

CASE REPORT

Orbital pseudotumor as a result of chronic sinusitis in an HIV-positive patient

Mitchell R. Gore 

Department of Otolaryngology, SUNY-Upstate Medical University, Physicians Office Building North, Suite 4P, 4900 Broad Road, Syracuse, New York 13215

Correspondence

Mitchell R. Gore, Department of Otolaryngology, SUNY-Upstate Medical University, Physicians Office Building North, Suite 4P, 4900 Broad Road, Syracuse, NY 13215. Tel: +1 315 492 3741; Fax: +1 315 492 5791; E-mail: mgoremdphd@gmail.com

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Introduction

Orbital pseudotumor or idiopathic orbital inflammatory disease is an idiopathic mass-like conglomeration of soft tissue involving the orbit [1]. It can present in any portion of the intraorbital contents and often involves extraocular muscles, causing proptosis and extraocular muscle restriction. Conditions such as orbital malignancy, congenital masses, infectious disease, and occult or past trauma may mimic orbital pseudotumor and must be considered in the differential. Orbital pseudotumor is often responsive to immunosuppressive therapy but may recur especially if immunosuppressive medications are discontinued. This report examines a case of orbital pseudotumor in an HIV-positive patient with the orbital inflammatory disease adjacent and ipsilateral to severe left chronic pansinusitis.

Case Presentation

A 56-year-old male with a medical history of HIV infection, type II diabetes, and hypertension presented with a 2-month history of headaches. He described the headache

Key Clinical Message

Orbital inflammatory pseudotumor is an infrequent lesion that may mimic malignancy and may be associated with autoimmune disease. Orbital pseudotumor is in the differential in any patient with an orbital mass, especially in the setting of chronic infection or immunosuppression. This HIV-positive patient presented with chronic sinusitis and orbital pseudotumor.

Keywords

Chronic sinusitis, endoscopic, HIV, orbital pseudotumor.

as occurring in the left frontal/temporal area as an intermittent stabbing pain. He also noticed twitching in the left eye. He stated that the headache was present during the day and prominent at night. The eye examination showed normal visual acuity, and the extraocular muscle examination showed mild left medial gaze restriction. The eye examination also showed mild left proptosis. Head CT without contrast and maxillofacial CT with contrast (Fig. 1A and B) were ordered that showed a round, circumscribed left intraorbital mass projecting into the center of the orbit measuring 28 × 18 × 19 mm with associated mass effect on the left optic nerve and left extraocular muscles. The mass appeared inseparable from the left medial rectus muscle and the left lamina papyracea. No lymphadenopathy was demonstrated in the neck. Interestingly, the CT scans also demonstrated left pansinusitis and an incidental massive left middle turbinate concha bullosa. The differential was noted to include neoplasms such as lymphoma/lymphoproliferative lesions, sarcoma or metastasis to the left medial rectus muscle, and ethmoid mucocele. An MRI (Fig. 2A and B) was ordered that showed a T1 + contrast enhancing left intraorbital mass with T2 hyperintense sinusitis on the

left. The patient underwent endoscopic sinus surgery and endoscopic trans-lamina papyracea biopsy of the mass. Intraoperatively, the left paranasal sinuses were noted to be filled with yellow purulent material and bulky sinonasal polyposis, including a large left middle turbinate concha bullosa filled with purulent debris. After endoscopically removing the left lamina papyracea, a white, fibrous medial intraorbital mass was noted (Fig. 3), and generous biopsies were taken, while staying medial to the left medial rectus muscle. Intraoperative frozen sections and routine pathology of the sinus polyposis showed benign inflammatory sinonasal polyps with associated chronic sinusitis. The left intraorbital mass showed benign chronically inflamed fibrous tissue with no atypia or malignancy identified, consistent with an inflammatory pseudotumor. Culture of the purulent material showed no growth.

