POSTERIOR CONICAL CORNEA*

BY

H. B. JACOBS

London

SINCE this condition was first described (Butler, 1930), only a handful of cases has been added to the literature. In view of its infrequency, some cases are presented and compared with previous descriptions.

Perhaps the name, although anatomically descriptive, is unfortunate, for it would imply that the condition has some relationship with the better-known conical cornea. In fact, there is no evident relationship beyond the unsubstantiated statement that the posterior form is a precursor of the ordinary form. The two conditions are so named merely because the cornea tends to become cone-shaped—throughout its thickness in ordinary keratoconus and solely in its posterior surface in keratoconus posticus. Two forms of the latter condition occur:

- (A) Localized—keratoconus posticus circumscriptus.
- (B) Generalized—keratoconus posticus totalis.

(A) **Localized.**—Seven cases have been recorded. Stallard (1930), Butler (1932), Goldsmith (1943), Leopold (1943), Wise (1944), Greene (1945), and Guimarães (1953) all describe a localized concavity of varying size in a part of the back of the cornea. Goldsmith's case (Fig. 1) is typical.

FIG. 1.—Case 5, slit-lamp appearance and sketch showing a typical posterior conus.

* Received for publication October 25, 1956.

Stallard (1930) makes no mention of a nebula in his patient but the other authors all comment on either a localized nebula or haze. The American authors (Leopold, 1943; Wise, 1944; and Greene, 1945) could obtain no history of trauma in any of their cases, and Leopold suggests that, since the condition has never been seen to develop after trauma, it is unjustifiable to assume that it is so caused. On the other hand, the case reported by Butler (1930) had a definite history of trauma, and there was reason to believe that in the case of Stallard's patient, a railway worker, the lesion was at any rate acquired. Six of the cases were men but Leopold's was a Negress. Five lesions were unilateral, one bilateral, one was in an only eye (Guimarães), and only one was associated with other possibly relevant ocular pathology. This (Greene's case) took the form of definite lens changes which were in line with the corneal lesion. Greene considered that, on the basis of the position of the lenticular opacities in the superficial layers of the foetal nucleus and their correspondence in position with the corneal lesion, the congenital origin of the latter could not be doubted.

Case Reports

Case 1, a woman aged 55, was admitted to hospital in June, 1953, for the removal of a cataract in the left eye. She had been observed in the out-patient department for 5 years and it had been noted that she had a corneal opacity in the left eye. On more detailed inspection, this was noticed to be associated with a posterior increase of curvature localized to that area (Fig. 2). This patient gave a history of trauma to the left eye in childhood. The right cornea was normal.

FIG. 2.—Case 1, slit-lamp appearance and painting of left cornea showing localized posterior conus and opacity situated in superficial layers of substantia propria.

Case 2, a woman aged 51, attended hospital in September, 1953, complaining of watering and blurring of the vision of the right eye for 5 days and blurring of the vision of the left eye for one day only.

On examination, some old pigment deposits were seen on the posterior corneal surface of the right eye, and a corneal opacity in the 8 o'clock meridian, deep to which was a localized posterior keratoconus. There were some cells in the anterior chamber. The left eye had acute glaucoma, presumably secondary to uveitis, and the patient was admitted to Moorfields Eye Hospital. During the course of investigation for the cause of the uveitis, the Wassermann reaction was found to be strongly positive.

The posterior conical cornea was localized in the situation described above. No similar lesion was present in the left eye.

Subsequently, a broad iridectomy was performed on the right eye for secondary glaucoma, and a sclerectomy on the left eye.

Case 3, a man aged 25, attended hospital complaining of discomfort in the right eye. A superficial corneal foreign body was removed and the cornea healed smoothly. In the course of routine examination, it was noted that this patient had a reduplicated lamina of lens opacity, white in the illumination of the slit lamp, and conjoined peripherally in the intermediate layers of the cortex. It was situated in the region of 6-7 o'clock and directly in front of it and a few millimetres from the limbus there was an unusual guttering of the posterior surface of the cornea (Fig. 3).

FIG. 3.—Case 3, diagram of unusual guttering of posterior surface of right cornea.

This was unlike other cases of posterior keratoconus described in this paper and elsewhere in that, instead of the lesion being oval or circular, it was merely a groove concentric with the corneal margin, approximately 5 mm. long and 1 mm. wide. This case is unusual in that it is the only one, other than that of Greene (1945), to show an associated congenital lenticular abnormality. Unfortunately no painting of this patient was done and only a diagram of the corneal condition can be presented.

Case 4, a short hunch-backed woman aged 55, attended hospital in July, 1952, with a 5-day history of pain and loss of vision in the right eye and also of vomiting. She had right acute glaucoma with a hypermature cataract. She was treated with eserine but after temporary improvement relapsed, so that an iridencleisis had to be performed. A few days later signs of infection developed, but these responded to intensive antibiotic therapy. 2 weeks after her discharge the eye was very red, with many keratic precipitates. Intensive treatment was advised but the patient failed to attend for a further 7 weeks when an enucleation was advised and duly performed.

