A 50-year-old man was referred for management of a long-standing, large-angle exotropia and limitation of adduction in the left eye. He had a history of poor vision in the left eye since childhood and 4 previous strabismus procedures. The most recent strabismus surgery was 38 years before presentation. Prior records were unavailable, and the patient could not recall any details pertaining to his earlier procedures or alignment in childhood. His wife did not report any recent change in his appearance or alignment.

Examination

Best-corrected visual acuity was 20/15 in the right eye and 20/200 in the left eye. There was no afferent pupillary defect, and color vision was full and symmetric. The patient had an exotropia of 40 prism diopters (PD) at distance and 45 PD at near, with a −3 adduction deficit of the left eye greatly increasing the exotropia in right gaze to 60 PD. There was a 2 mm ptosis of the left upper eyelid.

Anterior segment examination revealed conjunctival scarring over the medial and lateral fornices of both eyes. A palpable, painless, soft, purple-gray mass, 8 mm in horizontal diameter and of indeterminate depth was detected in the superonasal aspect of the orbit of the left eye. It extended subconjunctivally posterior to the caruncle and superonasally toward the upper lid. One large superficial conjunctival vessel penetrated the lesion (Figure 1). The bulk of the mass was apparent through closed lids. The patient was unaware of the presence of this mass. The remainder of the ophthalmological examination was unremarkable, including anterior chamber evaluation, intraocular pressure, fundus examination, and visual fields.

Ancillary Testing

Due to the heterogeneous color of the mass, the penetrating vessel, and the lack of adequate history, magnetic resonance imaging (MRI) of the orbits was performed to rule-out malignancy. The MRI showed a well-encapsulated cyst overlying the medial globe, with no evidence of erosion or invasion of surrounding structures. The medial rectus muscle appeared to insert at the posterior pole of the cyst (Figure 2).
Treatment

Planned strabismus repair targeted the recovery and advancement of the “apparently slipped” medial rectus muscle, recession of the lateral rectus muscle and excision of the mass in toto, if possible. Intraoperative unroofing of a superficial layer of conjunctiva over the mass revealed a chocolate-colored cyst filled with serosanguinous fluid, with strands of flaccid extraocular muscle straddling its surface (Figure 3). The color of the fluid was attributed to prior hemorrhage. Although excision in one piece proved technically challenging, an excisional biopsy of one large section of the mass included up to 10 mm of the flaccid medial rectus fibers. A gentle hand-over-hand technique was required to reach the posterior extension of the cyst, enabling recovery of the medial rectus muscle, well posterior to the equator and still attached to the posterior surface of the remaining wall of the cyst via a thin thread of muscle fiber. The body of the medial rectus muscle was recovered, advanced, and reattached to the anatomical insertion with an adjustable-suture technique after all visible remaining tissue from the cyst was surgically excised. The lateral rectus muscle was recessed on an adjustable suture. In the recovery room, the muscles were adjusted to a resultant esotropia of <10 PD at distance and near. Mild limitation of adduction remained. Three months later, the patient’s sensorimotor examination demonstrated an exotropia of 8 PD at distance and 8–10 PD at near, with −1 limitation of adduction and −1.5 limitation of abduction. There has been no recurrence of the mass over a one-year follow-up period, and alignment has been stable.

Differential Diagnosis

The differential diagnosis of a non-invasive cystic mass in the medial orbit includes a variety of simple epithelial cyst, dermoid cyst, cystic teratoma, neural cyst, mucocele, encephalocele, lymphangioma, inflammatory cyst, parasitic cyst, hematic cyst, and cystic tumors.

Diagnosis and Discussion

Histopathological examination of the excised lesion showed a cystic structure with a thin wall lined by a double layer of non-keratinizing cuboidal epithelium. The cyst content was acellular, proteinaceous material. No skin appendages or goblet cells were identified, and the epithelial cells did not have cilia. In some areas the cells appeared to have luminal projections, suggestive of apical apocrine snouts (Figure 4A). Immunohistochemistry for pan keratin highlighted the epithelial lining (Figure 4B). Immunostaining performed to identify apocrine differentiation was negative, and epithelial cells were immuno-negative for gross cystic disease fluid protein-15. Actin immunostaining was negative for actin positive myoepithelial cells.

The lining of the cyst in the present case was composed of epithelial cells, and this limited the differential to epidermal cyst, conjunctival cyst, respiratory epithelial cyst, and apocrine gland cyst. The lack of keratin production and of squamous differentiation excluded the possibility of epidermal cyst. The lack of cilia and gob-
let cells made respiratory epithelial cyst unlikely reducing the differential to an apocrine gland cyst (sudoriferous cyst) or a conjunctival cyst. Apocrine gland cysts, which may (rarely) occur in the anterior portion of the orbit, are congenital in nature and thought to develop from entrapped epithelial cells destined to form the glands of Moll. Most conjunctival cysts develop secondary to implantation following ocular trauma or surgery. A primary, congenital form has been described that arises along the common sheath of the superior rectus muscle and levator palpebrae superioris muscle that is thought to result from misdirected cleavage of mesoderm.2

Development of a clinically significant giant conjunctival cyst as a complication of strabismus surgery has been reported, albeit rarely.3−6 This complication has a reported incidence of 0.25% after strabismus procedures.7 The purple/blue grayish hue of such cysts, noted in our patient, has been previously cited along with the frequent delay in growth, secondary strabismus, and clinical presentation for evaluation.6,8

Although thermal cauterization or intraliesional injection with isopropyl alcohol have been successfully used to shrink some conjunctival cysts,9,10 these techniques do not address the large secondary incommitant strabismus associated with many giant conjunctival cysts. Management by marsupialization of the cyst has been reported to correct a simple epithelial cyst adjacent to a muscle.11 In cases such as the present one, where the cyst has encapsulated muscle fibers and is associated with strabismus, careful surgical excision remains the mainstay of therapy because it allows salvaging the healthy rectus muscle and recreating a new attachment to the globe.6,8,9

When examining a patient with an orbital mass with or without associated strabismus, it is vital to take into account the history of previous ocular surgery or trauma and to consider the possibility of a giant conjunctival cyst. The use of imaging modalities, such as orbital MRI or ultrasound, can help provide useful information regarding the consistency of the mass and its relationship to adjacent structures. An isolated cyst without adjacent bony erosion or soft tissue invasion is likely to be benign histologically, although it is not benign if it causes secondary strabismus, displacement, or compression of surrounding structures. Imaging in the present case provided invaluable insight into the cystic structure of the mass, and the extreme displacement of the medial rectus insertion; understanding this anatomical relationship prevented inadvertent loss of the medial rectus at the time of repair. Histopathological examination confirmed the nature of the lesion in this case, reassuring patient and surgeon alike.

References
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