Isolated mucocele is very rare in sphenoid sinus. It presents with varied symptoms in the form of vision loss, proptosis, and headache. We report a case of isolated sphenoidal mucocele that presented with vision loss. Although the vision loss was not sudden, immediate evacuation of pus and marsupialization was done on an emergency basis based on the clinical and radiological findings rather than relying on the history of insidious vision loss. The patient’s vision improved immediately after surgery. This underscores the importance of emergency drainage of this condition.

**Keywords:**
endoscopic approach, mucocele, sphenoid sinus, vision loss

**Introduction**

A mucocele is a mucus-containing cavity lined by the epithelium conforming to the dimensions of the paranasal sinus and which has a propensity for local expansile growth [1]. Mucoceles are commonly located in frontoethmoidal regions [2] but they can also occur scarcely in maxillary and sphenoid sinuses. Because of their close anatomical proximity to the optic nerve, sphenoid mucoceles tend to present with ophthalmological complaints primarily. This condition mimics an intracranial tumor or cavernous sinus lesion because of its clinical features [1]. This case report describes an isolated sphenoidal mucopyocele with vision loss.

**Case report**

This case was studied in a tertiary care hospital in South India in the month of May 2012. A 52-year-old woman presented to our outpatient department with progressive loss of vision in the right eye for the past 1 month. The patient was apparently asymptomatic 6 months before this when she developed right-sided retro-orbital pain, which was dull and progressive in nature and relieved by analgesics. It was for the past 1 month that she developed worsening loss of vision. She was not known to be suffering from diabetes mellitus, hypertension, or any comorbid conditions.

Clinical examination at the time of admission revealed that her visual acuity was reduced to perception of light in the right eye. The optic disc and ocular motility were normal. Anterior rhinoscopy was unremarkable. A diagnostic nasal endoscopy was performed on her, which showed a mucosa-covered bulge in the left sphenoid recess (Fig. 1); the right sphenoid recess was free. Left middle turbinate was bulky with high septal deviation to the left. Bilateral middle meatus was free. There was no evidence of polyp or discharge.

Computed tomography (CT) scan axial section (Fig. 2) shows expansion of the sphenoid sinus filled with enhancing soft-tissue density, with circumferential thinning of the sinus walls, with dehiscence in the right optic canal. MRI showed a lobulated mucosal thickening showing variable signal intensity with a mild expansion of sphenoid sinus, with minimal fluid surrounding right optic nerve.

Options were discussed with the patient, and she agreed for endoscopic management of the disease. Endoscopic drainage was performed on an emergency basis. The disease was approached through the left side as sphenoid sinus findings were on the left side, although the patient had complaints on the right side. Through the midline approach, the mucosa-covered bulge over the sphenoid recess was opened using microdebrider, and purulent discharge was drained from the sphenoid sinus (Fig. 3). Sphenoidal ostium was widened (Fig. 4). The intersphenoidal septum was lateralized to the right which was removed and the entire sphenoidal sac was marsupialized. The right optic nerve canal was dehiscent.

The specimen was sent for Gram staining and KOH mount. There were few pus cells and no bacteria in the...
Gram stain. KOH mount did not show any fungal elements. Cultures did not show any organisms.

Postoperatively, she had significant visual improvement the same day, from perception of light to finger counting at a 1-foot distance. At discharge, her vision improved and the optic disc and ocular motility were normal.

**Discussion**

As the orbit is in proximity to sphenoid and ethmoid sinuses, pathology in these can quickly spread to the orbit. In addition to its proximity, the intermediate bony wall is thin and porous because of many foramina in it. The thickness has been found to be around 300 μm, especially near the optic canal area [3]. Dehiscence of the canal has been shown in previous studies to be around 12% [3]. The optic nerve is not surrounded by any soft tissues but only by its sheath and thin bony covering. This makes it susceptible to compression by an expanding mucocele [4].

