Rhinology, 40, 34-40, 2002

Infectious and neoplastic diseases of the sphenoid sinus – a report of 10 cases*

Zan Mra[1], Jeffrey C. Roach[1] and Allan L. Brook[2]

[1] Departments of Otorhinolaryngology-Head & Neck Surgery, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, New York, NY, USA

[2] Department of Radiology, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, New York, NY, USA

SUMMARY

Sphenoid opacifications may be discovered during the radiological work up of patients presenting with fever, headache, or neurological changes. While most of these patients do not require surgical intervention, prompt assessment and management is nevertheless required. Ten patients who underwent sphenoidotomy for drainage or biopsy at Montefiore Hospital during a 4-year period from September 1995 through January 2000 are presented. Nine out of 10 patients had predisposing factors such as AIDS, diabetes, leukemia, and end-stage renal disease. The most common presentation was altered mental status. One patient rapidly developed cavernous sinus thrombosis. Microbiology of sphenoid cultures included various fungi, Mycobacterium avium intracellulare, coagulase negative Staphylococci, and Corynebacterium. Neoplastic processes included non-Hodgkin's lymphoma and sinonasal undifferentiated carcinoma. When evaluating hospitalized patients with sphenoid sinus disease, a thorough history and a bedside nasal endoscopy should be performed. Conservative management in the form of intravenous antibiotics and topical decongestion should always be the first line of treatment. Those patients with clinical or radiological evidence of disease extending beyond the confines of the sphenoid sinus require immediate surgical intervention.

Key words: sphenoid, paranasal sinus, sphenoidotomy, infection, neoplasm

INTRODUCTION

Otorhinolaryngologists are frequently consulted for the radiologic finding of pansinusitis during the fever work up of an intensive care unit patient. The management can simply involve the removal of all tubes from the nasal cavity and topical decongestion. In selective cases, a bedside maxillary antral lavage can be performed for diagnostic and therapeutic purposes. Less commonly, sphenoid opacifications are discovered during the workup of headache, fever, or altered mental status in a hospitalized patient. When the otorhinolaryngologist is called upon, a decision needs to be made whether surgical intervention is appropriate.

MATERIALS AND METHODS

The medical records of Montefiore Hospital during a 4-year period from September 1995 through January 2000 were reviewed. Inclusion criteria included patients with a diagnosis of sphenoid sinus infection or tumor. Excluded were those patients who had involvement of all sinuses (pansinusitis), and those who did not require a spehnoidotomy for diagnosis or treatment. A total of 10 cases met the criteria. This included 5 where the author (ZM) was the surgeon. Information gathered from the medical records included the patient's age, sex, symptoms at the time of presentation, predisposing factors, management, findings from bacteriology, pathology and radiology, and outcome. These 10 cases are summarized in Table 1 and are described in detailed below. Since the number of the cases is small, a compilation of information from literature is also presented in Tables 2, 3 and 4.

RESULTS (CASE PRESENTATIONS)

Case #1:

H.E. was a 56-year-old previously healthy woman who presented to the emergency department with headache, fever, elevated white count, and a preseptal cellulitis of her left eye. She was admitted to the hospital and treated with intravenous ampicillin/sulbactam. Within 24 hours, she developed ptosis, pupillary dilation, and lateral gaze paralysis in that eye. A computerized axial tomogram (CT scan) showed bilateral sphenoid air fluid levels and bilateral superior ophthalmic vein enlargement with peripheral enhancement suspicious for cavernous sinus thrombosis (Figure 1A). Magnetic resonance imaging

* Received for publication: May 14, 2001; accepted: September 28, 2001

Footnote: Presented at the IX Congress of the International Rhinologic Society, September 21, 2000, Washington D.C., U.S.A.

Table 1. Summary of findings in 10 hospitalized patients with sphenoid sinus disease.

