Respiratory epithelium lined cysts presenting in the orbit without associated mucocele formation

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SUMMARY Three patients presented with orbital cysts lined with upper respiratory tract epithelium. In each case there was no evidence of an associated mucocele. Two patients had a past history of orbital trauma, and the third had had preceding sinus surgery. It is postulated that these cysts were caused by traumatic herniation of nasal sinus epithelium cells into the orbit.

The usual pathological route for upper respiratory tract epithelium to find its way into the orbit is in association with mucocele formation. Mucoceles account for up to 10% of unilateral proptosis.^{1,2} They commonly present in the fourth and fifth decades of life in association with chronic sinus disease, though they may also occur in children with cystic fibrosis.^{3,4}

It is generally accepted that mucoceles form as a result of blockage of the normal drainage pathway of a sinus. Increased pressure, from continuing secretion by the lining of respiratory epithelium, causes sinus expansion and may eventually be associated with osteoclastic resorption of its bony wall. This allows the enlarging respiratory epithelium lined cyst to enter the orbit, with ensuing ocular displacement and proptosis (Fig. 1).

When the diagnosis of mucocele is suspected clinically, radiology is the most useful investigation. The classical signs are well described by Lloyd.⁵ As well as radiological evidence of an increase in size, which may not have been obvious clinically, there may also be opacification of the sinus. The characteristic scalloped margin of the frontal sinus is often lost, and in the ethmoid sinus the typical septate pattern may be eroded. Finally the bony deficit in the sinus wall may become visible.

Successful treatment involves a combined sinus and orbital approach, for it is vital to strip the respiratory epithelium from the wall of the offending sinus and to re-establish a drainage pathway to the nose in order to prevent recurrence.⁶ However, our three patients with orbital respiratory epithelium lined cysts had no evidence of a mucocele or underlying sinus disease at the time of presentation, and no evidence of bony dehiscence or destruction was

found radiologically or at the time of surgery. Effective treatment consisted in excision without recourse to sinus surgery, and there have been no

Fig. 1 Proposed pathogenesis of post-traumatic respiratory epithelium lined cysts in the orbit contrasted with orbital mucocele development. (a) Normal orbit and nasal sinuses. (b) Frontal sinus mucocele development following blockage of the sinus drainage pathway. (c) Trauma to the sinus region results in the transfer of sinus epithelial cells to the orbit through a small dehiscence/fracture of the orbital wall, (d) with the subsequent development of a respiratory epithelium lined cyst.

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Fig. 2 Case 1. Right eye with mass in the superonasal quadrant of the orbit.

recurrences. We consider that another mechanism must be invoked to explain the presence of respiratory epithelium in the orbit.

Case reports

CASE 1

A 26-year-old man presented with a faintly blue cystic mass in the superonasal quadrant of his right orbit. At the age of 12 he had received a blow to his face requiring sutures to a laceration of his right eyebrow. Since then he had noticed a small lump beneath the scar. During the year before he presented the lump had slowly increased in size (Fig. 2). His visual acuity was 6/5. There was no proptosis or ocular displacement. Orbital and sinus x-rays were normal. At surgery a postseptal brown cystic lesion was revealed which leaked oily brown fluid as it was being removed. Posteromedially there was a thin 'tail' from the cyst to the medial wall of the orbit, but no bony defect was seen. Postoperative recovery was uncomplicated.

CASE 2

A 23-year-old man presented with a four-year history of a slowly increasing mass in the superomedial quadrant of his left orbit. Ten years previously he had received a blow to the left brow requiring sutures. The visual acuity was 6/6, and the left eye was displaced inferolaterally but not proptosed. A palpable cystic lesion was present superomedially. Orbital and sinus x-ray views were normal. A CT scan showed an ill defined soft tissue mass in the left orbit (Fig. 3).

At surgery a subperiosteal cystic lesion was revealed overlying the ethmoid sinus. It was dissected free and found to contain brown oily fluid. There was

no obvious bony defect. The patient made an uncomplicated recovery.

CASE 3

A 20-year-old man presented with a 10-month history of a slowly increasing mass in the inferomedial quadrant of his right orbit. Eighteen months previously he had had a right sided maxillary sinus washout procedure. There had been several episodes of painful increase in size of the mass over the 10 months before presentation. The visual acuity was 6/6 and the right eye was displaced superolaterally and proptosed 2 mm. A faintly blue cystic lesion was present inferomedially (Fig. 4).

Orbital and sinus x-rays were normal; in particular

Fig. 3 Case 2. Axial CT scan with ill-defined soft tissue mass medial to the left eye (arrowed).

Fig. 4 Case 3. Right eye with mass in the lower inner quadrant of the orbit.

there was no sinus clouding. CT scans revealed a soft tissue mass in the right orbit. At surgery a postseptal brown cystic lesion was revealed. It contained brown oily fluid and there was a thin 'tail' from the cyst to the medial wall, but no bony defect was seen. Post-operative recovery was uncomplicated.

PATHOLOGY

The histopathological examination of the three orbital lesions showed each to be a thin walled cyst lined by a pseudostratified columnar epithelium containing scattered mucin-producing cells (Fig. 5).

There was evidence of focal epithelial atrophy, possibly due to pressure effects. In case 2 there were subepithelial foci of active chronic inflammation, with many macrophages containing lipoid material, indicating a response to leaked cyst contents (Fig. 6). In areas of the epithelium associated with a heavy chronic inflammatory response there was metaplasia

of the pseudostratified columnar epithelium to squamous epithelium. No epidermis or dermal appendages were present within the wall of any of these cysts, ruling out a diagnosis of dermal inclusion cyst or dermoid.

Discussion

The preoperative differential diagnosis in our first two cases included orbital varices or an implantation cyst, and we were surprised to find cysts lined by upper respiratory tract epithelium in the absence of an underlying mucocele. However, the characteristic clinical picture of a young man presenting with a post-traumatic, faintly blue cystic lesion lying in the medial orbit, with normal orbital x-rays, enabled us provisionally to diagnose our third case pre-operatively. In the two cases preceded by trauma to the region our hypothesis is that a tiny fracture or dehiscence allowed herniation of nasal sinus epi-

Fig. 5 Microscopic appearance of the cyst lining (case 3), comprising pseudostratified columnar epithelium with an occasional mucin-secreting goblet cell (arrow) (H and E, $\times 122$).

Fig. 6 Cyst lining (case 2) showing squamous metaplasia, and dysplasia of the epithelium, with underlying chronic inflammation which includes many macrophages (arrowed). (H and E, $\times 128$).

thelial cells into the orbit, with the subsequent development of a slowly growing cyst (Fig. 1). The third case could be related to underlying sinus disease which had subsequently resolved by the time of ophthalmic presentation. However, we feel the trauma of the maxillary antral washout may have had the same herniating effect as we have proposed for accidental trauma.

In summary, while cysts lined with upper respiratory tract epithelium classically occur in the orbit as a result of mucocoele development, we have described three young men with orbital cysts lined with upper respiratory tract epithelium following trauma in whom no evidence of mucocoele was found. Straight-forward excision resulted in a good prognosis, and there have been no recurrences.

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