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Case Report

Acute Unilateral Vision Loss and Optic Perineuritis (OPN) Associated with Mucormycosis Sinusitis in Geriatric Patient

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Abstract

Background: Optic Perineuritis (OPN) is an uncommon condition that has been reported with many systemic disorders.

Methods: We report a geriatric patient who suffered from acute unilateral vision loss and OPN in association with mucormycosis. We also included a brief literature review pertinent to OPN.

Results/Discussion: OPN has been reported in association with many systemic disorders, it has infrequently been reported with mucormycosis sinusitis.

Conclusions: OPN may develop secondary to mucormycosis sinusitis.

Keywords: Optic Perineuritis (OPN); Pathophysiology; Relative Afferent Pupillary Defect (RAPD)

Background

OPN is an uncommon condition that can be primary-idiopathic- or secondary to other conditions. In this case report: we report a geriatric patient who suffered from acute unilateral vision loss and OPN in association with mucormycosis. The patient developed OPN in the context of fungal sinusitis due to *Mucor* sps. Most Mucor species are commonly found in soil and are unable to infect humans. Thermotolerant species such as Mucor indicus sometimes cause opportunistic, and often rapidly spreading, necrotizing infections known as Mucormycosis (previously called zygomycosis). Mucor mycosis (zygomycosis) can cause a rare but yet aggressive fungal sinusitis in immunosuppressed patients like diabetics, HIV patients, or solid organ transplant patients. Prognosis is generally poor even on the most aggressive intervention, debridement, and intravenous potent antifungal (amphotericin B) [1].

Methods

This is a case report of acute unilateral vision loss and Optic Perineuritis (OPN) in association with fungal sinusitis due to Mucor mycosis, which has infrequently been reported to be associated with OPN in the past. We also included a brief literature review pertinent to OPN.

Case Presentation: Invasive Mucormycosis Causing OPN

A 75-year-old female with a history of Type 2 DM, COPD, HTN, HLD and CKD presented to the ED after a witnessed fall. Her daughter stated that prior to coming to the ED, her mom was confused and fell to the floor. At baseline, the patient is independent and cares for her own medications. Notably, she did admit to a poor appetite over the last few weeks prior to admission and has been struggling with increased wheezing and shortness of breath due to her asthma. The patient reports taking prednisone 5 mg as needed. On admission to the hospital, she was found to be in diabetic ketoacidosis for which she was treated with an insulin drip. Additionally, she was found to have leukocytosis and proptosis of her right eye for which ophthalmology was consulted.

On initial ophthalmologic evaluation, visual acuity at near was 20/70 right eye (OD), 20/40 left eye (OS). Intraocular pressures were 29 OD, 19 OS. Pupils were unequal and minimally reactive to light. Extraocular movements OD were limited (-3) on all directions of gaze; and OS were limited (-1) in elevation only. External exam was significant for prominence of right globe, complete right ptosis at rest and mild conjunctival chemosis. Dilated fundus exam revealed normal appearing optic nerves bilaterally, with flushing of vessels at the optic disc with slight globe pressure. Optic cup-to-disc ratio was 0.5 bilaterally. Fundus exam was otherwise unremarkable.

Contrast-enhanced MRI of the orbits on showed diffuse paranasal sinus disease in a pattern suggestive of acute sinusitis concerning fungal infection and right OPN figure 1. There was apparent dehiscence of the lateral nasal wall/medial orbital wall posteriorly and right orbital infection including 1 x 0.4 x 1.4 cm abscess in the right orbit extraconally and medial to the medial rectus muscle. Additionally, mild intraorbital fat infiltration, diffuse hyperenhancement and enlargement of the right-sided extraocular muscles, and proptosis were seen. ENT was consulted for surgical evaluation. They performed a bilateral functional endoscopic sinus surgery with cultures showing MRSA, Klebsiella pneumoniae, and Enterobactor cloacae. Pathology revealed invasive fungal infection, figure 2. The patient was started on Amphotericin B. After the initial surgery, the patient did not wish to proceed with future debridements. Instead, she wished to continue with Amphotericin B administration and repeat imaging every 2 weeks. Patient reported a loss of vision in her right eye three days after surgery. Repeat MRI revealed post-surgical swelling compressing her optic nerve. Ophthalmology and ENT deferred from starting steroids at this time due to extensive infection. Due to poor prognosis and patient and daughter preference, the patient was transitioned to comfort measures and was discharged home on hospice.

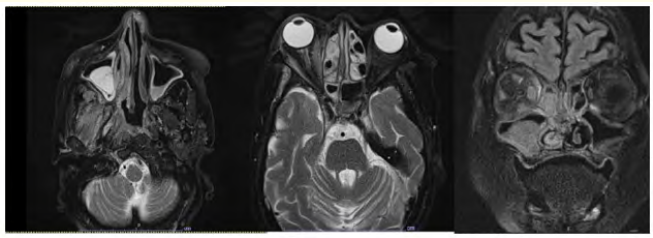

Figure 1: MRI of the Orbits shows right OPN and invasive fungal sinus infection.

