



Original Article

Isolated Sphenoid Sinus Disease: Analysis of 11 Cases

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Abstract

Objective: To evaluate the presenting symptoms, imaging data, surgical treatment, and clinical outcomes associated with isolated sphenoid sinus disease.

Patients and Methods: From July 2003 to February 2008, the authors performed surgery on 11 patients with isolated sphenoid sinus diseases. The presenting signs and symptoms, radiological studies, operative findings, and clinical outcomes were retrospectively reviewed and analyzed.

Results: Five patients presented with a fungal process, three with sphenoid sinusitis, one with mucocele, one with an angiofibroma, and one with fibrous dysplasia. The most common presenting symptom was headache, followed by rhinorrhea and visual symptoms. Ten patients were treated endoscopically and one patient was treated with an external approach.

Conclusion: Because of the proximity of the sphenoid sinus to important and vulnerable structures of the skull base, delay in diagnosis and treatment can lead to the development of serious intracranial and orbital complications. The presenting symptoms of patients with an isolated sphenoid sinus lesion are often vague and nonspecific, with headache being the only reliable finding. Endoscopic sinus surgery is a safe, effective, and direct approach to the sphenoid sinus. (*Tzu Chi Med J* 2009;21(3):227–232)

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1. Introduction

Paranasal sinusitis is a common disease diagnosed in otolaryngology clinics. However, the sphenoid sinus is uncommonly involved in inflammatory diseases and tumors. The sphenoid sinus is located deep in the skull base and the early symptoms of sphenoid sinus diseases are often nonspecific; examination with a headlight and plain radiographs offer little diagnostic information on sphenoid pathology (1). The sphenoid sinus lies in proximity to 13 vital structures—including the dura, pituitary gland, optic nerve, cranial nerves III,

IV, V, and VI, cavernous sinus, internal carotid artery, sphenopalatine ganglion, sphenopalatine artery, and pterygoid canal (2). Because of this, once infection spreads beyond the sphenoid sinus to these neighboring vital structures, serious intracranial and orbital complications may develop. This will result in irreversible neurological sequelae, and even life-threatening conditions. These risks should not be overlooked.

In recent years, advances in endoscopy and imaging techniques have allowed better diagnosis and safe management of sphenoid sinus diseases. In this study, we report on 11 cases of pathological changes

primarily involving the sphenoid sinus; these changes are identified and reviewed.

2. Patients and methods

We retrospectively reviewed the cases of 11 patients who had undergone surgery for isolated sphenoid sinus disease between July 2003 and February 2008 at our otorhinolaryngology department. Their age range was 13–67 years, with a mean age of 45 years. There were four men and seven women. The diagnosis of isolated sphenoid sinus disease was based on characteristic signs and symptoms, routine ear, nose, and throat examinations, radiographic imaging, and histopathological and microbiological examinations of the resected specimens. All specimens suggested isolated disease of the unilateral or bilateral sphenoid sinus. Tumors from adjacent intracranial structures or tumors primarily involving other sinuses were not included. The presenting signs and symptoms, radiological studies, operative findings, and clinical outcomes were retrospectively reviewed from the patients' medical records.

3. Results

Among the 11 patients, five presented with a fungal process, three with sphenoid sinusitis, one with mucocele, one with an angiofibroma, and one with

fibrous dysplasia (Table 1). The symptomatology was often nonspecific. Headache was the most frequent symptom presenting in seven (64%) of the 11 patients. Rhinorrhea was noted in five patients (45%). Visual symptoms were noted in three patients (27%). The visual symptoms occurred in the form of blurring of vision (67%), diplopia (67%), and proptosis (33%). Nasal obstruction was the presenting symptom in three patients (27%), and epistaxis in one (9%). All patients underwent coronal and axial computed tomography (CT) scanning of the paranasal sinuses. Magnetic resonance imaging (MRI) was performed in four patients (36%).

