Allergic Fungal Sinusitis With Unilateral Eye Involvement

We describe 2 patients with ocular signs and symptoms who were subsequently diagnosed as having allergic fungal sinusitis (AFS). This disease is characterized primarily by chronic rhinosinusitis, nasal polyposis, allergic mucin, and the presence of fungal organisms according to a culture and/or histologic examination. Clinical features are those associated with chronic rhinosinusitis, which include facial pressure, nasal obstruction, and rhinorrhea. Propositis, ptoisis, and diplopia are the most common ocular symptoms; however, these conditions rarely represent the initial manifestation of the disease. The nasal mucus specimens of patients with AFS are designated as allergic mucin because of the abundance of eosinophils and their degradation products within the mucus. Although standardized treatment for AFS is not well defined, surgical debridement and systemic corticosteroid therapy are commonly recommended.

The typical patient with AFS is young and immunocompetent with a history of asthma or atopy. Orbital involvement in AFS is caused by the direct extension of sinus inflammation and can result in compressive ocular symptoms. Although well described in the medical literature, AFS has rarely been described with opthalmic involvement. We found only 5 such patients discussed in the ophthalmologic literature and take this opportunity to add our case series to this list.2,3

Report of Cases. Case 1. A 15-year-old African American boy came to our clinic with mild proptosis, pain and tearing of the left eye, and a history of having been “elbowed” in that eye 4 months previously (Figure 1). Medications included ibuprofen, fexofenadine hydrochloride, and an artificial tear preparation. Medical and family histories were noncontributory. His visual acuity was 20/20 OU. Slight hypoglobus and 5-mm proptosis were present in the left eye. The remainder of the ocular examination results were unremarkable.

Figure 1. A 15-year-old African American boy with proptosis, hypoglobus, and mild blepharoptosis of the left eye.

Downloaded From: by a Non-Human Traffic (NHT) User on 01/14/2019
addition, a left frontal sinus soft tissue abnormality was noted extending into the left anterior cranial fossa and superior left orbit (Figure 2). Initial treatment included a daily nasal preparation with fluticasone propionate and a methylprednisolone taper.

Endoscopic sinus surgery included left polypectomy, ethmoidectomy, maxillary antrostomy, frontal sinus trephination, and right frontal sinus exploration. The operation revealed thick, “claylike” fungal material and thick mucinous secretions throughout the sinuses. The sinus contents were removed and submitted for histologic examination. The patient was instructed to self-administer saline irrigation with a bulb syringe and to continue taking fluticasone. A 1-month postoperative evaluation showed resolution of periorbital edema with a return of facial symmetry.

Histologic specimens revealed polypoid fragments of respiratory mucosa and bone admixed with sheets of hyaline eosinophilic material containing numerous cellular aggregates. The respiratory mucosa was largely denuded of epithelium, and the stroma was edematous and contained a chronic inflammatory infiltrate composed predominantly of lymphocytes and plasma cells but also with patchy aggregates of eosinophils. The cellular aggregates within the mucus were composed almost entirely of intact and degranulated eosinophils. Charcot-Leyden crystals were abundant (Figure 3). Fungal elements were not visible on a routine hematoxylin-eosin stain, but numerous branching septate hyphae with degenerative features were identified within the mucus on a Grocott methenamine silver stain (Figure 4). No invasive fungal elements were noted. Microbiological cultures of the specimens were not performed.

Case 2. A 22-year-old man came to our otolaryngology clinic with a 2-year history of progressive symptoms of left eye fullness and nasal airway obstruction. His medical history was significant for mild asthma, nasal airway obstruction, and nasal polyps requiring multiple endoscopic sinus operations since age 17 years. No double vision or other visual abnormalities were noted. Medication consisted of an over-the-counter inhaler. An examination revealed mild left proptosis and slight hypoglobus. An intranasal examination revealed polyps filling both nasal cavities with extension into the nasal vestibule. The remaining physical examination findings were normal. A coronal CT scan of the head, without contrast, revealed extensive sinusitis with soft tissue densities of the frontal, ethmoid, and maxillary sinuses (Figure 5). The medial walls of the maxillary antra were obscured, and involvement of the sphenoid sinuses was also noted. Focal areas within the soft tissue densities suggested the possibility of fungal disease. The patient was given 40 mg/d of prednisone for 5 days prior to endoscopic sinus surgery.
Surgery consisted of bilateral maxillary antrostomy, bilateral total ethmoidectomy, and left frontal sinus trephination. The operation revealed thick, “claylike” fungal material and edematous polypoid mucosa. The sinus mucosa and its contents were submitted for pathologic examination. The patient was instructed to self-administer saline irrigation using a bulb syringe. Three weeks postoperatively, he noted a resolution of discharge and facial pressure symptoms. An endoscopic evaluation showed mild residual sinus edema, and treatment was instituted with a regimen of oral corticosteroids for 10 days.

The results of histopathologic examination of the sinus contents and excised mucosa were identical to those in case 1; namely, edematous respiratory mucosa and mucoid sinus contents, both containing numerous eosinophils, and fungal hyphae within the sinus contents on a Gomori methenamine silver stain (Figure 6 and Figure 7). Microbiological cultures were not performed.

