Miller Fisher Variant of Guillain-Barre Syndrome Masquerading as Acute Sphenoid Sinusitis with Orbital Apex Syndrome

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Abstract

Background: Miller-Fisher syndrome (MFS), a variant of Guillain-Barre syndrome, is a rare disorder typically characterized by a triad of ataxia, areflexia, and ophthalmoplegia, which may have a highly variable clinical presentation.

Methods: Case report

Results: We report a case of MFS in a 45-year-old female presenting with sphenoid sinusitis and sixth-nerve palsy. She underwent endoscopic sphenoid sinusotomy without improvement, had postoperative deterioration, was diagnosed with MFS, and was treated with intravenous immunoglobulin with complete response.

Conclusion: Because of the potential severity of Guillain-Barre syndrome, great vigilance should be taken when examining sixth-nerve palsies to prevent misdiagnosis and delay in treatment of the MFS variant of this disease.

Introduction

Miller-Barre Syndrome (GBS) is a relatively uncommon disease, with an incidence rate of approximately 1-2 cases per 100,000 people annually. Currently, GBS has six known subtypes, resulting in variable clinical presentation. Symptoms range from cranial nerve involvement, to neuropathy, ataxia, progressive paralysis, or death. Emerging treatment is paramount to prevent permanent morbidity or death. We present a unique case in which a patient with GBS, specifically Miller Fisher Syndrome (MFS), presented with symptoms suggesting of sphenoiditis.

Case Report

A 45-year-old female was emergently transferred to our tertiary care center with a 2-day history of progressive diplopia, ataxia, left-sided headache, and left sphenoid sinus opacification. She reported upper respiratory tract infection (URI) symptoms for the past 7 days that were treated with amoxicillin without improvement. Initial physical examination revealed a left abducens nerve palsy, unsteady gait, general weakness, slurred speech and decreased hand sensation. Nasal endoscopy showed mild congestion. Computed tomography (CT) scanning of the orbits and paranasal sinus revealed opacification of the left sphenoid sinus (Fig. 1 A & B). Gadolinium-enhanced magnetic resonance imaging (MRI) showed opacification of the left sphenoid sinus with unremarkable orbits, cavernous sinus, and brain parenchyma (Fig. 2 A & B). The diagnosis of isolated left acute sphenoid sinusitis with possible cavernous sinus involvement was entertained, and the patient underwent an uneventful left sphenoid sinusotomy. Intraoperatively, turbid serous fluid was drained from the left sphenoid sinus and a biopsy of the sphenoid mucosa was taken. Histopathological analysis showed fragments of inflamed respiratory mucosa without organisms. The patient was subsequently started on Vancomycin and ceftriaxone.

Postoperatively, her neurological symptoms rapidly worsened. Within 7 hours of the surgery, she progressed to bilateral ophthalmoplegia with ptosis, and mild facial droop. Her speech was fluent, but mildly dysarthric. She developed sensory loss, motor weakness, and severe limb ataxia. Emergent neuro-ophtalmic and neurologic consultations were obtained. Complete areflexia, progressive bulbar palsy with headaches, decreased respiratory vital capacity, and difficulty swallowing were noted. The triad of areflexia, ataxia, and ophthalmoplegia allowed the clinical diagnosis of MFS. Lumbar puncture was remarkable only for increased cerebrospinal fluid glucose; the protein was 18, there were 6 white cells (79% lymphocytes).

Discussion

Guillain-Barre Syndrome is an autoimmune disorder with different clinical subtypes. These subtypes include MFS, acute inflammatory demyelinating polyradiculoneuropathy, acute motor axonal neuropathy, motor sensory axonal neuropathy, acute motor sensory axonal neuropathy, and acute panautonomic neuropathy. Since GBS is an autoimmune disorder, a majority of cases are associated with an antecedent infection. In fact, Gozzi and Granieri reported that in populations studied in Minnesota (USA) and Italy, antecedent infections prior to the presentation of GBS symptoms were present in 65% and 59% of patients respectively. Furthermore, most of these antecedent infections were URIs. Our present case also suffered from a URI for seven days prior to admission. Miller Fisher Syndrome represents only 5% of GBS cases, and usually presents with the clinical triad of ophthalmoplegia, ataxia, and areflexia, all of which our patient demonstrated. Abducens nerves are usually affected bilaterally in MFS. However, some researchers have previously linked isolated sixth nerve palsy as suffered by our patient, to MFS. In 2006, Tatsumoto et al examined 100 cases of isolated sixth nerve palsy. Of the 100 cases, 65 had symptoms of an antecedent infection, 27 had absent or reduced tendon reflexes, and 25 had positive serum anti-GQ1b antibodies, suggesting GBS. In fact, the authors concluded that in certain cases, isolated sixth nerve palsy could be labeled as regional variant of GBS, or a form of MFS. In 2007, Smith et al presented a case of a 32-year-old man with MFS, who presented only with left unilateral abducens nerve involvement.

Nonetheless, it is exceedingly more common for an isolated unilateral sixth nerve palsy to be found in the context of sphenoid sinusitis than in MFS. Ada et al reported a case in which a 12-year-old female presented with isolated sphenoid sinusitis, which resulted in unilateral sixth nerve palsy. The sixth nerve palsy improved with antibiotic therapy and endoscopic sphenoidotomy. Shukla et al reported a similar case in which a 38-year-old man presented with acute sinusitis, who later developed diplopia as a result of isolated sixth nerve palsy. The is also implicated optic, oculomotor, and trigeminal nerves subsequently became involved as the condition progressed. The patient made a full recovery after emergent endoscopic surgical drainage and antibiotic therapy.

Although unilateral sphenoid sinusitis with associated abducens nerve palsy is not uncommon, given the possibility of a catastrophic missed diagnosis of MFS variant of GBS, otolaryngologists should be aware of this entity and keep a high level of suspicion in patients presenting with isolated sphenoid sinusitis with abducens nerve palsy, and inquire about ataxia beyond what was allowed for by diplopia, as well as testing for areflexia. The laterality of the sixth nerve palsy may be useful in determining the presence or absence of underlying sphenoid sinusitis. If a patient's clinical course is rapidly deteriorating after adequate sphenoid drainage and broad-spectrum antibiotics, and no pain is found intraoperatively in the opacified sinus cavity, emergent evaluation is warranted and other potential diagnoses (such as MFS) entertained and investigated.

Conclusions

Miller Fisher syndrome, a variant of GBS, is a rare disorder with a highly variable clinical presentation. One such presentation is unilateral abducens nerve palsy which may mimic sphenoid sinusitis with cavernous sinus disease and abducens nerve palsy. While abducens nerve palsy secondary to sphenoid sinusitis is usually successfully treated with urgent sphenoidotomy with appropriate antibiotics, this management strategy is ineffective in GBS, and may cause significant delay in treatment with potential devastating outcomes. Otolaryngologists should maintain a high level of suspicion in patients presenting with unilateral sixth nerve palsy in the setting of an opacified sphenoid sinus as this may mask a potentially more devastating condition.

References