	Patient	Age	Sex	Presentation	Predisposing Factors	Microbiology/Pathology
1	H.E.	56	F	headache, fever	none	Cavernous Sinus Thrombosis *Mycobacterium avium intracellulare*
2	A.B.	38	M	altered mental status	AIDS	Aspergillus
3	L.B.	45	M	fever, altered mental status	AIDS, IDDM, ESRD	Candida albicans, Staphylococcus aureus
4	J.E.	63	F	fever, altered mental status	leukemia	Candida albicans
5	F.B.	56	M	altered mental status	acute renal failure	coagulase negative Staphylococcus aureus
6	L.W.	45	M	altered mental status, ophthalmoplegia, diabetes insipidus	multiple sclerosis	non-necrotizing granuloma
7	K.S.	60	F	CN-III palsy, midfacial hypesthesia	hereditary spherocytosis s/p splenectomy	sinonasal undifferentiated carcinoma
8	M.D.	33	F	diplopia, decreased visual acuity	AIDS	non-Hodgkin's lymphoma
9	C.D.	30	F	fever, altered mental status	AIDS	Corynebacterium species
10	G.S.	65	M	proptosis, headache, decreased vision,	ESRD s/p renal transplant	Rhizopus and Mucor species

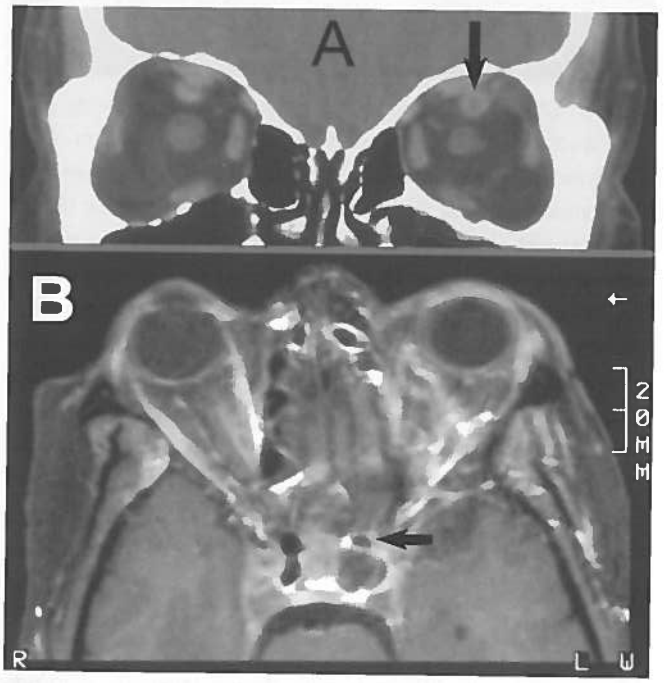

Figure 1. (A) coronal CT with contrast through the orbits denoting bilaterally enlarged superior ophthalmic veins (arrow on left one) with filling defects denoting thrombosis; (B) axial T1-weighted MRI with contrast showing cavernous sinus thrombosis bilaterally with marked narrowing of the left cavernous portion of the internal carotid artery (arrow); also note infiltrated orbital fat signifying congestion due to sinus thrombosis.

(MRI) confirmed the cavernous sinus and bilateral superior ophthalmic vein thrombosis (Figure 1B). The patient was immediately taken to the operating theatre, where the sphenoid sinuses were drained and cultures taken. Despite receiving multiple antibiotics and being heparinized, her symptoms worsened with cranial nerve 3rd and 6th palsies extending to the right eye. Follow-up MRI and magnetic resonance angiography (MRA) showed total occlusion of the cavernous portion of the left internal carotid artery (Figure 2). The left sphenoid culture eventually became positive for Mycobacterium avium complex by DNA probe. She did not test positive for the human immune deficiency virus (HIV). The patient received antimycobacterial therapy in addition to antibacterials and was switched to oral warfarin. She was discharged after at least a month of therapy with residual ptosis and extraocular muscle dysfunction of her left eye. She continues to show gradual improvement of her eye after almost a year.

Case #2:

A.B. was a 37-year-old hospitalized man with acquired immune deficiency syndrome (AIDS), who had deterioration of his mental status. A head CT showed, in addition to generalized cerebral atrophy, opacification of the sphenoid sinuses with bony erosion (Figure 3A). An MRI showed a left sphenoid mass eroding into the middle cranial fossa (Figure 3B). The patient was taken to the operating theatre, where a left sphenoid osteotomy and biopsy was performed mainly to get a diagnosis. Intraoperatively, there were no signs of necrotic

