


Sphenoid Sinus Cholesteatoma—Complications and Skull Base Osteomyelitis: Case Report and Review of Literature

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ABSTRACT

INTRODUCTION: Cholesteatoma of the paranasal sinuses is uncommon. Its clinical characteristics are an expanding growth of the affected paranasal sinuses consisting of keratinizing squamous epithelium with bony wall destruction. Among involved paranasal sinuses, sphenoid sinus cholesteatoma is the least common.

CASE PRESENTATION: An 82-year-old female diabetic patient presented with subacute onset of fever after experiencing chronic progressive headaches for more than 20 years. Nasal endoscopy found purulent discharge from left sphenoidal recess. Computed tomography (CT) scan of the paranasal sinus showed soft tissue lesions that totally filled the left sphenoid sinus with posterior and inferior wall destruction. There was no evidence of connection to the left mastoid cavity.

MANAGEMENT AND OUTCOME: Left sphenoidotomy was performed. Histopathology revealed cholesteatoma. Two months after surgery, she became worse and CT showed extensive skull base destruction. The patient underwent bilateral sphenoidectomy and craniotomy with surgical debridement of osteomyelitis of the skull base. She received long-term intravenous ertapenam and sitafloxacin for treating drug-resistant *Klebsiella* infection. The osteomyelitis could not be controlled, and she died.

DISCUSSION: Progressive headache can be caused by an uncommon disease such as sphenoid sinus cholesteatoma, which is a surgical condition. Complicating osteomyelitis of the skull base requires extensive debridement surgery and should be anticipated.

KEYWORDS: cholesteatoma, sphenoid, skull base, osteomyelitis, endoscopic sinus surgery

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Introduction

Cholesteatoma, also called keratoma, keratocyst, or epidermoid cyst,^{1,2} is characterized by a mass formed by keratin layers within hyper-keratinized squamous epithelium that replaces normal mucus membrane.³ Cholesteatoma is commonly found in mastoids and middle-ear cavities, but has been described in kidney, testicle, skin, breast, central nervous system (CNS), cranial vault, orbit, and mandible.¹ Less than 30 cases had been reported in paranasal sinus cavity in the English literature.^{4–6} Frontal sinus is the most common paranasal sinus involved, followed by maxillary and ethmoid sinuses.⁶ The first case in a frontal sinus was reported in 1930 by Spencer.⁷ In 1941, the first case in maxillary sinus was reported by Hutcheon.⁸ Ten more cases of maxillary sinus cholesteatoma were reported in the English language after 1980s.⁹ Cholesteatomas originated from sphenoid sinus, excluding those extending from the temporal bone to the sphenoid bone are extremely rare. To date, there are 4 cases reported.^{10–13} Three cases describing as sphenoid sinus cholesteatoma, extended to the sellar or clivus

region.^{10,11,13} Most presented with headache, diplopia, and cranial nerve involvement. One case was found incidentally.¹²

Case Presentation

Initial clinical presentation

An 82-year-old, bedridden woman with underlying Alzheimer disease, renal insufficiency, hypertension, and diabetes mellitus type 2, with poor blood sugar control, presented with 1 month of fever. A direct reliable clinical history could not be obtained. Her daughters stated that she complained of chronic headaches for more than 20 years, but progressing during the past 6 months. She did not experience rhinorrhea, nasal obstruction, anosmia, or facial pain. There was no history of head trauma and temporal region fracture. Nasal endoscopy found purulent discharge from the left sphenoidal recess. Otoscopy showed central perforation of left tympanic membrane. Clear mucoid discharge was present in the middle ear. There was no attic retraction or cholesteatoma seen. Computed tomography



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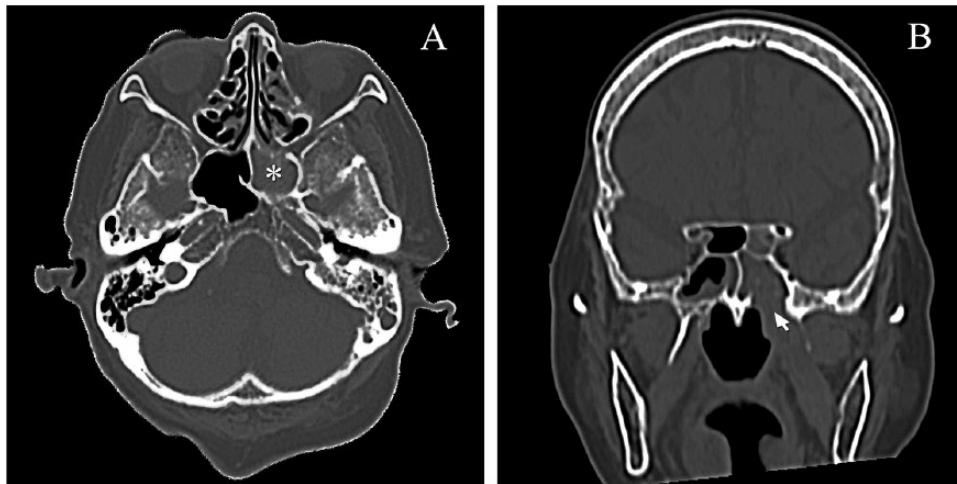


