Spheniodal mucocele causing bilateral optic neuropathy and ophthalmoplegia

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Sphenoid sinus mucocele comprises only 2% of all paranasal sinus mucoceles. In literature, there is a case report on sphenoidal mucocele causing bilateral optic neuropathy, with unilateral partial recovery and cranial nerve palsy, but we did not come across any literature with bilateral optic neuropathy and ophthalmoplegia together caused by spheno-ethmoidal mucocele. We present such a rare case of spheno-ethmoidal mucocele causing bilateral optic neuropathy and unilateral sixth nerve palsy who had postsurgery, unilateral good vision recovery, and complete resolution of sixth nerve palsy.

Key words: Cranial nerve palsy, ophthalmoplegia, sphenoidal mucocele, trans-sphenoidal surgery

Sphenoidal mucoceles occur rarely and have an incidence of 1% of all paranasal sinus mucoceles,[1,2] Anterior clinoid mucoceles causing optic neuropathy[3] and cranial nerve palsies[4] are reported in literature, but sphenoid mucoceles causing both optic neuropathy and ophthalmoplegia are very rare. Visual loss from sphenoid sinus mucoceles is usually associated with poor prognosis, if surgical treatment is delayed more than 7–10 days. The etiology of the mucocele is not clear. It is said to be a retention cyst caused by obstruction of normal drainage from the sinus. The presence of a chronic low-grade infection may be a causative factor. As the mucocele expands, surrounding bony structures undergo osteoclastic absorption. If the necrosis of bone continues, pressure on dura and adjacent neural elements may produce compression signs.

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Case Report

A 17-year-old boy presented with severe headache of 2 weeks duration and sudden decrease in vision in both eyes for 10 days, right eye followed by left within 5 days. On examination, right eye had no perception of light and left eye had of counting fingers close to face vision. Extraocular movements revealed abduction limitation in right eye. Pupil was fixed dilated and nonreacting to light in right eye and left eye pupil was normal in size, but had ill-sustained reaction to light. Anterior segment examination and intraocular pressure were normal. Fundus examination revealed subtle temporal pallor of optic disc in both eyes [Fig. 1]. He had undergone computed tomography of the brain and orbit which revealed a lesion in the spheno-ethmoid sinus which was slightly hyperdense. Bone windows showed erosion of the roof of ethmoidal sinus [Fig. 2]. MRI of the brain revealed a lesion in the spheno-ethmoid sinuses and located between the two internal carotid arteries which was hyperintense on T1W images and iso- to hyperintense on T2W images [Fig. 3].

Patient was immediately referred to neurosurgeon for surgical intervention, but there was a delay of 2 days by the patient. When he presented to the neurosurgeon, he had developed right oculomotor nerve paresis in addition to the sixth nerve paresis. The patient underwent an emergency decompression—right trans-ethmo-sphenoidal approach and total evacuation of the mucocele under general anesthesia. Histopathology was suggestive of mucocele lined with pseudo-stratified columnar epithelium.

One week after surgery, the right eye condition was same, but left eye showed vision improvement to counting fingers at 4 m; pupil was sluggishly reacting to light. One month after surgery the right eye showed only abducent nerve paresis, while oculomotor nerve paresis was completely resolved; pupil showed relative afferent pupillary defect. There was no PL in the right eye. Left eye showed improvement in vision of 6/6, N6, and pupillary reaction was normal. On 2 months follow-up, both eyes had full range of ocular movements [Fig. 4]. Right eye had no PL but the left eye regained vision to 6/6 N6. The optic disc showed temporal pallor more in right than left. Postoperative CT scan of the brain showed complete evacuation of the lesion [Fig. 5].

