FUCHS'S HETEROCHROMIC CYCLITIS WITH UNILATERAL SIMPLE GLAUCOMA*†

REPORT OF A CASE

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HETEROCHROMIC cyclitis was described by Fuchs (1906), who noticed, apparently without the aid of a tonometer, that the tension was raised in the affected eye in a proportion of cases. Subsequent observers confirmed this finding and established that the incidence of raised tension in the affected eye was about 5 to 15 per cent. Although heterochromic cyclitis is characteristically confined to one eye it is sometimes bilateral, and in such cases the incidence of raised tension has been reported as about 20 to 35 per cent. (François, 1954; Franceschetti, 1955; Huber, 1961; Velický, 1964; Becker and Shaffer, 1965).

The purpose of this paper is to describe a case of unilateral heterochromic cyclitis in a normotensive eye, and advanced simple glaucoma in the other eye. This is a combination that does not appear to have been reported before.

Case Report

A 69-year-old white woman was noticed to have a cupped disc and was referred for an opinion. She had no subjective complaints. The right eye showed the typical appearances of heterochromic cyclitis, while the left showed those of simple glaucoma.

There were discrete precipitates all over the corneal endothelium of the right eye but more so in the lower half, the anterior chamber containing a few cells. The lens had early anterior and posterior cortical opacities and there was a large anterior vitreous floater; there was no peripheral choroiditis. The right iris was depigmented in comparison with the left and showed peri-pupillary atrophy. The heterochromia had been present for at least 15 years, according to the history given.

By contrast, the anterior segment of the left eye was normal.

The pupils were equal in size, being 3 to 4 mm. in diameter. After instillation of gutt. cocaine 4 per cent. into the conjunctival sacs, the pupils became semi-dilated, the right being slightly smaller than the left. The tensions did not rise after mydriasis. The corrected visual acuity was 6/6 in each eye, there being a low degree of hypermetropic astigmatism.

The more interesting features are compared in the Table (opposite).

Discussion

The occurrence of raised tension in the more pigmented eye of a patient with heterochromia iridum may suggest, according to Becker and Shaffer (1965), the diagnoses of diffuse iris melanoma, haemosiderosis, or siderosis bulbi. In the case described here, these possibilities for the left eye could quite clearly be ruled out and a diagnosis of simple glaucoma was the only one acceptable. Moreover, the history and appearance of the

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TABLE
COMPARISON OF CLINICAL FINDINGS IN THE RIGHT AND LEFT EYES

Eye		Right	Left
Diagnosis		Heterochromic cyclitis	Simple glaucoma
Anterior Chamber	(a) Axial depth (mm.) (b) Angle	3.7 Wide	3.2 Medium to wide
Optic Disc		Normal	Pathological cupping
Visual Fields	(a) Perimeter 3/330 (b) Bjerrum 2/2000	Full	Loss of upper half
Tonometry (mm. Hg: Goldmann Applanation)	19.1.67 6.2.67 21.3.67 4.5.67	18 14 12 13	28 26 22 27
Tonography		$C = 0.25$	$C = 0.15$

right eye were so typical of heterochromic cyclitis, that other types of heterochromia—simple, atrophic, sympathetic, and abiotrophic (Duke-Elder and Perkins, 1966)—could be similarly dismissed.

In considering the significance of the occurrence of these two conditions in different eyes of this patient, it should be borne in mind that the one, heterochromic cyclitis, is characteristically unilateral but can be bilateral, whereas the other, simple glaucoma, is typically bilateral but can sometimes appear to affect one eye only.

The possibilities regarding this patient may be summarized in general terms as follows:

(1) There is a chance coincidence of heterochromic cyclitis in one eye and simple glaucoma in the other.

(2) Simple glaucoma would have been manifest in both eyes, but some factor prevented it in the eye with heterochromic cyclitis.

(3) Heterochromic cyclitis would have been present in both eyes with raised tension in the left eye, but some unknown factor suppressed the cyclitic and atrophic manifestations in that eye.

