INTRODUCTION

Mucoceles are benign, encapsulated cystic lesions filled with mucus that may be locally expansile or lead to the destruction of the bony structure surrounding it. The majority of paranasal sinus mucoceles commonly occur in the frontal sinus (60%) and ethmoid sinus (30%), while these are rarely found in the sphenoid sinus (1%-3%). However, the diagnostic rates of sphenoid mucoceles have been gradually increasing due to the development of imaging technologies. Even though sphenoid mucoceles have been reported to have a higher incidence of intracranial or orbital complications than those of other sinuses, these can be treated safely and easily owing to the well-established endoscopic sinus surgery. Moreover, it is vital to perform the surgery promptly with early diagnosis for quick relief of mucocele related symptoms.

In this article, we report a case of sphenoid mucocele in a Korean woman who had visited different clinics to find out the reason of her visual disturbance prior to undergoing endoscopic sinus surgery in the author’s hospital.

CASE

A 62-year-old female with a history of a one-year, slowly progressive disturbance of vision in the right eye, visited the department of ophthalmology in the author’s hospital. She claimed that she had visited several local eye clinics all over the city over a period of eight months to treat the symptom. However, the patient, who was only diagnosed with unilateral optic atrophy, could not know the reason for this finding.

On ophthalmologic examination, a relative afferent pupillary defect and light reflex tests were normal, and she did not complain of ocular pain, diplopia, and epiphora. Her best-corrected visual acuity (BCVA) was 0.05 in the right eye and 0.8 in the left eye. The result of the color vision test was 3/12 in the right eye and 12/12 in the left eye. Fundus examination of the right eye showed mild optic disc atrophy and a central scotoma was also identified at the same side (Fig. 1A). Retinal nerve fiber loss was observed in 2 quadrants in the right eye with optical coherence tomography (Cirrus OCT; Carl Zeiss Meditec, Jena, Germany) (Fig. 2A). Orbital MRI was immediately performed to look for the possible anatomical problem for ophthalmologic abnormality, and a huge mass lesion was...

ABSTRACT

A mucocele of the sphenoid sinus is relatively rare, and it has nonspecific clinical features. Hence, it is hard to diagnose it with the history of the illness alone. However, if the sphenoid mucocele is not detected and treated in early stage, its sequelae could be very serious or permanent, such as intracranial complication or blindness. Recently, the authors experienced a case of a sphenoid mucocele with unilateral visual disturbance for a period of one year. The patient underwent endoscopic sinus surgery-marsupialization successfully, but remnant visual disturbance as a sequelae did not fully recover. Thus high index of suspicion, prompt diagnosis using imaging studies, and urgent surgical intervention are crucial for promising of complete recovery from the disease.

KEY WORDS: Sphenoid Sinus · Mucocele · Optic Neuropathy · Marsupialization.

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identified in the right sphenoid sinus.

Complementary imaging and ENT examination were requested. There was no history of previous trauma, sinus surgery, or chronic rhinosinusitis. She had no specific ENT complaints, and laboratory studies were within normal limits. Nasal endoscopic examination did not find any bulging lesion, discharge, or anatomical abnormality. A contrast CT scan showed a huge expansion of the sphenoid sinus, suggestive of a mucocele. We could clearly identify the shape and size of the lesion including a sporadic skull
base defect. It dilated the bony wall of sphenoid sinus like a balloon and pressed the right optic nerve (Fig. 3). MRI disclosed a lobulated mass lesion, measuring 36-50 mm, in the sphenoid sinus extending into ipsilateral posterior ethmoid cells (Fig. 4). Because the internal contents of the lesion were not enhanced by contrast of the CT scan and revealed high signal intensity on T1-weighted images and low signal intensity on T2-weighted images in the MRI, the mass was determined to be consistent with sphenoid mucocele with a high protein content.

Soon after, the patient underwent endoscopic sinus surgery under local anesthesia at our same day surgery center. Using the transethmoidal sphenoidotomy technique, we extensively removed the anterior bony face of the mucocele. Through the wide marsupialization site, high amounts of mud color, thick viscous and purulent discharge were evacuated. Serial irrigation with warm normal saline solution was also performed. In order to avoid damaging vital organs, such as the cavernous sinus, optic nerve, internal carotid artery, and brain cortex, some mucosal lining of the mucocele was left in situ.

The pathology was composed of chronic inflammation of stratified respiratory epithelium and cholesterol granuloma. There was no atypia or pleomorphism in the tissue.

Two or three days after the surgery, her visual disturbance started to recover subjectively, and there were no surgery-related neurologic deficits. Two months after surgery, the symptom of visual acuity recovered fifty percent subjectively. However, since then, the patient has not yet reported further recovery.

At present, four months after the surgery, her BCVA is 0.32 in right eye and Goldmann visual field is slightly improved (Fig. 1B). However, central scotoma is still persistent and OCT reveals the decreased retinal nerve fiber layer thickness in the right eye (Fig. 2B).