Discussion

Mucoceles are cyst-like lesions lined with respiratory epithelium that most commonly produce bone destruction within the paranasal sinuses. Approximately two-thirds of all mucoceles involve the frontal sinuses, and the majority of the remainder involve the ethmoidal sinus. Sphenoidal mucoceles occur rarely and have an incidence of 1% of all

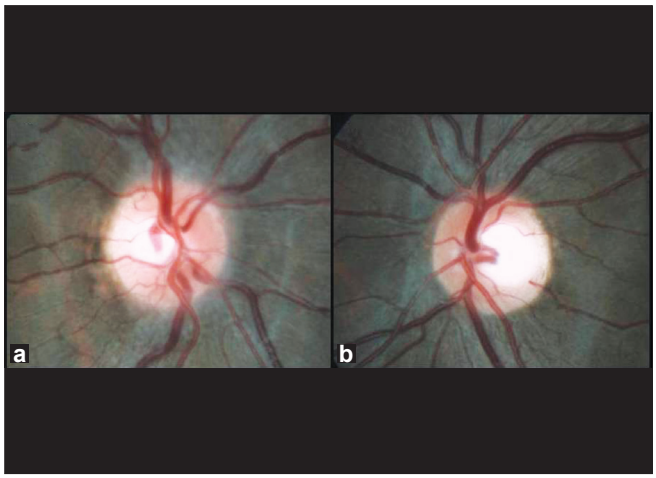

Figure 1: Picture of the optic nerve head right (a) and left (b)

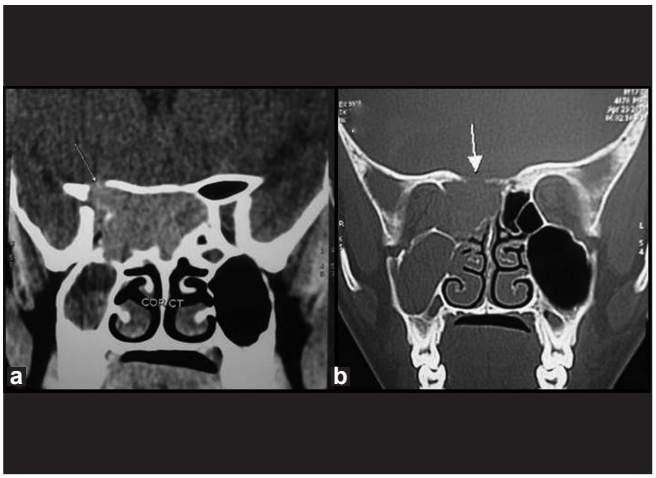

Figure 2: Preoperative CT scan of the brain and paranasal sinuses—coronal section (a) with bone cuts (b) shows lesion in the spheno-ethmoid sinus which is slightly hyperdense and erosion of bone in the roof of the ethmoidal sinus

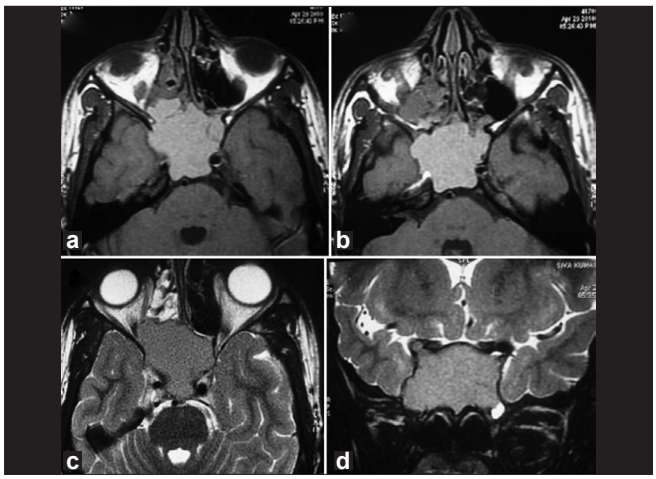

Figure 3: Preoperative MRI of the brain T1W (a and b): axial sections showing hyperintense lesion in the spheno-ethmoid sinuses and located between the two internal carotid arteries. T2W (c and d): axial and coronal sections showing iso- to hyperintense lesion

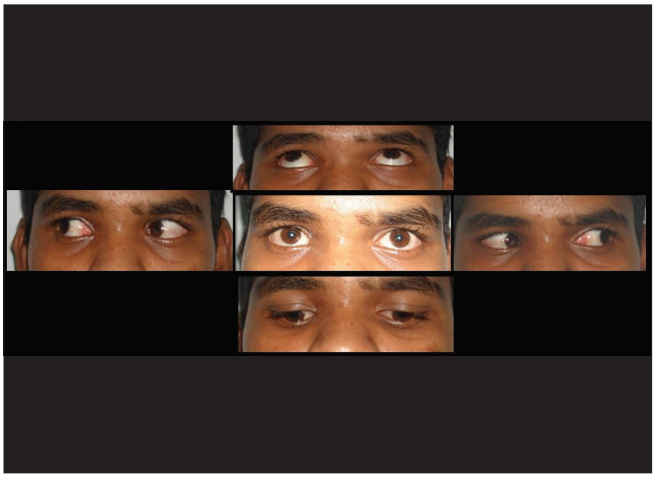

Figure 4: Postoperative clinical picture of the patient showing full range of ocular movements