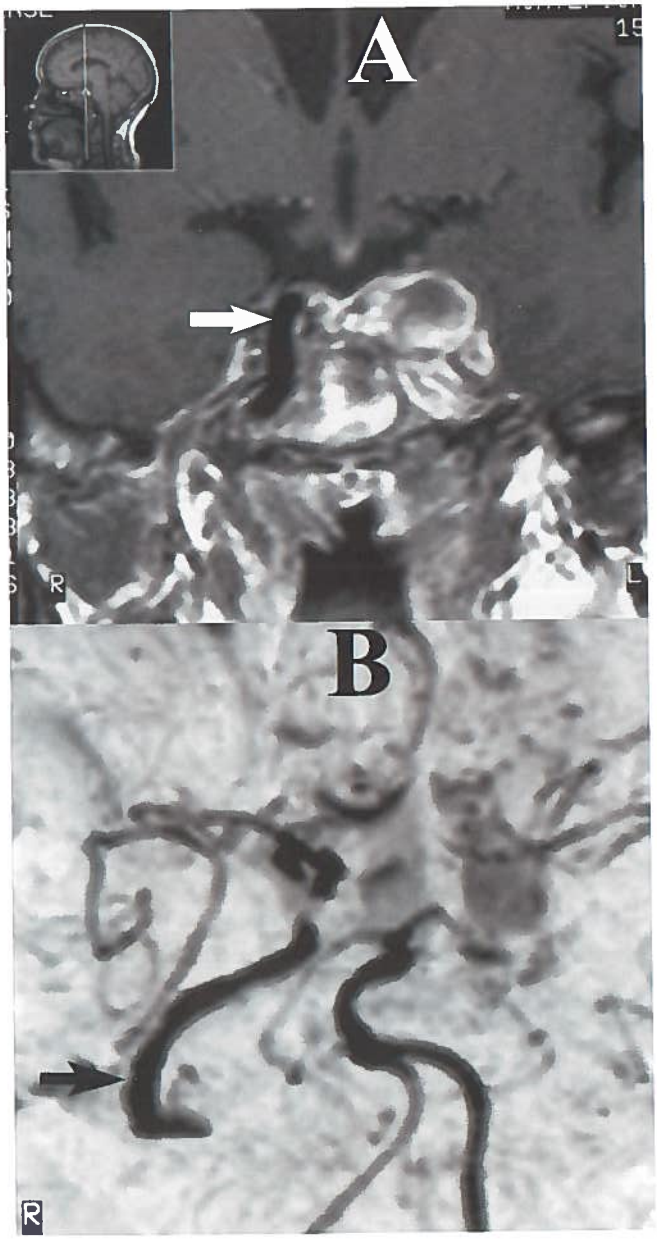

Figure 2. (A) T1-weighted coronal MRI with contrast and (B) MRA of the circle of Willis showing blood flow through the right internal carotid artery (arrow) and complete occlusion of the internal carotid artery within the cavernous sinus.

mucosa or bone. The mass appeared to be a mucocœle and was marsupialized since complete removal would have been potentially lethal. The pathology specimen was heavily laden with parallel-walled septate branching fungal hyphae, with the morphology of Aspergillus species. The patient's condition deteriorated from multiple systemic conditions and he soon expired.

Case #3:

L.B. was a 45-year-old hospitalized man with AIDS, insulin-dependent diabetes mellitus (IDDM), and end-stage renal disease (ESRD). He developed fever of unknown origin and altered mental status. CT and MRI showed air fluid level in

