Evaluation and Surgical Management of Isolated Sphenoid Sinus Disease

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Objectives: To evaluate the pathologic conditions, preoperative evaluation, treatment, and clinical outcomes associated with sphenoid sinus disease.

Design: Retrospective study.

Setting: Tertiary university-based referral center.

Patients: All patients with isolated sphenoid sinus disease managed surgically or in which surgery was considered a primary treatment option.

Main Outcome Measures: Demographic data, presenting signs and symptoms, endoscopic and imaging findings, surgical management, surgical pathology, and clinical outcomes were investigated in patients presenting with sphenoid sinus disease to the Medical College of Wisconsin, Milwaukee, between January 1, 1991, and December 31, 2001.

Results: The study population included 17 women and 12 men with a mean age of 52.3 years (range, 15-82 years). The most common presenting symptom was headache (20 patients [69%]). Imaging evaluation included computed tomography and/or magnetic resonance imaging studies in all cases. Sphenoid sinus abnormality was variable and included sinusitis (11 patients [38%]), tumor (7 [24%]), mucocele (5 [17%]), fungal process (3 [10%]), and cerebrospinal fluid fistula (3 [10%]). Twenty-one cases (72%) were managed endoscopically and 4 (14%) were managed with a transseptal approach. One patient (3%) underwent combined extracranial-endoscopic transnasal approach, while another (3%) underwent a midface degloving approach. The remaining 2 patients (7%) did not undergo surgical intervention.

Conclusions: Given the high prevalence of noninflammatory lesions within the sphenoid sinus, thorough preoperative evaluation is imperative. Initially, this should include nasal endoscopy and computed tomography to help define the location, extent, and character of the lesion. In some cases, magnetic resonance imaging may help further define the nature and extent of a lesion. Angiography should be considered if a vascular lesion is suspected. The clinical and imaging findings should all be taken into consideration when the surgical approach is planned.


INITIAL SYMPTOMS of sphenoid sinus disease are often vague and difficult to characterize. Despite the presence of several critical structures surrounding the sphenoid sinus, including the carotid artery, optic nerves, dura mater, and cranial nerves III to VI, symptoms related to these structures occur less frequently than nonspecific complaints, such as headache.1-4 The most common associated symptoms reported in the literature are headache and visual compromise, which may occur with both inflammatory and neoplastic processes. Sphenoid sinus disease is frequently detected radiographically as an incidental finding in the evaluation of some other abnormality. Varied pathologic findings in the region, including vascular neoplasms, vascular malformations, and encephaloceles, can increase the risks of surgery in this region.5-7 Thus, a thorough preoperative workup, including endoscopic evaluation and advanced imaging techniques, is helpful to allow safe management of lesions located in this region.

In this report, 29 cases of pathologic changes primarily involving the sphenoid sinus are identified and reviewed. The purpose of our investigation is to review our experience with lesions located in this region. Special attention is given to the preoperative evaluation, with an emphasis on imaging and its role in treatment planning.

METHODS

A retrospective study was undertaken to identify all patients treated at the Medical College of Wisconsin, Milwaukee, with a diagnosis of disease involving the sphenoid sinus. All of these cases underwent careful chart review to identify cases of presumed isolated sphenoid sinus disease. Only cases that were managed...
surgically or in which surgery was considered as a primary treatment option were included. Preoperative symptoms and physical findings, including cranial nerve deficits, imaging studies, operative procedures, surgical findings, pathological diagnosis, and postoperative results, were reviewed. Care was taken during the review process to include only patients with endoscopic and/or imaging evidence of abnormality primarily involving the sphenoid sinus. Tumors from adjacent intracranial structures that could be misconstrued as primary sphenoid processes were included. Tumors primarily involving other sinuses were excluded from this report.

