

From Sinus Congestion to Vision Loss

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A 58-year-old man presents with vision loss in his right eye. He reports having blurry vision for the last two weeks as well as rhinorrhea, sinus congestion, and subjective fevers. He is evaluated at an urgent care clinic for these symptoms; diagnosed with sinusitis; and prescribed diphenhydramine, loratadine, and antibiotics. His symptoms do not improve with the antibiotics. The morning of presentation, he awakes with loss of vision in his right eye and right-sided headache.

The differential diagnosis of vision loss is broad. Vision requires an image to pass through refractive structures of the eye, reach the retina, transmit the image via the optic nerve, and process the image through the occipital lobe of the brain. Problems anywhere along this pathway can cause vision loss. Pathology can be grouped into problems with the media, retina, optic nerve, and brain (Table 1).¹

The absence of eye pain is important. Most local problems like trauma, keratitis, ulcerations, uveitis, and acute angle closure glaucoma cause pain and redness in addition to vision loss. The absence of a red painful eye points away from these. A common culprit for painless acute vision loss is ischemia, but this most often occurs in the setting of risk factors for vascular disease such as smoking or hyperlipidemia.

The sinus congestion and rhinorrhea that preceded the vision loss are concerning for infections in the sinuses and orbital cellulitis. Alternately, overuse of anticholinergic medications can lead to mydriasis, loss of accommodation, and even angle closure glaucoma. The physical exam can help narrow the differential.

He denies any eye pain associated with the vision loss or trauma to the head. He reports no past medical history, does not take any medications regularly (other than as mentioned above), and has never smoked.

He is afebrile but hypertensive, with a blood pressure of 180/95 mm Hg. He is alert and oriented, follows commands, and is able to disclose details of his history without difficulty. Frontal and maxillary sinuses are not tender but fail to illuminate. Both the nasal mucosa and oropharynx appear normal.

He has normal visual acuity, intact extraocular movements, and normal pupillary reflex in his left eye. With his right eye, he is unable to count fingers or discern light. He has ptosis and impaired adduction, elevation, and abduction of the right eye. The right pupil is dilated greater than the left, and it sluggishly contracts with light. Ophthalmoscopic examination reveals no hemorrhage.

The eye exam is one of the most daunting parts of the physical exam to the internist. Decreased visual acuity in the right eye suggests pathology anterior to the optic chiasm (i.e. the eye or optic nerve). Pathology posterior to the optic chiasm usually presents with hemianopia (visual field loss that respects the vertical midline) and is detected with visual field testing.

An afferent pupillary defect is detected by swinging a light from one eye to the other and watching for pupillary response. A positive test means that pupillary response is impaired in the affected eye. An afferent pupillary defect is specific for optic nerve pathology. Ptosis and impaired extraocular movements suggest pathology affecting cranial nerves III and VI.

These findings help localize the pathology to the orbital apex, which contains the neurovascular structures of the eye including cranial nerves III, IV, and VI; the trigeminal nerve; the optic nerve; and the superior ophthalmic vein. Pathology at this site yields a constellation of signs and symptoms collectively termed orbital apex syndrome. Hallmarks of orbital apex syndrome are vision loss, ophthalmoplegia, anisocoria, and pupillary defects.²

A variety of conditions can present with orbital apex syndrome. They include mass lesions like head and neck or neural tumors; inflammatory diseases like sarcoidosis, lupus, and vasculitides; vascular lesions like an aneurysm; traumatic causes following surgery, injury, or fracture; and infections. Imaging and labs would be helpful to discern the cause.

Laboratory studies reveal a blood sugar above 400 mg/dL, creatinine of 2.3 mg/dL, and normal bicarbonate and anion gap. White blood cell count was $11.9 \times 10^9/L$. Further testing reveals a hemoglobin A1c greater than 12%. He is diagnosed with diabetes and started on insulin therapy.

CT scan of the orbit and brain reveals mucosal thickening in the maxillary, frontal, sphenoid, and ethmoid sinuses with air fluid levels consistent with sinusitis. MRI reveals paranasal sinusitis and vascular engorgement of the right superior ophthalmic vein concerning for cavernous sinus thrombosis. He is started on antibiotics for sinusitis.

Uncontrolled infection can definitely lead to orbital apex syndrome. Most likely, infection spread from his sinuses to the orbit. Cavernous

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sinus thrombosis is a complication of this pattern of infection via retrograde flow from the superior and inferior ophthalmic veins.

Staphylococcus aureus is the most common pathogen followed by Streptococcus, gram-negative rods, and anaerobes. Fungal infections occur less commonly. However, the setting of a new diabetes diagnosis and poorly controlled blood sugar raises suspicion for a fungal infection from Aspergillus or Rhizopus.

Nasal cultures grow coagulase-negative staphylococci. He undergoes endoscopic evaluation of the sinuses, and biopsy reveals Rhizopus. He is started on Amphotericin B and undergoes surgical debridement, including maxillary enterostomy, ethmoidectomy, and sphenoidotomy.

He continues antifungal therapy for three months. After completing this course, the cranial nerve palsies resolve; however, his visual acuity remains impaired. He is still on insulin therapy and reports better glycemic control.

Mucormycosis is fungal infection that typically affects immunocompromised patients.³ It is frequently cited in patients with diabetes with poor glycemic control. Other predisposing conditions include hematologic malignancies, neutropenia, trauma, iron overload, and use of immunosuppressants or illicit intravenous drugs.

Table 1: Pathology of the Eye

<i>Location</i>	<i>Pathology</i>
Media (cornea, lens, and vitreous humor)	Cataracts, glaucoma, corneal edema from hyperglycemia, endophthalmitis
Retina	Vascular occlusions, detachment, ischemia, macular degeneration
Optic nerve	Optic neuritis, papilledema, mass lesions that compress the optic tract
Brain (occipital lobe)	Stroke, masses, or hemorrhage

Typical sites of infection are the sinuses, lungs, and skin. Vascular invasion, tissue necrosis, and septic thrombosis are hallmarks of the infection. Dissemination as well as direct extension to the orbits and brain can occur. The mortality rate of mucormycosis in diabetic patients has been cited as high as 44%.

Evidence of infection of sinuses and orbital soft tissues may be seen on CT. Nasal swab and culture is insufficient to rule out mucormycosis. Diagnosis requires histopathological evidence of fungal tissue invasion.

Given its invasive nature and high mortality, diabetic patients with suspicious symptoms should undergo imaging and endoscopy to evaluate possible mucormycosis. Treatment includes a prolonged course of antifungal therapy and often surgical resection of infected necrotic tissue. Aggressive glycemic control is also imperative. Delay in diagnosis and treatment may result in tissue necrosis, permanent ophthalmologic deficits, and even death.

Key Points

1. Vision loss has a broad differential. History and physical exam can help localize the pathology.
2. Orbital apex syndrome arises from pathology in the orbital apex that contains the important neurovascular structures of the eye.
3. Fungal infections can spread from sinuses to the orbit leading to neurologic impairment, septic thrombosis, and tissue necrosis.

References

1. Leveque T. Approach to the adult with acute persistent visual loss. *Uptodate* 28 Nov 2011 (Web 28 April 2012).
2. Yeh S, Foroozan R. Orbital apex syndrome. *Curr Opin Ophthalmol* 2004; 15(6):490-8.
3. Petrikos G, Skiada A, Lortholary O, Roilides E, Walsh TJ, Kontoyiannis DP. Epidemiology and clinical manifestations of mucormycosis. *Clin Infect Dis* 2012; 54(1):S23-S34. **SGIM**