

SPONTANEOUS CYSTS OF THE CILIARY BODY*

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ALTHOUGH comparatively rare, spontaneous cysts of the ciliary body have evoked numerous contributions to the literature. The diagnosis is essentially clinical and differentiation from a melanosarcoma may be very difficult. These cysts were well named pseudo-melanosarcomata by Pagenstecher (1910). Before the report of Fischer (1920), the diagnosis of such cysts had been established only by histological examination of enucleated eyes. Since then, Elschmig (1925), Olsson (1944), Scheie (1954), and Grignolo (1954) have also reported cysts of the ciliary body as a clinical finding in otherwise normal eyes.

Case Reports

Case 1, a man aged 21, was admitted to hospital on May 4, 1958 for treatment of psoriasis. He was referred on account of a complaint of difficulty with close work. There was no history of injury to either eye.

The visual acuity was 6/5 in each eye. He was emmetropic and, apart from a near point of convergence of 12 cm., orthophoric. His pupils were equal and circular, reacting directly and consensually to light and on accommodation. Examination with both the slit lamp and the ophthalmoscope revealed no abnormality in the left eye.

In an otherwise normal right eye, examination with the ophthalmoscope revealed a cystic mass presenting at the inferior temporal quadrant of the posterior chamber (Fig. 1A, overleaf).

When the pupil was fully dilated, slit-lamp examination (Fig. 1B, overleaf) revealed a smooth cyst containing a clear fluid and enclosed by a thin transparent membrane which was bespeckled with fine iridescent particles on its posterior surface, *i.e.* the surface presented to the zonular fibres and vitreous face. (This fine pigment has been incorrectly exaggerated in the artist's drawing, as has been the distortion of the lens.) The cyst had insinuated itself between the posterior surface of the iris and the anterior surface of the lens. Where the cyst was in contact with the anterior lens capsule, there were a few subjacent opaque lens fibres. The cyst was tremulous on eye movements. The Goldmann lens showed that the cyst arose from the anterior part of the ciliary body. The rest of the ciliary body appeared normal. The angle of the anterior chamber, including the sector overlying the cyst, was of normal depth. There was no disturbance of pigmentation in the overlying iris. The tension in both eyes varied between 20–25 mm. Hg (Schiotz) over several serial readings. There was no evidence of any penetrating injury.

This patient was observed for 6 months until he left the country, and during that time

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there was no variation in the appearance and size of the cyst. The intra-ocular pressure remained at 20–25 mm. Hg (Schiötz). The convergence insufficiency responded to orthoptic exercises, and his symptoms were relieved.

Case 2, a man aged 19, was referred on July 14, 1958, with the complaint of a painful left eye. There was no history of injury to either eye.

The visual acuity was 6/5 in each eye and he was emmetropic. The right eye was normal. There was a patch of scleritis involving the left eye at the upper nasal quadrant, 6 mm. from the limbus. Slit-lamp examination revealed no flare, no cells in the anterior chamber and no keratic precipitates.

At the inferior temporal quadrant a cystic mass was seen with the ophthalmoscope at the extreme periphery of the fundus (Fig. 2A, opposite). It projected medially and slightly forwards, and the fundal vessels did not pass over it. Examination with the Hruby lens (Fig. 2B) showed the mass to be definitely cystic and tremulous. The cyst had a multilocular appearance and was a light reddish-brown colour. It transilluminated well. Indirect ophthalmoscopy and the Goldmann lens confirmed the suspicion that the cyst arose from the posterior part of the ciliary body. The angle of the anterior chamber was normal, as was the iris, and the ocular tension remained within 22–25 mm. Hg (Schiötz) over several serial readings. There was no evidence of a penetrating injury.

The scleritis responded well to treatment with topical corticosteroids.

This patient has been watched up to the time of writing and there has been no alteration in the appearance or size of the cyst. The intra-ocular pressure has remained normal.