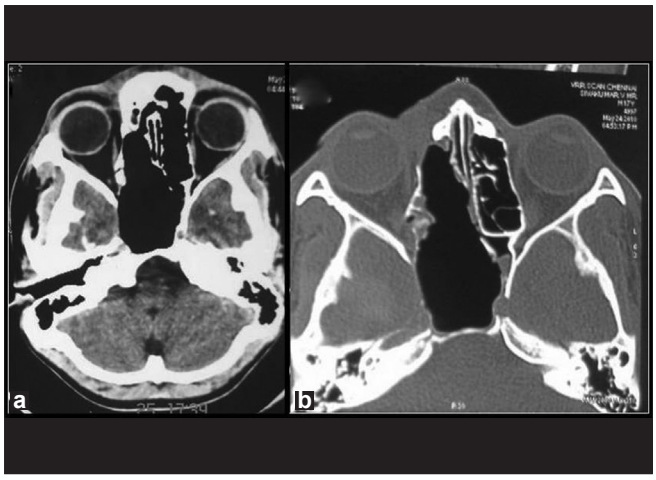

Figure 5: Postoperative CT scan of the brain and paranasal sinuses—axial section (a) with bone window (b) showing complete excision of the lesion

paranasal sinus mucoceles.[1,2] Anterior clinoid mucoceles causing optic neuropathy[3] and cranial nerve palsies[4] are reported in the literature, but sphenoid mucoceles causing both optic neuropathy and ophthalmoplegia are very rare and we could not find any similar report in the literature. The most common is headache (frontal or retroorbital; 70% of patient) and second is visual disturbance (65% of patients). Other visual symptoms include diplopia, ocular muscle paresis, exophthalmus, and complete visual loss. Rarely can they present with multiple cranial nerve palsies. The slow and silent expansion of a mucocele may be unsuspected until bone is eroded and it impinges on adjacent structures. Proposed theories for development of mucocele include chronic infection, allergic sinonasal disease, trauma, previous surgery, and in some cases the cause remains uncertain. Lund and Milroy[5] proposed that the obstruction to sinus outflow in combination with superimposed infection caused the release of cytokines from lymphocytes and monocytes. The cytokine release would stimulate fibroblasts to secrete prostoglandins

and collagenases, which in turn could stimulate bone resorption leading to expansion of the mucocele.

A large mucocele produces a classic radiographic appearance of an enlarged distorted sinus with a bony defect representing a breakthrough into the adjacent structures. The CT features include a homogeneous iso- to hyperdense expansile mass, which completely fills the sinus cavity and surrounding structures. On contrast administration, there is no increase in the density of mucocele due to its avascular mucoid content. Differential diagnosis includes hypophyseal, tumors, craniopharyngioma, meningioma, chordoma, dysgerminoma, cholesteatoma, and tumors arising from nasopharynx, sinuses, and base of the skull.

Sphenoid surgery was first introduced by Schloffer in 1885. Surgical treatment options consists of trans-nasal endoscopic, trans-ethmoidal, trans-antral, trans-palatal, and trans-septal. The degree of improvement in visual acuity after an operation depends on visual acuity before the operation, the mode of development of the mucocele, and the time from onset of the disease until surgery. Casteels[6] *et al*. have described that if surgery is delayed for more than 6–10 days after the start of visual loss and if there is optic atrophy, the visual prognosis seems to be poor.[7] The above presented case had symptoms in the right eye for 15 days and in the left eye for 10 days. Although there was no optic atrophy in the right eye, the vision did not improve. Poor recovery of the right eye could be due to comparative delay in presentation and intervention.

Conclusion

Mucoceles, though rare, have to be considered in the differential of visual deterioration. Computerized tomography delineates the characteristic features of a mucocele. Early surgery of these benign lesions will lead to complete recovery even in patients with poor vision.

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