Endoscopic surgery was performed in 10 of the patients (91%), and a lateral rhinotomy in one (9%). On sphenoid exploration, five patients had inspissated secretions and fungal debris consistent with fungoma. Two patients had chronically inflamed sinus mucosa and one patient had mucopurulent secretions filling the sphenoid cavity. One patient had a cyst with serous fluid content. One patient had a heterogeneous tumor mass that occupied the medial aspect of the sphenoid sinus. The follow-up period was between 1 and 5 years (mean, 3.5 years).

3.1. Inflammatory lesions

3.1.1. Fungal process

Fungal sinusitis was the most common isolated sphenoid lesion occurring in five patients. The most

Table 1 — Presenting symptoms and surgical findings of 11 patients with isolated sphenoid sinus disease

Case number	Age (yr)	Sex	Symptoms	Surgical findings
Fungal process				
1	67	F	Headache	Fungal debris
2	53	F	Headache Rhinorrhea	Fungal debris
3	41	F	Ocular signs: visual loss, diplopia Rhinorrhea Nasal obstruction	Fungal debris
4	41	M	Headache	Fungal debris
5	55	F	Rhinorrhea	Fungal debris
Sphenoid sinusitis				
6	49	F	Headache Rhinorrhea Ocular sign: visual loss	Mucopus
7	50	M	Headache	Polypoid
8	26	M	Headache Rhinorrhea Nasal obstruction	Polypoid
Mucocele				
9	39	F	Ocular signs: proptosis, diplopia	Cyst with serous fluids
Angiofibroma				
10	66	M	Epistaxis Nasal obstruction	Tumor
Fibrous dysplasia				
11	13	F	Headache	Whitish bony tumor

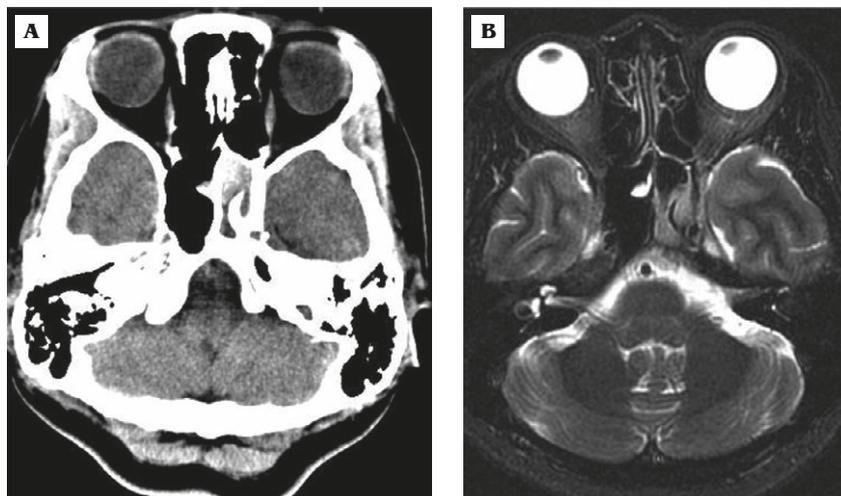


Fig. 1 — Case 2: a patient with fungal process (mycetoma). (A) Axial computed tomography image obtained with the soft-tissue window technique, demonstrates opacification of the left sphenoid sinus with marked thickening of the walls of the sinus. This suggests a long-standing inflammatory process. Foci of increased density within the material suggest chronic inspissated secretions or the presence of fungal elements. (B) Axial T2-weighted magnetic resonance imaging of the same patient reveals a hypointense signal within the left sphenoid sinus surrounded by a soft-tissue signal. Low signal foci are frequently noted in cases of fungal disease and may be related to the presence of hyphae.

common symptom was headache, which occurred in three patients. Two patients presented with retro-orbital headache and one with vertex headache. Three patients presented with rhinorrhea. One patient reported visual symptoms (visual loss and diplopia). In three patients, CT scanning showed a sphenoid mass with a speckled pattern of high attenuation on soft-tissue windows (Fig. 1). Unilateral sphenoid opacification was seen in the others. MRI was performed in one patient with unilateral visual disorders. Endoscopic sphenoidotomy showed a fungal mass within the sphenoid sinus; the contents confirmed aspergilloma. Postoperatively, all patients experienced relief of their symptoms. The patient with visual symptoms (visual loss and diplopia) also improved postoperatively.

