A 62-year-old male presented with 1 year of distorted vision OS. His medical history included alcohol and cigarette consumption.

Best corrected visual acuity was 20/30 and IOP 16 mm Hg. The anterior chamber and vitreous were clear other than mild asteroid hyalosis. Diffuse retinal pigment epithelium (RPE) changes and macular thickening were present. The right eye was normal (Fig. 1A, B).

Intravenous fluorescein angiogram showed nonspecific leakage at the level of the RPE and macular edema. B-scan demonstrated diffuse choroidal thickening. Optical coherence tomography confirmed subretinal fluid (Fig. 1C-F).

Magnetic resonance imaging of the head and orbits confirmed the choroidal thickening, and revealed a lobulated, elongated soft tissue mass encasing the optic nerve (Fig. 2A, B). Involvement of the intraforaminal optic nerve could not be excluded. The muscles and lacrimal gland were clear and there was no intracranial mass. Staging investigations, including computed tomography scans, chest x-ray, and CSF cytology were normal. Serum protein electrophoresis revealed IgM monoclonal gammopathy, in keeping with a lymphoproliferative disorder.
Referral was made for orbital biopsy. On examination, an irregular area of conjunctiva was discovered, which on biopsy revealed a dense infiltrate of small lymphocytes with positive staining for CD20 and CD79, with coexpression of BCL2 and focally CD10 (Fig. 2C, D). There was no coexpression of cyclin D1. Background T-cells were present. The diagnosis was marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT).

External beam radiotherapy was delivered to the whole left orbit (36 Gy in 20 fractions). Six months later, an MRI demonstrated a significant decrease in tumor size with residual thickening thought to be scar tissue.

Discussion

This is an atypical presentation of a primary MALT lymphoma. On fundus examination, the leopard spot appearance was suggestive of a lymphoma or metastatic carcinoma of unknown origin. The extent of the extraocular involvement with encasement of the optic nerve was also unusual.

Primary marginal zone B-cell lymphomas can originate from the choroid or orbit. Primary choroidal lymphomas tend to be low-grade MALT subtypes with an indolent course. Blurred vision, clear vitreous, choroidal thickening, unilateralarity, and no CNS involvement are characteristic. Episodes of blurred vision can be attributed to macular serous retinal detachments. Multifocal yellow lesions are present early, and can progress to diffuse choroidal thickening and extraocular extension. This correlates with the findings in our patient, suggesting a primary choroidal origin. In a review of 13 cases, 11 had extraocular extension, and only 3 developed systemic disease. Biopsy of conjunctival nodules can be useful for diagnosis, and is less invasive than a biopsy of the choroid or orbit.

An orbital origin is less likely because orbital lymphomas rarely invade the choroid as they mold around ocular structures. Nevertheless, 1 recent report described vitreitis secondary to this tumor.

Our case agrees with the current literature that the majority of uveal lymphomas have extraocular involvement. However, the extent of the intraocular and extraocular extension with tracking along the optic nerve was unusual. Biopsy of the conjunctival nodule allowed for prompt diagnosis and treatment without more invasive procedures.

Julia Baryla,* Larry H. Allen,* Keith Kwan,§ Michael Ong,* Tom Sheidow*

*Ivey Eye Institute, University of Western Ontario, London, Ont.; and the Departments of Pathology and Oncology, London Health Sciences, London, Ont.

Correspondence to:
Julia Baryla, MD; julia@baryla@gmail.com

Fig. 2—(A) Orbital MRI showing choroidal thickening with orbital mass. (B) Lesion encasing optic nerve. (C) Conjunctival Biopsy. H & E stain. Medium power magnification (10x) showing the infiltrate of malignant cells. (D) High power magnification (40x) of the conjunctival biopsy showing an infiltrate composed of atypical lymphocytes with small irregular nuclei with nucleoli. Long Arrow: Choroidal thickening. Short arrows: Mass encasing the optic nerve.
RE: Bilateral subconjunctival masses due to orbital fat prolapse

A 41-year-old male presented with 6 months of bilateral symmetrical subconjunctival lesions that appeared after a motor vehicle collision (Fig. 1). Injuries included a unilateral zygomatic fracture repaired without incision around the orbit. Ocular examination revealed a visual acuity of 20/20 OU and bilateral, soft, yellow masses in the temporal bulbar conjunctiva which were easily displaced posteriorly with a cotton-tipped swab. Computed tomography disclosed continuity of both subconjunctival lesions with the intraconal space. Both masses were excised without complication.

Both masses had a similar histologic appearance of uniform hypovascular lobules containing mature adipocytes separated by fibrovascular septa (Fig. 2). Focal areas showed increased cellularity, including inflammatory cells and frequent multinucleated giant cells within fibrous septa with their nuclei arranged in a “floretlike” pattern (Fig. 2, inset).

Orbital fat is divided into 2 compartments: extraconal fat lies outside the extraocular muscles and Tenon’s capsule, whereas intraconal fat lies within the extraocular muscle cone.1,2 Various mechanisms (e.g., trauma, surgery, degeneration) can lead to disruption of the connective tissue support of orbital fat leading to herniation.1 Unlike the more common extraconal fat prolapse frequently seen as an aging change, intraconal fat prolapse is rarely reported.1-4 Intraconal fat prolapse may be unilateral, or bilateral in one-third of cases, and generally is located superiorly and temporally, as in our case, secondary to the thin intermuscular septum and abundant fat in this region.1,3

Intraconal fat prolapse may be confused clinically with dermolioma, conjunctival lymphoma, and lacrimal gland prolapse.1,3,4 However, it can be distinguished by the ease with which it can be pushed posteriorly, and also histopathologically. CT and MRI may identify fat prolapse by demonstrating continuity with the intraconal space, as in our case.2,4

Recently, there has been confusion and debate concerning the differentiation between intraconal fat prolapse and pleomorphic lipoma.1,2,5 Pleomorphic lipoma is a rare subtype of lipoma most often seen in the soft tissues of the head and neck, on a morphologic continuum with spindle cell lipoma.1,5 In 2003, Daniel et al. published a series of 6 cases believed to represent pleomorphic lipoma of the orbit. Histopathological features of these cases included large multinucleated cells often arranged in a “floretlike” pattern. Although thought to represent a distinguishing feature of pleomorphic lipoma and other neoplasms, similar cells, but usually with small normochromatic nuclei, have been recently identified in both prolapsed and in situ intraconal orbital fat, suggesting they also may result from a reactive or degenerative process.1,2 As immunohistochemical markers have not been shown to differentiate

Fig. 1—Presentation with bilateral symmetrical conjunctival masses (arrows).

Fig. 2—Subconjunctival mass shows typical prolapsed orbital fat, with uniform hypovascular lobules, and fibrous septa (H&E 100×). Inset: high power view of a multinucleated, “floretlike” giant cell in the fibrous septum of the prolapsed orbital fat (H&E 640×).