Primary tumours of the ciliary body include adenoma, Fuch's adenoma, and adenocarcinoma. The adenoma can be easily distinguished by lack of infiltrative behaviour and relatively rare mitosis.[6] Similarly, ciliary body adenocarcinomas—including those arising from NPCE—are rare and have been reported in all age groups.[7] It has been believed that these arise as a reactive proliferation secondary to trauma or inflammation. These ciliary body masses are nonpigmented and irregular, have the tendency to cause localized cataract changes, and can be associated with sentinel vessels.[7] Differential diagnosis of this tumour includes medulloepitheloma, adenoma, melanoma, and metastasis. IHC studies have shown that these tumours stain positive with S-100 protein and vimentin as they originate from NPCE. They can show positivity with Kermix, CAM 5.2, and CK7.[7,8]

Since we have encountered the aforementioned 2 cases in which the adenocarcinoma had originated from 2 different intraocular structures, we have summarized the differentiating features between those tumours arising from the NPCE and the ones arising from RPE in Table 1.

In conclusion, intraocular adenocarcinomas may constitute diagnostic challenge to the ophthalmic pathologist. Good knowledge of the characteristic features and careful examination of routine evisceration tissue are a must to avoid overlooking such tumours or misdiagnosis especially when an underlying neoplasm is unsuspected.

Hind Manaa Alkatan, [*,†] **Abdullah A. Al Qahtani,** [‡] **Azza M.Y. Maktabi** [†]

[*]Ophthalmology Department, College of Medicine, King Saud University, Riyadh, Saudi Arabia; [†]Pathology & Laboratory Medicine Department, King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia; [‡]Department of Ophthalmology, College of Medicine, University of Dammam, Saudi Arabia.

Correspondence to:
Hind Manaa Alkatan, MD: hindkatan@yahoo.com

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Can J Ophthalmol 2016;51:e67–e70
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http://dx.doi.org/10.1016/j.jcjo.2015.12.010

IgG4-related orbital disease mass lesion

IgG4-related disease (IgG4-RD) is a systemic, tumefactive, inflammatory disease. It is increasingly recognized in the orbit (IgG4-related orbital disease [IgG4-ROD]), where it may affect the lacrimal glands, extraocular muscles, orbital fat, trigeminal nerve branches, orbital septum, sclera, optic nerve, eyelid, nasolacrimal drainage system, and bulbar conjunctiva.[1–3] We present a case of a solitary posterior orbital mass, histologically proven to be IgG4-RD.

A 42-year-old male presented with a 3-month history of increasing, painless reduction of vision in his right eye. The presenting right visual acuity was 6/60 unaided, 6/24 with pinhole, and 6/5 unaided in the left eye. Ishihara colour vision testing was 10/11 both eyes, with subtle right red desaturation. There was no globe displacement. Funduscopy found choroidal folds, confirmed on optical coherence tomography (Fig. 1). Magnetic resonance imaging (MRI) demonstrated a well-demarcated, contrast-enhancing lesion spanning the intra- and extraconal spaces in the right superotemporal orbit (Fig. 2). The extraocular muscles, optic nerve, lacrimal gland, and supra- and infraorbital nerves were normal. The findings of an MRI of the head and orbit performed 2 years previously for an unrelated condition were normal.

A latera orbitotomy was used for surgical excision. A large, firm, spherical lesion was excised intact. Histopathology revealed a densely sclerotic lesion with storiform fibrosis and obliterative fibrosis. The average IgG4 count per high-power field was 78, and the IgG4:IgG cell ratio was 41.4% (10 fields counted) (Fig. 3).

No other foci of disease were identified on computed tomography scan of the chest, abdomen, and pelvis. The findings of blood tests, including inflammatory markers and serum IgG4, were all normal. Six months postoperatively, the vision had improved to 6/24 unaided and 6/12 pinhole despite persistent choroidal folds, and there was no clinical or radiological evidence of disease recurrence.