3.1.2. Sphenoid sinusitis

Sphenoid sinusitis was the second most commonly isolated sphenoid lesion, occurring in three patients. The main complaint of all three patients was headache, and the second most common symptom was rhinorrhea. One patient suffered from an acute onset of unilateral blurred vision. CT scanning demonstrated isolated opacification in the sphenoid sinus in all three patients. MRI was performed in one patient with unilateral visual symptoms. All patients underwent endoscopic transnasal sphenoidotomy, which showed polypoid mucosa in the sphenoid recess in two patients and mucopus in one. There was no bacterial growth in the culture of the mucopus obtained on drainage. No symptoms of recurrence

were revealed within the postoperative follow-up period, which averaged 3.1 years. The patient in case 6—with blurred vision—did not improve during the 4-year follow-up period.

3.1.3. Mucocele

A 39-year-old woman (case 9) presented with diplopia and proptosis. CT scanning demonstrated an expansile, homogeneous opacity with bone remodeling. A T2-weighted MRI scan revealed a hyperintense mass within the left sphenoid sinus (Fig. 2). Mucocele marsupialization was performed by a direct endoscopic transnasal approach. Intraoperative and histopathological findings were consistent with mucocele. Unfortunately, although proptosis had improved gradually, diplopia did not improve during the 3-year follow-up period.

3.2. Noninflammatory lesions

3.2.1. Angiofibroma

A 66-year-old man (case 10) presented with frequent epistaxis and nasal obstruction. Endoscopic examination revealed a dark-red, polypoid mass extending out of the sphenoid sinus into the sphenoid recess. It was found to have no connection with the nasopharynx. CT scanning demonstrated a dense soft-tissue mass with contrast enhancement, complete sphenoid sinus opacification, erosion of the anterior sphenoid wall, and extension into the nasal cavity. Preoperative transarterial embolization was

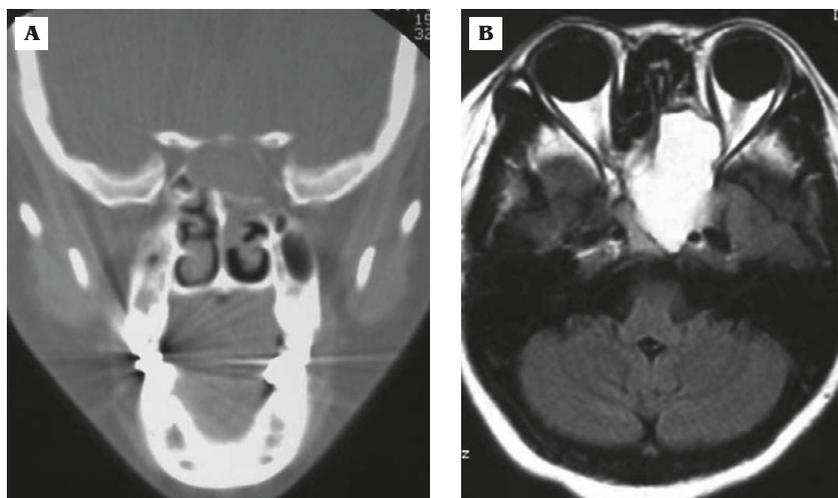


Fig. 2 — Case 9: a patient with a mucocele. (A) Coronal computed tomography imaging shows an opacification of the left sphenoid sinus that caused pressure erosion and remodeling of the adjacent bone. (B) T2-weighted magnetic resonance imaging shows a hyperintense mass within the left sphenoid sinus.



Fig. 3 — Case 11: a patient with fibrous dysplasia. (A) Axial computed tomography imaging shows a well-defined, heterogeneous tumor mass occupying the medial aspect of the right sphenoid sinus. (B) Intraoperative finding of a smooth-surfaced, whitish mass within the right medial sphenoid sinus.

performed. The patient then underwent tumor excision via a lateral rhinotomy. The patient showed no evidence of recurrent disease at the 2.5-year follow-up examination.