### RESULTS

Twenty-nine cases of isolated sphenoid sinus disease were identified. Seventeen patients were female and 12 were male, with an age range of 15 to 82 years (mean, 52.3 years). Clinical follow-up ranged from 2 months to 10 years (mean, 19 months). Headache was the most common symptom (20 patients [69%]) among all patients. The headaches were most frequently characterized as retro-orbital or occipital. Other symptoms noted at presentation included facial pain (5 [17%]), decreased visual acuity or diplopia (6 [21%]), purulent rhinorrhea (3 [10%]), and unilateral nasal obstruction (7 [24%]). Two patients presented with symptoms consistent with meningitis, 3 presented with cerebrospinal fluid (CSF) rhinorrhea, and 2 presented with endocrine abnormalities. One patient was asymptomatic and the lesion was found incidentally on imaging performed for another reason. All 29 patients underwent computed tomographic (CT) imaging, while 20 underwent both CT and magnetic resonance (MR) imaging, and 1 patient underwent CT, MR imaging, and cerebral arteriography.

Sphenoid sinus disease included sinusitis (11 patients [38%]), tumor (7 [24%]), mucocele (5 [17%]), fungal process (3 [10%]), and CSF fistula (3 [10%]). For descriptive purposes, lesions are divided into inflammatory and noninflammatory categories.

Surgical treatment of the patients was endoscopic in 21 (72%), transseptal in 4 (14%), midface degloving in 1 (3%), and a combined extracranial-endoscopic transnasal resection in 1 (3%). One patient chose primary radiation therapy (3%) and an additional patient (3%) declined surgical intervention.

### INFLAMMATORY LESIONS

#### Sinusitis

Table 1 highlights the presenting symptoms for the 11 patients found to have isolated sphenoid sinusitis. The most common presenting symptom was headache (8 of 11 patients). Clinical descriptions of the headache included retro-orbital (6 of 8 patients) and occipital (2 of 8 patients). Patients also reported, in increasing order of frequency, visual symptoms, purulent drainage, and nasal obstruction. Of the 2 patients reporting visual symptoms, one noted diplopia and the other described a unilateral decrease in visual acuity. On examination, the second patient was found to have asymmetric pupils, mild ptosis, and unilateral dry eye, consistent with Horner syndrome. The patient described in the “Other” category in Table 1 had findings consistent with meningitis (fever, mental status changes, headache, stiff neck) as well as positive blood cultures. On endoscopic examination, 6 of 11 patients were found to have purulent drainage in the sphenoidomaiual recess, while the remaining patients had nonspecific findings. Imaging studies in these patients showed findings consistent with inflammatory disease, such as air-fluid levels, mucosal thickening, opacification, and/or chronic osteitis. Bone erosion was not identified in any of these cases. The patient who presented with symptoms of meningitis demonstrated MR findings consistent with acute sphenoid sinusitis with intracranial findings of epidural empyema and dural inflammation (Figure 1).

Surgical therapy consisted of endoscopic sphenoidotomy in all patients, with the use of either an endoscopic transnasal or transethmoid approach in all cases. Postoperatively, 10 of 11 patients noted improvement of their symptoms. The remaining patient subsequently required a transseptal sphenoidotomy with complete removal of the anterior sphenoid face because of severe osteoneogenesis and was symptom free 28 months postoperatively. One patient required a revision endoscopic sphenoidotomy 6 years later for recurrent symptoms. One case was complicated by a CSF leak, which occurred as a result of injury to the posterior cribiform plate. This was noted intraoperatively and repaired with a free mucosal graft and no lumbar drainage. The patient recovered and the CSF leak resolved without incident. Both patients with visual compromise had resolution of their visual symptoms.

