THE PHENOMENON OF GLAUCOMATOCYCLITIC CRISES

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It was in April 1952 that Frederick H. Theodore of New York, writing in the British Journal of Ophthalmology, emphasized the importance to ophthalmologist and patient of recognizing this fascinating syndrome, first adequately described by Adolf Posner and Abraham Schlossman of New York in 1948. Even today its importance is not fully recognized and indeed it is described in extremely few of the text-books of ophthalmology.

I have seen 3 cases in the last 18 months, but particular interest was stimulated by a case I recently saw in Johannesburg. This case presented some features which to my knowledge are not mentioned in the literature. I was fortunate in obtaining a very accurate history, extending back 2 years, from the patient's husband, who is an experienced medical practitioner. Before describing the case I shall give a brief resumé of this clear-cut syndrome.

Gross Diagnostic Characteristics

The name is well chosen and represents the main features, namely, (1) raised intra-ocular pressure varying from 30 mm. to 90 mm. Hg, (2) evidence of cyclitis which is, however, confined to large, flat, white keratic precipitates (KP) and occasional cells in the anterior chamber and, (3) recurrent attacks of acute onset which always affect the same eye, the syndrome being strictly unilateral.

Additional Features

It is extremely rare in acute glaucoma (*a*) to get repeated attacks with no concurrent deterioration of the visual fields or cupping of the optic disc. (*b*) It is equally rare to get a high intra-ocular pressure with such little deterioration of visual acuity during an acute attack. (*c*) It is practically unknown for acute congestive glaucoma to subside spontaneously without treatment. These 3 features, however, are seen in glaucomatocyclitic crises and they are the points which provide the finesse in the diagnosis of this syndrome. Between attacks no evidence of previous ocular disturbance is seen and all provocative tests for glaucoma are negative, no signs of previous cyclitis being present. Paradoxically the treatment most effective in acute glaucoma is least effective in the treatment of this syndrome, since Diamox and meiotics have no effect and surgical intervention has never once been reported to control the attacks; indeed it is strongly contra-indicated. Mydriatics often increase the symptoms and raise the pressure.

CASE REPORT

Past History

In December 1953 the patient, a lady then aged 24 years, experienced het first attack. It began with a burning sensation in the left eye, blurring of vision and definite coloured haloes around lights. The doctor husband, taking these symptoms into account and noticing the left pupil larger than the right and feeling the pressure in the eye to be raised, diagnosed glaucoma and immediately put eserine drops into the eye. Although the pupil soon became pin-point in size, the symptoms remained unchanged, and the following day an ophthalmologist was consulted. His diagnosis was iridocyclitis with keratic precipitates and he prescribed 1% atropine drops. The attack finally cleared up after 3 weeks.

In an endeavour to locate a possible trigger focus, the patient's teeth, sinuses and chest were X-rayed and a Wassermann test and full blood-count and urinalysis performed. The results were all within normal limits. The ophthalmologist advised the patient to carry 2% homatropine constantly and instill it immediately if symptoms recurred. This opportunity arose 2 months later when a similar attack occurred. This attack was treated with the homatropine drops and cleared in 2 weeks.

The patient had for many years suffered from pyelitis with her menstrual periods and a particularly bad attack occured soon after her second ocular episode. A urologist was consulted, who advised a right nephropexy to abolish a definite kinking of the right ureter shown on intravenous pyelogram, because it was felt that the chronic pyelitis might have had some effect on the iridocyclitis.

A right nephropexy was performed, and it was indeed disappointing when the third ocular episode occurred only one month afterwards, in March 1954, and the fourth and fifth attacks in May and August 1954, all affecting the left eye. These attacks lasted about 2 weeks and were treated with homatropine 2% drops. The husband and wife moved to the Orange Free State and after a 6 months' period free of symptoms, the sixth left ocular attack occurred. Haloes were a particular feature this time and after an attack of almost projectile vomiting at 2 a.m. an ophthalmologist in the vicinity was consulted.