In the course of clinical examination, the left cornea was noted to have three localized nebulae, and one of these showed a related posterior increase in curvature (Fig. 4). When the tension in the right eye first subsided a localized posterior conical cornea was seen similar in size and position to that in the left eye. There were no other opacities in the right cornea. A previous history of recurrent soreness of the left eye in childhood but no trouble with the right until the glaucomatous episode suggests that the two nebulae in the left eye not associated with the conus were the result of ulceration in childhood, but gives no clue as to the time of onset of the bilateral lesion. The previous history also suggests a certain lack of observation in the patient. Apart from the corneal condition, no other abnormality was seen in the left eye. The vision was 6/18 pt.

FIG. 4.—Case 4, left cornea showing localized posterior keratoconus posticus (depth of cone exaggerated and schematized). This lesion was related to the largest nebula, the others being the result of ulceration in childhood.

The enucleated right eye was sent for section, but the keratoconus posticus could not be demonstrated histologically. It is difficult to explain this, but possibly the depth of the posterior excavation was too slight to show, bearing in mind the unavoidable distortion that must occur in the preparation of histological sections. If and when a similar opportunity arises alternative techniques will be tried.

Case 5, a man now aged 70, is that recorded by Goldsmith in 1943 (Fig. 1). He was examined again by me in 1955. A description is appended for the sake of completeness.

He was first seen in January, 1942, with a foreign body in the left eye. He showed bilateral symmetrical lesions of keratoconus posticus circumscriptus. He has been seen at irregular intervals since, the last time being in November, 1955.

There appears to have been no alteration in the corneal condition but the refraction has changed.

January, 1942: Right eye + 5.5 D sph. 6/9

Left eye + 5.5 D sph. + 1.00 D cyl., axis 95° 6/12

September, 1954: Right eye + 6.00 D sph. + 1.50 D cyl., axis 95° 6/9

Left eye + 7.00 D sph. + 1.00 D cyl., axis 95° 6/12

An intervening refraction in 1949 showed + 1.00 D cyl. in the right eye.

The clinical appearance with a loupe and focal illumination was of a localized symmetrical nebula in each eye. Slit-lamp examination revealed a posterior concavity, an opacity of the superficial substantia propria, a Hudson's line below, and some Hassall Henle bodies in the endothelium and localized to the site of the lesion.

***Case 6, a girl aged 13**, gave a rather vague history of longstanding visual defect; her vision could be improved only to 6/18 in each eye with -6.00 D cyl. There was a nebula in each cornea, the lesion being larger in the left eye than the right; the slit lamp showed posterior conical corneae similar to the previous case but larger and more centrally placed.

Keratotomy showed a marked distortion of the rings consistent with a regular corneal astigmatism, no localized defect being noted in the region of the conus defects.

This patient was fitted with contact lenses in Mr. Frederick Ridley's clinic; with them vision was improved to 6/12 in each eye.

Case 7, a man aged 55, attended hospital with bilateral cataracts, both of which were eventually extracted by Mr. Goldsmith. Faint, greyish, localized, eccentric corneal nebulae, bilaterally and not quite symmetrically placed, had been noted. Each nebula was related to a localized posterior concavity, the opacity being superficially placed in the substantia propria and showing some irregularly-disposed greenish pigmentation. Descemet's membrane in the region of the conus showed a slight brownish tinge. Corrected vision was 6/9 in each eye and no history relevant to the corneal lesions was elicited. The corneal appearance was similar to that in Case 5.

Case 8, a boy aged 15, had lived in Ireland until the age of 7. He attended school in England from that age till 15 and, during that time, had an eye test, but did not remember anything about it. On leaving school at 15 and applying for a job, his vision was found to be poor in both eyes, the left being worse than the right, so he sought hospital advice.

Questioning revealed that he had never been troubled subjectively and that he had no memory of any redness or discomfort in either eye.

The eyes were white and the visual acuity was 6/24 in the right eye and counting fingers at 1 foot uncorrected in the left. The right vision could be improved to 6/18 with -1 D cyl., axis 45° , and to 6/12 with a stenopoeic aperture. Retinoscopy on the left side indicated the need of -7.25 D sph., -3 D cyl., axis horizontal, but no significant improvement was obtained with such a lens and the eye was presumably amblyopic.

The right cornea showed a diffuse central haze and a denser opacity to one side. In relation to the former there was a posterior concavity and a Hudson's line, and the opacity was in the anterior part of the substantia propria. The endothelium was golden in colour. A brush of deep vessels ran to the denser opacity. The left cornea had many vessels in the deep substantia—all round—penetrating the cornea but avoiding a faint nebula in the central area. Related to this was a posterior corneal concavity; the opacity was in the middle part of the substantia propria.