Figure 1. Computed tomography of the first presentation. Bone window axial (A) and coronal (B) computed tomography shows total opacification with multiple density in left sphenoid sinus (asterisk) and left Onodi cell, osteolytic and hyperplastic lesion involving posterior and inferior wall of left sphenoid sinus (arrow).

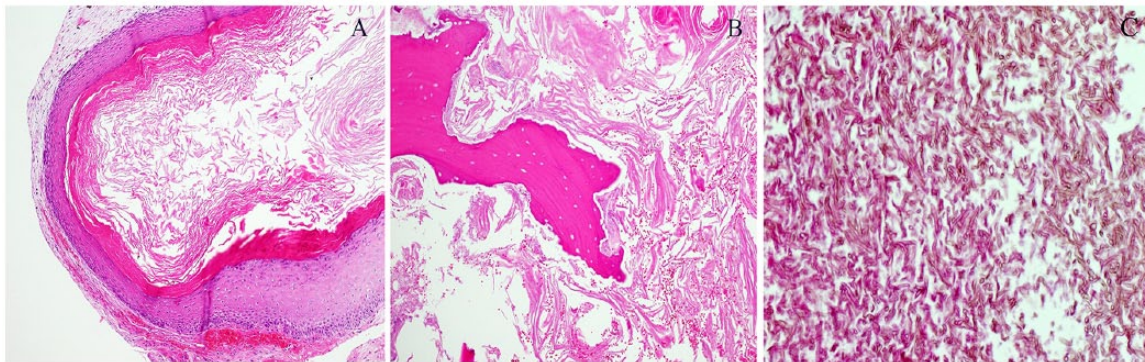


Figure 2. Histopathology of the lesions from the left sphenoid sinuses. Cyst-like spaces, lined by keratinizing benign squamous epithelium (A) (magnification: 100 \times); bone erosion, characterized by fragmented bony elements admixed with keratin materials, is also present (B) (magnification: 100 \times); and uniform regular-shaped hyphae, morphologically consistent with *Aspergillus spp.* (C) (magnification: 600 \times).

(CT) scan of the paranasal sinus showed soft tissue lesions of mixed high and low density that totally filled the left sphenoid sinus, left Onodi cell, and left sphenoidal recess. There was presence of high-density foci involving sinus, like bony flakes produced by fungi. Sinus wall destruction was seen, involving posterior and inferior walls of the left sphenoid sinus, left-sided clivus, and left pterygoid plate (Figure 1), and we suspected chronic invasive fungus rhinosinusitis. Soft tissue density was seen in the left mastoid cavity without evidence of bony destruction. Endoscopic left sphenoidotomy was performed. The accumulation of dense fungal collection was found intraoperatively within the left sphenoid sinus with overlying keratin material. The sphenoid sinus was widely opened, and all fungal balls were removed. Histopathology revealed cyst-like spaces, lined by keratinizing benign squamous epithelium compatible with sphenoid sinus cholesteatoma (Figure 2A). Bone erosion, characterized by fragmented bony elements admixed with keratin materials, were observed (Figure 2B). Fungal balls, consisting of uniform, regular-shaped hyphae,

morphologically consistent with *Aspergillus spp.* (Figure 2C), were detected in the left sphenoid sinus without evidence of tissue invasion. Headache and fever improved after the surgery. Nevertheless, clinical management became increasingly difficult due to the severe Alzheimer disease and multidrug-resistant bacterial infection. Nasal irrigation and topical therapies were not adequate.