As appeared by nasal endoscopy and postoperative CT scan, there is no evidence of recurrence, and the sphenoid sinus has remained well opened (Fig. 5). The patient is under close observation about remnant visual problems for 4 months after the surgery.

**DISCUSSION**

Mucoceles, demarcated by sinus mucosal lining, are expansile and cystic lesions filled with mucus.\(^1\)

The pathogenesis of mucoceles is still unknown, but several hypotheses have been proposed. The primary hypothesis postulates that a mucocele is induced by an inflammatory reaction, such as allergic rhinitis, chronic sinusitis, or nonspecific inflammation leading to submucosal edema or natural ostium’s obstruction. The secondary hypothesis assumes that the disease is caused by craniofacial trauma, sinus surgery, or radiation therapy resulting in cystic lesions with fluid retention.\(^2\)\(^5\)\(^6\) In our case, the patient could be considered to have primary mucocele because of no previous specific history and normal appearance in nasal endoscopy.

The reason why a sphenoid mucocele is extremely rare is that the sphenoid sinus has fewer opportunities to come in contact with pathogens because it is deeply located near the nasopharynx.\(^7\) Through a retrospective study of thirty-one cases of mucoceles, Kim et al. reported that primary mucoceles show the highest frequency in the ethmoid sinus and the lowest frequency in the sphenoid sinus.\(^8\)

In sphenoid sinus mucoceles, there could be no or nonspecific clinical features. However, the most common presenting symptom is a headache, which is most often described as frontal or retro-orbital in nature. The second most common symptom is a visual disturbance, which was noted in six out of fifteen patients by Benninger et al.\(^9\) A visual disturbance could be presented with other complaints including ocular movement dysfunction and
in our case, marsupialization was performed successfully which may be injured during the operation.\footnote{15} A prompt surgical treatment is the best way to avoid permanent visual loss or its sequelae.

Since the first description of a sphenoid sinus mucocele by Berg in 1889,\footnote{11} to date, only about 140 cases have been reported in the English literature. It is likely that sphenoid sinus mucoceles have been under-reported, as diagnosis through a physical examination is difficult due to nonspecific presenting symptoms and an inaccessibility of the sinus. However, recently, the detection rates of sphenoid sinus lesions have been increasing thanks to the development of CT/MR imaging technologies.\footnote{6} The sphenoid sinus lesion in our case was also caught by orbital MRI.

The most important clue in the diagnosis of mucocele is radiologic findings.\footnote{10,12} High-resolution computed tomography (HRCT) can reveal that a mucocele has multiple cystic fractions and sharp boundaries without contrast agent enhancement. Furthermore, it shows that a potential sinus cavity is homogeneously and completely filled with mucus by a mucocele, with expansion and remodeling rather than destruction of the bony sinus wall. When the same CT findings are detected as the ones listed above in a sphenoid sinus, a sphenoid mucocele can be diagnosed without hesitation.\footnote{13} MRI generally shows moderate or low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, and does not enhance the mucocele after administration of contrast media.\footnote{14} However, as our MRI findings, some cases could show opposite MRI features; high signal intensity on T1-weighted images and low signal intensity on T2-weighted images. This may be attributed to the contents of a mucocele; high proteins and thick viscous mucus by infection, over time. As discussed earlier, because this case had a high amount of mucus and had reasonable features in radiologic studies and histologic examinations, the patient was confidently diagnosed as having a mucocele.

Endonasal endoscopic sinus surgery has been advocated as the treatment of choice with lower morbidity and easier accessibility compared to the external approach. Both a marsupialization and an enucleation are available for the treatment of a mucocele via endoscopic sinus surgery. However, recently, endoscopic marsupialization is preferred over an enucleation.\footnote{5,15} The reason is that marsupialization alone is sufficient to achieve drainage and aeration, and the remnant mucosal lining of the mucocele can act as a protector for underlying structures, including the dura, carotid artery, brain cortex or optic nerve, which may be injured during the operation.\footnote{15} Likewise, in our case, marsupialization was performed successfully in a short time while keeping the surrounding structures safely.

Obviously, the major concern of a mucocele treatment is total recovery from the disease. However, in a sphenoid mucocele, the gradual decline of a vision is caused by the mass effect of the expanding mucocele leading to vascular compromise of the optic nerve, and it may terminate in optic atrophy as our case. Thus, complete regression of the visual disturbance depends on how severe the symptom is and on the time from onset of the symptom to surgical marsupialization.\footnote{16} The detection rates of sphenoid mucoceles are clearly increasing along with the development of imaging technologies and the establishment of the proof that a visual disturbance is a common symptom in subjects with sphenoid mucocele. Nonetheless, this case shows that several clinicians did not even suspect the possibility of a sphenoid mucocele. The fact, mucoceles may cause ophthalmic problems without any specific presenting nasal symptoms, is the point of this case. Therefore, careful attention with a good knowledge of this disease by several clinicians including otorhinolaryngologists is crucial for early diagnosis and prompt surgical treatment.

REFERENCES

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