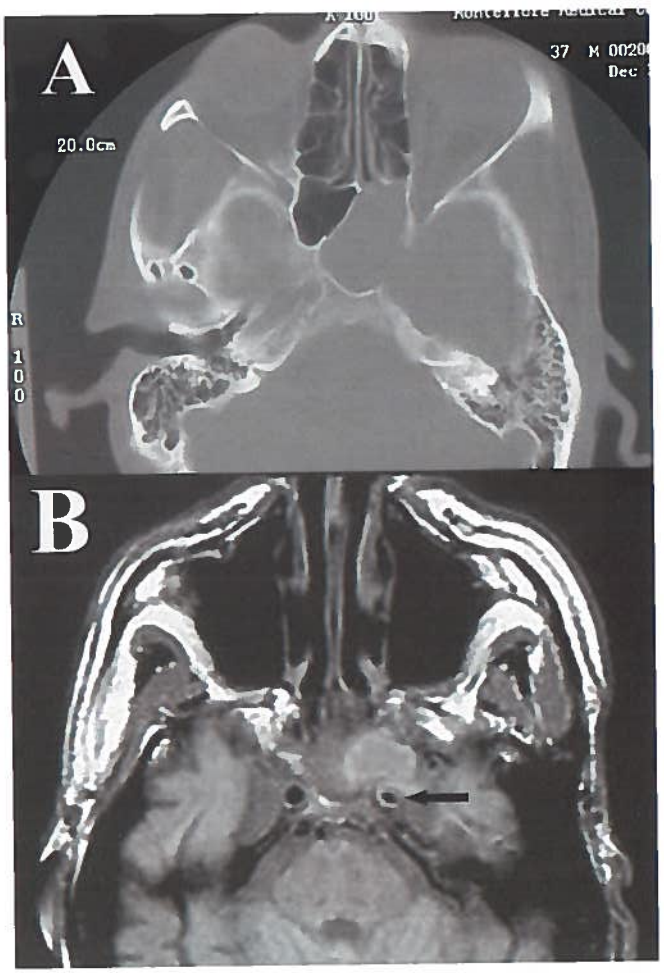

Figure 3. (A) Axial CT scan of the paranasal sinuses showing complete opacification of the left sphenoid sinus with destruction of the left lateral wall and communication into the middle cranial fossa; (B) T1-weighted axial MRI depicting left sphenoid mass invading the left middle cranial causing dural enhancement and surrounding the left cavernous carotid artery at 180 degrees (arrow); the high signal intensity is characteristic of proteinaceous material produced by fungi.

the sphenoid sinuses, which appeared to be inspissated mucus. Since the patient was already scheduled to undergo a tracheotomy, endoscopic sphenoid osteotomies were performed at the same time and cultures were sent. They showed Staphylococcus aureus and superinfection with Candida albicans. Despite appropriate anti microbial therapy, he died from systemic complications.

Case #4:

J.E. was a 63-year-old woman with chronic leukemia, who presented with waxing and waning level of consciousness and a fever of unknown origin, which did not resolve with different intravenous antibiotics. A CT scan was performed to rule out leukemic infiltration of the CNS. It showed opacification of the left sphenoid sinus. This was drained under general anesthesia. Fungal stains of the specimen were negative but Candida albicans was the final isolate. She improved on the proper therapy.

Case #5:

F.B. was a nursing home resident with multiple sclerosis admitted to the hospital for lethargy and a fever of 40.4 degrees Celsius. A head CT showed left sphenoid opacification. Purulent material was found at the time of sphenoidotomy. Cultures were positive for coagulase negative Staphylococcus aureus. The patient gradually recovered after being placed on nasal decongestants and intravenous ticarcillin/clavulanate.

