Dermoid cyst of the iris

Report of an unusual case

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The appearance in an eye of a visible mass reported to be increasing in size over a short period of time must always give rise to thoughts of malignancy. The diagnosis before any definitive treatment is begun may present difficulty in deciding on the specific surgical approach, if any, and on the extent of any surgery.

Case report

A white man aged 24 years arrived in Cape Town in October, 1970, from a distant part of Africa for the purpose of obtaining advice and treatment for his right eye. He had noticed a white nodule in the eye for the first time about 4 weeks earlier and he was certain that it had been increasing in size since then. There was no pain, nor was there interference with the vision of that eye, such as it was, but the appearance was worrying him. He insisted that the duration was no more than 4 weeks.

He volunteered the information that in March, 1968, he had sustained an injury to the eye which necessitated an operation, after which he was advised to have a contact lens fitted.

Examination

The left eye was found to be normal in all respects. The right eye was aphakic. With + 12 D sph., the visual acuity was 6/24. There was a coloboma of the pupil, presumably surgical, stretching from 2 to 6 o'clock on the limbus. Displacing the upper part of the free pupil margin was a large white rounded mass which occupied about one-quarter to one-fifth of the anterior chamber. It appeared to be attached to and continuous with the iris, which was of a dark brown colour, but the mass itself was almost dead white and semi-translucent (Fig. 1).

FIG. 1 Appearance of white mass in the eye
The intraocular pressure was not raised. There was no obvious sign of an operation wound. The fundus appearance was normal and so was the remainder of the iris.

Slit-lamp examination revealed a semi-solid structure with little transillumination (Fig. 2). There were no vessels to be noted, either superficial or deep. X-ray examination revealed no opaque foreign body in the eye or orbit.

I photographed the eye and observed the appearance on several subsequent occasions; 2 weeks later I thought that the mass was a little more prominent.

He was admitted to Groote Schuur Hospital for further examination.

The diagnosis was that of an implantation cyst, but the differential diagnosis also considered a granuloma pyogenicum and a neoplasm. It was generally agreed that the mass should be removed.

Operation

On October 23, 1970, an operation was performed under general anaesthesia (pethidine, gas oxygen, and halothane). 500 mg. Diamox was administered intravenously 10 minutes before induction.

A large limbus-based conjunctival flap was raised above and medially to allow a limbus incision from 10 to 3 o'clock. After entering the anterior chamber at 1 o'clock, the incision was extended medially and laterally for about 6 to 8 mm. and two precautionary 6/0 black silk edge-to-edge sutures were placed before extending the incision further. The swelling presented in the wound and its free medial edge was delivered upwards and laterally. The medial suture was tied and the swelling was then gripped with non-toothed forceps and extracted intact so that the iris pedicle could be severed. The pillar of iris was replaced in the anterior chamber and the second silk suture was secured. The wound was then further secured by a number of edge-to-edge sutures, using 8/0 virgin silk. The two black sutures, now redundant, were removed, air was introduced into the anterior chamber, and the conjunctival flap sutured with continuous 7/0 chromic catgut.

Result

There was no vitreous loss in this procedure. The eye recovered uneventfully; 10 days later examination revealed a clear view of the fundus. The vitreous face, not unexpectedly, was no longer intact and was mixed with aqueous. The iris coloboma was even larger than ever, and there was only about one-third of the iris left (Fig. 3). The visual acuity, with +12 D sph., was 6/18.

The specimen proved to be a rounded soft mass measuring about 6 × 4 mm.

Biopsy report

A well-defined dermoid cyst lined by squamous epithelium and containing keratin material. Fig. 4 (low power × 80) and Fig. 5 (high power × 500) show uveal pigmented tissue (a), squamous epithelium (b), and keratin (c).
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Prof. Uys of the Department of Pathology pointed out that this cyst could be developmental. Dermoid cysts usually occur in the eye on the limbus, but have been described in the iris and ciliary body, originating from the iris ciliary epithelium. More commonly, they come from the conjunctiva through implantation, and in this case the history makes it the more likely, but the histological appearance may not give any clue.

FIG. 3 Postoperative appearance of the eye

FIG. 4 Section of mass \( \times 60 \)

FIG. 5 Section of mass \( \times 200 \)
Discussion

Although a diagnosis of a cyst of the iris was never seriously challenged, there were some unusual features which made it necessary to exclude a neoplasm. Its growth seemed unusually rapid. It did not appear cystic on slit-lamp illumination.