Aetiology

Spontaneous cysts of the ciliary body show a separation of the pigmented and non-pigmented layers of the epithelium of the ciliary body. Their pathogenesis has excited a wealth of theories, *e.g.* inflammation (Collins, 1890); persistence of the annular sinus of von Szily (Wintersteiner, 1906); choroiditis causing adhesion of ciliary processes to one another (Coats, 1907); foetal iritis causing synechiae with resultant separation of the two layers of the secondary optic vesicle (Loewenstein and Foster, 1947).

Vail and Merz (1952), in an excellent paper which includes a review of the literature, suggest that the cysts are embryonic in origin. They put forward evidence suggesting that these cysts might be formed by the traction of the zonule on the ciliary epithelium with consequent separation of the two layers of the secondary optic cup, thus opening up what is normally only a potential space. The action of the zonule during accommodation would aggravate this condition. This theory accounts for the fact that smaller ciliary body cysts are found in the valleys between the ciliary processes (Reese, 1950; Garron, 1953; Scheie, 1954) where the greatest number of zonular fibres arise, (Wolff, 1954).

On the basis of a histological examination of a grossly abnormal eye and clinical examination of another equally abnormal eye (which he had treated surgically), Purtscher (1940) also suggested prenatal factors as the cause. He maintained that pigment cysts of the posterior chamber are caused by an early and intimate adhesion of the posterior pigment epithelium of the iris and the tunica vasculosa lentis, in the presence of a disturbance in the

