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Case Report

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Primary cholesteatoma of the sphenoidal sinus - surgical treatment with video

Abstract

Nasal sinus primary cholesteatomas are quite uncommon. Only five cases of sphenoid sinus cholesteatoma have been documented in the literature, with frontal sinus involvement being the most common. Our goal is to further scientific knowledge by describing the sixth case of primary cholesteatoma of the sphenoid sinus without extra-sinus extension and demonstrating the disease's surgical treatment in video. We discuss the case of a healthy women in her twenties who complained of a developing headache for years that worsened when combined with nasal symptoms. A bilateral sphenoidotomy was performed, with the whole cholesteatomatous matrix removed (partially preserved in areas with dehiscence). At 3 years, a sustained remission of the illness is noted. Progressively worsening headache may be a manifestation of sphenoid sinus pathology that may require surgical treatment. In this case, a rare disease such as sphenoidal cholesteatoma was noted. Surgical debridement must be carried out to avoid complications.

Keywords: cholesteatoma, sinus surgery, sphenoid sinus, video, case report

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Main points

Chronic headaches, especially those linked with nasal complaints, could be a sign of sinonasal pathology Primary cholesteatomas are uncommon in the nasosinusal cavity. The sphenoid sinus location is much more uncommon, with only five occurrences recorded in the literature. Surgical intervention is necessary. To properly manage the patient, the disease does not need to be completely removed

Introduction

Cholesteatomas, otherwise known as epidermal inclusion cysts, keratomas,[1] keratocyst or epidermoid cysts[2,3] are characterized by an accumulation of keratin debris surrounded by squamous epithelial cells.[1] Cholesteatomas are most commonly detected in the middle ear and mastoid, although they can also occur in the kidney, testicular, skin, breast, central nervous system (CNS), cranial vault, orbit, and mandibles.[2,4] The frontal sinus is the most frequent site of origin for the less than thirty cases of primary (congenital) involving the paranasal sinus that have been reported.[5–7] The ethmoidal, maxilar, and sphenoid sinuses are the next most common places of origin.[1] There have been multiple reports of secondary (acquired) cholesteatomas of the sphenoid sinus caused by direct expansion of mastoid cells.[8] Primary cholesteatomas of the sphenoid sinus are extremely rare, with only five instances reported in the literature.[4,9–12] This case report was written according to the CARE criteria.[13]

Clinical case

We discuss the case of a healthy woman in her twenties who was sent to our unit with a progressive headache that had been growing for years, exacerbated when combined with nasal complaints that was difficult to control with medical treatment. The patient denied having any visual deficits, other nasal issues or any other complaints. She had no known history of traumatic brain injury, temporal fracture or previous head and neck surgery. While an anterior rhinoscopy revealed nothing abnormal, nasal endoscopy revealed sparse non-purulent rhinorrhea of the left spheno-ethmoidal recess.

No medical treatment was prescribed because the patient wasn't in an acute or aggravated phase, with only an occasional headache; instead, a computer tomography (CT scan) and magnetic resonance imaging (MRI) was requested, and acetaminophen was given for pain management in case it developed. It was advised to get reevaluated right away if any new symptoms or alarm signs appeared.

Investigations

The CT scan (Figure 1) showed a complete bilateral homogenous filling of the sphenoid sinuses, suggestive of an expansile lesion with soft-tissue density with multifocal bony areas of dehiscence along the left carotid canal and optic canal, without any signs of continuity with the mastoid cavities or other accompanying problems. MRI findings were similar.

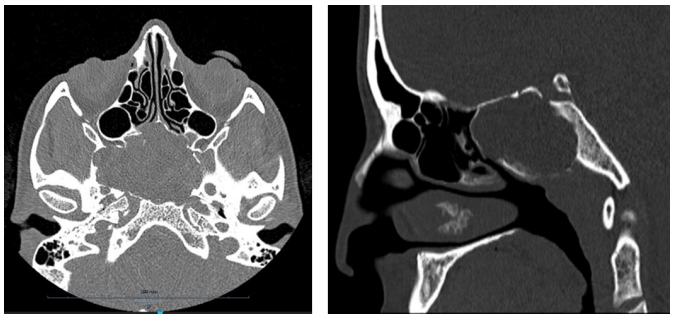

Figure 1 Soft tissue density lesion in the sphenoid sinus.

