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Idiopathic Inflammation of the Orbit and Contiguous Structures

Roman Shinder, M.D.^{1,2}, Qasim J. Nasser, M.D.¹, Shelly Brejt, B.S.², Justin Gutman, M.D.², Michelle Williams, M.D.³, and Bitá Esmaeli, M.D.¹

¹Section of Ophthalmology, Department of Head and Neck Surgery, The University of Texas M. D. Anderson Cancer Center, Houston, Texas

²SUNY Downstate Medical Center, Department of Ophthalmology, New York, NY

³Department of Pathology, The University of Texas M. D. Anderson Cancer Center, Houston, Texas

INTRODUCTION

Idiopathic inflammatory pseudotumor is a well known cause of unilateral proptosis and orbital pain in adults. Review of the literature documents a few isolated case reports of this disease process affecting both the orbit and contiguous structures such as the paranasal sinuses, skull base and brain.^{1–6} We herein report 3 such biopsy proven cases that involved the maxillary sinus in 2 patients, and the temporal fossa in a third patient. In all 3 cases, the initial radiographic findings were worrisome for malignancy, and patients were referred to our orbital oncology service to evaluate that suspicion. Clinical presentation, radiographic findings, histopathologic evaluation, and management strategy for each case will be reviewed.

Patient 1

An 18 year-old man presented to a community otolaryngologist with a 6 week history of rapidly progressive and painful right proptosis. He was otherwise healthy and denied prior history of cancer. Imaging studies confirmed a right maxillary sinus mass extending into the right orbit, and endoscopic biopsy was performed and found to be non-diagnostic. He was then placed on high dose hydrocodone and referred to our institution for evaluation by the Head and Neck and Orbital Oncology services. On exam, there was 3mm of right proptosis with increased resistance to retropulsion, hyperglobus, right facial hypesthesia involving V1, V2, and V3, limited eye movements in all directions with diplopia in primary and extremes of gaze, right upper lid fullness causing mechanical blepharoptosis, tenderness in the lacrimal fossa, and subjectively decreased visual acuity of 20–70 (Figure 1A). There was no relative afferent pupillary defect, and no adenopathy was appreciated. Slit lamp exam was significant for inferior conjunctival injection and chemosis with dilated episcleral vessels, and choroidal folds were observed in the posterior pole on retinal exam.

Repeat orbital imaging was performed and showed a right maxillary sinus mass with orbital invasion extending to the lacrimal fossa (Figure 1B). Computed tomography findings were significant for extension of the mass to the orbital apex, along with erosion of the medial wall of the maxillary sinus. The radiologic and clinical findings were worrisome for

Corresponding author: Bitá Esmaeli, M.D., Section of Ophthalmology, Department of Head and Neck Surgery, The University of Texas M. D. Anderson Cancer Center, 1515 Holcombe Blvd, Unit 1445, Houston, Texas 77030. Tel: 713-792-4457. Fax: 713-794-4662. besmaeli@mdanderson.org.

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malignancy. After multiple biopsies of the maxillary sinus mass through endoscopic and Caldwell-Luc approaches showed: “inflammation and fibrosis, no tumor present”, a simultaneous incisional biopsy through an inferior transconjunctival anterior orbitotomy was performed. A piece of the very firm mass was obtained, and on frozen section histopathologic analysis revealed a mixture of chronic inflammatory cells with a large fibrotic component (Figure 1C) consistent with a sclerosing orbital pseudotumor. The patient was started on 80 mg of prednisone to avoid potential compressive optic neuropathy. In the postoperative period the patient subjectively improved, but when prednisone was tapered to 60 mg, a flare of orbital symptoms ensued. Rheumatology evaluation was requested to consider steroid sparing treatments, and methotrexate 7.5 mg weekly was thus initiated. Prednisone taper ensued and at one year follow up he was taking 5 mg of prednisone daily and 12.5 mg of methotrexate weekly. Visual acuity improved to 20/25; hyperglobus and periorbital edema improved; episcleral injection and chemosis as well as macular striae resolved; ocular movements and subjective diplopia improved; and the patient no longer required narcotics for pain control and instead took oral ibuprofen on an as needed basis. Proptosis and imaging findings remained stable at last follow up 15 months after initial presentation.

