CASE REPORT

Skull Base Allergic Fungal Sinusitis with Abducens Palsy in the Third Trimester

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ABSTRACT—Allergic fungal sinusitis (AFS) usually follows a slow course, but bone erosion including that of the skull base can be seen. Patients may present with intracranial extension mimicking a cranial base neoplasm. We describe a 21-year-old pregnant female initially seen at 27 weeks gestation with a complete right sixth nerve paralysis. MR imaging showed an apparent nasopharyngeal neoplasm invading both temporal lobes. Further evaluation revealed typical findings of fungal sinusitis on both CT and MR images. Biopsy results fulfilled the diagnostic criteria of AFS, and after endoscopic treatment the sixth nerve palsy quickly resolved. Although abducens palsy has been reported with acute bacterial sinusitis and with mucocoele, and limitation of eye motility has been described with orbital involvement by AFS, we are unaware of any prior reports of true cranial nerve palsy associated with proven AFS. Our patient had no evidence of mucocoele or of bacterial infection. The palsy in this case was presumably the result of compression of the cavernous sinus. The patient's pregnancy made decision making regarding imaging and surgical intervention complex. Mother and child are alive and well 31 months after surgery. (*Skull Base Surgery*, 6(4):253–258, 1996)

CASE REPORT

A 21-year-old pregnant female was referred to the neurosurgical division at 27 weeks gestation with a 2-week history of diplopia and an intracranial mass on MR imaging. Otolaryngology consultation was obtained due to suspicion of carcinoma. The patient complained of chronic nasal obstruction, rhinorrhea, frontal headaches, and seasonal sneezing and itching since childhood. Ophthalmologic examination confirmed a complete right sixth nerve paralysis (Fig. 1). Intranasally there was bilateral polyposis, worse on the left with brownish debris in the osteomeatal complex. The remainder of the neurologic and general physical examination was unremarkable. Obstetrical evaluation revealed the fetus to be

healthy and appropriately developed for gestational age. The obstetricians thought the fetus would be viable if labor were induced, but would suffer from several features of prematurity including inadequate lung development.

MR showed a large mass in the paranasal sinuses with intracranial extension bilaterally in the sphenoid (Fig. 2). The mass was very low intensity on T2-weighted images (Fig. 3). CT scan showed an inhomogeneous mass filling all the paranasal sinuses on the left, and the frontal, sphenoid, and ethmoid on the right (Fig. 4). There was obvious bone erosion of the cribriform plate (Fig. 4) and of the superior and posterolateral walls of the sphenoid sinuses with extension of the disease into the prepontine cistern and the middle and posterior cranial fossae (Figs. 5–7). White blood cell count was $9.7 \times 10^9/l$ with 5%

Figure 1. Preoperative photograph demonstrating inability to move the right eye laterally on right lateral gaze.

Figure 2. Coronal T1 MR image demonstrating central areas of low signal corresponding to surgical findings of allergic mucin surrounded by areas of high signal corresponding to inflamed mucosa at surgery. Note extension into middle cranial fossa bilaterally (arrows).

Figure 3. Coronal MR showing the characteristic signal void on T2 imaging sequence.

Figure 4. Coronal bone algorithm CT scan demonstrating erosion of the roof of the posterior ethmoid. Note the characteristic "double density" (inhomogeneous areas of high attenuation) of the opacified sinuses.

Figure 6. Axial bone algorithm CT showing destruction of the posterolateral wall of the sphenoid sinus in the region of the right internal carotid artery. (arrow = normal left internal carotid artery for comparison).

Figure 5. Coronal bone algorithm CT showing extension through the inferolateral wall of the left sphenoid (large arrow) and erosion of the superolateral wall of the right sphenoid (small arrow).

Figure 7. Axial CT with contrast demonstrating enhancement in the left middle cranial fossa (three small arrows) and extension across the petroclival synchondrosis into the right posterior cranial fossa (large arrow).

eosinophils. Allergic fungal sinusitis was strongly suspected and treatment with topical steroids and antibiotics was given.

Over the next 3 weeks the white cell count rose to $12.7 \times 10^9/l$, and the right sixth nerve palsy did not improve. Following reconsultation with Obstetrics, Neuro-

surgery, and Ophthalmology, and careful discussion with the patient regarding the risks of induction of labor, she was taken to the operating room at 30 weeks gestation and endoscopic sinus surgery was performed with fetal monitoring. It was thought that the additional time since diagnosis allowed a better chance for the baby if labor occurred and could not be suppressed. There was now a significant risk to the mother if surgery was not performed because the WBC was increasing (threatening a bacterial cavernous sinus infection). At surgery, viscid yellow-brown debris was encountered in all involved sinuses. Cytopathologic evaluation revealed eosinophilic mucin and hyphae. Total sphenoidectomy and bilateral middle meatal antrostomies were performed endoscopically. Mucosal preservation technique was used and removal of bony intersinus partitions was less complete than usual for allergic fungal sinusitis to shorten the surgery and decrease fetal exposure to general anesthesia. The primary goal was biopsy, culture, aeration, and drainage. It was anticipated that the patient would require major revision surgery after delivery. Bone erosion in the sphenoid was confirmed, but the dura was not disrupted. Tissue cultures grew *Bipolaris spicifera*. Histopathology confirmed the cytopathologic diagnosis (Fig. 8). There was no evidence of tissue invasion. No antifungal agents or systemic steroids were given. A craniotomy was not