There is no communication with the middle ear

Obviously, our main diagnosis hypothesis was not a primary sphenoid sinus cholesteatoma, and the most likely diagnosis at this point was a mucocele. Endoscopic nasal surgery was offered to marsupialize the lesion and confirm the diagnosis.

Treatment

Following a period of preoperative tapered-dose prednisone, the patient was taken to the operating room for endoscopic nasal surgery. After nasal decongestion with topical adrenaline (1:1000), we

performed a wide sphenoidotomy with partial posterior septectomy for wide exposure after a bilateral total ethmoidectomy in reverse Trendelenburg position under total intravenous anesthesia with permissive hypotension. A cholesteatoma was diagnosed clinically after an expansile lesion with typical white keratin debris was found in both sphenoid sinuses during surgery (Figure 2).

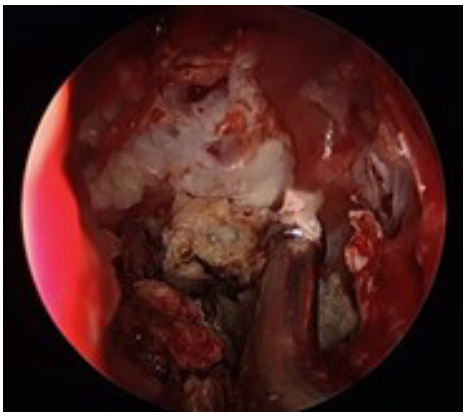

Figure 2 Intraoperative image of the cholesteatoma matrix in the sphenoid sinus

The keratin debris was completely removed, and an attempt was made to remove the entire cholesteatomatous matrix, however it was partially preserved in specific locations with carotid artery or optic nerve dehiscence, as shown in the video (Video 1)**.** There were no complications during the surgery. The pathologic results revealed extensive keratin debris and regions with active moderate chronic inflammation, supporting the surgical diagnosis of cholesteatoma.

Outcome and follow-up

The patient was discharged home the day after surgery and given a topical corticosteroid twice a day. She was directed to perform nasal saline irrigation three times per day. There were no neurological or ophthalmic complications. During the follow-up, the patient underwent successive in-office debridements using rigid endoscopy, followed by washing of residual crusting. Figure 3 and Video 2 show the wide sphenoidotomy one month after surgery.

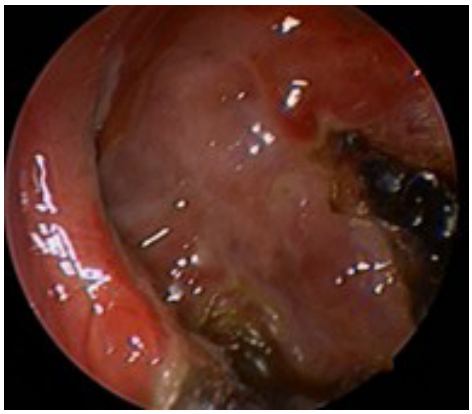

Figure 3 Sphenoid sinus 1 month after surgery

At the one-year follow-up, a single bacterial infection was treated with amoxicillin and clavulanic acid. At her most recent follow-up appointment, which took place three years after her surgery, a nasal endoscopy revealed some moderate inflammation in the left pterygoid recess. This inflammation is being treated with washout and outpatient monitoring. The primary concerns of persistent nasal discomfort and headache were resolved.