Clinical Course

After review of the culture and pathology, the patient was treated with doxycycline for 21 days for the chronic sinusitis and a high-dose prednisone taper for the inflammatory pseudotumor. Complete excision of the pseudotumor was not attempted due to the close proximity of the medial rectus to the pseudotumor and given the benign nature of the mass. The patient had some mild left preseptal swelling that improved with the doxycycline and a high-dose steroid taper (60 mg tapered over 12 days), and had improvement in his headaches and chronic sinusitis symptoms. The patient had some recurrent left medial gaze restriction and headaches and was referred to Rheumatology and started on maintenance suppression with 20 mg of prednisone

and 50 mg of azathioprine daily with resolution of his symptoms.

Discussion

This patient presented with an infiltrative left orbital mass with CT and MRI appearance worrisome for primary malignancy. Lymphoma or sarcoma, among other neoplastic etiologies, was considered in the differential. The differential also includes Tolosa–Hunt syndrome (a related condition with involvement of the cranial nerves in the cavernous sinus and resulting ophthalmoplegia), thyroid-associated orbitopathy, orbital sarcoidosis, orbital metastases, orbital cellulitis, and orbital venous malformation. Colletti and Deganello [2] reviewed venous malformations of the orbit, formerly known as cavernous hemangiomas, which are the most common vascular lesion of the orbit in adults. They noted that newer nomenclature for classification of vascular anomalies recommends discontinuing the use of the older, historical term cavernous hemangiomas, and referring to these slow flow vascular lesions as orbital venous malformations. Orbital venous malformations account for approximately 5% of all orbital tumors, typically present in middle-aged females, and may present as a slowly growing orbital mass. If orbital venous malformations cause no visual symptoms, they can often be surveilled with periodic MRI. If symptomatic or expansive, they can be surgically removed.

While malignancy should always be ruled out, pseudotumor should be considered as a possible etiology, especially in cases of immunosuppression or chronic inflammatory disease. Orbital pseudotumor is an idiopathic inflammatory mass most commonly affecting the

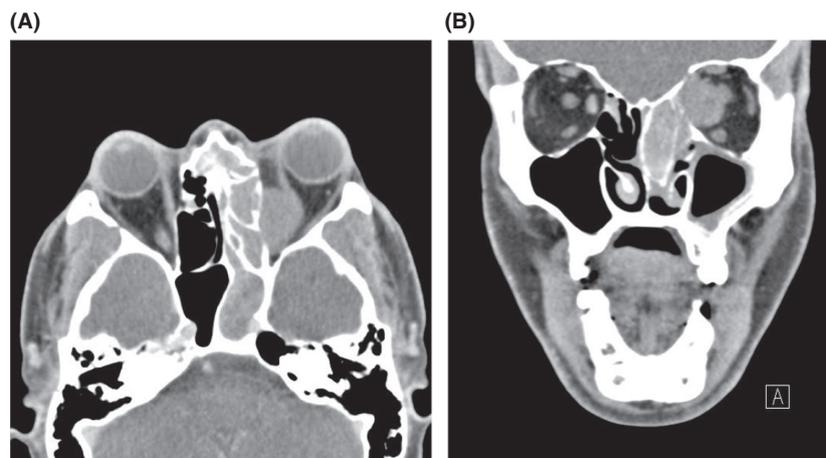


Figure 1. Axial (A) and coronal (B) computed tomography images of left orbital pseudotumor. Note the intranasal sinusitis along with the adjacent intraorbital pseudotumor.

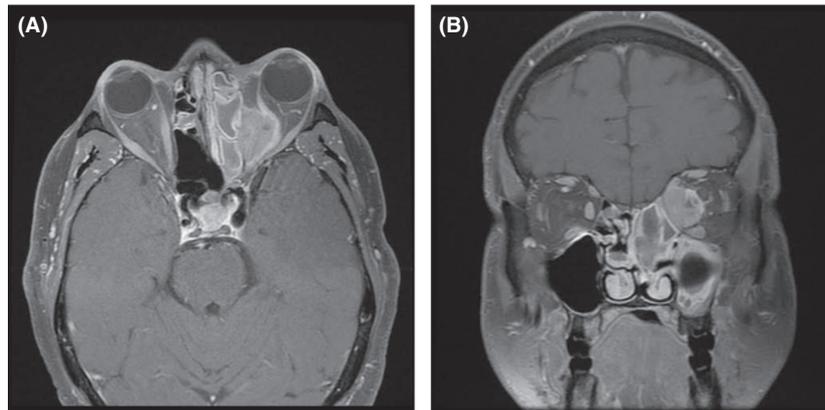


Figure 2. T1-weighted axial (A) and coronal (B) images of left orbital pseudotumor. Note the medial bowing of the left medial rectus caused by the orbital pseudotumor, as well as the intranasal sinusitis adjacent to the intraorbital pseudotumor.