#### Fungal Lesions

All 3 patients diagnosed as having fungal lesions presented with headache. The headache was described in 2 cases as retro-orbital. One patient experienced unilateral decrease in visual acuity along with purulent rhinorrhea (Table 1). Nasal endoscopy showed purulent sphenoidomaiual recess drainage in all 3 patients. The CT and MR findings were consistent with fungal dis-

<table>
<thead>
<tr>
<th>Table 1. Presenting Symptoms for Inflammatory Lesions of the Sphenoid Sinus</th>
<th>No. (%)</th>
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<tbody>
<tr>
<td><strong>Facial Pain</strong></td>
<td>Headache</td>
</tr>
<tr>
<td>Total Inflammatory Group (n = 19)</td>
<td>15 (79)</td>
</tr>
<tr>
<td>Fungal (n = 3)</td>
<td>3 (100)</td>
</tr>
<tr>
<td>Mucocele (n = 5)</td>
<td>4 (80)</td>
</tr>
<tr>
<td>Sinusitis (n = 11)</td>
<td>8 (73)</td>
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</table>
ease isolated to the sphenoid sinus (Figure 2). The patient with decreased visual acuity was noted to have bony erosion over the optic nerve on CT imaging, and MR imaging showed optic nerve inflammation.

Surgical management consisted of endoscopic transnasal sphenoidotomy with findings of fungal mycetoma (Aspergillus species) and mucosal thickening. Histopathologic examination disclosed noninvasive Aspergillus species in all cases. Postoperatively, all patients had relief of their symptoms. The patient with decreased visual acuity experienced partial recovery with continued symptoms that were difficult to distinguish from an unrelated dementia. None of the patients had recurrence of the disease based on postoperative endoscopy and CT.

**Mucoceles**

In our series, 5 patients had mucoceles or mucopyoceles (Table 1). Headache was the most common symptom (4 of 5 cases). Other symptoms included decreased visual acuity, nasal obstruction, and purulent rhinorrhea. Endoscopic findings were nonspecific in 4 of 5 cases. In the remaining case, the mucocele displaced the face of the sphenoid sinus anteriorly. The CT and MR findings were consistent with mucocele in all 5 patients, with opacification and expansion of the sinus walls.

All 5 patients underwent endoscopic marsupialization and partial resection of the mucocele by an endoscopic transethmoid approach. Intraoperative and histopathologic findings were consistent with mucocele in all cases. Postoperatively, all patients experienced symptom resolution, and no revision surgery has been required. Follow-up evaluations, including nasal endoscopy and CT, showed no evidence of recurrent or persistent disease at a mean follow-up period of 37 months (range, 24-60 months).

**NONINFLAMMATORY LESIONS**

**CSF Leak**

Three patients were identified with sphenoid sinus CSF fistula confirmed by β2-transferrin studies. The cause of the leaks was not apparent on the basis of history and physi-
Neurone patient was found to have herniation of arachnoid through the bony defect in the sinus was not identified by MR imaging in any of the cases.

The MR findings included hyperintense signal on T2-weighted magnetic resonance image in the same patient demonstrates hyperintense signal consistent with fluid in the left sphenoid sinus. No herniation of brain parenchyma into the sinus through the bony dehiscence seen on the computed tomographic examination was identified.

Figure 3. Cerebrospinal fluid leak. A, Coronal bone window computed tomographic image demonstrates a prominent air-fluid level within the dependent portion of the left sphenoid sinus. A defect in the roof of the left sphenoid sinus (arrow) is noted laterally. The lateral position of the defect in a somewhat hyperpneumatized sphenoid sinus is suggestive of an erosive arachnoid granulation as the cause of the defect. B, Coronal T2-weighted magnetic resonance image in the same patient demonstrates hyperintense signal consistent with fluid in the left sphenoid sinus. No herniation of brain parenchyma into the sinus through the bony dehiscence seen on the computed tomographic examination was identified.

Sphenoid Sinus Tumors

Two groups of patients with neoplasms involving the sphenoid sinus were identified. Three patients were identified with primary sphenoid sinus tumors, including hemangioma, squamous cell carcinoma, and adenoid cystic carcinoma. Four patients had neoplasms arising from adjacent intracranial structures that resulted in complete opacification of the sphenoid sinus. These lesions included a meningioma and 3 pituitary adenomas. As a group, the patients with tumors involving the sphenoid sinus most commonly presented with nasal congestion or visual symptoms.