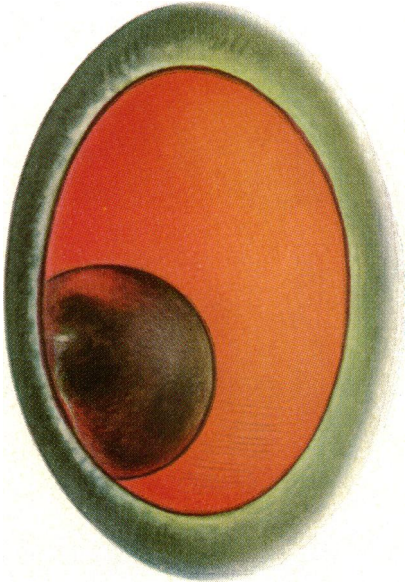


FIG. 1A.—Cystic mass seen with the ophthalmoscope

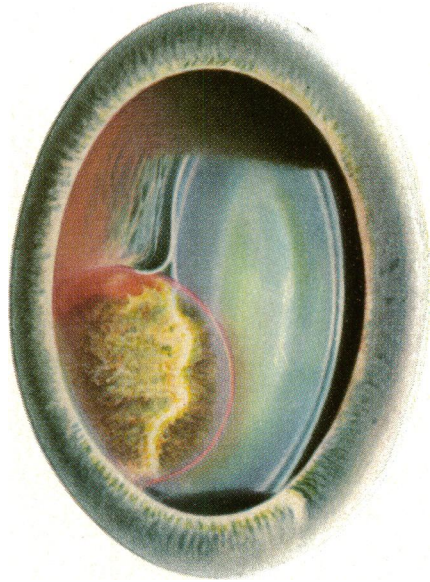


FIG. 1B.—Cyst seen with the slit lamp

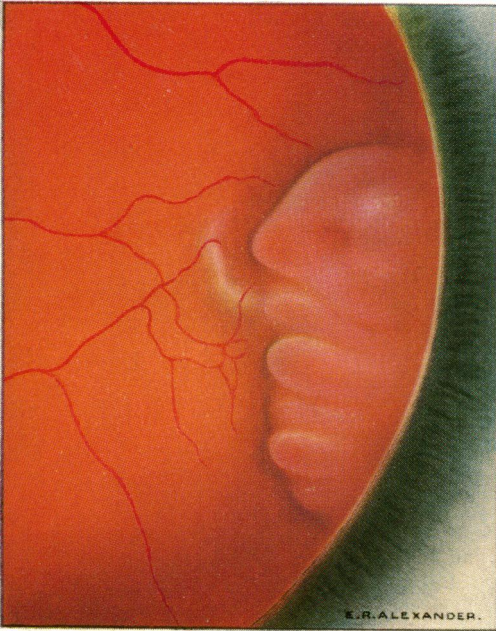
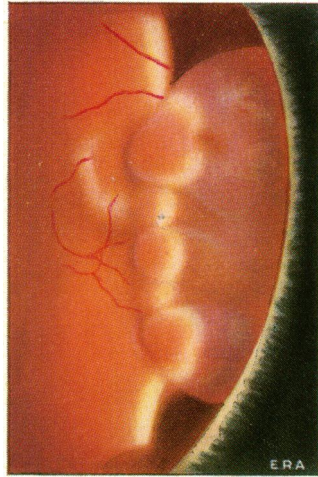


FIG. 2A.—Cystic mass seen with the ophthalmoscope.



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FIG. 2B.—Cystic mass seen with the Hruby lens.

regression of the tunica vasculosa lentis, especially in the neighbourhood of the pupillary membrane. He suggested that hypertrophy of the ciliary processes occurred as a result of tension upon them when the growing globe slowly pulled them from the fixed lens.

Purtscher's theory correlated with that of Loewenstein and Foster (1947) may explain the occurrence of ciliary body cysts in association with other developmental defects and abnormalities, such as those reported by McCrea (1936), Badeaux (1936), Trevor-Roper (1948), and Bonaccolto (1957).

Vail and Merz (1952) do not explain why the separation of the two layers of the ciliary body epithelium does not normally occur with the tension of the zonular fibres on accommodation. Attempting to reconcile the views of Purtscher (1940) and Vail and Merz (1952), one might envisage a temporary adhesion between the embryonic ciliary processes and the lens, this attachment breaking down as the differential growth of the foetal eye proceeds. There will now be a weakness in the union of pigmented and non-pigmented layers of the ciliary epithelium. In post-natal life, the contraction and relaxation of the zonular fibres during accommodation will act on this point of weakness, and the apposition of the two layers of epithelium will give way to form a cyst in the valley between the ciliary processes where the zonular fibres are attached.

Garron (1953) emphasized the fact that most of these cysts were situated in the inferior temporal quadrant and cited several cases taken from the literature. Scheie (1954) and Grignolo (1954) have since reported similar findings. The two cases described in this paper were also situated in the inferior temporal quadrant, but no embryological explanation for this can be suggested.

Clinical Features

The clinical features of spontaneous cysts of the ciliary body have been emphasized by Reese (1950) and Garron (1953). Any history of trauma should first be eliminated and evidence of a penetrating wound excluded by a meticulous examination of the suspected eye.

Schieck (1904) first remarked on the difficulty in differentiating clinically between a cyst of the uveal tract and a melanosarcoma, and stressed the greater frequency of melanosarcomata of the uveal tract. However, only 9 per cent. of melanosarcomata are found in the ciliary body and 6 per cent. in the iris (Duke-Elder, 1940). One need only instance cases taken from the literature where the affected eye was mistakenly enucleated to realize the importance of making the diagnosis (Schieck, 1904; Wintersteiner, 1906; Coats, 1907; Pagenstecher, 1910; Stephenson, 1916; Remky, 1923; Roth and Geiger, 1925; Meek, 1932; Vail and Merz, 1952; Garron, 1953).

These cysts are often characterized by multiplicity (Olsson, 1944; Reese, 1950; Scheie, 1954; Grignolo, 1954), and this should be confirmed or excluded

by the Goldmann lens and indirect ophthalmoscopy. These methods of investigation will also confirm the site of origin of the cyst; Scheie (1954) emphasized the value of gonioscopy when the cysts are small and indenting the iris root. In an interesting paper, Grignolo (1954) described his observations on spontaneous cysts of the ciliary body using the Schepens binocular indirect ophthalmoscope. He formed the impression that these cysts occurred quite frequently and that, contrary to previous descriptions, they were situated in the pars plana and never in the ciliary processes. (He also noted a detachment of the adjoining retina in one case and, in other patients, the co-existence of cysts of the pars plana in one sector and a flat detachment of the pars plana in another sector of the same eye or of the fellow eye).