Figure 3. Endoscopic view of the left medial orbital mass using a 45° endoscope. Note the orbital fat in addition to the pseudotumor mass.

extraocular musculature. It may also cause inflammatory change in the uvea, sclera, lacrimal gland or the soft tissues surrounding the globe/orbit. Onset is usually rapid and unilateral, and it can cause pain, proptosis, and double vision. The presence of an associated comorbid inflammatory or autoimmune condition may help narrow the differential to orbital pseudotumor.

Image guidance was instrumental in this case to ensure that biopsies were taken from the substance of the mass, making false-negative diagnosis less likely. Leo et al. [3] investigated the possible role of Epstein–Barr virus (EBV) in idiopathic orbital inflammatory pseudotumor (IOIP) in Caucasian patients, but found no EBV DNA in blood or orbital tissue samples of patients with IOIP. Rico et al. [4] noted a case of inflammatory orbital pseudotumor in a patient with systemic lupus erythematosus with gaze restriction and thickening of the superior rectus that

resolved with high-dose corticosteroids followed by suppressive therapy with Rituxan. Similarly, Brunelle et al. noted a case of orbital inflammatory pseudotumor due to sarcoidosis [5]. Orbital inflammatory pseudotumor has also been noted as an extra-abdominal manifestation of hepatitis C infection [6] and as a result of rheumatoid arthritis [7]. Interestingly, Li et al. [8] noted an ~30% concomitance between orbital pseudotumor and sinusitis in their group of 46 patients with IgG4+ dacryoadenitis subtype orbital inflammatory pseudotumor. Feng et al. [9] noted that dynamic contrast enhanced MRI and diffusion weighted MRI could be used to differentiate lacrimal gland inflammatory pseudotumor and lacrimal gland lymphoma, noting that several signal characteristics were consistently different between the two diagnoses. On MRI, orbital pseudotumor typically shows an iso- to hypointense affected region on T1 sequence without contrast. On T1 sequence with gadolinium contrast, the affected region typically shows diffuse enhancement. On the T2 MRI sequence, the affected area is usually hypointense and becomes more hypointense as the pseudotumor fibrosis progresses. In contrast, orbital venous malformation is isointense to the extraocular muscles on T1, but shows gradual irregular enhancement with delayed wash-out on T1 with contrast. On T2 MRI sequence, orbital venous malformation is hyperintense relative to the extraocular muscles with low-density septations and pseudocapsule [10].

Conclusion

While the orbit and lacrimal gland appear to be the most frequent head and neck sites of inflammatory pseudotumor, this lesion should be in the differential of any head and neck mass. Malignancy must of course be ruled out regardless of the location. This patient presented with

symptoms and imaging worrisome for malignancy, and the patient responded to steroid treatment once the diagnosis of orbital pseudotumor was made. While surgical excision is a viable option, it must be weighed against the risk of morbidity given the often intimate attachment of the pseudotumor to the surrounding anatomy, especially given the good response to steroid and immunosuppressive treatment noted in many series and case reports. In this patient, endoscopic techniques were invaluable in obtaining targeted biopsies for pathology, while avoiding highly morbid procedures or putting structures such as the periorbital muscles or the optic nerve at high risk, and avoiding potentially higher morbidity and recovery times with open techniques. It is also notable that this patient had symptoms (headaches and eye pain) that could easily have been ignored or been deferred. Prompt imaging was crucial to obtaining a diagnosis. Fine cut imaging suitable for image guidance should also be considered given the need for accurate biopsies and given the delicate surrounding anatomy.

Conflict of Interest

None declared.

Authorship

MG: prepared manuscript, provided images, compiled literature review, edited the final manuscript.

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