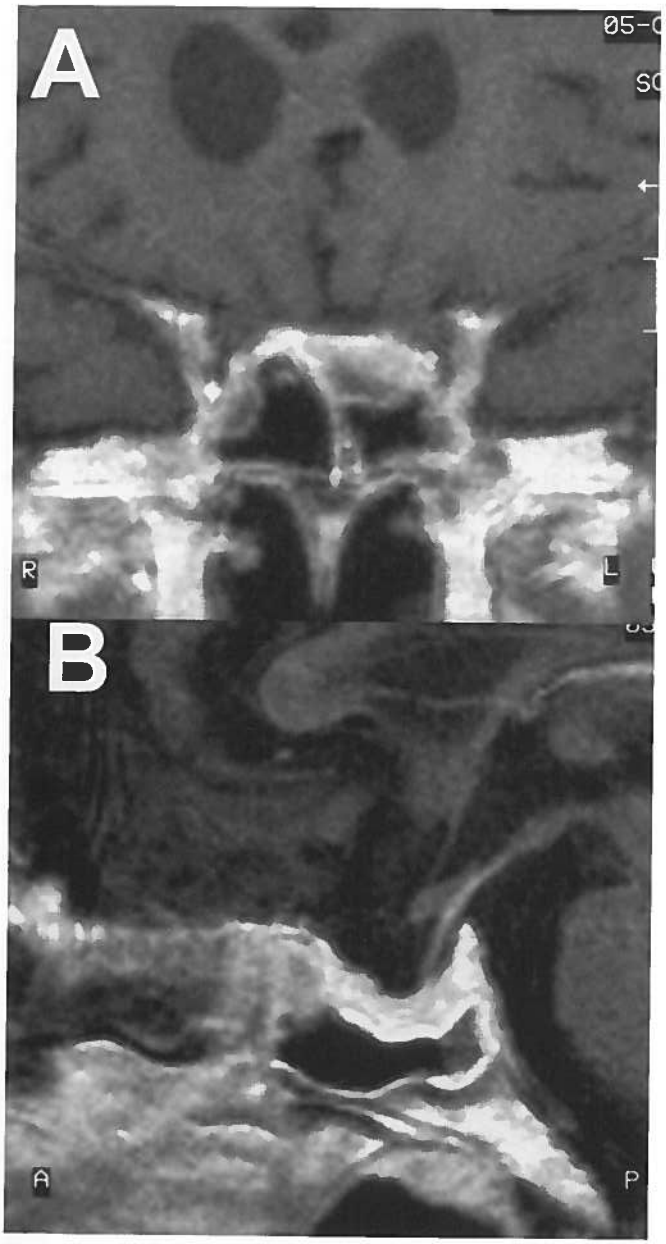

Figure 4. (A) T1-weighted coronal MRI with contrast showing inflammatory process in the left spheenoid sinus; (B) T1-weighted sagittal MRI with contrast showing decreased signal within the sphenoid portion of the clivus representing diffuse marrow replacement, and abnormal enhancement in the region of the sella, with dural enhancement also noted.

Case #6:

L.W. was a 45-year-old man with multiple sclerosis, who was hospitalized after developing mental status changes, and onset of ophthalmoplegia and diabetes insipidus. Initially, CT scan of the paranasal sinuses showed opacification of the sphenoid (Figure 4A) and posterior ethmoid cells, and he was treated with intravenous antibiotics. MRI performed later showed improved pneumatization of the sphenoid sinus but also picked up a diffuse marrow replacement of the sphenoid portion of the clivus, with abnormal enhancement and dural enhancement (Figure 4B). Endocopic left spenoidotomy and biopsy with CT guidance was performed. It showed respiratory mucosa with one non-necrotizing granuloma in the subepithelial tissue. Additional studies including cerebrospinal fluid studies failed to reveal the exact cause of the granuloma. The family elected to transfer him to a nursing home where he received supportive treatment.

Case #7:

K.S. was a 60-year-old woman who presented with headache, right eyelid ptosis, right 3rd cranial nerve palsy and right midfacial hypœsthesia. Past history was significant for breast carcinoma and congenital spherocytosis, for which she had had a splenectomy. CT Scan of the sinuses showed a right sphenoid sinus lesion with erosion of the right lateral sphenoid sinus wall and invasion of the right cavernous sinus. MRI showed the lesion to be a solid mass (Figure 5). Endoscopic right sphenoidotomy and biopsy was performed using the Instatrak® (Visualization Technology Inc.) image guidance system. Pathological diagnosis was that of a high grade carcinoma with features of sinonasal undifferentiated carcinoma.

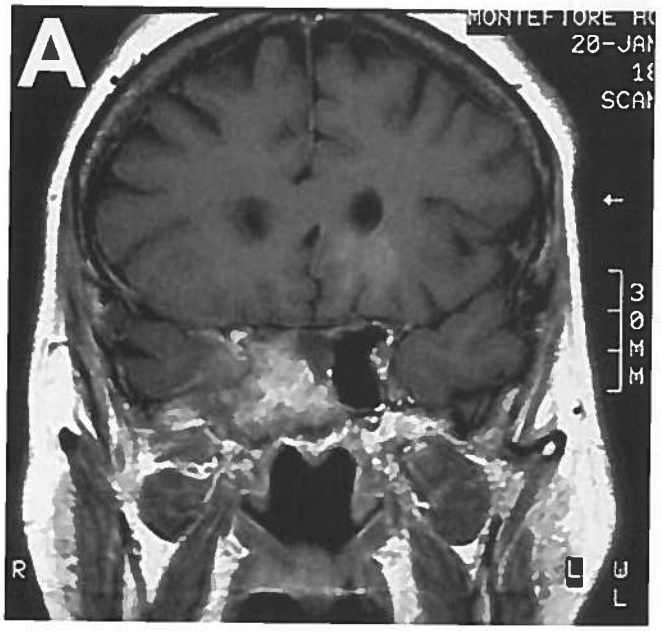

Figure 5. (A) T1-weighted coronal MRI with contrast.