As the non-pigmented layer of epithelium usually forms the major part of the cyst wall, spontaneous cysts of the ciliary body are usually translucent (as in Case 1), or they may have a reddish appearance due to reflected light from the underlying uveal vessels (as in Case 2). Most of these cysts have a smooth surface and trans-illuminate well and evenly. A valuable diagnostic sign is the tremulous appearance of the cyst on eye movements, which was first described by Eales and Sinclair (1896). If the cyst is in contact with the lens, the subjacent lens fibres are often opaque.

The cyst may occlude the angle of the anterior chamber where it pushes the iris root forwards. This may be sufficient to cause a secondary glaucoma (as in the cases of Wintersteiner, 1906, and Garron, 1953), making the diagnosis of malignant melanoma more likely. As the iris root is pushed forward, the anterior surface of the iris may come into contact with the posterior surface of the cornea. This may provoke a pigmentary disturbance in the affected sector of the iris and further increase the difficulty in differentiating between these cysts and malignant tumours. Reese (1956) observed that the non-pigmented layer is able to produce melanin:

“Although normally the one epithelial layer is pigmented and the other is not, even the nonpigmented layer, continuous as it is with the pigment epithelium of the iris, is potentially a melanin-producing tissue; the ability to fabricate melanin could not be a constant trait of the one layer and never manifest itself in the other. As a matter of fact, in otherwise normal ciliary epithelium, the nonpigment layer may be pigmented and thus may or may not contain melanin in its cells.”

Treatment

Surgical interference is not indicated unless the cyst gives rise to complications such as secondary glaucoma or serious opacity of the lens. Fuchs (1911) and Castroviejo (1949) have proposed diagnostic puncture of suspected cysts. (This may result in a hyphaema as in a case reported by Juler (1911) where enucleation finally revealed a melanosarcoma). However, puncture of the cyst, preferably with a diathermy needle (Villard and Dejean, 1933) and repeated if necessary, may be efficient therapeutically.

The introduction of phenol into the sac of a cyst of the iris, followed by repeated irrigation, has been successful (Wright, 1925). Two needles were inserted into the cyst, one for injection and the other for aspiration. Similarly a large implantation cyst of the iris has been treated, using one needle, by aspiration followed by injection of iodine (Alger, 1932).

Elschnig (1925) excised a cyst of the ciliary body by performing an iridectomy, removing the anterior portion of the cyst wall. Later, through a limbal incision (having turned a conjunctival flap), he removed the rest of the affected ciliary epithelium. In the majority of cases, a broad iridectomy and excision of as much of the anterior wall of the cyst as possible would probably suffice and, since degeneration of epithelial cells of experimentally produced cysts of the iris has been shown to occur with x rays (Suzuki, 1934), it would not seem unreasonable to follow this with irradiation.

Summary

Two cases of spontaneous cysts of the ciliary body are described.

It is suggested that these cysts may arise as a result of a transitory adhesion of the immature ciliary processes to the lens in the foetal eye. This soon breaks down, but in later life traction on the zonular fibres during accommodation at the former site of adhesion produces a separation of the pigmented and non-pigmented epithelium of the ciliary body to form a cyst.

The clinical features of such cysts are noted and the differential diagnosis from melanosarcomata discussed. Their treatment is reviewed.