The eyes were otherwise apparently healthy. The Wassermann reaction was negative.

Case 9, a man aged 49, was the father of Case 8. He had no ocular complaints and no history of any previous eye troubles. Vision in the right and left eyes was 6/12 and 6/9 respectively, and in the centre of each cornea a faint greyish nebula was visible. In the right eye, there was a related posterior concavity and an excess of Hassall Henle bodies, and in the left a mere posterior dimpling was related to the nebula. The endothelium on this side was of a golden hue. The opacity in both corneae was in the anterior part of the substantia propria.

The occurrence of these lesions in Case 9 suggested that the lesions in Case 8 might be hereditary rather than the result of previous attacks of interstitial keratitis.

* The author is indebted to Mr. A. J. B. Goldsmith for the description of this case.

(B) Generalized.—Only three cases of keratoconus posticus totalis have been mentioned in the literature.

The first was described by Butler (1927), who stressed the following points:

- (1) No nebula.
- (2) No tears in Descemet's membrane.
- (3) No increased number of corneal nerves.
- (4) No Fleischer's ring.
- (5) A perfectly regular, geometrically precise increase in curvature of the posterior surface of the cornea.
- (6) Reflections of Placido's disc normal but elliptical.
- (7) Structure of cornea normal but thinned.
- (8) Sex female.
- (9) Lesion unilateral.

In Butler's case the visual acuity was 6/18, and was only slightly improved by a cylinder of "plus 2, 3, or 4 dioptries at 45°".

In the discussion on Butler's case (shown at a meeting of the Ophthalmological Society of the United Kingdom in 1930) a further case was mentioned by Mr. P. L. Stallard.

A basin-shaped depression seen in the posterior surface of the cornea. This increase in concavity formed a perfect curve, involved one half of the cornea, and was fairly centrally situated. The spherical regularity of the anterior surface as evidenced by the keratometer mires and by the reflections of a Placido's disc was noted. No comment on the presence or absence of a nebula was made. The patient was a male Indian; the lesion was unilateral and thought possibly to be traumatic.

This case is probably better viewed as a keratoconus posticus circumscriptus; but is mentioned here because it was described as a companion case to that of Butler. (It is referred to in the preceding section.)

The second case (Ingram, 1936) showed a regularly increased curve of the posterior corneal surface similar to the case described by Butler.

Other points were as follows:

- (1) A central haze was visible with a loupe.
- (2) No Fleischer's ring.
- (3) Some pigment deposits on the posterior corneal surface.
- (4) Unilaterality.
- (5) Sex female.

The visual acuity was less than 6/60 and unimprovable. No mention was made of the structure of the cornea, but presumably there must have been some change otherwise no haze would have been seen.

The third case (Ross, 1950) showed the following characteristics:

- (1) Bilaterality.
- (2) Sex female.
- (3) Uniform increase of posterior corneal curvature. Anterior curvature was normal.
- (4) Horizontal superficial Hudson's lines.
- (5) Thinned but normal corneal stroma.
- (6) Mixed astigmatism (right eye +3.25 D sph., - 4 D cyl., axis 108°; left eye +2.75 D sph., - 4 D cyl., axis 72°), correction of which did not increase visual acuity which was 20/40 and 20/50 in the right and left eyes respectively.

Thus all three typical cases of this condition so far described in the literature were seen in women, who had had impaired vision for many years, and in all the condition was probably congenital. Ross remarks on the resemblance of the shape to the developing cornea.

Case Report

Case 10, a woman aged 49, presented with a history of poor vision in the right eye for as long as she could remember. She thought the condition was static but was unsure.

Examination.—Visual acuity was 1/60 in the right eye and 6/12 in the left unaided. The lids, conjunctivae, and ocular tension were normal.

The right cornea showed a faintly hazy appearance; the left cornea was clear apart from a small nebula near the limbus. The pupils reacted normally, the lens and vitreous in each eye were clear, the fundi showed no abnormality. The slit-lamp appearance of the right cornea showed marked thinning of the substantia propria associated with and apparently due to an increased curvature of the posterior corneal surface. The apex of the posterior "cone" and therefore the thinnest area of the cornea was just below and temporal to its centre (Fig. 5). The left cornea was normal.

FIG. 5.—Case 10, keratoconus posticus totalis. View with a loupe and focal illumination to demonstrate the faint corneal haze and a ring of greenish pigmentation. Slit-lamp appearance showing thinning of cornea due to generalized increase in curvature of its posterior surface. Left cornea with a small incidental and unrelated nebula.

Placido's disc showed an irregularity just off centre towards 9 o'clock in the right eye. The reflections in the left eye were normal.

