Allergic Fungal Sinusitis Presenting as Vision Loss and Strabismus in a Teenage Girl

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A 16-year-old girl with a medical history significant for chronic allergies and asthma presented to the emergency department (ED) for strabismus and visual changes.

History

The patient had been in her usual state of health until approximately 2 months prior to arrival, when she noted the development of sinus pressure. Approximately 1 month later, she began to experience slight strabismus, followed by worsening vision 1 to 2 weeks later. The patient noted that when her left eye was covered, she was unable to see with the right eye. Magnetic resonance imaging (MRI) was done at an outside facility, which detected a right frontal sinus mass impinging on the optic nerve. The patient was then emergently transferred to our ED for management.

Physical examination.

On physical examination, the patient's vital signs were within normal limits. She was in no acute distress and was not ill-appearing. An enlarged nasal polyp was noted in the right lateral nostril. Mild exotropia of the right eye also was noted. The pupils were unequal but reactive to light. A grade 3+ relative afferent pupillary defect was present in the right pupil. The right eye tracked and was unable to focus when the left eye was covered. In the right eye, visual acuity was impaired (hand motion was detected, along with counting fingers in a small temporal area). Color vision and red saturation were unable to be tested as the patient's vision was so poor. In the left eye, visual acuity was 20/20. The remainder of the examination findings were unremarkable.

Diagnostic tests.

Upon arrival to the ED, the patient's MRI scans were reviewed, and a computed tomography (CT) scan of her orbits was obtained, the results of which showed mass effect in the right sphenoethmoidal sinus complex and right clinoid process, both of which were expanded with hyperdense material (**Figures 1 and 2**). Multiple erosions were also noted involving the medial orbital wall, the roof and medial wall of the right anterior clinoid process, the bilateral optic nerve canals, the right superior orbital fissure, and the bilateral

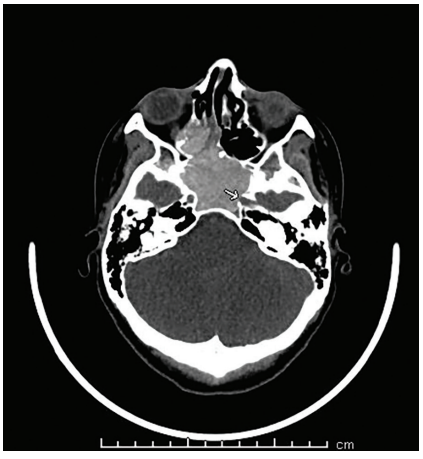

Figure 1. Transverse-view maxillofacial CT image without contrast showing homogenous opacification of hyperdense material filling the ethmoid and sphenoid sinuses.

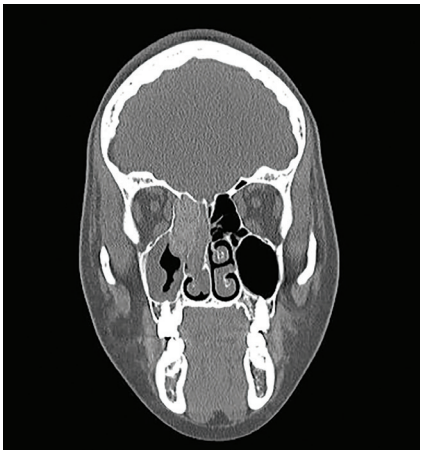

Figure 2. Coronal-view maxillofacial CT image without contrast showing expansive hyperdense material filling the paranasal sinuses, as well as erosion of bone walls.

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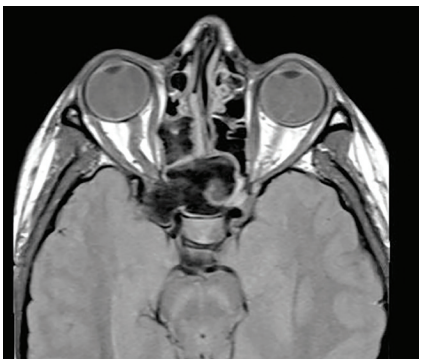

Figure 3. Axial-view T2-weighted MRI image of the brain showing compression of the right optic nerve.

foramen rotundum, carotid canals, and Vidian nerve canals (**Figure 2**). Additionally, the right optic nerve was noted to be compressed (**Figure 3**).