Primary Sphenoid Sinus Neoplasms. Three patients were identified with primary sphenoid neoplasms, 2 of which were malignant (Table 2). Nasal congestion was the most common complaint (3 [100%]). Neither malignant neoplasm resulted in cranial nerve deficits. Endoscopic findings included soft tissue lesions in the sphenoid sinus recess, one of which was pulsatile. Soft tissue-density masses with variable enhancement, partial to complete sinus opacification, and erosion of 1 or more of the sphenoid walls were noted with CT imaging. The MR findings varied with tumor type but in all cases helped further characterize the tumor and define its extent. Prominent enhancement on the CT and MR studies in the patient with the cavernous hemangioma suggested a vascular lesion, although flow voids were not identified on the MR study (Figure 4). The patient underwent conventional cerebral angiography that demonstrated a dense tumor blush and multiple arterial feeders from the internal and external carotid arteries. Preoperative transcatheter embolization with balloon test occlusion of the internal carotid artery was performed in this patient.

Treatment included surgical excision in 2 patients, while 1 patient with squamous cell carcinoma of the sphenoid sinus and invasion of the cavernous sinuses underwent primary radiation therapy. Excision of the cavernous hemangioma required a midface degloving approach, while the adenoid cystic carcinoma was resected by means of a transthyroid endoscopic approach. In this case, the tumor was located at the anterior face of the sphenoid. An ethmoidectomy was performed for the purpose of exposure and to facilitate postoperative endoscopic monitoring. Then the anterior face of the sphenoid as well as the posterior septum was resected en bloc with the tumor. There were no treatment complications, and all 3 patients were disease free on the basis of postoperative endoscopic and radiographic evaluation at varying periods of follow-up (Table 2).

Tumors From Surrounding Structures. In our series, 4 tumors arose from surrounding structures (Table 3). Presenting symptoms and endoscopic findings were variable. The CT scan showed partial or complete sphenoid sinus opacification with erosion of the sphenoid walls.
presenting with CSF rhinorrhea, nasal endoscopy can be
fining any extension into the nasal cavity. For patients
examination of patients with primary sphenoid sinus dis-
inflammatory and noninflammatory lesions.

cists, although important to document, may occur with both
related to decreased visual acuity or oculomotor dysfunc-
headache as the primary present-
cases (21%), which is
sphenoid sinuses disease. Computed to-
mass can be catastrophic, and death has
inflammatory and noninflammatory lesions.

Nasal endoscopy should be performed as part of the
of patients with primary sphenoid sinus dis-
ease. It is helpful in obtaining culture material and de-
fine any extension into the nasal cavity. For patients
helpful in localizing the site of the leak to the sphenoid
Endoscopy can also be helpful in identifying any
anatomic obstructions that may need to be addressed at
the time of surgery. However, even in cases where the
lesion extends into the nasal cavity, minimal informa-
treatment plan is in place.

Sinonasal CT is indicated in all cases of isolated sphen-
oid sinus disease. Computed tomographic imaging in the
axial plane, in addition to standard coronal scanning, can
provide important information. The axial plane clearly
demonstrates the relationship of the optic nerves and internal
carotid arteries to the sphenoid sinus walls. Computed to-
mography is helpful for defining the extent of the lesion
and for identifying local dehiscences within the sinus walls.
Although frequently associated with malignant neoplastic
processes, bony erosion may be associated with a wide va-
riety of pathologic conditions. In addition, lesions within
the sphenoid sinus that originate from adjacent structures
almost invariably demonstrate bony destruction. Computed
tomography is somewhat limited when compared with
MR imaging, and certainly angiography, in its ability
to determine the vascularity of a lesion. Biopsy in the case
of hypervascular mass can be catastrophic, and death has
been reported as a complication of diagnostic biopsy. If a
lesion is suspected to be vascular, gadolinium-enhanced
MR imaging, MR angiography, and/or conventional angi-
genish process may be obtained regarding the cause of the process.
Given the possible devastating consequences, endo-
scopic biopsy should be avoided until the workup is com-
plete and a treatment plan is in place.