Fig. 1—Right-eye choroidal folds shown on (a) fundus photograph and (b) macular optical coherence tomography scan.

IgG4-ROD most commonly affects the extraocular tissue and the lacrimal gland.[4,5] Cases of IgG4-RD orbital mass lesions that are distinct from other structures have been described but are typically bilateral and associated with extraorbital disease.[6]

The diagnosis criteria of IgG4-RD are still evolving. However, the high IgG4 count, high IgG4:IgG ratio, and characteristic histology in the present case fulfils all current criteria, other than the normal serum IgG4 level, which is in fact frequently observed in discrete ocular lesions.[2,7–9]

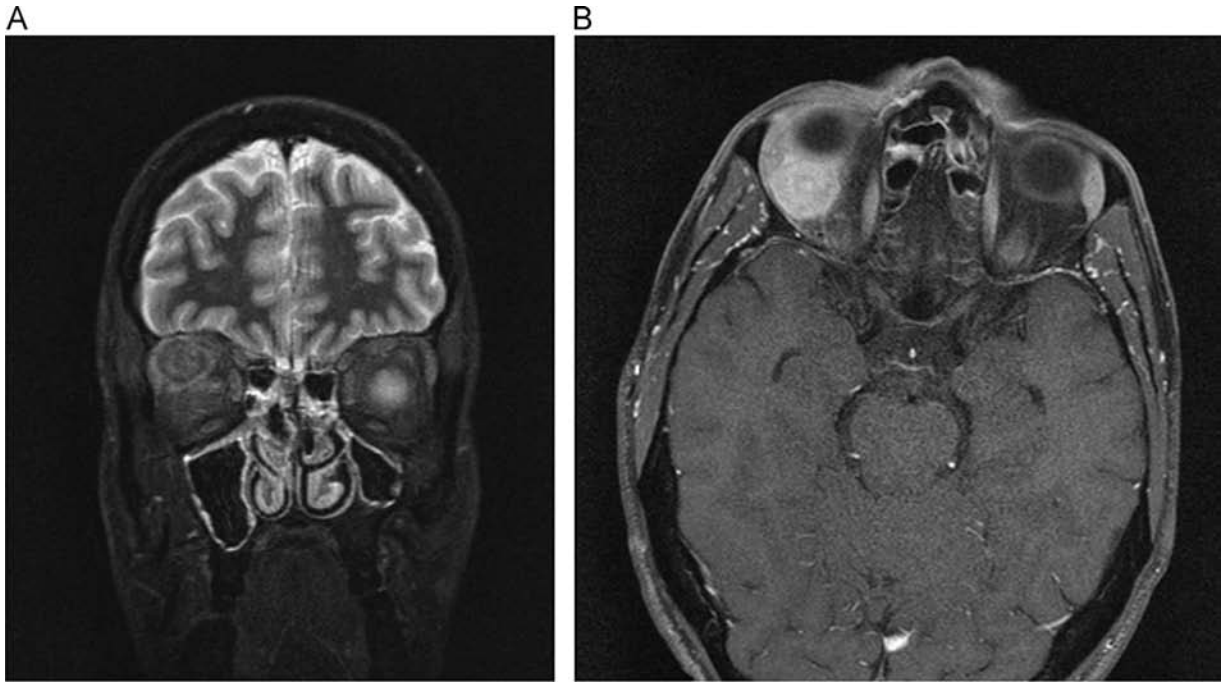

Fig. 2—Coronal (a) and axial (b) magnetic resonance imaging images showing a well-defined mass in the superoposterior region of the right orbit.