Differential diagnosis.

It is quite uncommon to find a mass in the nasal sinuses of a pediatric patient. The differential diagnosis includes benign vs malignant tumor, mucosal disease, cyst, infectious etiology, or congenital anomaly. It is also important to rule out a number of common disorders in a pediatric patient who presents with visual changes and evidence of sinus-related disease. Orbital cellulitis can develop as a complication of acute bacterial sinusitis and can present with vision changes. This was less likely with our patient, since she had no fever and lacked the typical examination findings associated with orbital cellulitis (periorbital edema, erythema, and tenderness). Furthermore, on CT scans, orbital cellulitis would present with evidence of inflammation rather than a hyperdense mass in the nasal sinuses.

CT imaging is critical to further narrow the differential diagnoses. In this patient's case, the hyperdense material seen on imaging ruled out tumor, mucosal disease, and cysts, since these processes typically appear hypodense. The hyperdensity of the mass suggested a process involving blood, inspissated mucus, or fungus. Given the patient's history of chronic allergies, coupled with the imaging findings of an expansive hyperdense mass, the differential was narrowed to a chronic fungal process with an allergic component, most likely allergic fungal sinusitis (AFS). Invasive fungal infection was also on the differential, although it was unlikely given the patient's immunocompetency and lack of fever or other systemic symptoms.

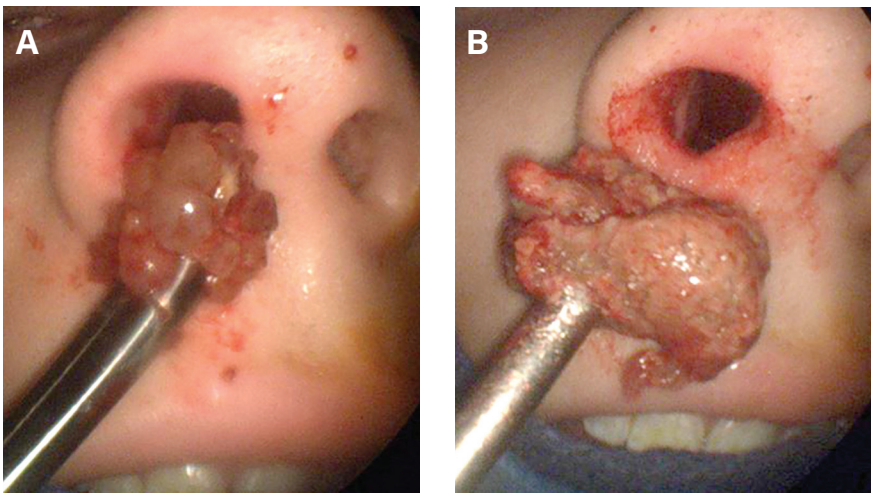

Figure 4. Photos of mucoid material removed from the sinuses during functional endoscopic sinus surgery.

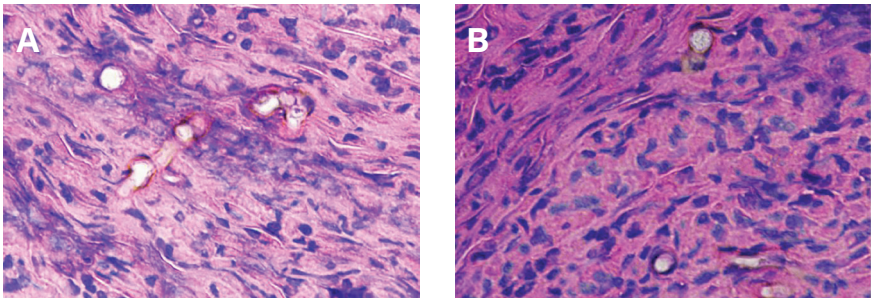

Figure 5. Pathology test results of specimens from the nasal septum showing eosinophils, allergic mucin, and fungal hyphae elements

Hospital course.

