Case report

Primary adenocarcinoma of the orbit initially diagnosed as idiopathic sclerosing orbital inflammation

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ABSTRACT

Purpose: Differentiating idiopathic sclerosing orbital inflammation from orbital inflammation secondary to neoplasia may be challenging, as both processes can present similarly. Neoplasms in the orbit may induce inflammation with accompanying fibrosis. Limited sections of histopathological specimens may demonstrate nonspecific inflammation and lead to an inaccurate diagnosis.

Observations: The authors present a case of infiltrating adenocarcinoma of the orbit with mucinous features which was misdiagnosed as idiopathic sclerosing orbital inflammation due to three separate benign biopsy specimens.

Conclusions and Importance: The ophthalmologist must remain suspicious of malignancy in cases of suspected idiopathic orbital inflammation with an atypical clinical course, regardless of apparently benign biopsy results.

1. Introduction

Idiopathic orbital inflammation (IOI) is an immune mediated infiltrative condition that has distinct variants, a non-sclerosing and a sclerosing type, which is characterized by a polymorphous lymphoid infiltrate or a predominance of fibrosis, respectively.1-2 The non-sclerosing type is fairly common, representing the majority of orbital inflammatory syndromes.3 The differentiation of idiopathic orbital inflammation and orbital inflammation secondary to neoplasia can be challenging, as both processes may manifest with similar clinical presentations.7

2. Case report

The patient discussed is an 86-year-old Hispanic male who presented with complaints of left eye irritation. His past ocular history was significant for a left central retinal vein occlusion, managed over several years with panretinal photocoagulation and multiple anti-VEGF injections. He also had advanced glaucoma with a superotemporal Ahmed valve tube shunt in the left eye, and his left eye was pseudophakic. Additional medical history includes prostate cancer treated with a radical prostatectomy in 1998, coronary artery disease treated with coronary artery bypass grafting in 2002, atrial fibrillation, hypertension, and chronic kidney disease. He has no family history of cancer and his social history is unremarkable. Of note, the collection and evaluation of protected patient health information was HIPAA-compliant.

Fig. 1. CT orbit without contrast, coronal cut, demonstrating the large orbital mass surrounding and indenting the left globe.

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complaint.

On the day of his initial presentation he was scheduled to have an anti-VEGF injection for macular edema for his retinal vein occlusion, but this was deferred because slit lamp examination revealed an elevated inferotemporal subconjunctival mass. The patient denied pain or facial cutaneous sensory changes. An MRI orbit with and without contrast showed soft tissue enhancement in the temporal aspect of the left orbit between the sclera and the lacrimal gland without evidence of bony involvement. A biopsy of the orbital lesion demonstrated chronic inflammation and subepithelial fibrosis but no sign of malignancy. Because of the findings of orbital inflammation without malignant changes, the patient was started on 40mg oral prednisone with a gradual taper.

Four months later he developed a new subconjunctival lesion adjacent to the initial area of concern, but this time it did not resolve with corticosteroids. Another biopsy of the lesion was performed, and was consistent with an admixture of active and chronic inflammation and fibrosis but negative for malignancy. Over the next several months the patient was noted to have a relapsing, remitting course of inflammation and ultimately developed periorbital pain, symblepharon in the area of the biopsies, worsening blepharoptosis, and diplopia from a restrictive strabismus. A more extensive biopsy of the orbital mass was performed which again showed signs of acute and chronic inflammation but no sign of malignancy. After consultation with rheumatology the patient

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**Fig. 2.** A) H&E stain demonstrating cords of epithelial and ductal tissue with fibrotic inflammatory background. B) PAS stain demonstrating pockets of mucinous material. C) Mucicarmine stain, highlighting mucin, which appears pink on a yellow background. D) Pancytokeratin immunostain highlighting surface ectoderm relative to fibrotic background. E) H&E stain of the anterior chamber angle demonstrating epithelial tumor infiltrating the ciliary body and coursing across the anterior surface of the iris and cornea.
was started on steroid-sparing anti-inflammatory therapy with myco-
phenolate mofetil.

Despite treatment of his inflammation, his pain continued to worsen
and vision declined gradually to no light perception, with complete
restriction of the left globe but intact facial cutaneous sensation. The
patient then developed marked hypotony and his exam showed the
Ahmed valve plate had eroded through the conjunctiva. This was
treated by removal of the Ahmed valve and placement of an amniotic
membrane graft. On a postoperative fundus exam, the patient was
noted to have inferotemporal elevation of the retina and choroid con-
cerning for choroidal effusion from the hypotony which had not been
noted on prior fundus exams, but B-scan ultrasound demonstrated in-
dentation of the inferotemporal aspect of the globe secondary to orbital
mass effect. A computed tomography scan of the orbits demonstrated
marked increase in size of the orbital mass in the temporal orbit in-
volving both intraconal and extraconal spaces but without bony in-
volvement (Fig. 1), and biopsy of the orbital mass was scheduled.

A subtotal exenteration was ultimately performed given the ex-
tensive tumor burden noted intraoperatively after the surgical speci-
mens were sent for histopathologic evaluation (Fig. 2). The final pa-
thologic diagnosis was infiltrating adenocarcinoma, not otherwise
specified, with some features suggestive of mucoepidermoid carcinoma.
The tumor was determined to be fairly well-differentiated and did not
appear cytologically aggressive, but with infiltrative growth pattern
lining the internal globe suggestive of a high-grade tumor, T4N0M0.
The patient was provided the option for additional surgery with or
without radiation and chemotherapy, and ultimately decided to un-
dergo adjuvant radiation therapy. The patient has finished adjuvant
radiation therapy, and neuroimaging at six months post exenteration
showed no signs of recurrence.

3. Discussion

Neoplasia may induce inflammation with varying clinical pre-
sentations to include involvement of the extraocular muscles, lacrimal
gland, or sclera. Histopathological specimens may reveal non-specific
chronic inflammation, consisting of mature lymphocytes and fibrosis
leading to an incorrect diagnosis.1 Additionally, as in the case demon-
strated, there exists the possibility initial biopsies performed may not be
representative of the main body of the tumor and may be related to
prior trauma or surgery. Furthermore, orbital and ocular inflammation
may be present irrespective of an underlying malignancy. A review of
the literature revealed other cases of malignancies initially diagnosed as
idiopathic orbital inflammation to include metastatic breast carcinoma
and metastatic orbital carcinoid tumor.4–8 However, the cases pre-
tened were metastatic to the orbit. The authors did not find any other
case report of primary adenocarcinoma of the orbit masquerading as
idiopathic orbital inflammation.

Typically, idiopathic orbital inflammation presents with abrupt
onset of pain and inflammatory signs and responds robustly to admin-
istration of systemic corticosteroids.9 The case presented was atypical
in that the patient had a painless initial presentation, and his in-
flammation recurred following administration of corticosteroid
therapy. Atypical cases of idiopathic orbital inflammation warrant
extensive evaluation for alternative causes. As demonstrated in this
case, the clinician must remain vigilant to the possibility of malignancy
in cases of suspected idiopathic orbital inflammation, regardless of
apparently benign biopsy results. This is particularly true in the pre-
sence of worsening clinical signs and symptoms or an atypical initial
presentation.

Patient consent

Written consent to publish case details was obtained from the pa-
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Conflicts of interest

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Authorship

All authors attest that they meet the current ICMJE criteria for
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Appendix A. Supplementary data

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