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.

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 fibres involved (motor, sensory, sensory and motor, cranial or autonomic), method of fibre 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].

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, 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

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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

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 is the motor or sensory
neurons. Gangliosides are complex glycosphingolipids containing
sialic acid and are 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. 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 post
gadolinium 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 polynueiritis cranialis forms of GBS [5]. This
enhancement occurs because of the breakdown of blood brain barrier
due to inflammation [5].

Discussion

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 patients.

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References

1. Dimachkie MM, Barohn RJ. Guillain-Barre syndrome and variants.
2. Ray S, Jain PC. Acute bulbar palsy plus syndrome: a rare variant of
3. Hughes RA, Hadden RD, Gregson NA, Smith KJ. Pathogenesis of Guillain-
pathological roles in Guillain-Barré syndrome and related diseases. Infect.
Barré syndrome with unilateral peripheral facial and bulbar palsy in a