Isolated Sphenoid Sinus Mucocele Presenting as Third Nerve Palsy

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A sphenoid mucocele often presents late due to its deep seated anatomical site. And it has varied presentation due to its loose relationship to the cavernous sinus and the base of the skull. We describe a case of large sphenoid sinus mucocele. A middle aged old man suddenly developed third cranial nerve palsy. Brain imaging study revealed an isolated sphenoid sinus mucocele, compressing right cavernous sinus. Endoscopic marsupialization of the mucocele via transnasal approach led to complete resolution of the third cranial nerve palsy. Involvement of the third cranial nerve in isolated mucocele is rare but important neurosurgical implications which must be excluded. In addition, proper and timely treatment must be performed to avoid permanent neurologic deficit.

KEY WORDS: Sphenoid sinus mucocele · Third nerve palsy · Transnasal approach.

INTRODUCTION

Mucoceles are benign, encapsulated, expansile, locally invasive masses within a paraanalar sinus filled with mucus and lined by epithelium. Mucoceles occur in all of the paraanalar sinuses but arise most commonly in the frontal sinus. Isolated sphenoid sinus mucocele is rare in paraanalar sinus mucoceles. The mechanism in the development of mucocele is not clear, but postulated as the obstruction of the sinus initiates its development. There are other theories about its etiology, including cystic degeneration of epithelial mucus glands, or cystic degeneration of an inflammatory polyp.

The presenting symptoms are primarily caused by compression on the adjacent structures. Mucoceles expand gradually and result in resorption and sometimes erosion of the bony walls of the sinus. Because of its locally invasive nature, sphenoid sinus mucocele might mimic a more serious condition or be associated with a tumor or tumor-like conditions.

We present our successful management of rare case of a huge isolated sphenoid sinus mucocele compressing third cranial nerve using minimally invasive endoscopic surgery.

CASE REPORT

A 49-year-old male presented a three-day history of drooping of his right eyelid and diplopia (Fig. 1A). He also gave a one-month history of headache. Examination revealed isolated right-sided oculomotor nerve palsy with sparing of the ipsilateral pupil. Physical examination revealed moderate reduction of elevation, depression, and adduction in the right eye.

An urgent computed tomography (CT) scan of the head and orbit was performed before and after intravenous contrast agent administration (Fig. 1B, C). The CT scan revealed a non-enhancing well-defined, lobulated space-occupying lesion expanding the right sphenoid sinus, measured as 37 × 39 × 37 mm. The lesion was causing marked thinning of the posterolateral bony wall of the sphenoid sinus with areas of bony erosion. Magnetic resonance image (MRI) was performed subsequently (Fig. 2). The lesion was hyperintense on both T1- and T2-weighted images. There was no enhancement with gadolinium DTPA. It was causing a mass effect on the adjacent structures, especially on right cavernous sinus. The imaging features suggested that the lesion was predominantly cystic in nature.

The mucocele was decompressed by transnasal sphenoidotomy and marsupialization utilizing endoscopy. Endonasal sphenoidotomy and drainage of the sinus content produced...
mucus-like fluid with some internal granularity. Bone overlying the middle cranial fossa and periorbita was found to be dehiscent but there was no CSF leakage evident. Microscopic sections of the mucocele showed a cyst lined by columnar epithelium, with acute and chronic inflammation. Bacterial cultures showed no growth.

His ptosis was markedly reduced the following day. The headache was also improved. One week after surgery, his third nerve palsy had completely resolved (Fig. 3). A postoperative CT scan demonstrated markedly decreased partially air-filled bony cavity replacing the cystic lesion. There was no recurrence and neurologically intact without any visual symptoms at the time of his 12 months follow-up examination.

**DISCUSSION**

Mucocles are the most common lesions causing expansion of the paranasal sinuses, lined with respiratory epithelium. More than half of these lesions are located in the frontal sinuses. However, isolated sphenoid sinus mucocles are rare, representing 1-2% of all cases of paranasal sinus mucocles. They usually start unilaterally, but by the time of presentation, the entire sphenoid sinus complex may be opacified and expanded with thinning of its bony walls.

Mucocele of the sphenoid sinus act as benign neoplasm and can result in bony erosion extending from the sinus into the cranium and the orbit. Some tumors and tumor-like conditions, such as carcinoma, fibrous dysplasia, osteoma and ossifying fibroma, might be associated with a mucocele of the sphenoid sinus\(^9\). Characteristic radiographic findings can suggest the preoperative diagnosis. The bony changes are best demonstrated by CT scans. The secretions in the mucocele can be serous, proteinaceous, thick and pasty or dry and desiccated. Depending on their biochemical constituents,
mucoceles can be hypo-, iso-, or hyperintense or signal void on the MR images. They do not show contrast enhancement centrally.

The clinical manifestations are variable and related to the direction of extension towards neighboring structures. The most common clinical symptom is headache which is present in 70-80% of cases. This is typically retro-orbital. Visual disturbance is the second commonest group of symptoms making surgery necessary. The optic nerve is the most frequently involved cranial nerve with reduction in visual acuity. Palsies of eye movement with diplopia occur in 30-50% of cases. The oculomotor nerve is affected more frequently than the trochlear and abducens nerve. Oculomotor nerve involvement accounted for 70% of oculomotor palsies.

As a result of the peripheral location of the pupillary fibers in the oculomotor nerve, compressive lesions such as aneurysms or tumors that produce third nerve palsy usually produce ocular motility disturbance and ptosis. In contrast, isolated oculomotor nerve palsy with papillary sparing is characteristic of diabetic ophthalmoplegia. Pupillary sparing is believed to result from microvascular ischemia of the central portion of the nerve with sparing of the more peripherally placed parasympathetic fibers.

The treatment of sphenoid mucocele is surgical. Traditionally, the management of sphenoid mucoceles was complete removal via transfrontal or transcranial approach. However, transnasal sphenoidotomy has largely replaced the conventional open method with excellent results. The recommended management of a mucocele of the sphenoid sinus is endonasal sphenoidotomy with sufficient removal of the anterior and inferior wall of the sinus along with drainage of the mucocele. The present study supports the treatment of isolated sphenoid sinus disease with visual disturbances by endoscopic sinus surgery, especially in cranial nerve palsy.

Proper and timely treatment usually results in rapid regression of the ophthalmic manifestation, but vision seldom returns to normal in the case of visual dysfunction. The cause of visual dysfunction may be optic nerve compression with vascular compromise or extension of the sinus infection and inflammation to the optic nerve. A delay in surgery of more than seven to ten days after the onset of visual dysfunction is often associated with poor visual prognosis. Thus, the appropriate diagnosis must be made early.

CONCLUSION

We emphasize an awareness of sphenoid sinus mucocele as a cause of isolated oculomotor nerve palsy. In addition, surgical approach must be attempted as soon as possible.

References