He diagnosed acute congestive glaucoma and said there were no signs at all of previous iridocyclitis. The Schiotz tenometer reading was L.E. 45 mm. Hg and R.E. 20 mm. Hg. An intensive course of 1% eserine brought no relief and 24 hours later the Schiotz reading remained unchanged. Diamox and eserine were then combined and, although side-effects of paraesthesia of the fingers and feet were produced, the symptoms and signs were unaffected and after 3 days the tension was 50 mm. Hg in the left eye and remained so for the next 5 days. The ophthalmologist now advised a drainage operation as the only alternative. As a last resort the husband decided to try an idea of his own. He produced a marked diuresis by giving his wife large doses of ammonum chloride followed by an injection of 2c.c. of Mersalyl. The patient felt much better the following day and for the first time in 2 weeks the tension in the left eye had fallen to 25 mm. Hg (Schiotz). On this occasion the ophthalmologist suggested using neocortef ointment to treat the patchy shredding of oedematous corneal epithelium. Three days later all signs and symptoms had disappeared. Three months later, when back in Johannesburg, the patient had an allergic reaction to an intramuscular injection of 100 mg. of Pethidine given for the treatment of a miscarriage. This manifested itself as oedema glottidis and slow respiration, but recovery was satisfactory. The seventh attack in the left eye began soon after this allergic episode and another ophthalmologist was consulted. He diagnosed acute congestive glaucoma and commented that there were no signs of any previous iridocyclitis. He prescribed 1% pilocarpine drops, 4 hourly. Mersalyl was again tried and that night 2 c.c. were given intramuscularly. The next morning symptoms had lessened and the tension was reported to be normal. All signs and symptoms disappeared within the next 3 days.

This story of medical misfortune was soon to be continued, for after taking 2 aspirins for dysmenorrhoea while on holiday in Rhodesia, the patient developed the worst urticarial rash the husband had ever seen. Luckily, however, there was a rapid response to ACTH. The eight left ocular attack occurred soon after their return to Johannesburg and another ophthalmologist's opinion was sought. He thought acute congestive glaucoma to be the basis of the trouble and prescribed 1% pilocarpine drops 4 hourly. On a second visit 5 days later he saw keratic precipitates and changed his diagnosis to secondary glaucoma. The visual

fields were tested and found normal and the attack had cleared in another 5 days.

The last two Attacks

At 11.30 p.m. in mid-December 1955 I received a telephone call from the husband, who explained that his wife had nausea and was seeing haloes with her left eye. I examined her about an hour later.

On Examination. Unaided vision of right eye 6/9, left eye 6/12. There was a partial ptosis of the left upper lid and slight swelling of the lateral part of both the left upper and lower lids. There was mild congestion of the conjunctival vessels. The cornea appeared bright, but some small white specks could be seen naked-eye, just below the centre of the cornea. The pupils were equal in size and the pupillary reflexes were normal. There was no heterochromia. The media, discs and fund were normal.

Slit-Lamp Examination. The cornea showed no evidence of oedema. There were 8 white, flat, discrete, irregular keratic precipitates, all aggregated in a small area about 2 mm. below and slightly nasal from the centre of the cornea. Occasional cells were seen in the anterior chamber, but no flare. The pupillary margins of the iris and the anterior lens-capsule were normal and showed no signs of past or present iridocyclitis.

Visual Fields and Tension. The visual fields were full and normal The tension was 20 mm. Hg (Schiotz) in both eyes. Since I felt sure the diagnosis was glaucomatocyclitic crises, I prescribed cortisone ointment 1·5% hourly. Two days later the KP began to fade and 3 days after commencing treatment the left eye was normal. The vision was now 6/6 R and L the eyes appeared normal on examination. The tension was measured many times during this attack, and was always found to be normal.

The missing link in the diagnosis, namely a proven raised tension, was provided by the next attack some 5 weeks later. I then saw the patient at 5 p.m. on 26 January 1956, 12 hours after the attack had begun. Haloes were a predominant symptom.

On Examination. Right eye, vision 6/6, normal on examination; left eye, vision 6/9.

Examination of Left Eye. There was a slight drooping of the left upper lid. Ocular movements were full and normal. The moderately congested conjunctival and ciliary vessels had a definite cyanotic tinge. The periphery of the cornea appeared dull, but the centre part was normally bright. In exactly the same position as before, the little white specks could be seen. The left pupil was slightly larger than the right, and was round but sluggish in its reaction to light and accommodation. The media, disc and fundus were normal.