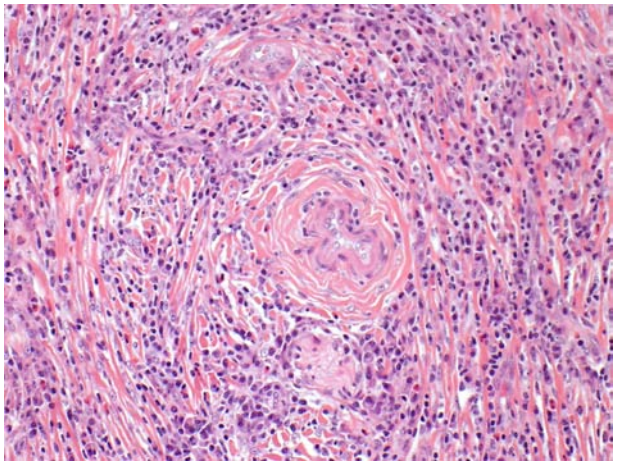

Fig. 3—Tissue sample stained with haematoxylin and eosin illustrating keloid-like fibrosis in a small vessel with abundant plasma cells.

This case of a unilateral isolated mass with neither local nor systemic involvement adds to the ever-enlarging spectrum of IgG4-ROD presentations.

Disclosure: The authors have no proprietary or commercial interest in any materials discussed in this article.

Saul N. Rajak, [*,†] **Thomas A. Eldredge,** [‡] **Faiza Rashid,** [§] **Geoffrey P Brittain** [*]

[*]Brighton and Sussex University Hospital, Brighton, United Kingdom; [†]The International Centre for Eye Health, London School of Hygiene and Tropical Medicine, London, United Kingdom; [‡]The Royal Adelaide Hospital, Adelaide, Australia; [§]Surrey and Sussex Healthcare NHS Trust, Surrey, United Kingdom.

Correspondence to:
Saul N. Rajak, FRCOphth, PhD, The Sussex Eye Hospital, Eastern Road, Brighton BN2 5BE, United Kingdom: saul.rajak@lsthm.ac.uk

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Can J Ophthalmol 2016;51:e70–e72
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http://dx.doi.org/10.1016/j.jcjo.2015.12.009

Histopathology of endocrine mucin-producing sweat gland carcinoma of the eyelid

A 74-year-old male presented to the oculoplastic surgery service with a left lower eyelid margin lesion that had enlarged over the previous 8 months. Results from an incisional biopsy performed by his plastic surgeon suggested dermal carcinoma. A second opinion on the histopathology of the lesion resulted in a diagnosis of eccrine adenocarcinoma. The patient had a history of a similar left lower eyelid lesion that had been resected 20 years previously with no signs of recurrence for 15 years. Histopathology from this past surgery was not available.

The patient reported noticing subtle changes in the affected eyelid margin over the past 5 years. Over the 8 months before his presentation, the area began to enlarge more rapidly. The patient was an ex-smoker of 40 pack-years with a history of colon cancer and benign prostatic hyperplasia. Findings of ophthalmic and orbital examinations, including visual acuity, intraocular pressure, pupil reactivity, and eye motility, were all within normal limits.

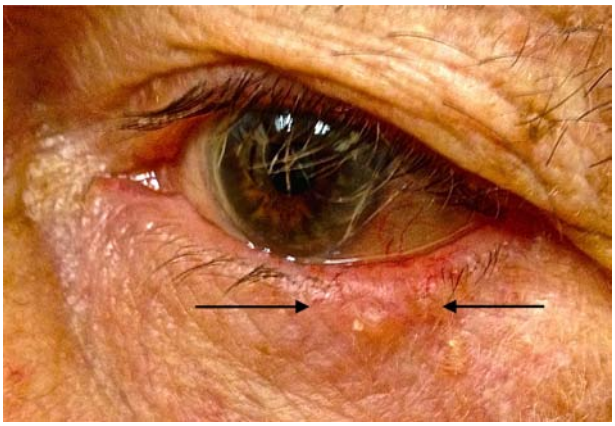

Fig. 1—Left eyelid papule (3.4 mm) with associated madarosis, telangiectasia, and distortion of the eyelid margin architecture.