(4) Heterochromic cyclitis is sometimes complicated by raised tension, cupping of the disc, and field loss without closure of the angle, *i.e.* by "simple glaucoma". If this complication is regarded as one component of the condition, it is possible that, since the condition can be bilateral, the "cyclitic component" has appeared in one eye and the "glaucomatous component" in the other.

Each of these possibilities will be considered in turn.

(1) Coincidence by Chance

The probability of heterochromic cyclitis occurring in one eye and simple glaucoma in the other may be calculated on the assumption that the one condition is completely independent of the other. Such calculation involves the reported incidence of various conditions, and since the figures vary from observer to observer, the computed probability can only be approximate. In general the maximum incidence of each condition has been calculated so that the coincidence of the two by chance has probably been overestimated.

The incidence of uveitis is about 1 in 1,000 over all age groups (Darrell, Wagener, and Kurland, 1962), heterochromic cyclitis comprising 2 to 3 per cent. of these cases (Kimura, Hogan, and Thygeson, 1955; Perkins, 1961; Duke-Elder and Perkins, 1966). Without making any allowance for bilateral cases, these figures give a maximum incidence of 3 in 100,000 (0.003 per cent.) for heterochromic cyclitis in one eye.

The incidence of primary glaucoma in the population over 40 years old is generally agreed to be in the region of 1 to 2 per cent. (Sugar, 1964). There is considerable disagreement regarding the relative frequency of the various types of primary glaucoma but, according to Smith (1965), simple glaucoma makes up about 40 per cent. of the total. The overall incidence of simple glaucoma may thus be taken to be about 0.8 per cent. There is further disagreement on the incidence of apparently unilateral simple glaucoma. Chandler and Grant (1965) regarded this as being so low as to suggest a mistaken diagnosis. On the other hand, of a random selection of 212 cases diagnosed as simple glaucoma in the Glaucoma Clinic at Moorfields Eye Hospital, eighteen showed no abnormality of the apparently healthy eye on applanation tonometry, ophthalmoscopy, examination of the visual fields and, in some cases, on tonography; according to these findings 8.5 per cent. of cases of simple glaucoma may appear to be unilateral. On this basis, the incidence of unilateral simple glaucoma would be 8.5 per cent. of 0.8 per cent., *i.e.* 0.068 per cent.

A coincidence by chance of heterochromic cyclitis and unilateral simple glaucoma could therefore be expected in 0.003 per cent. of 0.068 per cent., *i.e.* in 0.000204 per cent. of the population. This will include the coincidence of the two conditions in the same eye; the frequency of their occurrence by chance in opposite eyes, as in the patient described here, will be half that given above, *i.e.* 0.000102 per cent. or about 1 in 1,000,000. Although this is a low incidence, it does not rule out an association by chance of these two conditions in the patient described here.

(2) Suppression of Glaucoma in the Cyclitic Eye

This could have occurred in the right eye if: (a) the fall in facility of outflow, characteristic of simple glaucoma, had been prevented, or (b) the facility had fallen but the intra-ocular pressure had not risen because there had been a concomitant reduction in the rate of aqueous formation.

With regard to the first possibility, no sound suggestion can be made as to the nature of any protective mechanism because the pathology of the glaucomatous fall in facility of outflow is not understood. It can only be pointed out that a single tonographic result indicated a better outflow in the right eye than in the left.

The second possibility leads to the conjecture that reduction of aqueous formation was brought about either directly by damage to the ciliary body or indirectly by diminished sympathetic activity. At a late stage in various forms of uveitis, sufficient damage can be sustained by the ciliary body for hypotension to ensue while, in heterochromic cyclitis, stromal fibrosis and muscular atrophy in the ciliary body have been demonstrated histologically (Kimura and others, 1955).