3.2.2. Fibrous dysplasia

A 13-year-old girl (case 11) underwent CT scanning because of the recent onset of severe headache. The scan demonstrated a heterogeneous hyperdense mass within the right sphenoid sinus. The patient was treated with endoscopic sphenoidotomy and tumor removal (Fig. 3). Histopathological examination confirmed fibrous dysplasia. Her headaches ceased during the 1-year follow-up period.

4. Discussion

The sphenoid sinus is located deep in the skull base within the sphenoid bone. Isolated sphenoid diseases are rare and account for approximately 2–3% of all sinus disease (3,4). In our case series, fungal sinusitis was present in most cases, a finding contrasting with Lawson and Reino's report (3), in which sphenoid sinusitis was the most commonly isolated sphenoid lesion. The higher percentage of fungal sinusitis in our cases may be due to the moist weather in Taiwan, which favors the growth of fungus in the nasal cavity and paranasal sinus. The presenting symptoms of the isolated sphenoid sinus lesion are

often vague and nonspecific; thus, delay in diagnosis is rather frequent (5). The most common symptom of isolated sphenoid sinus lesion is headache that is refractory to medical management; it occurs in 70–90% of patients (2,3,6). In this series of 11 patients, the incidence was 64% (7 of 11). Six patients were first referred to neurosurgery for severe headache, but after radiographic imaging was completed, isolated sphenoid sinus lesion was eventually detected. Sethi (1) reported that retro-orbital headaches were typical in patients with an isolated sphenoid sinus lesion. In the present study, however, the predominant site of headache was nonspecific. As in previous reports (7), headache, followed by postnasal drip, were the most common presenting symptoms in our case series.

Visual changes were the third most common presenting symptom in our series (27%), which is similar to the incidence found in other series (3,5,6). Although reported more commonly in patients with malignant neoplastic processes (3), visual changes occur in inflammatory and benign neoplastic processes as well (6), as was reported in our series. Lee et al (8) reported that the results of endoscopic sinus surgery for patients with abducens palsy and aspergillosis were excellent. As in the patient with fungal process (case 2), visual symptoms were improved postoperatively. However, in case 6 (sphenoid sinusitis) and case 9 (mucocele), visual symptoms did not improve. The visual disturbance may have resulted from irreversible cranial neuritis, pressure ischemia attributable to compression by an expanding lesion, or ischemic infarction from thrombophlebitis or vasculitis. Recovery of vision depends on the length of time before surgery, the severity of the visual disturbance, and the severity of sphenoid sinus disease. Visual disturbances lasting for more than 6 months were regarded as irreversible and were indicative of a poor prognosis (8).

When the sphenoid sinus is involved with a disease process, the history and physical examination do little to establish the correct diagnosis. Nasal endoscopy and imaging studies (CT scanning and/or MRI) are essential for a thorough evaluation and management (3,9). Even though the presenting symptoms of isolated sphenoid sinus disease tend to be vague and nonspecific, clinical suspicion of it warrants imaging because of its serious complications. In the present series, CT was performed in all patients and provided valuable information in relation to bony erosion. MRI was performed for further evaluation in patients who had space-occupying lesions with bony erosion, visual symptoms, or cranial nerve palsy (36%). MRI permitted a more precise diagnosis of sphenoid sinus disease by providing valuable information regarding its relationship to the dura, pituitary gland, cavernous sinus, optic nerve, and carotid artery. When visual

symptoms appear and a clouded sinus is revealed on CT or MRI, it becomes imperative that surgery is performed as soon as possible (8,10).

Approaches to the sphenoid sinus include the use of both open and endoscopic techniques. Currently, endoscopic sinus surgery has become a mainstream treatment for sphenoid sinus diseases. The endoscopic approach can proceed in a transethmoidal fashion or directly, through the anterior sphenoid sinus wall at the site of the natural ostium (4). Each of these may be used in conjunction with an open approach if necessary. In cases involving an inflammatory process, performing transnasal endoscopic sphenoidotomy by using the superior turbinate as a key landmark has been reported to be a safe and effective technique (4). It enables excellent visualization of the whole sinus, which induces less surgical trauma and results in less morbidity (4,7). Ten of our patients were treated with a direct endoscopic transnasal approach and no morbidity was reported.