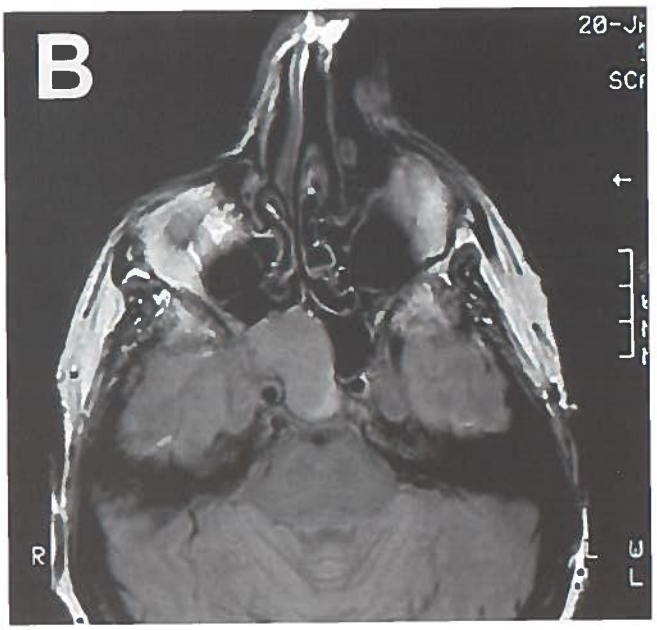

Figure 5. (B) T2-weighted axial MRI showing solid right sphenoid mass, extending into the right cavernous sinus with enhancement of the temporal lobe dura.

Case #8:
D.M. was a 33-year-old woman with AIDS, who presented with right eye ptosis, ophthalmoplegia, photophobia, decreased visual acuity and right-sided facial pain. CT Scan showed sphenoid and right ethmoid opacification with displacement of the medial rectus by soft tissue. The patient did not improve with decongestants and intravenous ceftriaxone. A right sphenoethmoidectomy and orbital decompression was performed. The tissue removed revealed a non-Hodgkin's lymphoma of the diffuse large cell variety. The patient became stable enough to be discharged from the hospital after the appropriate therapy was instituted.

Case #9:
C.D. was a 30-year-old woman with AIDS, who presented with fever and altered mental status. CT scan showed an air fluid level in the left sphenoid sinus with no bony destruction or erosion. She did not improve with decongestants and intravenous clindamycin. The sphenoid sinus was drained endoscopically. Although her blood and sputum cultures grew the more common Staphylococcus and Hemoplilus bacteria, the sphenoid culture was notable for growth Gram positive bacilli of the Corynebacterium species.

Case #10:
G.S. was a 65-year-old man transferred from a small community hospital with fever, severe headache, proptosis and echymosis of the left eye, and decreased vision in that eye. He was on immunosuppression, having received a renal transplant for ESRD 4 months ago. CT scan showed sphenoethmoid disease. Intranasal exam showed necrotic mucosa posteriorly. He was immediately taken to the operating theatre, where extensive debridement was performed. Frozen section of the specimens obtained during surgery showed tissue invasion by fungal elements. Final isolates were Rhizopus and Mucor species. The patient received intravenous amphotericin B and recovered.

DISCUSSION

Since the number of cases presented here is too small for drawing any conclusions, a literature review was included in order to better discuss isolated sphenoid disease.

Presentation: In a large series by Lawson and Reino (1997) the most common presenting symptom of sphenoid sinus disease was headache, followed by visual changes and cranial nerve palsies. Mild seizure disorders (Holt et al., 1984) and pain in the trigeminal distribution (Lew et al., 1983) have also been described. Fever is not a reliable sign since it is non-localizing. In already hospitalized patients from other causes, altered mental status may be the only manifestation.

Predisposing Factors: Since our hospital is a tertiary care center, AIDS, ESRD, and immunosuppression were the predominant factors. Contact with sea water (Holt et al.,1984; Lopes et al., 1993), maxillofacial trauma, cocaine sniffing (Lew et al., 1983), and radiotherapy to the region (Rothfield et al., 1991) have been reported to lead to infection of the sphenoid sinus.