Patient 2

An otherwise healthy 5 year-old African American girl presented with a 3 month history of progressive and painful right periorbital swelling and proptosis. The mother reported that initially a small papule developed in the lateral right upper eyelid that rapidly progressed to edema of the entire right upper lid. She was initially diagnosed with a “chalazion” by her pediatrician and given a short course of cephalexin for 3 days. After progression of periorbital edema, the pediatrician and a community ophthalmologist agreed to switch her to amoxicillin/clavulonic acid for a 10 day course. She experienced no improvement in her symptoms or signs, and a subsequent computed tomography (CT) scan showed an extensive right lacrimal fossa mass with bony erosion of the lateral orbital wall. The child underwent a lacrimal gland biopsy with histologic evaluation consistent with chronic dacryoadenitis displaying acute and chronic inflammatory cells admixed with fibrous tissue.

The patient was then referred to our orbital oncology service for a second opinion 3 months after symptoms began, with progressive periorbital edema and proptosis. Given the child’s age, relatively rapid onset of symptoms, and radiographic findings, concerns for orbital rhabdomyosarcoma were raised. On exam, her visual acuity in the right eye was 20/30, and there was no relative afferent pupillary defect. She had 9 mm of right globe proptosis, along with diminished supra- and infraduction with diplopia in both up- and downgaze. There was a firm subcutaneous, immobile mass in the right temporal and lacrimal fossae giving mechanical blepharoptosis and inferonasal globe dystopia (Figure 2A). Ishihara color vision was 11/13 on the right. There was no evidence of lymphadenopathy. Repeat orbital CT with contrast displayed a progressive, enhancing, poorly circumscribed mass centered at the right lateral orbital wall with lytic changes, and involvement of the superolateral orbit and temporalis muscle (Figure 2B). The differential diagnosis considered was inflammatory pseudotumor, eosinophilic granuloma, and rhabdomyosarcoma. A repeat anterior orbitotomy through a lid crease incision was performed and the lacrimal gland mass was again biopsied. Additionally, part of the bony lateral orbital wall was taken for histologic investigation. Intralesional injection of triamcinolone was given after frozen section evaluation failed to show a malignant lesion. Histologic evaluation of the biopsy specimen showed an inflammatory lesion with scattered moderate chronic inflammatory cells along with fibrotic bands dissecting through the parenchyma, confirming a diagnosis of idiopathic pseudotumor. Investigation of the zygomatic bone specimen showed bone with marked re-

modeling and ectatic vessels, edema, and chronic inflammation in the intertrabecular spaces, favoring a reactive process.

Postoperatively, the patient had brisk improvement of the periorbital edema, and was started on systemic glucocorticoids. The prednisone was slowly tapered over 5 months. At last follow up 10 months after initial presentation, visual acuity was 20/25, and there was significant improvement in pain, ocular movements, diplopia, and blepharoptosis. There was a residual 2 mm of proptosis. Repeat orbital CT showed almost complete resolution of the lesion.

Patient 3

A 35-year old otherwise healthy female presented with a 5 month history of progressive proptosis and hyperglobus of the left globe. She was initially given amoxicillin/clavulonic acid by her internist for what was thought to be sinusitis. With no improvement, she was evaluated by an otolaryngologist who performed CT imaging of the orbit & facial bones which showed an infiltrative process in the left maxillary sinus and inferior orbit. She then underwent an endoscopic maxillary sinus biopsy which showed: “chronic inflammation and fibrosis, no tumor present”. Due to progressive hyperglobus, periorbital pain, and erythema, she subsequently underwent repeat endoscopy with maxillary antrostomy and biopsy, as well as an anterior orbitotomy with biopsy through an infraciliary approach. Repeat histologic evaluation of the two specimens again confirmed chronic inflammation and fibrosis. Rheumatologic evaluation yielded negative results on extensive blood work including C-ANCA. A short course of oral prednisone was then given for three weeks beginning at 40 mg on a quick taper, which resulted in moderate improvement in her proptosis and hyperglobus. Given her residual symptoms of orbital pain and proptosis, she was referred to our orbital oncology service for a second opinion.

