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Case report

Invasive Streptococcus viridans sphenoethmoiditis leading to an orbital apex syndrome

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ABSTRACT

Purpose: Orbital apex syndrome due to spread of infectious sinusitis is a serious disease, often with an insidious presentation with few ophthalmic signs and symptoms. Failure to recognize and treat infectious orbital apex syndrome early portends a grave prognosis, including profound, permanent visual loss and potentially death. Herein we describe a representative case and discuss the relevant aspects of prompt diagnosis and treatment.

Observations: An unusual case of infectious orbital apex syndrome due to contiguous spread of Streptococcus viridans sphenoethmoiditis in a hospitalized, immunosuppressed patient with acute myelogenous leukemia is presented. Given the few clinic signs and subtle imaging findings, a delay in diagnosis occurred resulting in vision loss to light perception and internal carotid artery occlusion within the cavernous sinus. A brief literature review of orbital apex syndromes is presented.

Conclusion and importance: A high clinical suspicion for orbital apex syndrome must be maintained in the appropriate circumstance given the subtle clinical signs and imaging, as well as the potential devastating morbidity of the disease process. Prompt diagnosis and treatment is crucial to patient survival and preservation of vision.

1. Introduction

We present an unusual case of orbital apex syndrome due to sphenoethmoid sinusitis and review the relevant literature. Orbital apex syndrome due to local spread of an adjacent sphenoethmoid sinusitis is an uncommon, life threatening condition. Diagnosis can be difficult as patients often have very subtle symptoms, as well as a lack of common clinical and radiological signs seen in other orbital processes until the disease is advanced.

2. Case report

A 68-year-old Caucasian female with acute myelogenous leukemia (AML) was admitted to a community hospital with low white blood cell count, weakness, fatigue, and rectal pain and bleeding. The patient was also noted to have a strong history of migraine headaches. Chemotherapy with Idarubicin and Cytarabine were initiated.

15 days into her admission and chemotherapy treatment she developed intense, sharp right-sided headaches. The neurology service felt this was consistent with her history of migraines and there was some response noted with a migraine cocktail. Lumbar puncture was considered but not done due to the patient's low platelet count. Neurology suggested a contrasted magnetic resonance imaging (MRI) study of the brain, but this was not initially performed.

The patient continued to have severe headaches for 5 additional days, after which time neurology noted some possible mild ptosis of the right eye. They also believed she may have had some very mild transient periorbital edema of the right eye. At this time both an MRI brain, as well as a computed tomography angiogram (CTA) of the head and neck were ordered, both of which were noted to be relatively unremarkable other than some abnormal mucosal thickening of the sphenoid and ethmoid sinuses.

The patient's severe right sided headaches and ptosis persisted for two more days, at which point both a community comprehensive ophthalmologist and a stroke specialist were consulted. The community ophthalmologist noted no abnormalities other than a mild right-sided ptosis. The stroke specialist noted right-sided

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ptosis with pain to palpation of the right eye. A CTA head was then repeated. No stroke was noted, however, there was interval change in the thickening of the mucosa of the sphenoid and ethmoid sinuses, as well as opacification of the sinuses noted (Fig. 1). The stroke specialist performed angiography to rule out a carotid cavernous fistula, as well as a CT venogram (CTV) of the brain, and lumbar puncture, all of which were negative. She was subsequently discharged with continued headaches and ptosis of the right eye.

Two weeks later the patient presented to a university retina clinic with light perception vision of the right eye. She was also noted to have a cranial nerve III and VI nerve palsy of the right eye with subtle proptosis. The patient was seen by the orbital surgery service and underwent emergent orbital computed tomography (CT) imaging. She was found to have interval increase in the mucosal thickening and opacification of her sphenoethmoid sinuses with erosion through the bone into the orbital apex (Fig. 2). MRI further indicated erosion into the intracavernous portion of the internal carotid artery, with resultant thrombosis (Fig. 3).