Visual Fields and Tension. The visual fields were full and normal. The tension was 45 mm. Hg (Schiotz). (The right tension 20 mm. Hg.)

Slit-Lamp Examination. The right eye was normal. The left eye showed a peculiar corneal oedema confined to the periphery of the cornea and especially clustered in blebs around the loops of the limbal vessels. This was particularly noticeable on the medial side opposite a moderately vascular pinguecula. There were 9 large white flat discrete keratic percipitates in exactly the same position as in the last attack, and some peppering with minute precipitates of the small area between the closely aggregated keratic precipitates. There were occasional cells in the anterior chamber, but no flare. Iris and lens were normal.

Treatment and Result. 26 January: Cortisone, 1·5% drops, was started immediately in the left eye and instilled every 5 minutes; after 2 hours 1·5% cortisone ointment was used and the patient went to bed.

27 *January:* Symptoms and signs about the same, the tension of the left eye now being 36 mm. Hg.

29 *January:* The symptoms were still present and the patient had vomited at about 2 a.m. The keratic precipitates were beginning to fade, but the tension was still 40 mm. Hg (Schiotz). The husband suggested using Mersalyl and we gave the patient 2c.c. intramuscularly.

30 *January*: The patient volunteered that in the morning she had felt a peculiar sucking sensation in the eye. After this all symptoms disappeared. In the evening, the complete metamorphosis was striking, for now the left eye was completely normal. The tension was 20 mm. Hg in both the right and left eyes and the visual fields full and normal.

DISCUSSION

Here indeed is a fascinating case of the syndrome of glaucomatocyclitic crises with a 2-year history. During this time 10 attacks were experienced, the last 2 being observed by the author. It appears from the history and findings of colleagues that the syndrome was sometimes predominantly glaucomatous in appearance, with accompanying symptoms of haloes, burning of the eye and, on two occasions, projectile vomiting, whereas on other occasions the appearance was one similar to cyclitis with no raised tension.

Treatment. Although Diamox, eserine, pilocarpine, homatropine and atropine were all used on different occasions, the only effective remedy in shortening an attack appears to be cortisone, and especial note should be taken of the dramatic effect of the diuresis produced by Mersalyl if the tension is raised, aborting the attack and lowering the tension almost immediately to within normal limits.

Etiology. It is tempting to postulate on an allergic basis, as previously suggested in the literature, to account for these attacks. Supporting evidence is undoubtedly the acute onset and cessation, the self-limited attacks, an allergic history of the patient and a strong family history of allergy. The slit-lamp appearances of oedema localized to the vicinity of the limbal loops and the slight swelling of the lids with slight ptosis all support this hypothesis of an allergic reaction, with vascular dilatation and oedema. The keratic precipitates are peculiar, for their position was the same in each attack observed by me—they are closely aggregated together and hence do not appear to be deposited according to the influences of gravity, convection currents or generalized corneal endothelial disturbance. They never become pigmented, and disappear without leaving a trace. Their usual flat and white appearance suggest that they are not formed by inflammatory cells and debris adhering to the damaged endothelium, but are possibly protein products, the result of an allergic vascular intra-ocular anomaly. Indeed if the angle was blocked by oedema, it might well explain the dramatic effect of Mersalyl in carrying away the oedema, and the apparent lack of effect of Diamox, so useful in most types of glaucoma, since the action of Diamox does not seem to depend on its diuretic properties, but more on its effect on carbonic anhydrase, bicarbonate secretion into the eye and intra-ocular fluid production.

In the few cases reported on which surgical drainage operations were performed, no relief from attacks was obtained—very different from our experience of ordinary acute congestive glaucoma. It is puzzling that although the tension may remain high for as long as 2 weeks, no deterioration is seen in the visual field and no atrophy or cupping of the optic disc.

SUMMARY

A detailed history and case report of a patient suffering from glaucomatocyclitic crises is given. Treatment and

etiology are discussed and emphasis laid on the importance to patient and ophthalmologist of recognizing this syndrome as a distinct clinical entity. Cortisone is the treatment of choice in cases predominantly cyclitic, whereas cortisone eye-ointment and Mersalyl intramuscular injection appear most effective if the pressure is raised.

REFERENCES

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