Comment. Allergic fungal sinusitis was originally described by A. L. Katzenstein in 1983 and was categorized as allergic Aspergillus sinusitis. The dermatophagous family of fungi has been recognized as the most common etiology of AFS. This family includes Bipolaris species, Curvularia lunata, and Alternaria species. The mucous specimens of patients with AFS are designated as allergic mucosa owing to the marked presence of eosinophils and their degradation products, which include Charcot-Leyden crystals and a major basic protein. The hypothesized pathophysiologic mechanism of AFS is the presence of an eosinophilic reaction to fungal exposure of the sinuses. Elevated IgE levels have been noted in less than 33% of patients with AFS.

Allergic fungal sinusitis has occurred in patients ranging from age 8 to 56 years with a mean age of 26 years. This disease often occurs in patients with a marked allergic diathesis such as atopy and the presence of nasal polyps. Blunt trauma has been identified as an inciting factor for AFS. A recent study identified AFS in 93% of patients with chronic rhinosinusitis, suggesting that the disease may be more prevalent than originally estimated.

Clinical features are those associated with chronic rhinosinusitis, which include facial pressure, nasal obstruction, and rhinorrhea. Proptosis, ptosis, and diplopia are the most common ocular symptoms; however, these seem to occur late and rarely represent the initial manifestation of the disease. Epiphora has been noted, and loss of vision is rare.

Diagnosis of AFS is made primarily with a combination of characteristic findings on a CT scan or magnetic resonance imaging (MRI) that correlate with the presence of allergic mucin and positive fungi according to a culture or histologic examination. Classic sinus CT scan findings in AFS include central areas of hyperattenuation that correspond to a hypointense signal on T1-weighted MRI and a signal void with...
T2-weighted MRI. Sinus expansion and bony erosion with evidence of remodeling have also been reported. Results of fungal cultures are often negative and may be due to sampling difficulty.

Although no standard treatment for AFS has been defined, surgical debridement and systemic corticosteroid therapy are commonly recommended. Fungal desensitization with immunotherapy injection is finding an increasing role in the treatment of this disease. Corticosteroid therapy is based on similarities between the pathophysiology of AFS and those of allergic bronchopulmonary aspergillosis. Antifungal agents generally are not recommended, even in cases with intracranial extension.

Figure 6. Case 2. Edematous polypoid respiratory mucosa (bottom) and eosinophilic intraluminal mucoid material (top) comprise the pathologic specimen (hematoxylin-eosin; original magnification ×100).

Figure 7. Case 2. Eosinophils predominate in the lamina propria of the respiratory mucosa (hematoxylin-eosin; original magnification ×200).

Sequestration and Late Activation of Lenticular Candida Abscess in Premature Infants

Endogenous intraocular Candida infection typically presents as chorioretinitis with varying degrees of vitreous infiltration and inflammation. In patients with concurrent or recent candidemia, intralenticular fungal abscess is rare but has been reported previously in premature infants. We report a fourth and fifth case of this unusual syndrome, emphasizing its clinical signs, peculiar clinical course, and the possibility of a good visual outcome with appropriate therapy.

Report of Cases. Case 1. An infant girl was born at 24 weeks postconceptual age and weighed 750 g. The postnatal course was complicated by bronchopulmonary dysplasia, anemia, patent ductus arteriosis, hypertension, gastrointestinal dysfunction, and recurrent pneumonias. Bilateral chorioretinitis developed at 1 week of age. Computed tomography and magnetic resonance imaging of the brain revealed a large subependymal astrocytoma. The child died at 4 weeks of age. Microscopic examination of the right eye revealed an intraocular candida abscess. The left eye was normal. Intraocular Candida infection is rare in premature infants, and preferential visualization of the right eye explains the survival of the patient.

Case 2. A 3-month-old girl was born at 30 weeks gestation and weighed 1300 g. Chromosomal analysis was normal. She was delivered by cesarean section and treated for respiratory distress syndrome. The initial course was complicated by anemia, patent ductus arteriosis, and seizures. At 3 months of age, she began to develop vomiting and irritability, and a head MRI revealed a large subependymal astrocytoma. The patient was discharged home, and 2 weeks later, she developed left eye proptosis and photophobia, and her vision decreased to hand movements. A head MRI revealed a large subependymal astrocytoma that had grown significantly. The patient was rehospitalized and treated for meningitis and had an infectious workup. Cultures of her blood, urine, and cerebrospinal fluid were negative. Intravenous antifungals were initiated. The patient was discharged home, and a few days later, she returned with the same symptoms and was treated with oral antifungals. Two weeks later, she developed right eye pain, irritability, and visual loss. The patient was admitted to the hospital, and her vision decreased to light perception. A fundus examination revealed a white lesion in the right eye. A biopsy of the lesion revealed an intraocular candida abscess in a prematurity infant, and the patient was treated with intravitreal antifungals. The patient was discharged home with normal vision.

Blake N. Geren, BS
Harry H. Brown, MD
H. Graves Hearnsberger III, MD
Christopher T. Westfall, MD

This study was supported in part by unrestricted grants from Research to Prevent Blindness, New York, NY, and the Pat and Willard Walker Eye Research Center, Little Rock, Ark.

Correspondence: Dr Westfall, Department of Ophthalmology, Mall Slot 523, 4301 W Markham St, Little Rock, AR 72205-7199 (westfallchristophert@uams.edu).