Skull base osteomyelitis after endoscopic sinus surgery

Two months after surgery, she complained of worsening headache with abrupt onset of fever. The CT scan showed progression of disease with multiple permeative bony destructions, involving bony sinus walls and central skull base. This was likely skull base osteomyelitis (SBO). It involved left greater wing of the sphenoid bone, left medial pterygoid plate, clivus, medial border of left foramen ovale, left anterior part of foramen lacerum, medial part of left foramen spinosum, left petrous bone,

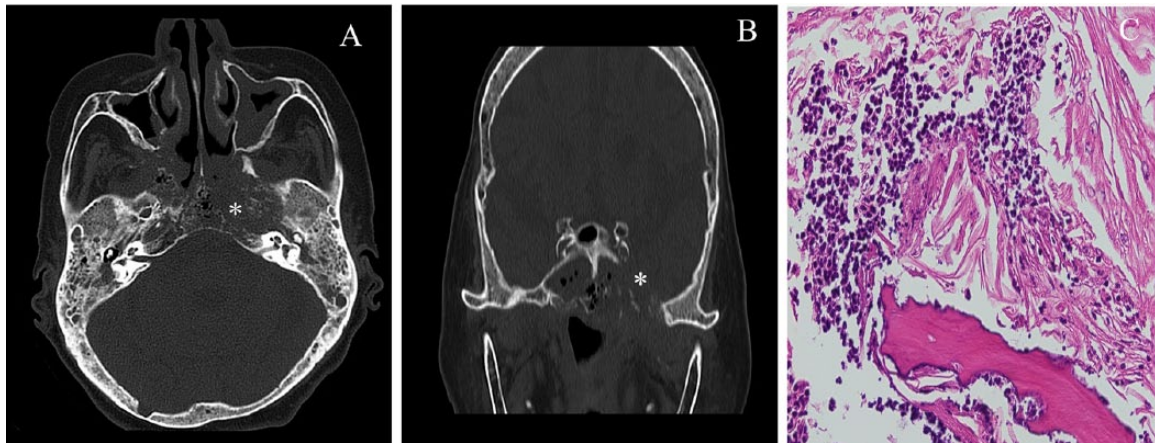


Figure 3. Computed tomography and histopathology of disease progression. Bone window axial (A) and bone window coronal (B) computed tomography shows progression of disease with multiple permeative bony destructions, involving bony sinus walls and central skull base (asterisk), involving left greater wing of the sphenoid bone, left medial pterygoid plate, clivus, medial border of left foramen ovale, left anterior part of foramen lacerum, medial part of left foramen spinosum, left petrous bone, and left carotid canal, encasing intrapetrous internal carotid artery and histopathology shows numerous inflammatory cells, mainly neutrophils admixed with keratin materials surrounding resorbed bony spicules, compatible with acute osteomyelitis (C) (magnification: 200 \times).

and left carotid canal, encasing intrapetrous internal carotid artery (Figure 3A and B). Soft tissue density lesions were visualized in left middle ear cavity and left mastoid, with preserved mastoid septae and ear ossicles, and without a connection with petrous soft tissue. This was likely otomastoiditis rather than cholesteatoma of the temporal bone. She was then scheduled for debridement of the infected bone. Left sphenoidotomy was maximally opened together with middle meatal antrostomy and ethmoidectomy. Right sphenoidotomy was performed and combined with left sphenoidotomy by removal of the sphenoid septum. Combined subtemporal and suboccipital craniotomy was performed. Intraoperative findings revealed keratin material with debris involving the skull base. Debridement of skull base was done for removal of cholesteatoma and infected bone. Cholesteatoma and infected bone were left around internal carotid artery and optic nerve area. Mastoidectomy was done. There was no cholesteatoma in the mastoid cavity. The histopathologic report revealed cholesteatoma with SBO. There were many neutrophilic infiltrates admixed with keratin, the features of which were compatible with acute osteomyelitis (Figure 3C). Culture reported *Klebsiella pneumoniae* (ESBL (Extended Spectrum Beta Lactamase)-producing strain). Ertapenam plus oral sitafloxacin was given for treatment of SBO. After the revision surgery, headache persisted with intermittent fever. Nasal endoscopy showed purulent discharge consisting of infected debris. CT scan showed disease progression eroding the central skull base. Risk for further surgery outweighed benefits and she succumbed to this disease.

Discussion

To the best of our knowledge, there have been only 3 case reports of sphenoid sinus cholesteatoma with cephalad extension to the sellar or clivus region.^{10,11,13} One showed lateral extension to temporal lobe.¹² All the cases had successful

marsupialization surgery by sphenoidotomy without report of complications. Cranial nerve and pituitary function returned to normal after treatment.

Our case of sphenoid cholesteatoma extended to the skull base and was complicated by osteomyelitis. Infection of the skull base was uncontrolled although receiving extensive surgery and culture-directed antibiotics. Without evidence of migration of squamous epithelium and metaplasia of epithelium, it is unclear whether this was a primary or secondary acquired cholesteatoma.¹ Having had chronic headache for more than 20 years, it is more likely that this was a congenital sphenoid cholesteatoma.

