARE PRESENTATION OF ACUTE INFLAMMATORY DEMYELINATING POLYRADICULOPATHY (AIDP)

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ABSTRACT

This is the description of a case report of a patient who presented very uniquely with AIDP (Acute Inflammatory Demyelinating Polyradiculopathy). It commonly presents as bilateral lower extremity weakness and areflexia with albuminocytologic dissociation on CSF findings. Sensory symptoms are not prominent presenting features in patients. However, Bulbar and Miller Fisher variants of AIDP are very rare. This case report highlights the unique presentation where unilateral facial nerve palsy along with perioral numbness and decreased taste sensation was the presenting symptom. A 33-year-old male with a past medical history of rhabdomyosarcoma s/p resection who presented with the chief complaint of numbness and dysphagia. He developed left-sided facial droop and decreased taste sensation following an episode of bronchitis six weeks ago. Pertinent positives include mild dysphagia, decreased appetite, congestion and rhinorrhea. Physical exam was not significant except for decreased sensation of the left facial droop, decreased sensation of the right lower face, decreased light touch of the RUE, 3+ reflexes of both upper and lower extremities bilaterally. This case report highlights the unique presentation of Bulbar AIDP with some symptoms of Miller Fisher syndrome. Predominant sensory symptoms such as perioral numbness and decreased taste sensation might be the predominant symptoms that bring the patient to attention. Hence, it is imperative to perform a lumbar puncture to support the diagnosis of variant AIDP when predominant sensory symptoms are present.

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Introduction

Guillain-Barré syndrome (GBS) is characterized by a triad of rapidly progressing ascending paralysis, mild sensory loss and hypo- or areflexia. CSF analysis reveals albuminocytologic dissociation in 90% of cases (1). Acute inflammatory demyelinating polyneuropathy (AIDP) is the most common form of GBS. In AIDP, the immune system targets the peripheral nerve myelin sheath with some axonal loss (1). There are different presentations of GBS based on the types of nerve fibers involved (motor, sensory, sensory and motor, cranial or autonomic), method of fiber injury (demyelinating versus axonal), and the presence of changing levels of consciousness (1). Miller Fisher Syndrome (MFS) is a variant of GBS which consists of ophthalmoplegia, ataxia, and areflexia without any weakness (1). Most patients have at least two of the above symptoms along with elevated CSF protein and autoantibody (1).

Atypical presentation of GBS such as the pharyngeal-cervical-brachial variant can occur in the form of unilateral or bilateral facial palsy, dysphagia like symptoms due to bulbar muscles being affected mostly with or without other cranial nerve involvement, extremity weakness or ataxia (2).

Materials and Methods:

Not applicable

Case presentation:

A 33-year-old male with a past medical history of rhabdomyosarcoma s/p resection who presented with the chief complaint of numbness. Patient admitted to a baseline level of numbness and tingling of the bilateral hands and feet as a side effect of the chemotherapy and radiation. However, he has noticed increased tingling of the right hand which is extending into the R forearm, perioral numbness and tingling with extension into the tongue, diminished sense of taste, left-sided facial droop and decreased ability to close the left eye. Patient admitted to having acute bronchitis six weeks ago for which the patient was given a course of Bactrim. Review of system were all negative except for the decreased appetite due to diminished taste sensation, blurry vision, mild dysphagia, right posterior neck pain, rhinorrhea, congestion and fatigue. Physical exam was negative except for decreased sensation of the right lower face, left facial droop with incomplete closure of the L eye, decreased light touch of the RUE, 3+ reflexes of both upper and lower extremities bilaterally. Vitals were stable except tachycardia of 123 on presentation. Labs were not significant, and an MRI of the brain and cervical spine was done to rule out the possibility of leptomeningeal carcinomatosis and they were both negative for any acute pathology. A lumbar puncture was performed which showed an elevated protein of 117 mg/dl. The team discussed the likely diagnosis of Bulbar variant of Acute Inflammatory Demyelinating Polyradiculopathy (AIDP) and the patient agreed to a five-day course of IVIG. Upon the completion of the five-day treatment, patient stated that his symptoms are stable.

Results and Discussion:

GBS can be caused by infectious, inflammatory, or systemic diseases. C. jejuni is the most common culprit with cytomegalovirus being the second most

common infectious precipitants of GBS. However, EBV and influenza vaccination has also been implicated in a minority of GBS cases.

The main cell type affected in GBS are the motor or sensory neurons. Gangliosides are complex glycosphingolipids containing sialic acid and is present in the cell membranes of many cells and are concentrated in neuronal membranes and processes (3). The immune system in GBS is mistakenly attacking myelin or axons which serve as the pathway for communication to and from the brain. This autoimmune phenomenon occur because of the surface of C. jejuni contains polysaccharides that resembles the glycoconjugates of the nerve tissue membranes. This is referred to "molecular mimicry," wherein a single B- or T-cell receptor recognizes a microbe's structure and an antigen of the host at the same time[d]. As a result, these activated lymphocytes migrate across the endoneurial capillary walls and attract macrophages to cause an inflammatory response of the peripheral nervous system (4). It is these macrophages that invade the basal lamina of the Schwann cell and degrade the myelinated axons. Location where this phenomenon takes place will determine the corresponding neurological deficit seen in the patient. The typical electrophysiological features seen include multifocal slowing of nerve conduction and partial conduction block (3).

There are very atypical presentations of GBS such as polycranial neuritis and acute bulbar palsy plus syndrome where patients present with oropharyngeal weakness with or without other cranial nerve involvement (5). In these rare forms of GBS with very nonspecific features, neuroimaging has become an important tool since the postgadolinium enhancement of the peripheral nerve roots and cauda equina is present on spinal MRI (5). The pattern of enhancement can provide diagnostic clues about the type of demyelination (5). The enhancement of the dorsal and ventral root is commonly seen in AIDP compared to anterior root enhancement seen in acute motor axonal neuropathy (AMAN) (5). Cranial nerve enhancement has been noted in MFS and polyneuritis cranialis forms of GBS (5). This enhancement occurs because of the breakdown of blood brain barrier due to inflammation (5).

This case report highlighted the importance of keeping a wide differential when patients present with oropharyngeal and facial weakness in the presence or absence of hypo/areflexia and other cranial nerve palsies. It is imperative that to think of bulbar AIDP variant and obtain a lumbar puncture to further help with diagnosis and management of these patient.

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References

Dimachkie MM, Barohn RJ. Guillain-Barre syndrome and variants. Neurol Clin 2013; 31:491.

Ray S, Jain PC. Acute bulbar palsy plus syndrome: a rare variant of Guillain-Barre syndrome. J Pediatr Neurosci 2016; 11:322–323.

Hughes RA, Hadden RD, Gregson NA, Smith KJ. Pathogenesis of Guillain-Barré Syndrome. J Neuroimmunology. 1999; 100:74–97.

Yu, R. K., Usuki, S. & Ariga, T. Ganglioside molecular mimicry and its pathological roles in Guillain–Barre syndrome and related diseases. Infect. Immun. 2006; 74:6517–6527.

Sharma, K., Tengsupakul, S., Sanchez, O., Phaltas, R., & Maertens, P. Guillain–Barré syndrome with unilateral peripheral facial and bulbar palsy in a child: A case report. SAGE Open Med Case Rep. 2019; 21:7.