Leiomyoma of the ciliary body

A clinico-pathological case report

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Although partial cyclectomy has become an established procedure for the excision of tumours of the ciliary body, difficult clinical decisions are required before embarking on this surgery. Anxiety is caused especially by the uncertainty of unduly disturbing a large malignant lesion and by the surgical risks to the eye in relation to the chances of success in maintaining the integrity of the eye and possible vision. The larger the tumour the more the uncertainty, and hence the interest of this case.

Case report

A housewife aged 24 years was seen by Dr. Veronica Hughes at the Angau Memorial Hospital, Lae, New Guinea, on August 6, 1968. She had accidentally discovered that the visual acuity of the right eye was poor only 2 weeks previously. At that time it was 6/24, with +2 D cyl., 90°. In the left eye the unaided vision was 6/5. She was referred to Melbourne for treatment.

Examination (December 4, 1968: R.F.L.)

The right eye was free of inflammation and showed a large, almost smooth, brown tumour behind the iris in the 12 o'clock position, considerably indenting the equator of the lens and presumably arising from the ciliary body. Gonioscopy showed the peripheral iris bulging in two places at the 12 o'clock position thereby closing the angle but apparently by iris apposition and not by adhesions or infiltration. Elsewhere the angle was open. Transillumination was negative indicating the absence of a cyst.

When the pupil was dilated with cyclopentolate eyedrops the iris moved freely over the mass exposing more of it and the adjacent lens which was deeply indented and contained heavy local opacities (Fig. 1). The tumour appeared confined to the ciliary body with no extension into the retina. The ocular tension was normal.

FIG. 1 Large tumour of ciliary body indenting the lens and causing extensive local opacities

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DIAGNOSIS
The patient was of the Phillipino race born in Manila. In view of her age and the localized appearance of the tumour, and because malignant tumours of the uveal tract are relatively rare in Asians, a clinical diagnosis of probably benign tumour of the ciliary body was made and removal by partial cyclectomy was recommended.

SURGERY
On December 18, 1968, after intensive administration of oral acetazolamide and intravenous mannitol, a partial cyclectomy was performed, using the method described by Stallard (1961) as a guide. At the 12 o'clock position a fornix-based conjunctival flap was reflected, a 180° limbal incision was cut with razor blade and scissors, and a Y-shaped incision was made extending back to the superior rectus tendon. Using a cyclodialysis spatula, flaps were mobilized and diathermy was applied to the edges of the ciliary body around the tumour.

The tumour could be vaguely identified and the iris was snipped across just anterior to the iris root. The tumour then presented and was held by an erisophake while it was lifted and the ciliary body incised around it, so that it could be totally removed. No vitreous was lost; the lens was not seen. The limbus incision was closed with virgin silk sutures, the sclera by white silk. The fornix-based flap was drawn down and sutured at the sides to cover the wound.

Postoperative progress was slow but uneventful. The anterior chamber reform ed slowly but remained clear, and the pupil was roughly circular. Atropine eyedrops were used to assist in tightening the zonule to minimize anterior lens displacement, but atropine sensitivity required their cessation after several weeks.

The patient returned to New Guinea under the supervision of Dr. Veronica Hughes. One month after operation the corrected visual acuity was still 6/24 although lens clouding had increased. The optic disc and retina could be seen vaguely and appeared intact.

REVIEW AFTER 12 MONTHS
The patient was re-examined (by R.F.L.) in Melbourne in January, 1970. The right eye was free from inflammation and cosmetically good in alignment and appearance. The upper eyelid hid the operation scars.

Apart from these scars the cornea was clear. The right pupil was semidilated and roughly circular and many posterior synechiae were present with the upper iris plastered to the lens. The lens was translucent white and tilted forwards above with the upper lens equator adherent to the operation scar (Fig. 2). The right anterior chamber was 2.4 mm. deep at the centre, whereas the left anterior chamber was 2.8 mm. deep. Gonioscopy showed the angle widely open in the lower quarter in the right eye, but in the other three-quarters, the angle was smothered by iris at the sides and by lens above (Fig. 3). Applanation tonometry showed that the ocular tension was 17 mm. Hg in each eye.

FIG. 2 Right eye one year after excision of ciliary body tumour

FIG. 3 Postoperative goniophotograph, showing iris and lens adherent to partial cyclectomy scar
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The visual acuity in the right eye was reduced to perception of hand movements, but light projection was very quick and accurate in all areas. The eye appeared generally healthy and in good condition.

Lens removal by irrigation appeared possible, but this was deferred because the patient was returning to New Guinea where ophthalmological attention would not be available. Contact lens wearing seemed too difficult.

Pathology report (C.H.G.)

Macroscopic appearances  The specimen was an ovoid tumour measuring 8 x 6 x 5 mm., with a smooth dark brown external surface and a yellowish-grey finely granular cut surface (Fig. 4).

Microscopic appearances  Sections showed a typical leiomyoma of neoplastic spindle cells (Fig. 5) which fulfilled all the histological criteria cited below. The outer aspect of the growth was invested by the compressed tissues of the ciliary body in which many seams of pigmented ciliary epithelium were evident (Fig. 6). Part of the iris was attached to one pole. The tumour was rich in myofibrils (Fig. 7). Scanty reticulin fibres and small numbers of uveal melanocytes were dispersed among the tumour cells. In the base of the growth, where it had been attached to the ciliary body, normal ciliary muscle was recognizable, the orderly arrangement of its cells contrasting with the more haphazard disposition of those constituting the tumour.
Discussion

Leiomyomata of the ciliary body are very rare tumours. A review of the world literature (Blodi, 1950) revealed only ten reported cases, none of which was entirely acceptable by modern histological criteria. These criteria comprise the following features:

(1) The tumour is composed of interlacing compact bundles of spindle-shaped cells which may exhibit palisading. There is very little intervening collagenous stroma.

(2) The tumour cells contain oval nuclei with rounded ends.

(3) The tumour cells have eosinophilic fibrillary cytoplasm due to the presence of longitudinally orientated intracellular myofibrils which stain purplish blue with phosphotungstic-acid haematoxylin.

Blodi (1950) presented a description of the first ciliary body leiomyoma to fulfil the above criteria. Ten additional authentic cases have since been reported, seven of which came from the files of the Armed Forces Institute of Pathology (Meyer, Fine, Font, and Zimmerman, 1968).
Summary

A large ciliary body mass diagnosed as a benign tumour was removed by partial cyclectomy. Clinically the dark brown tumour was presumed to be composed of pigment cells, but the dark colour was derived merely from ciliary epithelium stretched over a cream leiomyoma. Increased lens opacities prevented useful vision but the eye remained cosmetically good and generally healthy. The prospects of improving the vision by cataract removal and the wearing of a contact lens were deferred but not abandoned.

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References