Disease of the sphenoid sinus, although rare compared with
that of other sinuses, presents several diagnostic and ther-
apeutic challenges. The initial signs and symptoms of sphen-
oid sinus disease are often similar regardless of the ab-
normality. According to a review of the literature and our
experience, the most frequent presenting symptom is fa-
cial pain or headache. Cakmak et al, reporting on the larg-
est documented series of patients with isolated sphenoid
sinus disease, reported headache as the primary present-
ing complaint in 72.5% of patients. Similarly, our series
found headache in 69%. This pain is often vague and dif-
ficult to characterize regardless of the process involving
the region. Visual changes were the second most com-
mon presenting symptom in our series (21%), which is
similar to the incidence found in other series. Although
reported to be more common in malignant neoplastic pro-
cesses, visual changes occur in inflammatory and benign
neoplastic processes as well. Thus, visual changes, whether
related to decreased visual acuity or oculomotor dysfunc-
tion, do not provide reliable information with regard to
the pathologic nature of the lesion. Other cranial nerve de-
ficits, although important to document, may occur with both
inflammatory and noninflammatory lesions.

Nasal endoscopy should be performed as part of the
examination of patients with primary sphenoid sinus dis-
Table 2. Patients With Primary Tumors of the Sphenoid Sinus*

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Diagnosis</th>
<th>Presenting Symptoms</th>
<th>Endoscopic Findings</th>
<th>CT Findings</th>
<th>MR Findings</th>
<th>Treatment</th>
<th>Postoperative Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Cavernous hemangioma</td>
<td>Headache; diplopia; nasal congestion</td>
<td>Pulsatile mass extending out of sphenoid sinus into sphenethmoidal recess</td>
<td>Sinus opacification with multiple polypoid masses and bony dehiscence</td>
<td>Contrast enhancement without flow voids; extension into cavernous sinus</td>
<td>Subsellar midface degloving approach with excision of sphenoid tumor</td>
<td>No evidence of disease at 18 mo</td>
</tr>
<tr>
<td>2</td>
<td>Squamous cell carcinoma</td>
<td>Nasal congestion</td>
<td>Mass extending out of sphenoid sinus into sphenethmoidal recess</td>
<td>Sinus opacification with heterogeneous mass; bony dehiscence over carotid artery</td>
<td>Direct abutment to the carotid artery and invasion into cavernous sinus without dural involvement</td>
<td>Primary radiation therapy</td>
<td>No evidence of disease at 4 mo</td>
</tr>
<tr>
<td>3</td>
<td>Adenoid cystic carcinoma</td>
<td>Nasal congestion</td>
<td>Mass extending out of sphenoid sinus into sphenethmoidal recess</td>
<td>Sinus opacification and bony destruction anterior face of sphenoid</td>
<td>Soft tissue mass of anterior sphenoid sinus with postobstructive inflammation</td>
<td>Endoscopic resection of tumor with postoperative radiation therapy</td>
<td>No evidence of disease at 5 mo</td>
</tr>
</tbody>
</table>

*CT indicates computed tomography; MR, magnetic resonance.

Magnetic resonance imaging was helpful in identifying
the site of tumor origin and the relationship of the mass
to surrounding vital structures such as the optic nerve,
internal carotid artery, and cavernous sinuses.

A transseptal approach was used in the cases in-
volving pituitary macroadenomas. The meningioma was
resected with a combined intracranial-endoscopic trans-
nasal approach. No complications were encountered, and
all 4 patients were disease free at varying follow-up pe-
riods (Table 3).

**COMMENT**

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Vascular lesions, although rare, have been reported to occur in the sphenoid sinus and should be considered in the differential diagnosis of isolated sphenoid sinus disease. In addition to cavernous hemangiomas, internal carotid artery aneurysms, carotid-cavernous fistulas, and hemangiopericytomas have been reported to occur as isolated sphenoid sinus lesions.

