Orbital Apex Mucopyoceles Causing Sudden Blindness Despite Timely Maximal Surgical Treatment: Case Report and Review of the Literature

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Abstract

Mucoceles form by accumulation of mucus within a mucoperiosteum-lined cystic structure leading to bone remodeling and erosion. Mucoceles can become acutely infected, forming mucopyoceles that lead to significant intraorbital and intracranial complications. We report a case of a patient with sphenoid and posterior ethmoid mucopyoceles presenting with sudden blindness and remained blind despite timely maximal medical and surgical treatment.

Key words: blindness, mucocele, sphenoid and ethmoid mucopyoceles

Case Report

The patient is a 32-year-old man who underwent extensive sinus surgery with a bicoronal approach and osteoplastic flap for allergic fungal sinusitis more than 12 years ago, and presents with sudden blindness in the left eye. He first noted URI symptoms and subsequently pain behind the left eye for one week. Two days after completion of one week course of antibiotics and steroids, he presented to the ED with sudden onset blindness in the left eye. Ophthalmologic exam showed no light perception and loss of papillary reflex while extraocular movements, intraocular pressure, and slit lamp exam were grossly normal.

High resolution CT of the orbits showed a 3cm left posterior ethmoid and a 2cm left sphenoid sinus hypodense lesion causing mass effect on the optic nerve and 4mm proptosis of the globe. On MRI, the contents of the masses showed no enhancement and T1 and T2 signals were consistent with mucoceles. The patient was immediately taken to the operating room for endoscopic marsupialization of the mucoceles. Upon entering the masses, a large amount of purulent material projected out under high pressure Fig 2.

Post operatively, patient was continued on appropriate IV antibiotics and steroids and no improvement of the patient’s vision was seen in 36 hours. Decision was then made to perform a 360 degree decompression of the optic nerve via an open intradural approach.

Patient remained blind after optic nerve decompression and his hospital course was complicated by a CSF leak, which was managed conservatively with a lumbar drain. The patient was discharged in stable condition on post-op day 21 and the patient remained blind.

Discussion

Mucoceles are benign, expansile lesions of the paranasal sinuses that form over years from continued mucus production within the obstructed sinus. Mucoceles occur most commonly in the frontal, frontoethmoidal, and ethmoidal regions [1-3]. The time interval between inciting trauma and clinical presentation varies depending on the extent of mucosal injury. Mucoceles develop on average 5.3 years after functional endoscopic sinus surgery, 17.7 years after maxillofacial trauma with no history of surgery, and 18.1 years after open sinus surgery [3].

The reported incidence of intraorbital and intracranial complications secondary to mucoceles is 35-47% [4, 9-9]. Sphenoid sinus mucoceles and Onodi cell mucoceles have been reported as potential causes for blindness [8-9]. Two primary mechanisms have been described in the literature underlying vision loss in patients with spheno-ethmoidal mucoceles. The first theory hypothesizes a direct compression ischemia by the expansive mucocele causing compressive neuropathy, whereas the second theory involves ischemic infarction of the optic nerve secondary to thrombophlebitis or vasculitis [10-11]. Mucoceles can thus present as slow vision deterioration over time or sudden blindness secondary to a rapidly expanding mucocele or a vascular event of the optic nerve.

Severity and rapidity of vision loss and time interval between symptom onset and surgery tend to determine vision outcome [4, 6, 9, 10, 12-14]. Rapid onset of complete vision loss tends to have the worst prognosis where slow progressive or partial losses do better. Studies have found that patients presenting with blindness tend not to improve after endoscopic marsupialization [4, 10, 12, 19].

Conclusion

We present a young patient with a history of extensive allergic fungal rhinosinusitis that required open and endoscopic sinus surgery 13 years ago, lost to follow-up and subsequently presented with mucopyoceles after a week of URI symptoms that resulted in permanent blindness despite maximal emergent endoscopic marsupialization followed by optic nerve decompression through an open approach.

This case highlights a few key concepts in managing patients with a history of sinus surgery.

• This case stresses the importance of long-term follow-up after sinus surgery as mucopyoceles can form many years after the initial surgery.

• Clinicians should be aware of the possibility of a mucocele becoming secondarily infected in patients with URI symptoms, forming mucopyoceles that can rapidly expand leading to devastating intraorbital and/or intracranial complications.

• Clinicians should be aware that mucoceles arising from the sphenoid and posterior ethmoidal are more likely to result in orbital apex syndrome and permanent blindness.

• Orbital complication can present as slow vision deterioration over time or sudden blindness.

• Sudden complete blindness is associated with poor outcome despite maximal medical and surgical treatment which is important in counseling patients on presentation.

References