

Communications

Essential progressive iris atrophy

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Unilateral progressive iris atrophy is uncommon and only a few cases have been reported, although the clinical picture has been recognized since Harms (1903) gave the first description of this interesting atrophy, and reviewed other cases previously reported by Johnson (1886) and Hess (1892). Duke-Elder (1966) mentions ninety authors who have reported cases.

The condition occurs much more frequently in women than in men; it usually begins in the third decade, and is commonly unilateral. The bilateral cases have been seen mainly in males, but some have also been seen in females. Other diseases have been reported in association with it, and other organic changes have been described, but are probably coincidental (hypogonadism, myotonia atrophica, Gazala, 1960).

The clinical course of a typical case has an insidious onset with slight displacement of the pupil. The atrophy is visible in the "stretched" area of the iris, opposite the side of the displacement. Large holes appear, which tend to become larger, leaving only a few fine strands of tissue attached to the rather conspicuous sphincter pupillae (Fig. 1), which is unaffected by the atrophy. It seems that stromal atrophy is most pronounced, while the atrophic changes of the ectodermal pigment give rise to a clear red reflex on transillumination. The atrophic changes in essential iris atrophy do not differ from iris atrophy caused by other diseases, but the absence of other changes (such as posterior synechiae, keratitic precipitates, pigment deposits, flare, and other signs of inflammation) aids the diagnosis. Gonioscopy is most important in clinical examination and in following the development of the condition.

Troncoso first presented a gonioscopic description (see McKeown, 1937) followed by Sugar (1945), Löhlein (1951), and Daily and Daily (1957). The anterior stromal layer of the iris is drawn towards the angle of the anterior chamber, where tissue of a compact fibrous yellowish appearance is formed fixing the iris to the limbus with peripheral anterior synechiae. When the atrophy has advanced, the tension may rise and this glaucoma offers the most serious threat to the function of the eye. Medical treatment of the glaucoma seems to have very little effect, and surgery should be attempted before sight is impaired (Duke-Elder, 1966). A sclerectomy controlled the tension for 4 years (Waite, 1928), Preziosi's filtering operation was successful for 1½ years (Papierbuch and Kurz, 1960), cyclodialysis has helped (von Grósz, 1936; Thorpe-Thorne, 1949; Vancea, 1960), repeated trephinnings and radiation controlled the tension for 11 years (Friedenwald, 1950), and cyclodiathermy for some years (Castroviejo, 1953); a total removal of the atrophic iris was suggested by Clarke (1960).

Most of the eyes described with this disease have eventually become blind because of uncontrolled glaucoma, which is as it appears from the literature, recalcitrant and resistant.

Case report

The glaucoma in the following case of a woman now aged 54 has been treated successfully with early surgical intervention and the ocular tension has remained well controlled for 10 years.

History

The family history was without congenital anomalies and other inherited diseases except that the mother was diabetic in her seventies. Since childhood the patient had suffered from hemicrania of changing localization. This was successfully treated with ergotamine medication, and has been very rare since her early forties. At the age of 16 years, she had paraesthesia at varying sites, sometimes in one half of the tongue, sometimes changing localization and without relation to the hemicrania. These paraesthesiae increased during pregnancy at the age of 39, when she gave birth to a son, who is healthy and without ocular symptoms. Apart from a slight myogenic headache varying in intensity she had no further complaints. Blood tests, serum electrophoresis, metabolism, antibodies, liver function, and hormonal tests have shown normal values. X-ray examinations have revealed no abnormalities and no abnormalities of the connective tissue have been shown (Prof. G. Asboe-Hansen). A slight chronic vaginal inflammation has recently been treated (Dr. Find Andersen).

Examination

In April, 1956, the visual acuity in both eyes was 0.8 with +2 D cyl., axis 160°, in the right eye and +2 D cyl., axis 20°, in the left. The right pupil was round and central but with two atrophic areas which gave a slight reddish reflex by retro-illumination; 6 months later the atrophic area was more pronounced, now showing two narrow radial slits in the iris at 5 and 6 o'clock. A year later the iris had changed further with atrophic moth-eaten holes and the pupil was slightly updrawn towards the 12 o'clock position (Fig. 1). The visual acuity was 0.6, and the refraction unchanged.

Ocular tension

In 1958 the tension started to rise to 30–40 mm. Hg but was controlled with miotics. The iris atrophy progressed slowly during 1958 and 1959, and increased medication had no satisfactory effect.

FIG. 1 *Essential iris atrophy in the right eye in 1958. The complete absence of iris tissue is seen to extend from the sphincter to the limbus at 6 o'clock. The absence of the anterior layer is visible in two areas, a small one at 4 o'clock, and a larger one at 6–8 o'clock. The updrawn pupil makes the atrophy conspicuous*

FIG. 2 *Shortly after the Stallard type operation for glaucoma. Iridectomy at 11 o'clock. The conjunctiva covers the filtration area. and the subconjunctival oedema is visible*

FIG. 3 *Gonioscopy in 1968 shows the pupil displaced upwards and the iridectomy. Some opacities of the centre of the lens are seen just below the sphincter muscle*