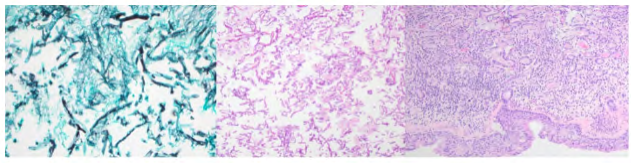

Figure 2: H&E and GMS sections demonstrate marked submucosal acute and chronic inflammation and fungal infection as evidenced by the presence of broad hyphae with variable angle branching.

Results/Discussion

Optic perineuritis (OPN) is an uncommon inflammatory disorder in which the inflammation is confined to the optic nerve sheath. Most cases are idiopathic (idiopathic optic perineuritis). OPN has been reported to be associated with many systemic disorders but to our knowledge, it has infrequently been reported to complicate mucormycosis infection of the sinuses. Systemic disorders reported being associated with OPN include but not limited to the following: sarcoidosis [2-4], IgG4-related disease [5], granulomatosis with polyangiitis (GPA) [3] [6], giant-cell arteritis (GCA) [7], Behcet's

disease [8], systemic lupus erythematosus [1], inflammatory bowel disease [9], syphilis [3], tuberculosis [3], herpes simplex virus [10], herpes zoster virus [3], leukemia [11], other viral encephalitides [12], and primary or metastatic malignancy [5]. It has also been documented to present simultaneously with neuroretinitis [13].

Pathophysiology

The pathophysiology of OPN may reflect the underlying cause and most cases remain idiopathic. Although the immune-mediated inflammation is primarily within the nerve sheath, axons of the optic nerve may also be affected to varying degrees. The variable degree and location of inflammation contribute to the range of symptoms and exam findings.

Signs/symptoms

The clinical presentation of OPN may mimic optic neuritis. Most patients present with acute visual loss, eye pain, or both. Like optic neuritis, the eye pain may be exacerbated by eye movement but the pain in OPN may be more severe or last longer compared to the typical optic neuritis. The visual loss in OPN is variable and may range from none to severe. Patients may describe visual blurring, dimming, "spots" in vision, or splotches [2]. A phenomenon known as "amaurosis fugax," translated as "fleeting dark," and interpreted as transient loss of vision is frequently associated with OPN as well.

Clinical diagnosis

OPN is often clinically difficult to distinguish from optic neuritis and, in some cases, from other orbital inflammatory or infectious conditions due to the overlap in symptomatology. However, it is important to identify underlying treatable causes.

Clinical similarities to optic neuritis include decreased visual acuity, the possible presence of a relative afferent pupillary defect (RAPD), visual field loss, and dyschromatopsia. However, there are often clinical differences between OPN and optic neuritis. These include faster onset in optic neuritis (days) compared to OPN (weeks). Central vision is more often spared in OPN, which also leads to milder dyschromatopsia and sporadic or more subtle RAPD. The diagnosis of OPN is typically based on a combination of clinical and radiographic findings. Gadolinium-enhanced, fat-saturated T1 magnetic resonance imaging (MRI) of the orbits is key to the diagnosis of OPN.

General treatment

Initial treatment for OPN generally includes high doses of systemic corticosteroids. The dosage of steroids may vary depending on the frequency of attacks, with higher frequency requiring a higher dosage [2]. Likewise, higher rates of recurrence were seen in case studies that used 30-40 mg/day versus a higher dosage of 60-80 mg/day.

Prognosis

OPN typically has a relatively good prognosis. Unlike demyelinating optic neuritis, there is no known association between OPN and multiple sclerosis [2]. The visual outcome tends to be excellent in patients with OPN, with most patients in clinical case studies returning to 20/20 vision or better [2]. However, the prognosis appears to be poor when OPN is associated with Mucormycosis (zygomycosis) sinusitis [14].

Conclusions

OPN is an uncommon condition that may be secondary to an underlying cause or idiopathic. OPN typically has a relatively good prognosis. Invasive infections involving the sinuses, as seen in Case 2 reported here, have infrequently been previously recognized as causes of OPN. Mucormycosis (zygomycosis) manifests in different ways in immunocompromised patients and those with diabetes mellitus. Devastating rhino-orbital-cerebral and pulmonary infections are the most common syndromes caused by these fungi. Prognosis is generally poor even with aggressive interventions, including debridement and IV amphotericin B. Multimodal therapy with surgical debridement and antifungal chemotherapy is required for an optimal outcome. Additionally, discontinuation of immunosuppressive therapy, if possible, is the cornerstone of management.

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