In most cases a rounded or lobular mass partially filling a sinus represents a mucous retention cyst or polyp. However, given the anatomic location of the sphenoid sinus and its relationship to surrounding vital structures, a superiorly or laterally based polyp may represent an encephalocele or an internal carotid artery aneurysm. The use of MR imaging has been recommended to differentiate these entities. Although not represented in our series, CSF rhinorrhea may be associated with sphenoid encephaloceles. Magnetic resonance imaging in cases of encephalocele readily demonstrates herniated brain parenchyma, meninges, and CSF within the sinus.

A sphenoid sinus mucocele appears as an opacified, expanded sinus on CT. Magnetic resonance imaging may be helpful for differentiating mucoceles from other lesions that expand the sinus and have similar density on CT. Magnetic resonance imaging typically demonstrates a homogeneous, expansile hypointense lesion on T1-weighted images with hyperintense T2 signal. Because of variable protein content within long-standing mucoceles, signal intensity can be highly variable on both T1- and T2-weighted sequences. Magnetic resonance imaging may also demonstrate neoplastic or inflammatory disease obstructing the sphenoid ostium, which may result in mucocele formation.

Squamous cell carcinoma is the most common primary malignant neoplasm of the sinuses, but it is uncommon in the sphenoid sinus. A variety of benign and malignant tumors have been reported to arise from or secondarily involve the sphenoid sinus. Metastatic disease has been reported to involve the sphenoid sinus, with prostate, renal, and lung cancers predominating. The use of MR imaging is helpful in the preoperative evaluation of these lesions, both in terms of providing information about the possible abnormality and to determine the relationship to surrounding vital structures. Furthermore, MR imaging is superior to CT in differentiating tumor from obstructed secretions.

The transnasal surgical approaches to isolated sphenoid lesions include the endoscopic transnasal, the endoscopic transethmoid, the transseptal, and the endoscopic pterygoid fossa. Each of these may be used in conjunction with an extracranial approach if necessary. In cases involving an inflammatory process, transnasal endoscopic sphenoidotomy using the superior turbinate as a key landmark has been reported to be safe and effective. A transethmoid approach may be considered in a narrow nose or if other sinuses are involved in the disease process or wider surgical exposure may be required. A transethmoid approach may also be considered in patients with a complication of sphenoiditis to facilitate postoperative ventilation and debridement. The transseptal approach provides wider exposure of the entire anterior face of the sphenoid and allows the use of binocular vision. At our institution, the transseptal approach is most commonly used for pituitary surgery, but it can be adapted for other processes involving the sphenoid sinus when necessary. It allows the use of both hands for dissection, which is especially helpful in the resection...
of vascular tumors or lesions and the repair of CSF leaks. The angled endoscopes may also be used through this approach to enhance visualization in the lateral recesses of the sinus. The endoscopic pterygoid fossa approach is most useful for approaching lesions at the lateral aspect of the sphenoid sinus, CSF fistulas in particular. In recent years, we have used computer-assisted sinus surgery in the resection of skull base tumors and repair of skull base defects. This technology allows for intraoperative localization and is helpful when the anatomy is distorted by the disease process or previous surgery. Objective documentation of improved safety and outcome, however, has not been established, and computer-assisted sinus surgery remains an adjunct to surgical experience and anatomic knowledge.

Isolated sphenoid sinus disease occurs relatively infrequently and presenting symptoms are often vague. Headache and visual symptoms are the most common presenting symptoms and occur with almost any abnormality involving the region. Cranial nerve deficits, although more commonly reported with malignant processes, may occur with benign and nonneoplastic processes as well. Thus, when these patients are examined, the entire differential diagnosis should be kept in mind to avoid potentially devastating surgical complications. A CT scan reliably demonstrates bony erosion, which may indicate spread to adjacent structures or, conversely, involvement of the sphenoid by a lesion originating from a surrounding structure. Magnetic resonance imaging is indicated in cases of bony erosion or dehiscence or when soft tissue masses of the superior or lateral portion of the sphenoid are identified. Angiographic studies and intervention may be required if vascular lesions are suggested on CT or MR imaging studies. Therapeutic options should take into account patient desires and endoscopic and radiographic findings to optimize patient safety and outcome.

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