Granulomatosis with polyangiitis presenting as vision loss and sinus disease

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Granulomatosis with Polyangiitis Presenting as Acute Vision Loss in a Young Cocaine User

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Abstract

Purpose: To report a case of acute vision loss due to granulomatosis with polyangiitis in a 32-year-old female with a history of cocaine abuse.

Methods: Interventional case report.

Results: A 32-year-old female with a past medical history of several years of intranasal cocaine abuse presented with acute vision loss in the right eye and mild disc edema. Orbital neuroimaging demonstrated bilateral diffuse panhypersensitivity, extensive bony erosion involving the posterior floor and medial wall of the right orbit, and a soft tissue mass extending from the maxillary and ethmoid sinuses into the orbital apex. Right ethmoid sinus and orbital apex biopsy demonstrated necrotizing granulomatous inflammation and vasculitis. Laboratory results included positive c-ANCA with positive anti-PR3, negative levamisole antibody, and negative HNE antibody.

Conclusion: Granulomatosis with polyangiitis can present in a patient of any age or demographic. The presentation is wide and varied and can be confounded by other etiologies.

Introduction

A 32-year-old Caucasian woman with a several year history of intranasal cocaine abuse presented with a history of sudden-onset of right eye vision decline. This progressively worsened over two weeks before the patient presented to Ophthalmology for evaluation. The patient denied any significant other past ophthalmic history.

Her ophthalmic examination demonstrated count fingers vision at 1 foot distance in the right eye and no letters could be distinguished on the near card. Other abnormal findings on ophthalmic examination included a right afferent pupillary defect, mild abduction deficit, diffuse gross visual field restriction, and no color vision. Intravascular pressure and anterior segment examination were normal in the right eye. Funduscopic examination demonstrated grade I optic disc edema and retinal venous dilation and tortuosity as shown in Figure 1A. Her left eye vision was 20/20 and the remaining ophthalmic exam was unremarkable. It was also noted that the patient demonstrated a saddle nose deformity, as depicted in Figure 2. Follow-up ophthalmic examination the next day demonstrated worsening of vision with no light perception vision (complete blindness) in the right eye.

Computed tomography (CT) demonstrated bilateral diffuse panhypersensitivity and extensive bony erosion including the posterior floor (Figure 1A) and medial wall (Figure 2B) of the orbit. Magnetic resonance imaging (MRI) demonstrated a soft tissue mass extending from the maxillary and ethmoid sinuses into the right orbital apex (Figure 4). Relevant laboratory results included positive cytoplasmic antineutrophil antibodies (c-ANCA) with positive anti-PR3 (PR3). Other pertinent labs included negative perinuclear antineutrophil antibodies (p-ANCA) and negative antineutrophil antibody (ANA). Sinus culture demonstrated methicillin-sensitive Staphylococcus aureus (MSSA).

Initial management included treating MSSA sinusitis with nafcillin along with initial management to decrease the mortality risk associated with GPA, whereas other conditions such as cyclophosphamide or rituximab due to the required long-term steroid-sparing agents such as cyclophosphamide or rituximab due to the required long-term immunosuppression.2,11,12 The necrotizing inflammation of GPA itself or the compressive optic neuropathy due to the mass effect at the right orbital apex likely resulted in vision loss for our patient.

Discussion

Acute vision loss in the setting of intranasal cocaine abuse can result from a variety of etiologies, thereby confounding a diagnosis of GPA in the patients. Infectious etiologies, such as orbital cellulitis (in this case due to MSSA sinusitis), can result in visual compromise. In immunocompromised patients, invasive necrotizing rhino-orbital-cerebral fungal infection, most notably mucormycosis, is important to consider as mortality rate has been reported to be 25 to 62 percent.2,5 Cultures and biopsy did not demonstrate any findings suggestive of fungal sinusitis in this patient.

Cocaine abuse can result in two syndromes related to this case: levamisole-induced vasculitis and cocaine-induced midline destructive lesions (CIMDL).2,3 Levamisole is a paralytic anticholinergic that is no longer prescribed for human use within the United States due to leukopenia, agranulocytosis, and skin vasculitis.5,6 It was estimated in 2009 that approximately 70 percent of cocaine within the United States is contaminated with levamisole.6,5,7 Snorting and smoking cocaine contaminated with levamisole can result in vasculitic purpuritic necrotic skin lesions and immunological changes.2,4 It is an often self-limiting condition upon drug cessation, although it can recur with reuse. Perinuclear antineutrophil antibodies (p-ANCA) are found in 86-100% with anti-myeloperoxidase in most of these cases.7,8,9 Anti-HNE can be positive in those having anti-PR3.10 Anti-HNE can be found in these patients. CIMDL may have similar laboratory findings; however, it presents as necrosis or erosion of midline facial structures including the nasal septum, turbinates, and/or lateral nasal wall including the lamina papyracea and orbital floor.1 A saddle-nose deformity can be seen in these patients. Most of these patients have positive anti-HNE,12 which is not seen in ANCA-vasculitides.

Immunological vasculitis caused by granulomatosis with polyangiitis (GPA) can result in granulomatous necrotizing inflammation and vasculitis as demonstrated on pathology. The most notable symptoms include constitutional symptoms, rhinosinusitis, nasal deformities, hemoptysis, cough, urinary abnormalities, renal dysfunction, and conjunctivitis.11,12 Laboratory results demonstrate positive c-ANCA with positive anti-PR3 in 82 to 94 percent of patients.11 This condition can have overlapping features with CIMDL. Extravascular microabscesses, necrotizing granulomas, and multinucleated giant cells may be few of the distinguishing features of GPA when compared to CIMDL. Disease management is imperative given the mortality rate of 90 percent of two years of GPA if left untreated.11 Treatment usually includes glucocorticoids and steroid-sparing agents such as cyclophosphamide or rituximab due to the required long-term immunosuppression.11,12 The necrotizing inflammation of GPA itself or the compressive optic neuropathy due to the mass effect at the right orbital apex likely resulted in vision loss for our patient.

Conclusion

Granulomatosis with polyangiitis can present in patients of any age or demographic and can result in acute vision loss. Its presentation is wide and varied and can be confounded by other etiologies, as exemplified in this case of a young patient with a history of cocaine abuse. Diagnosis is imperative to initiate immunosuppressive management to decrease the mortality risk associated with GPA, whereas other conditions associated with cocaine abuse are often self-limiting.

References

1. Cox, Gary M MD. Mucormycosis (zygomycosis). In: UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2020.