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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.[2]

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-

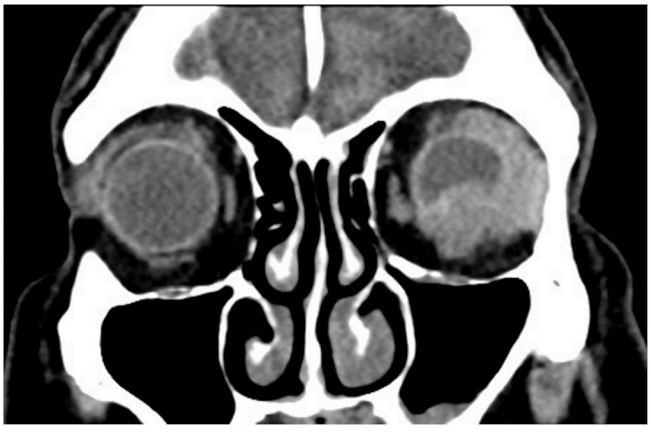

Fig. 1. CT orbit without contrast, coronal cut, demonstrating the large orbital mass surrounding and indenting the left globe.

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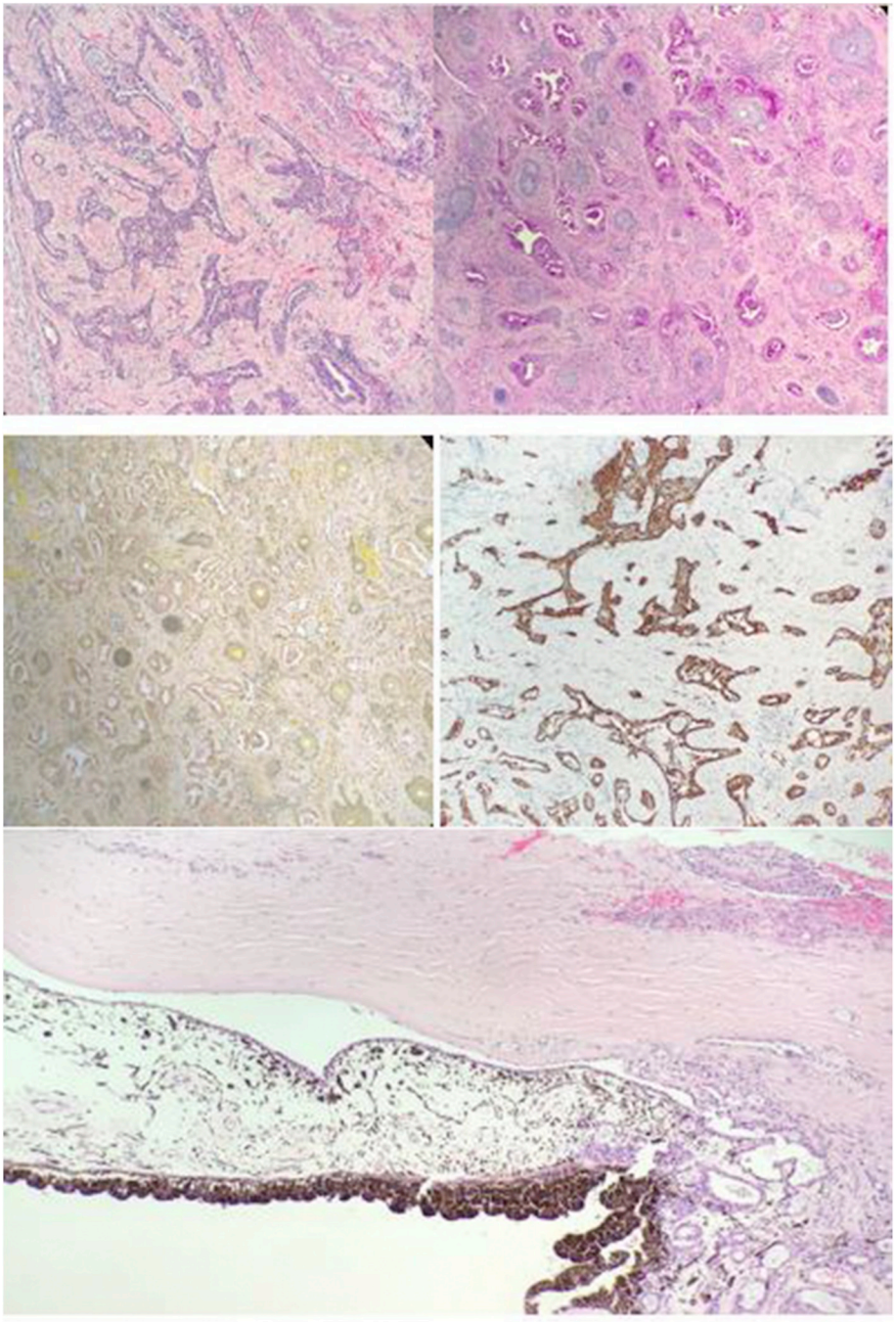

Fig. 2. A) H&E stain demonstrating cords of epithelial and ductal tissue with fibrotic inflammatory background. B) PAS stain demonstrating pockets of mucinous material. C) Mucicarmine stain, highlighting mucin, which appears pink on a yellow background. D) Pancytokeratin immunostain highlighting surface 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.

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

was started on steroid-sparing anti-inflammatory therapy with mycophenolate motefil.

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 hypotonyand 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 concerning for choroidal effusion from the hypotony which had not been noted on prior fundus exams, but B-scan ultrasound demonstrated indentation 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 involving both intraconal and extraconal spaces but without bony involvement (Fig. 1), and biopsy of the orbital mass was scheduled.

A subtotal exeneration was ultimately performed given the extensive tumor burden noted intraoperatively after the surgical specimens were sent for histopathologic evaluation (Fig. 2). The final pathologic 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 undergo 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 presentations 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 demonstrated, 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 presented 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 administration of systemic corticosteroids.[9] The case presented was atypical in that the patient had a painless initial presentation, and his inflammation 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 presence of worsening clinical signs and symptoms or an atypical initial presentation.

Patient consent

Written consent to publish case details was obtained from the patient.

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Conflicts of interest

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Authorship

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

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2019.100529.

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