FIG. 4 *Gonioscopy. The nasal area is less affected*

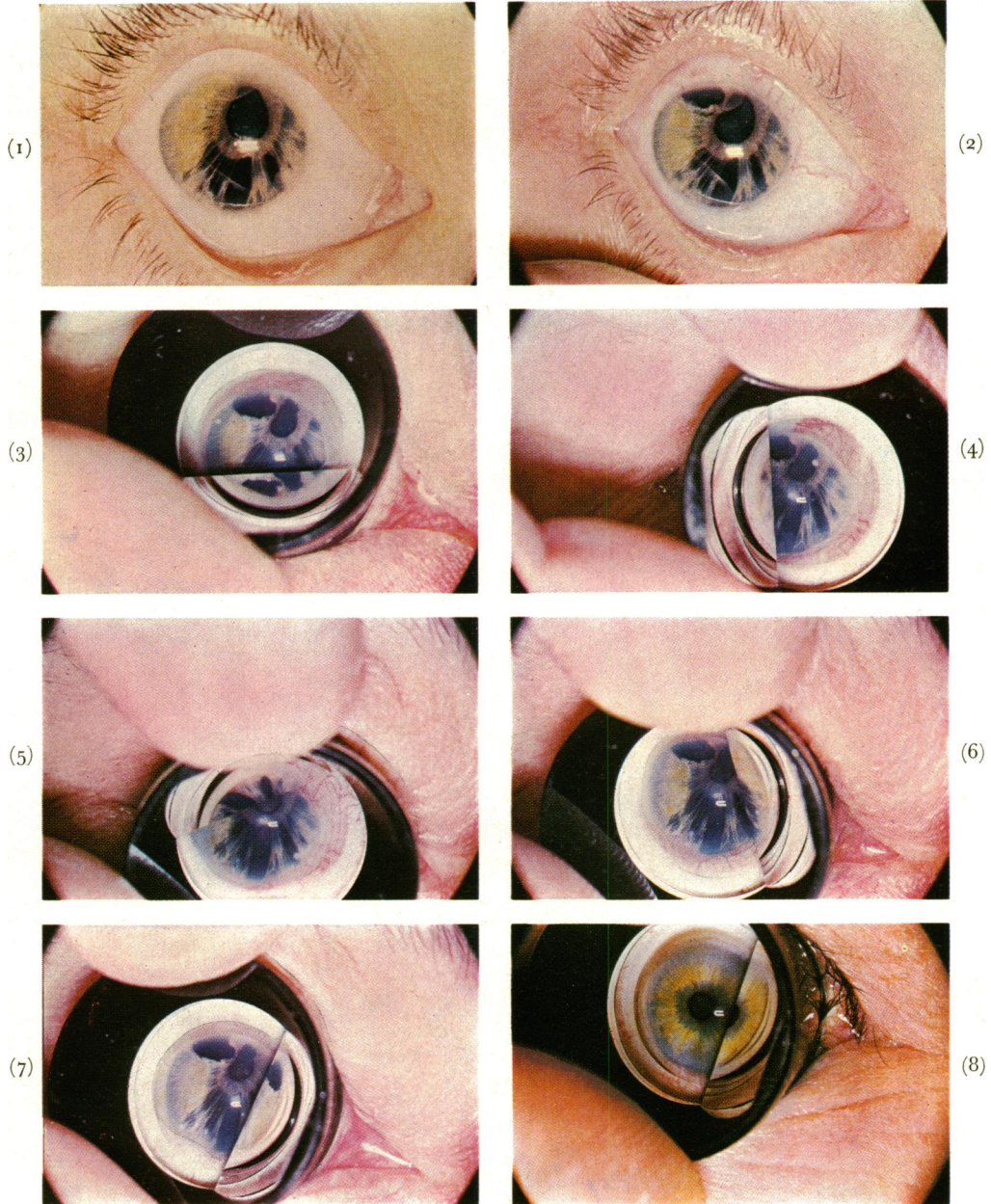
FIG. 5 *Inferior nasal quadrant, showing atrophy involving the angular structures*

FIG. 6 *Inferior temporal quadrant, showing the atrophic area below; a less atrophic area from 8 o'clock to the iridectomy*

FIG. 7 *Superior temporal quadrant, showing the result of the glaucoma operation*

FIG. 8 *Normal left eye with normal gonioscopy*

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Surgery

In November, 1959, it was necessary to perform a drainage operation, and the Stallard type of operation was chosen, *i.e.* sclerotomy, sclerectomy, partial cyclodialysis, and iridectomy, with iridencleisis (Fig. 2). The operation was placed as far away from the atrophic area as possible, *i.e.* close to the updrawn pupil at 11 o'clock. At the same time a partial iris transcision was attempted above the updrawn pupil in order to release the tension there. There were no complications.

Result

The tension has since remained under control at 15 to 18 mm. Hg. There have been periods of slight oedema of the cornea even with a tension less than 16 mm. Hg. This has disappeared completely with the use of local cortisone. As the atrophy has progressed it is conspicuous that the stromal atrophy precedes the atrophy of the pigment (Figs 4 to 8).

Present state

The visual acuity of the right eye is 0.4 with +0.50 D sph., 2.00 D cyl., axis 160°, and that of the left eye is 0.8 with +0.50 D sph., +1.00 D cyl., axis 10°. The ocular tension in both eyes is normal.

Comment

The case seems to indicate the beneficial effect of early filtering operation in cases of essential iris atrophy with raised intraocular pressure.

I have never been able to follow the more conservative attitude in the treatment of glaucoma, and especially not in cases of secondary glaucoma; in one way or another it should always be possible to keep the drainage open. Sometimes it may be difficult to close the drainage if the tension becomes too low, but that is another question.

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