Diagnosis: Although bedside endoscopy after adequate topical decongestion and anesthesia should always be attempted when evaluating a patient's sinuses, a CT scan and if necessary, an MRI are invaluable in excluding or assessing sphenoid sinus disease. Plain films of the sinuses are almost useless and are impractical in obtunded or intubated patients. When all other sources of fever have been ruled out, the otorhinolaryngologist is obligated to sample fluid or tissue from the sphenoid sinus for bacteriological diagnosis. If radiological assessment points towards a tumor, a biopsy should be performed promptly.

Organisms: Staphylococcus aureus and other common respiratory pathogens (Lew et al., 1983) are the usual bacteria cultured from the sphenoid sinus. Aspergillus fumigatus (Xenos et al., 1995) is the most common fungal pathogen involved. Aspergillosis can be in the form of allergic fungal sinusitis, aspergilloma, or invasive fungal sinusitis. Rhizopus and Mucor are the other lethal fungi. Since most patients have already been administered broad spectrum antibiotics soon after the onset of fever, cultures are often negative. Superinfection with Candida albicans is also likely to occur as seen in our series. Less common organisms isolated from the sphenoid sinus are listed in Table 2.

Table 2. Unusual organisms cultured from sphenoid sinus.

Fungi:	*Paecilomyces variotti*[7]
	Petriellidium (Pseudallescheria) boydii[2,8]
Chlamydia[2]	
Vibrio alginolyticus[4]	
Nocardia asteroides (acid-fast bacilli)[9]	
Corynebacteria	

Non-Infectious Etiologies: Table 3 lists the conditions that will cause radiological abnormalities in the sphenoid sinus.

Table 3. Non-Infectious Etiologies of Sphenoid Sinus Disease [10,17].

Mucocœle	
Inflammatory sphenochoanal polyp	
Inverting Papilloma	
Fibrous dysplasia	
Foreign Body	
Rhinoliths	
Encephalocœle	
Internal Carotid Artery Aneurysm	
Clival Cyst	
Neoplasms:	Benign
	Malignant
	Metastatic
Granulomatous Disease	
Pituitary Adenoma	

Complications: Due to the anatomic proximity of the sphenoid sinus to the orbit, cavernous sinus, pituitary fossa and the clivus, spread of infection or invasion by tumour can cause serious complications leading to permanent disabilities or death (Table 4).

Table 4 Complications of Sphenoid Sinus Disease [3,6,13-16,20].

Orbital Cellulitis and Abscess
Superior Orbital Fissure Syndrome
Orbital Apex Syndrome
Optic neuritis
Blindness
Cortical Vein Thrombosis
Cavernous Sinus Thrombosis
Carotid Artery Thrombosis
Hypopituitarism
Pituitary Abscess
Abscess of Clivus and Petrous Apices
Cerebral Cortical Irritation
Meningitis
Epidural Abscess
Infratentorial Subdural Empyema
Hemiplegia
Death

Management: Management will vary on the disease process, i.e., infectious vs. neoplastic. For infections, topical decongestion and appropriate antibiotics should be tried first. In the case of tumours, a rapid diagnosis by biopsy will initiate proper treatment. Various surgical approaches to the sphenoid sinus have been described in the literature. Performing an external ethmoidectomy through a Lynch incison will lead to the sphenoid sinus (Lew et al., 1983). The sphenoid sinus can also be approached via a sublabial (Rothfield et al.,1991), intranasal transseptal, transantral, or the transpalatal routes (Postma et al.,1995). Metson and Gliklich (1996) described the endoscopic approach as a safe technique in 34 patients. When the sphenoid ostium is difficult to reach due to anatomical constrictions, the ethmoid cells may need to be removed first. A transsuperior meatal approach to the sphenoid sinus has also been described (Min et al., 1995). In fact, 9 of the cases in our series were performed utilizing the endoscope. Important measurements are: the sphenoid ostium lies 7 cm from the anterior nasal spine at a 30 degree angle and 0.6-9.0 mm from the nasal septum. One should not chase the posterior ethmoid cells since anatomical variations can lead the inexperienced surgeon lateral or superior to the sphenoid sinus endangering vital structures. With the advent of image guidance systems such as the Instatrak®, open approaches are rarely warranted. However, even high-tech machines cannot replace surgical finesse since even the slightest bending of the probe or instrument in the tracking device can throw off the navigation significantly in crucial areas.

