Intracorneal and sclerocorneal cysts

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SUMMARY Two cases of corneal cysts are reported, one sclerocorneal and the other intracorneal. The two conditions differ in pathogenesis, appearance, course, and management.

The presence of a corneal cyst is a rare clinical condition. Only a few cases have been reported in the literature. Most of the corneal cysts have a scleral component from where they actually originate. These are described as congenital and progressive in course, although scleral cysts without corneal involvement have been reported. A small number of cysts are strictly corneal and of presumed congenital origin. Nevertheless, they can be a product of local trauma or follow ocular surgery.

There are differences in appearance, pathogenesis, treatment, and prognosis between the intracorneal and sclerocorneal variants of this cystic condition, and these are demonstrated by the following report of two such cases.

Case reports

CASE 1

A White baby girl was first taken to the local ophthalmologist at the age of 10 months because her

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mother had noticed a small spot in the right eye. This was diagnosed as a conjunctival cyst, and no specific treatment other than simple observation was undertaken. The cyst gradually increased in size, and when at the age of 2½ years the patient was referred to this unit an examination under anaesthetic was performed with the following findings.

A small scleral cyst was present at the superonasal quadrant beginning at 2 mm from the limbus and invading the cornea for a further 2 mm at the level of mid-stroma. The remaining cornea was normal, but faint scarring at the level of Bowman's membrane surrounded the corneal extension. Gonioscopy revealed a normal angle, and examination of the rest of the eye showed no abnormality.

No surgery was performed until 13 months later, when the cyst was much larger, involving the nasal edge of the pupillary axis, reducing the visual acuity to 6/36. In addition cell debris simulating a hypopyon level could be seen occupying the bottom of the cystic cavity (Fig. 1). At that stage aspiration was performed, and the aspirate was identified histo-

Fig. 1 Sclerocorneal cyst. Note the scleral origin of the cyst (black arrow) and the cell debris simulating a hypopyon level in the cavity of the corneal extension (white arrow)

logically as containing 'numerous degenerate epithelial cells' (Fig. 2).

During the next 6 months the cyst refilled and increased further in size. Thus, the anterior wall was removed surgically to the largest possible extent and the remaining portion was cauterised. The nasal part of the cornea was left, without graft, using the epithelial lining of the cyst as the covering layer. Histological examination confirmed the diagnosis of inclusion cyst resulting from congenital sequestration of epithelium (Fig. 3).

Four months later the cyst has not re-formed and the cornea has healed with a thin, even scar. The

visual acuity remains unchanged, and because of this a lamellar graft is planned for the near future.

CASE 2

A healthy White boy 10 years old presented to the clinic because his mother had noticed a spot on his right eye. There was some vague history of local injury some years ago but otherwise nothing significant. There was no family history of eye disease apart from a grandfather who apparently was blind from retinal disease.

Examination showed the visual acuity 6/6 in each eye, with no significant refractive error. On the right

Fig. 2 *Histological appearance of the cyst aspirate. Multiple epithelial cells can be seen, the darker ones representing the better preserved cells. H and E. $\times 125$*

Fig. 3 *Histology of the anterior wall of the cyst. Squamous epithelium covers both sides of the wall. H and E. $\times 50$*

side there was a limbal intracorneal cystic lesion in the superonasal quadrant (Fig. 4). There were no blood vessels, but a nebulous opacity outlining the lesion was present. The anterior wall of the cyst was formed by epithelial cells and perhaps Bowman's membrane. The posterior wall was formed by a thin layer of Descemet's membrane in the centre of which there was a pinpoint circular gap covered only with endothelial cells (Fig. 5). The adjacent sclera was normal, and gonioscopy revealed a wide open angle. The rest of the right eye and the entire left eye were normal. This cystic lesion has been observed for over 6 years, and no change has occurred at all.

Discussion

The origin of the sclerocorneal cyst is clear from the histological picture of the aspirate and the cyst wall. The presence of epithelial cells points towards a sequestration of conjunctival epithelium, and this is in complete agreement with most of the previously described cases (Fox, 1928; Huber, 1930; Lepri, 1947; Bischler, 1947; de Ferrari, 1950; Mandras, 1951). An alternative theory was put forward by Wernicke (1908) when, in describing a case, he suggested that the cyst was an extension of the anterior chamber. Custodis (1932) demonstrated that such cases were out-

Fig. 4 Slit-lamp appearance of intracorneal cyst. Note absence of scleral involvement

Fig. 5 Higher magnification reveals a pinpoint gap in the posterior wall of the cyst (large arrow), covered only by endothelium (small arrow)

growths of the canal of Schlemm, and Vrolijk (1941) favoured a similar theory. This was not the pathogenesis in the present case, because no continuation with the anterior chamber was present and gonioscopy showed a normal angle. Babel and Avanza (1966), reviewing 18 reported cases, stated that they usually become evident at the age of 5 or 6 years. The present case is rather unusual in becoming apparent at the age of 10 months, although the youngest patient reported was that of Custodis (5 months).

The fact that simple aspiration was followed by no significant improvement underlines the importance of excising the anterior wall if a more permanent cure is desirable. As the course is progressive, failure to treat will impair the sight by extending over the pupillary area. In that case corneal grafting may be necessary in order to restore maximum vision.

Intracorneal cysts without scleral involvement are very rare. Appia (1853) was the first to describe such a case, and authors of the majority of the approximately 30 cases reported since accept a congenital origin, analogous to that of the sclerocorneal cysts. However, some authors have suggested a traumatic origin, for example Ruedemann (1956) reporting a case associated with a foreign body, and Reed and Dohlman (1971) reporting 8 cases following either corneal surgery or undetected injury.

The traumatic theory is more attractive, and, although in many cases no history of injury is found, a trivial, undetected accident is sufficient to force a number of epithelial cells into the stroma, where they become isolated owing to closure of the surface wound. This theory is supported further by the normal appearance of the sclera and the anterior chamber, the clinical similarity with the traumatic cysts, and the non-progressive course of the condition.

The latter is significant in connection with the management of the intracorneal cysts. On diagnosing such a condition a conservative approach should be adopted, and only in the unlikely event of extension of the cyst should surgery be undertaken.

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