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REFERENCES

- ALGER, E. M. (1932). *Arch. Ophthal. (Chicago)*, **7**, 984.
 BADEAUX, F. (1936). *Bull. Soc. Ophtal. Paris*, p. 68.
 BONACCOLTO, G. (1957). *A.M.A. Arch. Ophthal.*, **57**, 18.
 CASTROVIEJO, R. C. (1949). *Trans. Amer. ophthal. Soc.*, **47**, 145 (quoted by Scheie, 1954).
 COATS, G. (1907). *Roy. Lond. ophthal. Hosp. Rep.*, **17**, 143.
 COLLINS, E. TREACHER (1890). *Ibid.*, **13**, 41.
 DUKE-ELDER, S. (1940). "Text-book of Ophthalmology", vol. 3, p. 2478. Kimpton, London.
 EALES, H., and SINCLAIR, W. W. (1896). *Trans. ophthal. Soc. U.K.*, **16**, 56.
 ELSCHNIG, H. H. (1925). *Klin. Mbl. Augenheilk.*, **74**, 476.
 FISCHER, M. A. (1920). *Ibid.*, **65**, 876.
 FUCHS, E. (1911). *Trans. Amer. ophthal. Soc.*, **12**, 884.
 GARRON, L. K. (1953). *Trans. Pacif. Cst oto-ophthal. Soc.*, **34**, 125.
 GRIGNOLO, A. (1954). *Ateneo parmense*, **25**, 163.
 JULER, F. A. (1911). *Trans. ophthal. Soc. U.K.*, **31**, 44, 131.
 LOEWENSTEIN, A., and FOSTER, J. (1947). *Arch. Ophthal. (Chicago)*, **37**, 8.
 MCCREA, W. B. (1936). *Trans. ophthal. Soc. U.K.*, **56**, 377.
 MEEK, R. E. (1932). *Arch. Ophthal. (Chicago)*, **8**, 864.
 OLSSON, G. F. (1944). *Acta ophthal. (Kbh.)*, **22**, 319.
 PAGENSTECHE, A. H. (1910). *v. Graefes Arch. Ophthal.*, **74**, 290.
 PURTSCHER, E. (1940). *Ibid.*, **141**, 569.

- REESE, A. B. (1950). *Amer. J. Ophthal.*, **33**, 1738.
——— (1956). "Atlas of Tumor Pathology", Section X, Fasc. 38: "Tumors of the Eye and Adnexa", p. 12. Armed Forces Institute of Pathology, Washington, D.C.
- REMKY, E. (1923). *Klin. Mbl. Augenheilk.*, **70**, 347.
- ROTH, R. H., and GEIGER, C. W. (1925). *Amer. J. Ophthal.*, **8**, 870.
- SCHEIE, H. G. (1954). *A.M.A. Arch. Ophthal.*, **51**, 288.
- SCHIECK, F. (1904). *Klin. Mbl. Augenheilk.*, **42**, (2), 341.
- STEPHENSON, S. (1916). *Trans. ophthal. Soc. U.K.*, **36**, 270.
- SUZUKI, E. (1934). *Acta Soc. ophthal. jap.*, **38**, 553 (Abs. in *Zbl. ges. Ophthal.*, **32**, 507, 1934).
(Cited by Vail, 1936).
- TREVOR-ROPER, P. D. (1948). *Trans. ophthal. Soc. U.K.*, **68**, 282.
- VAIL, D. (1936). *Arch. Ophthal. (Chicago)*, **15**, 270.
- , and MERZ, E. H. (1952). *Amer. J. Ophthal.*, **35**, 1676.
- VILLARD, H., and DEJEAN, C. (1933). *Arch. Ophthal.*, **50**, 91, 194, 272. (Cited by Duke-Elder, 1940, p. 2445).
- WINTERSTEINER, H. (1906). *Klin. Mbl. Augenheilk.*, **44**, (2) (n.s.2), 297.
- WOLFF, E. (1954). "The Anatomy of the Eye and Orbit", 4th ed., p. 144. Lewis, London.
- WRIGHT, R. E. (1925). *Brit. J. Ophthal.*, **9**, 454.