Otolaryngology was consulted and the patient underwent prompt sinus drainage. The patient was treated with broad antibiotic coverage with IV Vancomycin, Piperacillin-Tazobactam, Amphotericin B, and Voriconazole. Cultures grew Streptococcus viridans. All other cultures, including fungal cultures, were negative. Her antibiotics were narrowed to Piperacillin-Tazobactam alone, then to ampicillin-sulbactam after culture results were finalized. The patient remained light perception with a mild right cranial nerve III and VI palsy. Several months later she passed from unrelated complications of her malignancy.

3. Discussion

Orbital apex syndrome is rare and has a broad differential diagnosis, including inflammatory, infectious, vascular, or neoplastic conditions. By definition this syndrome involves the optic nerve, with variable involvement of cranial nerves IV, VI, and V1. Patients with orbital apex syndrome can present similarly but should be differentiated from those with syndromes of the superior orbital fissure or cavernous sinus, both of which do not involve the optic nerve. The latter characteristically involves early cranial nerve VI involvement and lower facial hypoesthesia due to the anatomy of the cavernous sinus with the centrally located VI nerve and V2 branch respectively.[1]

While orbital apex syndrome is very uncommon, those due to the adjacent spread of sphenoethmoid sinusitis are exceedingly rare, although there are a few case reports and series in the literature.[2–5] The thicker sphenoid bone and more adherent periorbita posteriorly make this much less likely compared to the more commonly seen anterior orbital cellulitis from adjacent sinus invasion.[2] As was seen in this case, when this rarely occurs the patients are often immunocompromised. As such, ruling out invasive fungal disease is very important, as this has both therapeutic and prognostic implications.[6]

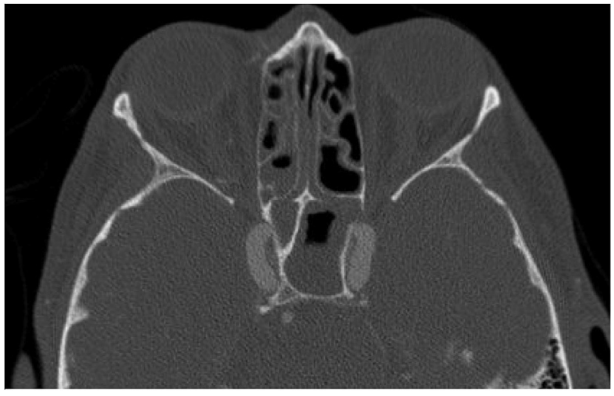

Fig. 1. Axial contrast-enhanced computed tomography displayed on bone window shows mucosal thickening throughout the paranasal sinuses, with multiple fluid levels indicating acute bacterial sinusitis. The walls of the sinuses are intact.

