Keratoconus

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Keratoconus as an entity is now so well documented that it is unlikely to engender great excitement in itself. However, the following case report involves such an unusual sequence of events that it would seem worth considering.

History

An active, apparently healthy, adult male, aged 25 years, had been 6 years earlier diagnosed at having bilateral keratoconus. Contact lenses were prescribed and comfortably worn. His past ophthalmic history goes back many years. Myopia was diagnosed at the age of 3 years and glasses were prescribed, and these were changed at intervals until the diagnosis of keratoconus was made.

There was nothing significant in the family history. The patient's general health was described as excellent throughout.

The first unusual ocular episode was a rupture of Descemet's membrane with gross corneal oedema affecting the right eye. Less than a year later a similar acute rupture of Descemet's membrane affected the left eye. After an interval of 3 months the visual acuity in the right eye, with correction was 6/12 and in the left 6/36. At this point it was decided to perform a 7 mm. full thickness graft on the left eye. The operation and postoperative period are recorded as being "without incident". The graft remained clear for a while with a visual acuity of 6/18 with contact lens correction.

Several months later an iris cyst appeared in the operated eye in the upper outer quadrant. This was excised 16 months after the initial graft procedure, through an external incision in the 2 o'clock meridian.

Postoperatively the eye never settled down and was always uncomfortable; a low-grade uveal reaction persisted and the visual acuity was never better than 6/24.

The patient was not seen again for almost a year, when he returned complaining of continued discomfort and the appearance of a small "blister" at the site of the incision for the removal of the iris cyst.

Clinical findings

The following features were noted:

- (1) A small "bleb" at the site of the incision for the removal of the cyst.
- (2) Lens opacity.
- (3) Marked uveal reaction.
- (4) Ocular hypertension of marked degree.
- (5) Deterioration of vision to doubtful perception of light.

THERAPY

In consultation with a colleague, antiglaucoma therapy was instituted, as the patient refused further surgical intervention of any kind. The bleb, at that time, was considered to be due to dehiscence of the wound from raised intraocular pressure.

PROGRESS

Nothing further was heard from the patient for 16 months. He returned with the complaint that the eye had become unsightly, in spite of his continued antiglaucoma therapy.

A large cystic swelling extended from 12 to 6 o'clock on the temporal side of the limbus (Fig. 1).

The graft showed band-shaped calcareous degeneration. The anterior chamber was quiet but with signs of previous uveitis. The lens was absent having been absorbed but remnants of lens capsule remained. There was no perception of light.

CYST

The patient refused to lose the eye and therefore the opportunity to explore this enormous cystic swelling presented itself.

It was found to be a large intrascleral cyst, communicating with the anterior chamber through the old incision for the removal of the iris cyst (Fig. 2). Aqueous had slowly and progressively split the sclera and forced its way round the temporal side to produce the picture seen above. Pathologically the cyst was lined with epithelium, and no additional information about the sclera was obtained from the many sections taken and stained with the various methods. Electron microscopy was not done.

TREATMENT

As permission for enucleation had not been granted, the cyst was cauterized, the redundant sclera excised, and a fistula created in the original wound area, so that drainage could occur subconjunctivally, as in a filtering procedure.

RESULT

One year later the patient reports that "the discomfort has been negligible and the eye is, cosmetically, far more acceptable. I sense that the pressure is more nearly what it should be."

Comment

No reference could be found in the literature to intrascleral splitting as noted in this case. The aqueous, once it had found its way into the *ab externo* incision, under the constant high intraocular pressure, continued to dissect the sclera rather than to find a subconjunctival outlet, as in the usual filtering procedure. The underlying pathology which had caused the keratoconus and two subsequent ruptures in Descemet's membrane may have had a bearing on this sequence of events.

It is interesting to link the two processes, as both could be explained on the basis of a weakness in resistance to the normal (or raised) intraocular pressure.

Before the sclera could be split and fluid accumulate in the space, the *ab externo* incision must have healed, at least in its anterior half, otherwise the anterior chamber would have communicated with the subconjunctival space. This was shown to be the case at operation. That the scleral space merely acted as a reservoir and not as a drainage system is evidenced by the persistently high intraocular pressure. It seems that this unremitting pressure continued to split the sclera.

It was established at operation that communication existed between the scleral cyst and the anterior chamber. From this it must be concluded either that the posterior part of the wound did not heal or, more likely, that the wound healed soon after the operation for the removal of the cyst but secondary glaucoma developed after the attack of uveitis. Under the force of increased pressure, the posterior layer ruptured at the site of incision—possibly in a previously weak area—and perhaps by a process comparable to the rupture of Descemet's membrane in keratoconus. Aqueous then tracked into the sclera and split it to the extent shown.

Nothing happens without reason—and in an attempt to find the reason the following explanation is offered:

FIG. 1 *Cystic swelling of firm consistency on temporal side of limbus*

FIG. 2 *Scleral splitting and communication of cyst with anterior chamber through original ab externo incision*

The inherent weakness in keratoconus exists in the posterior layers of the cornea. Evidence for this is found in the case of acute corneal hydrops which is caused by ruptures in Descemet's membrane. The exposure of sclera to aqueous occurs in the Elliott trephine procedure. No splitting of sclera seems to occur there—even if the trephine closes, and the

intraocular pressure continues to rise. It is suggested that in keratoconus the scleral structure may be affected as well as the cornea. In this case, the iridectomy incision predetermined the site of "rupture of Descemet's". The anterior layer healed well and held fast. Once the aqueous had penetrated the posterior wall, it continued to split the scleral lamellae—being prevented from reaching the subconjunctival space by the firm healing of the anterior layers. This conforms to the suggestion that the inner and not the outer layers are the weak links in the formation of the cone, in keratoconus.

Summary

A case of keratoconus is presented with a sequence of unusual complications that confirms the view that it is the inner layers of the cornea that are at fault, and that the scleral structure may be similarly involved. The case is presented not only for its interest, but in the hope of stimulating further study.

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