Tumours of the iris are uncommon, and non-pigmented tumours are very rare. In searching the available literature, including Duke-Elder (1954) and Duke-Elder and Perkins (1966) I could find no description of a similar tumour which was neoplastic. A malignant melanoma may present as a non-pigmented mass, but is never quite white. The leiomyoma and leiomyosarcoma have been described as pinkish or greyish-white and are always vascular on the surface.

In the differential diagnosis of a growing mass attached to the iris, every described uveal tumour must in theory be considered, but the clinical appearance in this case would certainly exclude a malignant growth: white, semi-translucent, avascular, and causing no pain or raised ocular tension, or any interference with vision at this stage. Enucleation is therefore not justified.

In a review of 7,877 eyes, Ferry (1965) found that 644 contained a malignant melanoma of the uvea, and of these 54 were of the iris, of which 49 were visible, and the diagnosis correct in 45; 66 eyes were enucleated for a supposed malignant melanoma of the iris and in some 35 per cent. of these there was, in fact, no tumour. In only one was an iris cyst mistakenly called a malignant melanoma. This was a cyst of the posterior chamber lined by stratified epithelium introduced from surface epithelium in the course of a previous injury. The diagnosis was “cystic melanoma of the iris and ciliary body”.

It would appear therefore that, of various lesions that simulate a malignant melanoma, an iris cyst is least likely to be so regarded.

Nauman and Green (1967) reported two cases of spontaneous non-pigmented iris cysts—both in infants—and in both cases the eyes were enucleated because of the rapid growth, in one because of suspected neoplasm. In neither case was the appearance that of a yellow swelling, and both were lined by cuboidal epithelium with goblet cells. They quoted Thiel as stating that it may be difficult to distinguish between an early malignant melanoma and a non-pigmented cyst deep in pigmented iris stroma, but Ferry (1965) in his study of pseudomelanomata encountered no such case. They concluded that the enucleation in one of the cases was unnecessary and emphasized that non-pigmented iris cysts should always be considered in the differential diagnosis of lesions of the iris in infants.

El-Bayadi and El-Defrawi (1965) described two cases of epithelial cyst. One, in a child of two, was lined by stratified columnar epithelium containing goblet cells. This was taken to prove that the cyst had developed from surface ectoderm.

In the other, in a woman aged 65, there was a history of lens extraction a year previously. The cyst was lined by stratified squamous epithelium and was attached to the anterior mesodermal layers of the iris.

Fine (1969) described a free-floating pigmented cyst in the anterior chamber in a two-year-old Indian male. There was no history of injury. Histology showed pigmented cells, presumably of neuro-ectoderm and derived from iris pigment epithelium in the posterior layers. The cyst increased over a period of 6 months and so obscured vision that it had to be removed.

Esposito (1965) recorded a similar free-floating cyst and its removal from the anterior chamber. It was reported simply as consisting of “iris pigment cells”.

Ferry and Naghdi (1967) described in detail the cryosurgical removal of an “epithelial
Dermoid cyst of the iris and anterior chamber", in a 75-year-old woman who had previously had an operation for lens extraction in that eye. This was therefore regarded as an implantation cyst.

Wilson (1964), in a review, wrote that spontaneous cysts of the iris are rare, whether pigmented or non-pigmented, and are probably in all cases derived from neuro-ectoderm of the rim of the optic cup. Traumatic cysts, however, are epithelial implants. They are pearly in colour and the walls are lined with stratified or cuboidal epithelium.

Reese (1968) described an “epidermoid cyst of the iris and ciliary body” pearly-white in appearance and present since birth in an infant girl aged 2 years. Excision was advised because of the possibility of a dictyoma and because the cyst appeared to be growing in size. The operation planned was a corneo-scleral iridocyclectomy using a 6 mm. trephine and removing the cyst-bearing uvea. The wound was closed by a corneal graft and conjunctival flap.

The microscopical examination revealed an epidermoid cyst containing flakes of keratin. He stated then (in 1965) that he had not been able to find any report in the literature of dermoid cysts of the iris or ciliary body, although Dr. Lorenz Zimmerman had told him that the Armed Forces Institute of Pathology had observed three such cases.

He went on to state that spontaneous epidermoid cysts arose from congenital remnants of primitive ectoderm at the closure site of a foetal cleft.

Such cysts must therefore appear early in life, as reported in several of the cases quoted. A cyst arising in adult life, with the history of trauma or surgery preceding the appearance, is very much more likely to be an implantation cyst.

Summary

(1) A cyst of the iris, of rapid growth and unusual appearance, is described.
(2) The operation for removal is described.
(3) The difficulty of preoperative diagnosis and the distinction between congenital and acquired iris cysts are discussed.

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