Upon admission, intravenous-combined ceftriaxone and dexamethasone was started, and the patient was immediately scheduled for functional endoscopic sinus surgery. During the procedure, polyploid material with fungal elements, as well as allergic mucin-like material described as very thick and tenacious, were removed from the sinuses (**Figure 4**). The optic nerve was noted to be partially dehiscent before proceeding with bony work. The surgeon theorized the optic nerve was susceptible to compression by fungal elements likely due to the dehiscence that was noted during the procedure. Once the optic nerve was able to be fully visualized, it was noted to have not been invaded by fungal elements. The nerve was decompressed successfully and was noted to look pink and healthy. Multiple specimens were sent to the laboratory for culture and staining.

Pathology test results of the surgical specimens showed polypoidal mucosa with overlying allergic mucin, consistent with the diagnosis of AFS (**Figure 5**). Cultures grew Bipolaris spp. The patient was subsequently stable for discharge and sent home with an oral steroid taper and voriconazole.

Discussion

AFS is a rare disorder that typically presents with rhinorrhea, facial pressure, and congestion. The disorder occurs when an allergic response occurs to otherwise benign inhaled environmental fungi. The allergic response is generated by type 2 helper T cells and results in the production of immunoglobulin E, mast cell degranulation, and the recruitment of eosinophils that further attack the antigen and release even more mediators, eventually resulting in the formation of allergic mucin and accumulation of debris that fills the sinuses.

Most reported cases of AFS have been in the United States, particularly in the South around the Mississippi River basin (which is where this patient's case occurred). AFS is typically treated with a course of glucocorticoids and, in some cases, endoscopic sinus surgery to remove the mucin and debris.[1] Systemic antifungal therapy is controversial, since there is a paucity of data supporting its benefit, although it may be beneficial in certain circumstances.[2]

In a study published in 2006, the various clinical presentations were evaluated in 20 children with AFS over 12 years.[3] In the vast majority of cases (90%-100%), the children had presented with typical nasal symptoms, nasal polyps, sinusitis, or atopy. In addition, the researchers found that in children, proptosis is a more common clinical presentation compared with adults, with approximately 50% of the children in the study presenting with it.[3] Other studies have echoed these findings, showing that children are more likely to present with obvious facial skeletal abnormalities, usually proptosis, compared with the adult population.[4] However, it is difficult to find reports investigating the incidence of optic neuropathy secondary to AFS in children, although some have been published on the adult population. In these studies, the various proposed mechanisms for the associated vision loss have been secondary to direct or indirect compression, secondary to inflammation, and secondary to increased orbital pressure.[5] In our patient, given the imaging findings and the lack of inflammation seen around the optic nerve, the vision loss was likely due to a compressive effect.

Furthermore, strabismus secondary to AFS has not previously been reported in the literature to our knowledge, which introduces another clinical presentation of this disorder. The only relevant article found during a literature review dates to 2004, in which the authors demonstrated a possible relationship between sinus-related disease and strabismus.[6] The proposed mechanism is that the sinus mass led to inflammation of the sinuses, which may have led the extraocular muscles to contract and cause strabismus.[6]

Outcome of the case.

Although the patient initially mentioned some subjective improvement in vision after having undergone functional endoscopic sinus surgery, no improvement has been noted objectively since discharge. The patient has followed up with numerous providers since discharge, all of whom have noted a persistent afferent pupillary defect, as well as poor vision (hand motion only). A recovery in vision is not expected at this time. The persistence of the visual defect could be due to the chronicity of the presenting illness.

In summary, this case demonstrates a rare clinical presentation of vision loss and strabismus secondary to AFS in a pediatric patient and highlights the importance of early detection and treatment to prevent irreversible vision loss.

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