The case of the cavernous sinus thrombosis deserves separate mention since she was the only patient without any predisposing factor and the disease progressed rapidly. Although she was heparinized, there is no solid evidence that anticoagulation serves as a useful therapeutic adjunct to antibiotic therapy. Steroids also have a limited use (Kriss et al., 1996).

In summary, sphenoid sinus disease has been shown to be a serious condition especially in debilitated hospitalized patients. As experts in this area, otorhinolaryngologists should orchestrate the ordering of proper radiological studies and intervene surgically when indicated for diagnostic or therapeutic reasons.

REFERENCES

1 Andaz C, Alsanjari N, Garth RJ, Dearnaley DP (1991) Metastatic seminoma of the sphenoid sinus. J Laryngol Otol 105: 1075-1078.

2 Bikhazi NB, Sloan SH (1998) Superior orbital fissure syndrome caused by indolent aspergillus sphenoid sinusitis. Otolaryngol Head Neck Surg 118: 102-104.

3 Holt GR, Standefer JA, Brown WE Jr, Gates GA (1984) Infectious diseases of the sphenoid sinus. Laryngoscope 94: 330-335.

4 Kriss TC, Kriss VM, Warf BC (1996) Cavernous sinus thrombophlebitis: case report. Neurosurgery 39: 385-389.

5 Lawson W, Reino AJ (1997) Isolated sphenoid sinus disease: an analysis of 132 cases. Laryngoscope 107:1 590-1595.

6 Lew D, Southwick FS, Montgomery WW, Weber AL, Baker AS (1983) Sphenoid sinusitis: a review of 30 cases. N Engl J Med 309: 1149-54.

7 Lopes CM, Rabadao EM, Ventura C, da Cunha S, Corte-Real R, Melico-Silvestre AA (1993) A case of vibrio alginolyticus bacteremia and probable sphenoiditis following a dive in the sea. Clin Inf Dis 17: 299-300.
8 Marks PV, Furneaux CE (1984) Pituitary abscess following asymptomatic sphenoid sinusitis. J Laryngol Otol 98: 1151-1155.
9 Metson R, Gliklich RE (1996) Endoscopic treatment of sphenoid sinusitis. Otolaryngol Head Neck Surg 114: 736-744.
10 Min YG, Shin JS, Lee CH (1995) Trans-superior meatal approach to the sphenoid sinus. ORL 57: 289-292.
11 Postma GN, Chole RA, Nemzek WR (1995) Reversible blindness secondary to acute sphenoid sinusitis. Otolaryngol Head Neck Surg 112: 742-746.
12 Roberts SA, Bartley J, Braatvedt G, Ellis-Pegler RB (1995) Nocardia asteroides as a cause of sphenoidal sinusitis: case report. Clin Inf Dis 21:1041-1042.
13 Rothfield RE, de Vries EJ, Rueger RG (1991) Isolated sphenoid sinus disease. Head & Neck 13: 208-212.
14 Sahjpaul RL, Lee DH (1999) Infratentorial subdural empymea, pituitary abscess, and septic cavernous sinus thrombophlebitis secondary to paranasal sinusitis: case report. Neurosurg 44(4): 864-868.
15 Sethi DS (1999) Isolated sphenoid lesions: diagnosis and management. Otolaryngol Head Neck Surg 120: 730-736.
16 Hoistad DL, Duvall III AJ (1999) Sinusitis with contiguous abscess involvement of the clivus and petrous apices. Ann Otol Rhinol Laryngol 108: 463-466.
17 Sofferman RA (1983) Cavernous sinus thrombophlebitis secondary to sphenoid sinusitis. Laryngoscope 93: 797-800.
18 Thompson RF, Bode RB, Rhodes JC, Gluckman JL (1988) Paecilomyces variotii: an unusual cause of isolated sphenoid sinusitis. Arch Otolaryngol Head Neck Surg 114: 567-569.
17 Watters GWR, Milford CA (1993) Isolated sphenoid sinusitis due to pseudallescheria boydii. J Laryngol Otol 107: 344-346.
18 Xenos C, Rosenfeld JV, Kleid SM (1995) Intracranial extension of sphenoid sinusitis. Head & Neck 17: 346-350.

Dr. Zan Mra
45 Weaver Street
Scarsdale
NY 10583
USA

Tel: +1- 914-722-9295
Fax: +1-914-722-9187