performed. The sixth nerve palsy started to improve within 24 hours after surgery and was completely resolved at 1 week postoperatively (Fig. 9). Her headaches and nasal obstruction were also resolved. The pregnancy went to term and a healthy boy was delivered. The patient had one subsequent procedure 6 months later to open a single undrained ethmoid air cell by CT on the left, and at that time the sphenoid was well aerated. There has been complete resolution of the disease in the sinuses as well as intracranially by CT (Figs. 10, 11) and nasal endoscopy (Fig. 12). The patient is currently asymptomatic and without recurrence at 31 months after her initial surgery. She remains on topical steroids only. Her mucosa continues to have a normal appearance except for mild pallor as in Figure 12.

DISCUSSION

Allergic fungal sinusitis (AFS) is one of the four major varieties of fungal sinusitis. The other forms include acute fulminating fungal sinusitis (usually mucormycosis in immunocompromised individuals), chronic indolent fungal sinusitis, and mycetoma. Typically, AFS is slowly progressive and not aggressive, but has been shown to destroy bone of the orbit,^{1,2} and skull base³⁻⁵ in

Figure 8. Photomicrograph of sinus contents: "allergic" mucin composed of dense mucin (light background) with masses of eosinophils (dark) and intact branching septate pigmented fungal hyphae (arrows) (Fontana-masson stain, 100 $\times$).

Figure 9. Postoperative photograph demonstrating resolution of the right sixth nerve palsy.

some cases. Recently, diagnostic criteria have been clarified^{1,4} based on experience gained since the original criteria were described by Katzenstein et al.⁶ Mandatory for the diagnosis histologically are:

1. allergic mucin, characterized by eosinophilia, cellular debris, and Charcot-Leyden crystals,
2. fungal hyphae scattered in the allergic mucin,
3. absence of tissue invasion by fungus.

Figure 10. Postoperative coronal bone algorithm CT in the sphenoid sinus region.

Figure 11. Postoperative axial bone algorithm CT at level of sphenoid sinus (arrow = right internal carotid artery).

Figure 12. Postoperative endoscopic photograph. The ball tip probe is placed into the sphenoid on the left and viewed through the sphenoid sinus on the right (small arrow = optic nerve, large arrow = internal carotid artery).

Positive fungal culture has not been considered mandatory. In the past, the disease was thought to be caused by *aspergillus*.⁶ Recent cases have shown many dematiaceous fungi, which may have a microscopic appearance similar to *aspergillus*.⁷

Some authors consider nasal polyposis and characteristic findings on CT and MR imaging to be part of the diagnostic criteria as well. On MR imaging, fungal sinusitis has a characteristic low signal intensity on T1-weighted images and signal void on T2-weighted images.⁸⁻¹⁰ We have employed the term "double density" to describe the inhomogeneous opacity on noncontrast CT images that is a characteristic finding in AFS.¹¹

Peripheral eosinophilia, elevated total IgE, and asthma occur frequently, but are not present in all patients.^{1,4} At least one indicator of type I hypersensitivity (history, elevated total IgE, specific antigen identification by RAST, ELISA, or SET, or peripheral eosinophilia) is always present.⁴ Manning et al¹² have reported positive IgE RAST to fungal antigen in 16 consecutive AFS patients.

Actual tissue invasion suggests that a different form of fungal sinusitis is present (either acute fulminating or chronic indolent).

Our patient manifested all of the typical clinical, radiographic, and histologic features of AFS, but presented in a very unusual way. Otolaryngologists, ophthal-

mologists, and neurosurgeons who treat skull base lesions must be aware of this disease.

AFS must be considered in the differential diagnosis of skull base masses even when cranial neuropathy is present. This will avoid unnecessary treatment for intracranial disease such as craniotomy, dural resection, and systemic antifungal agents. As this case illustrates even the most severe cases of skull base AFS may be managed successfully with endoscopic sinus surgery and topical nasal steroids. Traditional sinus surgery may be indicated for refractory maxillary or frontal sinus disease, just as in chronic nonfungal sinusitis. Systemic steroids and/or immunotherapy may prove efficacious, but have not yet been subjected to clinical trials. Biopsy to rule out neoplasia and more aggressive forms of fungal sinusitis must be performed.

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