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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]

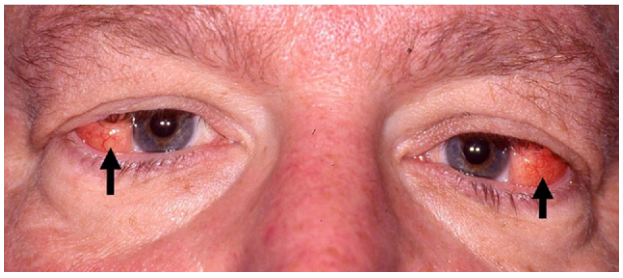

Fig. 1—Presentation with bilateral symmetrical conjunctival masses (arrows).

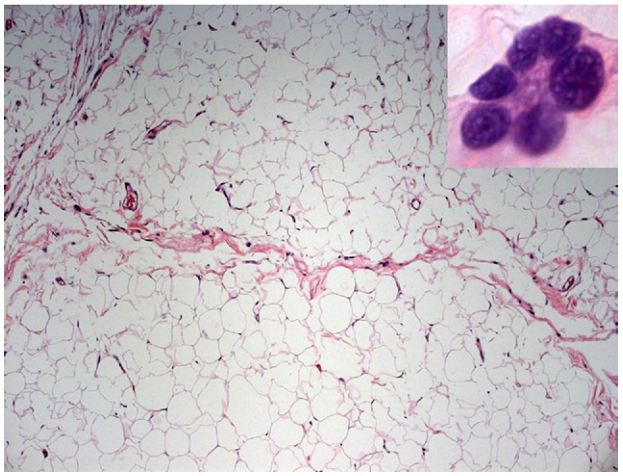

Fig. 2—Subconjunctival mass shows typical prolapsed orbital fat, with uniform hypovascular lobules, and fibrous septa (H&E 100×). Inset: high power view of a multinucleated, "floretlike" giant cell in the fibrous septum of the prolapsed orbital fat (H&E 640×).

Intraconal fat prolapse may be confused clinically with dermolipoma, 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.[5] 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

orbital fat from pleomorphic lipoma, recently proposed diagnostic criteria for pleomorphic lipoma include wire-like collagen and hyperchromatic multinucleated cells and pleomorphic spindle cells located within fat rather than within fibrous septa.[1,2]

In summary, we report a case of bilateral, symmetric intraconal fat prolapse. The relative rarity of intraconal fat prolapse has led to concern over the correct diagnosis of these lesions. Despite recent clarification of the histopathologic distinction between orbital fat prolapse and pleomorphic lipoma, the rarity of both entities necessitates careful clinical and histopathologic assessment of all suspected cases.

André Jastrzebski,*† Seymour Brownstein,*† David R. Jordan,* Bruce F. Burns†

*Department of Ophthalmology, †Department of Pathology, The Ottawa Hospital and the University of Ottawa, Ottawa, Ont.

Correspondence to:
Seymour Brownstein, MD, FRCSC: sbrownstein@ohri.ca

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Intravenous immunoglobulin (IVIG) for orbital Wegener's granulomatosis

A 73-year-old female was referred with a presumptive diagnosis of posterior scleritis based on a 2-month history of visual loss, ocular pain and injection, and serous retinal detachments. Her symptoms had responded initially to oral prednisone (60 mg daily), but discomfort recurred on attempted tapering. On our first encounter, her vision was 20/40 OD, 20/30 OS, and she had orbital pain and conjunctival injection in spite of prednisone 50 mg daily.

A computed tomography scan revealed enhancing, irregular soft tissue surrounding both globes, most prominent over the superior aspect, interpreted as a sclerosing inflammatory process. An orbital biopsy revealed necrotizing granulomatous inflammation with numerous giant cells but without obvious vasculitis (Fig. 1).

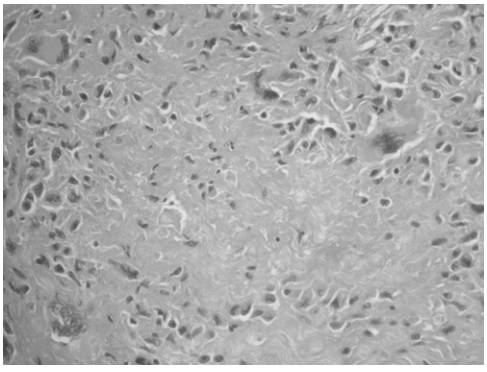

Fig. 1—High power view of the orbital biopsy showed necrotizing granulomatous inflammation with numerous giant cells and occasional neutrophils (H&E, ×40).

A rheumatologic evaluation found circulating immune complexes, elevated erythrocyte sedimentation rate and C-reactive protein, reduced C3 and C4 levels, hematuria without casts, and small bilateral pulmonary pleural effusions. Her renal function and centrally accentuated antineutrophil cytoplasmic antibody test (cANCA) were initially normal, but the cANCA became elevated within a year.

Based on a presumptive diagnosis of localized orbital Wegener's granulomatosis (WG), therapy was initiated with intravenous methylprednisolone (3 g over 1 week) and oral trimethoprim-sulfamethoxazole with a positive response, but with recurrent pain and scleritis on attempted cessation. Cyclophosphamide 100 mg daily and methotrexate were initiated to spare corticosteroid usage but were discontinued because of persistent leukopenia. Intravenous immunoglobulin (IVIG) was started at a dosage of 400 mg/kg for 4 consecutive days every 3 weeks that produced dramatic improvement without side-effects. Attempted reduction to 350 mg/kg 2 years later resulted in recurrent ocular pain and inflammation, requiring resumption of the initial dose. Her symptoms remain well controlled after 10 years of treatment.

WG is a systemic necrotizing vasculitis characteristically involving the upper and lower respiratory tracts and kidneys.[1,2] Orbital involvement may occur in 58% of patients[1,3] and may be isolated. Conventional treatment consists of combined corticosteroids and cyclophosphamide[2] for both systemic and ocular manifestations.[3] IVIG has been used when other therapies fail[3] although its use in isolated orbital WG has not been described previously.

IVIG contains monomeric immunoglobulin G and is prepared from pooled plasma from several thousand donors, accounting for its high cost.[4] It may modulate T- or B-cell-mediated immune responses and has been used in