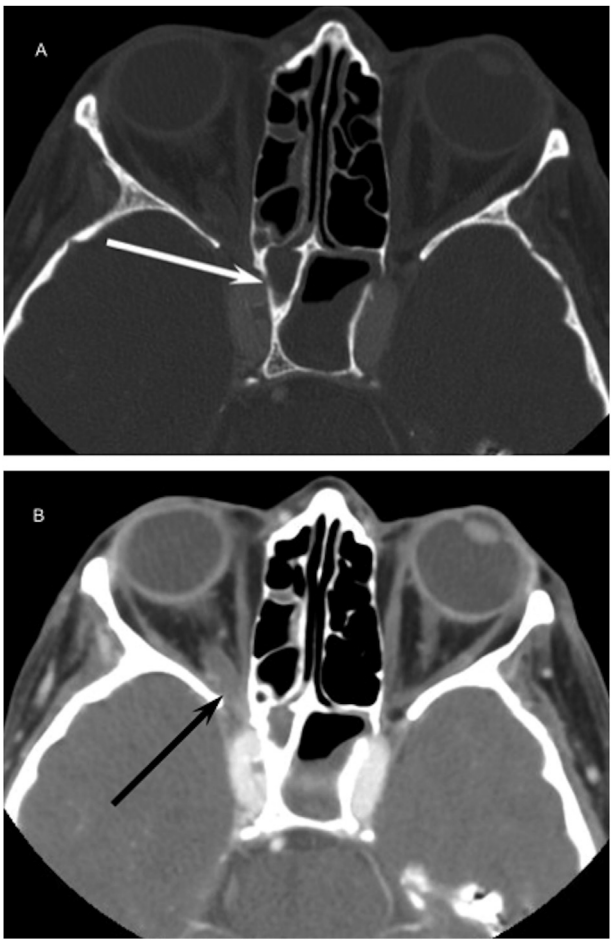

Fig. 2. Axial contrast-enhanced computed tomography displayed on bone window. A. shows a new focus of bone rarefaction (arrow) along the lateral wall of the right sphenoid sinus, overlying the cavernous segment of the internal carotid artery. B. Soft tissue window shows infiltration of the fat planes at the apex of the right orbit (arrow). Compare to the intact fat planes at the apex of the left orbit.

For both orbital apex syndrome due to bacterial and fungal infection, prompt identification of the process, with immediate broad antibiotic coverage, is essential for patient survival and possible visual and cranial nerve recovery. Delay in identification can increase the probability of permanent visual loss, which is exceedingly high in the reported cases, as well as increasing the incidence of mortality.[1,2,6] Neither corticosteroids nor orbital decompression have been shown to be effective.[1,2]

4. Conclusion

Given the potential morbidity and mortality, as well as the subtle presentation of the disease, physicians must have a high index of suspicion for orbital apex syndrome. This is especially true in immunocompromised patients. While this is essential for the orbital surgeon, other practitioners of facial and periorbital surgery should also familiarize themselves with the pattern and be prepared to consult orbital surgery, otolaryngology, or neurosurgery colleagues as appropriate.

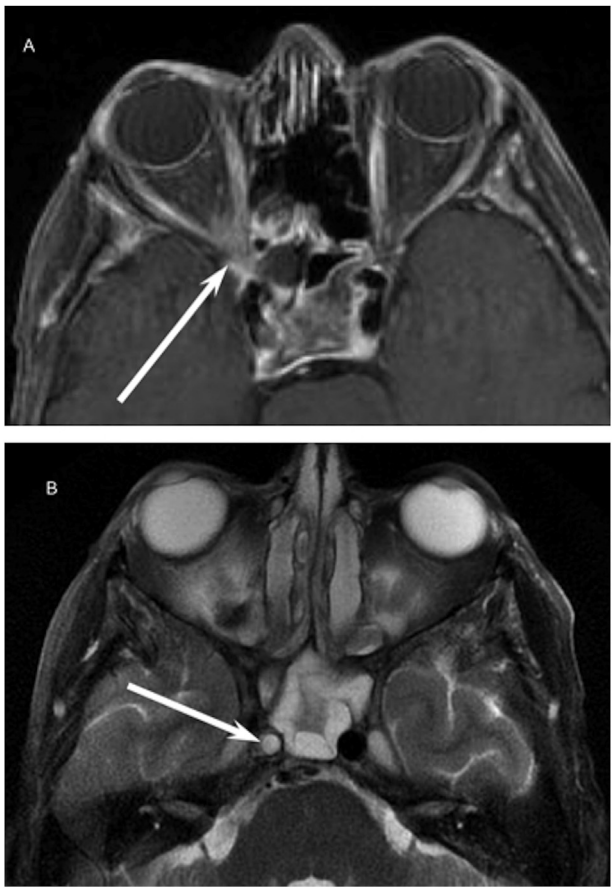

Fig. 3. Magnetic resonance imaging (MRI) showing erosion into the internal carotid. A. Axial fat-suppressed contrast-enhanced T1-weighted MRI shows enhancement at the right orbital apex (arrow), extending back into the cavernous sinus. B. Axial T2-weighted MRI shows normal flow void in the left internal carotid artery. Bright signal in the right internal carotid artery (arrow) suggests thrombosis.

4.1. Patient consent

Informed consent was obtained orally from this patient for publication of her case details. This report was conducted with IRB approval. All information was gathered with the consent of the patient, consistent with HIPAA.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

The following authors have nothing to disclose- Lance Bodily, Jenny Yu, Dante Sorrentino, Barton Branstetter.

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