On exam she admitted to mild left periorbital pain. Her visual acuity was 20/20 in the left eye, without relative afferent pupillary defect. Extraocular movements were full and there was no diplopia in primary or extremes of gaze. There was 4 mm of left globe proptosis, along with hyperglobus (Figure 3A). Otherwise, her ocular exam was within normal limits. Repeat CT of the orbits and facial bones displayed an infiltrative mass originating in the left maxillary sinus and extending into the extraconal left inferior orbit to the inferior and medial recti (Figure 3B). Based on histopathologic review of the prior biopsies, results of repeat imaging, and clinical response to steroids, a diagnosis of sclerosing inflammatory pseudotumor was made. The patient was treated with 80 mg of prednisone on a slow 4 month taper that resulted in significant subjective and objective improvement in her signs and symptoms. At last follow up, 26 months after presentation, the patient was asymptomatic and her proptosis and hyperglobus had resolved without recurrence. Repeat CT imaging confirmed radiographic resolution of her disease.

DISCUSSION

The three patients described in this report are unusual in that they presented with simultaneous involvement of the orbit and periorbital structures by an infiltrative aggressive inflammatory process that mimicked a malignancy based on the radiographic and clinical findings. In each case, biopsies were done to rule out malignancy. A mass involving the orbit and its contiguous structures, such as the paranasal sinuses, is rarely due to inflammatory pseudotumor and more frequently the result of a neoplasm or aggressive infection. Rare reports describe inflammatory pseudotumor originating in the orbit and invading contiguous structures (paranasal sinuses, brain, skull base), or originating from these contiguous structures and secondarily invading the orbit.¹⁻⁶ Two of the patients in our report had concurrent maxillary sinus involvement, and one patient had secondary

involvement of the temporal fossa. In 2 patients (cases 1, 2), there was bony orbital wall erosion, but in the third patient the orbital walls were intact. Two of the three patients had sclerosing pseudotumor with a significant fibrotic component, requiring a long course of steroid therapy with slow taper over several months. One patient with sclerosing pseudotumor involving the orbit, the maxillary sinus, and skull base required immunosuppression with methotrexate to allow successful steroid taper. At last follow up (median 15 months, range 10–26 months), all patients had improvement of clinical and radiographic findings without residual orbital pain.

Although rare, idiopathic inflammatory pseudotumor should be considered in any patient who presents with a mass involving the orbit and periorbital contiguous structures such as the maxillary sinus, temporal fossa, or skull base. Som *et al* have previously reported that maxillary sinus pseudotumor has a higher likelihood of causing lytic bony erosion and secondary invasion of nearby structures than pseudotumor that originates in the orbit.¹ Our report suggests that idiopathic inflammatory pseudotumor may simultaneously involve the orbit and contiguous periorbital structures such as paranasal sinuses and/or temporal fossa with or without bony erosion. Bony erosion of the orbital walls is worrisome for a malignant process or Langerhans cell histiocytosis, and in such cases, the diagnosis of idiopathic inflammatory pseudotumor may require multiple confirmatory biopsies to rule out a malignant process, and to insure representative tissue from the disease process is biopsied and evaluated histologically. Recognizing idiopathic inflammatory pseudotumor as a possible etiology of an infiltrative process involving the orbit and the paranasal sinuses or temporal fossa may prevent a delay in diagnosis and initiation of appropriate immunosuppressive treatments.

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Figure 1.

A. Patient 1 at presentation with right globe proptosis and hyperglobus, superior periorbital fullness with mechanical blepharoptosis, inferior conjunctival injection and chemosis with dilated episcleral vessels. B. Coronal T-1 weighted post gadolinium magnetic resonance imaging showing an enhancing right maxillary sinus mass with intra- and extraconal orbital invasion extending to the lacrimal fossa. C. Histologic section of the orbital biopsy specimen displaying a large fibrotic component on the left and chronic mixed inflammatory cells on the right (H & E, 10X).

Figure 2.

A. External photograph of patient 2 showing fullness in the temporal fossa, mechanical blepharoptosis, proptosis and inferonasal globe dystopia. B. Coronal orbital CT with contrast demonstrates an enhancing, poorly circumscribed mass in the superolateral orbit with bony erosion of the lateral wall of the orbit and extension into the temporal fossa.

Figure 3.

A. External photograph of Patient 3 demonstrates left proptosis and hyperglobus. B. Coronal orbital CT showing an infiltrative mass in the left maxillary sinus and extraconal left inferior orbit with involvement of the inferior and medial rectus muscles.