Clinical presentations of paranasal sinus cholesteatoma vary depending on the site of the originate location and its extension. Symptoms may include nasal obstruction, rhinorrhea, cheek pain, buccal swelling, and headache. Our patient's chronic headache was likely due to accumulation of keratin together with fungal residue and secondary infection causing progressive bony destruction with fever. Radiological imaging is useful for suggesting differential diagnosis, although it does not give a definite diagnosis. Cholesteatoma appears as sharply circumscribed round or ovoid bony defect with marginal sclerosis.⁴ It appears as homogeneous lesion with bony erosion and remodeling. On magnetic resonance imaging (MRI), it appears as low intensity in T1 and high intensity in T2.⁹ Diagnosis is confirmed by histology.

Treatment of choice of paranasal sinus cholesteatoma is surgery. Endoscopic surgery for paranasal sinus cholesteatoma is similar in approach as temporal bone surgery for middle ear and mastoid cholesteatoma. Goal is to eradicate all disease and to remove all the walls of cholesteatomas. If not possible, marsupialization would be preferred to promote drainage and ventilation to prevent further erosion of surrounding structures and disease progression.⁹ Advances in

instrument and technique and in endoscopic sinus surgery should allow more success in outcomes.

Intracranial complications secondary to obstructed sino-nasal cholesteatoma are rare. Up to date, of 30 English case reports of paranasal sinus cholesteatoma identified from Medline database, only 2 cases of intracranial complications were reported.^{14,15} Both cases were secondary to frontal sinus cholesteatoma. There was no complication reported secondary to maxillary, ethmoid, and sphenoid sinus cholesteatoma. There was no SBO reported. The first case¹⁴ presented with painful frontal-orbital mass, fever, and generalized seizure. *Streptococcus pneumoniae* was later identified by lumbar puncture. Meningoencephalitis as an aggravating factor of frontal sinusitis was diagnosed. Open frontal sinus surgery with long-term antibiotics was given. The second case¹⁵ presented with confusion and disorientation. Brain abscess in left frontal lobe was identified. Endoscopic frontal sinus drainage and abscess aspiration was done. Frontal sinus obliteration was performed on second-look operation. Both cases responded well to the treatment.

SBO is a rare condition. This life-threatening entity was historically described in elderly diabetics. Disease progression from otitis to osteomyelitis has been well-documented, the term SBO has been used interchangeably with necrotizing or malignant otitis externa, and *Pseudomonas aeruginosa* is the usual pathogen. Nevertheless, central or atypical SBO, unrelated to auricular cause, has been described in case reports and series. It may originate from paranasal sinus inflammatory disease or be hematogenous in origin.¹⁶ Diabetes mellitus is a predisposing factor due to defects in chemotaxis and phagocytosis of polymorphonuclear leukocytes, monocytes, and macrophages.¹⁷ It theoretically suggests that any conditions that adversely affect blood flow and oxygen delivery through bone may also predispose to infection of the central skull base.

Several factors may contribute to SBO. First, bone resorption and subsequent bone infection start easier through resorbed bone. Second, the immunocompromised state of patients also represents an important factor. Third, infection that originated in paranasal sinuses should be considered as source for invasion of the skull base. In our case, cholesteatoma was not expected at first. The first sphenoidotomy was wide enough for sphenoid fungal ball removal and for marsupialization of keratin from sphenoid cholesteatoma. When our patient was bedridden with severe Alzheimer disease, adequate postoperative debridement and topical care was difficult. The retained squamous epithelium producing keratin and enzymes became the source of aggressive infection and caused SBO. Aggressive debridement by craniotomy and extensive sinus surgery was performed, and long-term culture-directed antibiotics were used for treatment of this fatal complication.

Conclusions

Paranasal sinus cholesteatoma is a rare disease, and sphenoid sinus is the rarest location. Sphenoid cholesteatoma should be considered one of the causes of unexplained progressive headache. It is a

surgical condition. The sphenoid sinus should be maximally opened. The related fatal complication is osteomyelitis of the skull base. The location of cholesteatoma and the immunologic status of patient are key contributing factors to the development of this complication. Aggressive surgical debridement is necessary. Culture-directed antibiotics are required. Effective postoperative debridement and adequate topical therapies are essential.

Author Contributions

DK: literature review, data collection, drafting the article; PC: data collection, revising the article; SK: data collection, revising the article; KS: conception, study design, drafting the article and final approval.

Ethical Approval

This study was approved by the Bangkok Hospital Institutional Review Board for research involving human subject.

Informed Consent

Patient informed consent was obtained.

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