One aetiological theory of heterochromic cyclitis postulates a unilateral sympathetic paresis (François, 1949), a view which is supported by reports of the occurrence of the condition following stellate ganglionectomy (Calmettes, Déodati, and Amalric, 1953) or removal of the superior cervical ganglion (Makley and Abbott, 1965), but which is denied by others (Perkins, 1958). An effect of the sympathetic upon aqueous formation in man

is suggested by several observations; thus, decreased formation leading to ocular hypotension has been reported in Horner's syndrome (Swegmark, 1963), while a reduction in intra-ocular pressure has also been recorded after blocking the stellate ganglion (Miller, 1953) or after the administration of sympathetolytic drugs (Oosterhuis, 1962).

With regard to the case under consideration, it can only be pointed out that the smaller effect of cocaine on the right pupil as compared with the left may be of some significance.

(3) Suppression of Cyclitis in the Glaucomatous Eye

Although this is theoretically possible, it would seem improbable and no mechanism is suggested for it.

(4) Occurrence of a "Cyclitic Component" of Heterochromic Cyclitis in one Eye and a "Glaucomatous Component" in the Other

This possibility seems to us to have become more worthy of consideration since the publication of the interesting findings of Hart and Ward (1967). These observers studied sixteen patients with unilateral heterochromic cyclitis. Tensions were measured with the Goldmann applanation tonometer and, of the eyes not affected by cyclitis, three showed tensions of 21 mm. Hg, one of 22 mm. Hg, one of 24 mm. Hg, and one of 30 mm. Hg. On tonography, one unaffected eye had a coefficient of facility of outflow of 0.13, another of 0.12, two of 0.11, and another of 0.09. Hart and Ward interpreted their findings on the basis of the work of Becker and Shaffer (1965) and of Graham and Hollows (1966), concluding that only a third of the eyes unaffected by heterochromic cyclitis were completely normal. It is possible, of course, to disagree with the exact proportion to be considered abnormal, depending upon the values one is prepared to accept for normality. If we take the view that an applanation tension over 21 mm. Hg or a value of C less than 0.15 gives rise to the suspicion of glaucoma, and that a tension over 24 mm. Hg or C less than 0.12 can be regarded as pathological (Gloster, 1966), then we may say that two of the cases described by Hart and Ward were suspicious and four pathological. It would seem that, even with these different criteria of assessing the findings of Hart and Ward, there was a greater tendency towards glaucoma in the non-cyclitic eyes of their patients with unilateral heterochromic cyclitis than in the eyes of the general population.

Returning to the patient described here, it is impossible to decide which of the four suggested explanations accounts for the clinical findings, although we are inclined to regard the third explanation as highly improbable. An association of the two conditions by chance is possible, yet we are not aware of another such case having been reported in the literature; perhaps this is because previous observers have regarded an association by chance as being so likely that such cases are not noteworthy. The possibility that a cyclitic suppression of aqueous formation prevented the development of glaucoma in the right eye is interesting, but its consideration adds little to our understanding of the pathogenesis or control of glaucoma. On the other hand, the patient we have described may represent a more advanced condition than some of those recorded by Hart and Ward, in that there was frank glaucoma instead of a glaucomatous tendency in the non-cyclitic eye. If this is so, the concept of cyclitic and glaucomatous components in heterochromic cyclitis, the one component being sometimes separated from the other, becomes more worthy of consideration. Reports of similar cases may help in assessing the probability of this hypothesis, the justification of the present publication resting solely on the questions it raises.

Summary

A case is presented of unilateral Fuchs's heterochromic cyclitis occurring in association with simple glaucoma in the fellow eye.

Four explanations are offered:

- (1) Coincidence of the two conditions by chance;
- (2) Suppression of glaucoma by cyclitis;
- (3) Suppression of cyclitis in the glaucomatous eye;
- (4) "Cyclitic" and "glaucomatous" components of heterochromic cyclitis were manifested independently in opposite eyes.

Each explanation is considered in turn. In view of another recent publication, the fourth explanation was considered the most interesting.

This patient is under the care of Mr. Redmond Smith to whom we are grateful for his help and interest.

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