Discussion

Until the present date, to the best of our knowledge, there are only five primary cholesteatomas of the sphenoid sinus described in the literature. They all had persistent headaches, just like our patient. Unlike the other examples documented, ours did not show diplopia or accompanying cranial nerves impairments, which may indicate an earlier diagnosis of the disease in our case. Most of them (four) were successfully treated with sphenoidotomy and marsupialization via surgery, without associated complications. Only one case described skull base osteomyelitis, which proved fatal despite aggressive targeted antibiotic treatment and surgical intervention.[4] The carotid artery and optic nerve, two important sphenoid sinus components, showed areas of dehiscence in our instance, but there was no cerebral involvement or deficits of cranial nerves. All the related symptoms were resolved surgically and without any complications.

The abnormal implantation of ectodermal cells of the epidermis during the union of epidermal surfaces is thought to constitute the pathophysiology of primary cholesteatoma. This is believed to happen between the third and fifth weeks of embryonic development while the face is developing.[5,14] This was probably a primary congenital sphenoid cholesteatoma because the patient had a long history of headaches and no previous history that would have suggested a secondary cause.[1]

Its origin and infiltration of neighboring structures determine its symptoms and clinical presentation. It could resemble unilateral nasal mass presentation or chronic rhinosinusitis with headache, rhinorrhea, localized pain, and nasal blockage.[4] Keratin debris in cholesteatoma are normally surrounded by inflammatory tissue with secondary infection which explains the frequent aggravations described. It is also the main mechanism of surrounding bony destruction, apart from mass effect.[15] The pathological detection of a build-up of keratinizing stratified squamous epithelium leads to the final diagnosis. This is linked to inflammation, stromal fibrosis, and keratinous debris—all of which are indicative of benign, non-neoplastic lesions.[15]

The recognizable look of an intraoperative cholesteatoma strongly suggests it. Given how uncommon the diagnosis is in the perinasal region, it may also be suspected in CT or MRI scans, however this would likely be more challenging. Bony erosion is specifically demonstrated in these radiologic exams.[16] An MRI is necessary to differentiate a soft tissue lesion or tumor from sinusitis.[5] The primary differential diagnosis includes malignancy, chronic rhinosinusitis, dermoid cysts, and mucoceles.[1]

Surgery is the method used to treat cholesteatomas. The goal, similarly to middle ear and mastoid surgery, is to remove all the disease and its walls without damaging nearby structures. To stop the lesion from spreading, eroding the surrounding bone, and invading surrounding structures, marsupialization of the lesion is advised if total removal is not feasible.[17]

Nowadays, an endoscopic approach is most preferred to do the resection because improvements in endoscopic sinus surgery procedures can offer enough exposure to remove illness.[5,18] In sinus surgery, stereotactic image guidance is widely used to improve precision and reduce the risk of harm to important intracranial and extracranial structures.[1] This method also yields better cosmetic outcomes as compared with open surgery. Nevertheless, as one example[5] reports, open surgery is still a possibility, particularly in situations that are refractory or if endoscopic surgery provides a difficult approach. The method used in our instance was entirely endoscopic, involving the subtotal excision of the cholesteatoma

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matrix and marsupialization in the crucial structure dehiscent regions. It allowed the resolution of all the chronic symptoms with minimal morbidity. Despite the suggestion of some authors,[19] there was no need for a second look because the follow-up could be completed in the office with direct vision and debridement of the surgical region.

Conclusion

Progressively worsening headaches may indicate sphenoid sinus pathology, necessitating surgical intervention, or, as in this case, an extremely unusual condition such as primary sphenoidal cholesteatoma. Surgical debridement must be performed to avoid complications. Proper postoperative debridement alongside suitable topical treatments are crucial for success in managing this disease.

Institutional review board statement

The study was conducted in accordance with the Declaration of Helsinki and approved by the Ethics Committee of the hospital.

Informed consent statement

Informed consent was obtained from all subjects involved in the study. Written informed consent has been obtained from the patient to publish this paper.

Data availability statement

No new data were created or analyzed in this study. All relevant data are contained within the article.

Conflicts of interest

The authors declare that there are no conflicts of interest

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