Fungal disease necessitates surgical treatment to open the sphenoid sinus and allow complete removal of the fungal debris by suctioning and repeated washing. Histological examination is always performed to identify if the disease has spread through the mucous. All of our cases were the noninvasive form (fungus ball), and no further medical treatment was necessary. All patients with fungal sinusitis and headache experienced relief of their symptoms postoperatively. Case 2, a patient with visual symptoms, improved satisfactorily after the surgery.

Sphenoid sinusitis is categorized as acute (case 6) or chronic (cases 7 and 8). Risk factors for the development of acute sphenoid sinusitis include diving in contaminated water, cocaine or steroid use, diabetes, and nasal fractures. Chronic sphenoid sinusitis may result from obstructive polyposis of the sphenoid recess. Lew et al (11) demonstrated that for most patients, acute sphenoiditis was the result of Gram-positive infection, usually *Streptococcus* or *Staphylococcus* species, and that the cultured organisms in chronic sphenoiditis tended to be evenly divided between Gram-negative and Gram-positive species. Broad-spectrum antibiotics, often administered via an intravenous route, are the treatment for acute bacterial sphenoid sinusitis. For patients who are clinically ill, fail to respond to medical treatment, or show evidence of complications resulting from the spread of infection to neighboring structures, decompression of the sphenoid sinus is indicated, either via an endoscopic or an open approach. One patient (case 6) suffered from an acute onset of unilateral blurred vision and headache. CT demonstrated isolated sphenoid opacification. Although an emergency endoscopic transnasal sphenoidotomy was performed and mucopus was drained to decompress the sphenoid sinus, her vision did not recover

postoperatively. Chronic sphenoid sinusitis is treated most effectively by removal of the obstructing polyps and debris. This restores the patency of the natural ostium and returns the sinus to its healthy state of effective mucociliary clearance. As in cases 7 and 8, headache was relieved in the postoperative period.

A sphenoid sinus mucocele appears as an opacified expanded sinus on CT. MRI may be helpful for differentiating mucoceles from other lesions that expand the sinus and have a similar density on CT (6). The mucocele requires only marsupialization and subsequent drainage of the sinus. The bone layer overlying the mucocele is frequently thin or dehiscent (12). The area is identified by palpation with a suction tube; the mucocele is entered with a curette and is then enlarged with a circumferential punch. Lee et al (8) reported that 75% of their patients had improved visual acuity postoperatively; this was not the case in our patient. Our patient's lack of improvement might have been the result of irreversible compressive optic neuropathy.

Isolated sphenoid sinus angiofibroma is very rare; only three such cases have been reported (13–15). Endoscopy and imaging studies are essential to make an accurate diagnosis and to plan treatment. In the present case, it would not have been safe to perform *en bloc* removal of the tumor via a transnasal endoscopic technique because of its size and the potential risk for bleeding. Transarterial embolization of the feeding artery and extirpation of the tumor via an external approach was the best choice of treatment for angiofibroma in the sphenoid sinus.

Fibrous dysplasia restricted to the sphenoid sinus is a rare entity. They tend to be slow-growing tumors and are infrequently symptomatic. Often, it is difficult to say with certainty whether a patient's symptoms can be attributed to a fibro-osseous lesion. Most authors agree that resection is the treatment of choice when these lesions are symptomatic or when they exhibit rapid growth. The patient in case 11 had satisfactory resolution of headache after removal of the tumor. The correlation between headache and sphenoid fibrous dysplasia can be strongly ascertained.

Because of the anatomical proximity of the sphenoid sinus to important optic and intracranial structures, isolated sphenoid sinus diseases should be treated promptly. If left untreated, sphenoid sinus disease can result in serious intracranial and orbital

complications. The presenting symptoms of isolated sphenoid sinus disease tend to be vague and non-specific, with headache being the only reliable symptom. We emphasize that clinicians should keep in mind the possibility of sphenoid sinus disease when treating refractory headache and consider the success of surgical treatment in sphenoid fungal sinusitis. The majority of isolated sphenoid sinus opacifications associated with inflammatory or infectious disease can be managed with endoscopic techniques.

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