Paranasal sinus mucoceles predominantly occur in the frontoethmoidal region (64%), whereas the frequency in sphenoid sinus has been shown to be around 8.4% [5]. Primary sphenoid mucoceles are uncommon in isolation and usually occur with ethmoidal pathology or nasal polyposis [6]. Although rare, sphenoid mucoceles receive considerable attention from otolaryngologists, neurologists, and ophthalmologists, as they may present with nasal, visual, or neurological symptoms because of its close proximity to cranial nerves, pituitary fossa, and cavernous sinuses.

Headache, facial pain, anosmia, proptosis, ophthalmoplegia, and vision loss are the most common symptoms reported. Visual symptoms in patients with sphenoid sinus mucoceles include diplopia, ocular muscle paresis, exophthalmos, and complete visual loss.
The main indication of imaging is vision loss [1], while others being meningitis and suspected intracranial extension. CT scan of mucocele usually shows homogeneous expansive lesion with thinning/erosion of the bone. It is also helpful in determining the anatomical variations and important landmarks for surgery. Axial views are required optimally to show the optic nerve and carotid artery. Contrast enhanced computed tomography (CECT) shows ring enhancement in the case of a pyocele. However, in our patient, although ring enhancement was not present, purulent secretions was released on opening the sinus wall. This may be because of the early presentation of the patient to the hospital before encapsulation of the infective process had occurred, as encapsulation is manifested in the CT scan as ring enhancement usually.

Many possible mechanisms have been suggested for the cause of visual dysfunctions. The mucocele may compress the optic canal leading to loss of vision. In addition to direct pressure, ischemia or venous congestion around the optic nerve occurs [7]. Inflammation due to infection of the mucocele can spread to the nerve through zones of bony erosion or dehiscence [8]. This means that the vision loss may be due to a local inflammatory response that responds to steroid therapy. However, even with response to steroids, further diagnosis and immediate surgical drainage are imperative. In our patient, there was a bony dehiscence of the optic canal and also the mucocele was infected. Steroids were not used in our patient, but prompt surgical treatment was done in our case. The prognosis for the visual loss is poor if there is a sudden loss or if there is no light perception preoperatively. In our case, the patient had light perception at presentation and the vision loss was not sudden.

The differential diagnosis includes fungal disease, cholesterol granuloma, odontogenic cyst (maxilla), and neoplasia (either benign or malignant). Mucoceles are treated surgically. The location and expansion of the lesion determine appropriate surgical procedure [9]. The goal of surgery is to open the mucoceles widely to allow for drainage (marsupialization). Scarring must be prevented to avoid recurrence. An endoscopic approach is ideal in those mucoceles that can be accessed and widely marsupialized.

The sphenoid can be opened from the posterior ethmoid or by traversing the basal lamella of the middle turbinate or medial to the septum through the natural ostium. As mucociliary clearance occurs into the sphenethmoidal recess, it is safer to enlarge the natural ostium of the sinus rather than use the posterior ethmoid approach. That is why we used the midline approach in this case. Great care must be exercised in the sphenoid sinus, which is variable in anatomy and shape and intimately related to the carotid artery and optic nerve. In our case, the anomalies were that (a) the intersphenoidal septum was laterialized to the right and (b) the right optic canal was dehiscent.

This patient had insidious vision loss that had progressed faster during the past 2 weeks. At the time of presentation, the visual acuity on the right eye was only perception of light. In addition to direct pressure, ischemia or venous congestion around the optic nerve causes rapid deterioration of vision [7]. Therefore, emergency decompression of the sphenoid sinus takes paramount importance for recovery of vision. Moreover, this patient had a dehiscent right optic canal compounding the problem. The patient's vision improved as the surgical decompression was done on an emergency basis. Rather than planning the management based on the history of insidious vision loss, management was planned based on clinical and radiological findings, which resulted in recovery of vision.

Conclusion
Sphenoid sinus mucocele has a varied clinical presentation. In patients presenting with visual disturbances and proptosis, the possibility of sphenoid sinus mucoceles must be kept in mind. CT scan can provide a reliable clue to diagnosis. Prompt treatment can halt the progress of the disease.

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