Retinoscopy gave the following refraction: right eye +1.75 D sph., -5.5 D cyl., axis 90°, but no improvement was effected either with these lenses or with a stenopoeic aperture. The visual acuity in the left eye was improved to 6/5 with +0.25 D sph., +0.75 D cyl., axis 180°.

Keratometry—The right eye was complicated by slight irregularity of the surface, but the readings were as follows:

$$\left. \begin{array}{l} \text{At 15 } \left\{ \begin{array}{l} 7.52 \\ 7.65 \end{array} \right\} \text{mean } 7.58 \\ \text{At 130 } \left\{ \begin{array}{l} 6.55 \\ 6.79 \end{array} \right\} \text{mean } 6.72 \end{array} \right\} \text{cylinder—5.5}$$

In the left eye the readings were straightforward and corresponded with the refractive findings.

Discussion

(A) **Keratoconus posticus circumscriptus.**—Although little is known of the possible mechanism of formation of these corneal deformities, it is known that disturbances of Descemet's membrane and/or the corneal endothelium can give rise to opacities in the more anterior levels of the substantia propria. It would seem possible that such a disturbance in the very young could be accompanied either by a resorption of the deeper lamellae or failure in their formation, either of which might result in the formation of a localized posterior concavity.

The lesion of Descemet's membrane or the endothelium might take the form of a definite tear, some similar but subclinical loss of continuity, or merely a disorder of normal function of the endothelium in preserving the physiological status of the substantia propria.

This mechanism is consistent with prenatal or neonatal trauma or inflammation being responsible for the condition, while there are the additional possibilities that a failure in the normal development of either Descemet's membrane, the endothelium, or the substantia propria may be a causative factor.

Of the localized cases which constitute the first half of this paper, Cases 3, 4, 5, 6, 7, 8, and 9 are almost certainly congenital. Cases 1 and 2 could be congenital, but there is insufficient evidence to point one way or another. Of the five cases collected from the literature, three were probably of congenital origin and two possibly traumatic.

Cases 8 and 9 are of particular interest in that they represent the only instance so far recorded of a familial link in this condition.

(B) **Keratoconus posticus totalis.**—All these cases, including the one herein described, have a long history. All had had poor sight for as many years as they could remember. Ross's patient had fair vision, but it had not deteriorated at least during the previous 6 years. Harrison Butler stated

that he had watched two cases for 6 years and observed no change. All the cases failed to improve with correcting spectacle lenses; this makes it appear likely that all are amblyopic and argues for the congenital origin of the lesion.

None of the authors who have described these cases mentions a change in corneal structure, and Harrison Butler states that its absence is characteristic. The present case showed such a thinned appearance that it is difficult to make a point. All the authors comment on the regularity of the anterior surface.

The present case differs in small details from the generalizations made by Harrison Butler, but the other cases also show slight distinguishing features.

Ingram mentions a corneal haze and pigment deposits behind the cornea.

Ross noted a Hudson's line and a few Hassall Henle bodies peripherally. His is the only bilateral case on record.

In the present case, there was a definite faint haze, and the anterior corneal surface was slightly irregular as judged on the keratometer and with Placido's disc. No other author describes both these findings, and Harrison Butler states that in his case the reflections of the Placido's disc were regular.

All the cases were female.

No positive family history was given in the patients previously reported and none was obtained in the present case.

Only one of Harrison Butler's cases can be found in the literature though he gives the impression of having seen at least two. His generalizations would seem to exclude the present case from being grouped with the condition he originally described. However, there is certainly a general resemblance to his case and to the others referred to in the present report, and their many common features bear this out.

Summary

The literature concerning keratoconus posticus is reviewed. Some new cases are described. It is suggested that in nearly all cases the condition originates in early life and is probably present at birth.

It is a pleasure to record my debt to Mr. A. J. B. Goldsmith for his assistance in the preparation of this paper. For permission to publish their cases, I wish to record in addition my thanks to Mr. Ayoub, Mr. Gimblett, and Mr. A. Lister. Professor Sorsby kindly arranged for the keratometric readings.

REFERENCES

- BUTLER, T. HARRISON (1927), "An Illustrated Guide to the Slit-lamp", p. 39. Oxford University Press, London.
- (1930). *Trans. ophthalm. Soc. U. K.*, **50**, 551.
- (1932). *British Journal of Ophthalmology*, **16**, 33.
- GOLDSMITH, A. J. B. (1943). *Trans. ophthalm. Soc. U.K.*, **63**, 180.
- GREENE, P. B. (1945). *Arch. Ophthalm. (Chicago)*, **34**, 432.
- GUIMARÃES, W. (1953). *Arch. brasil. Oftal.*, **16**, 209.
- LEOPOLD, I. H. (1943). *Arch. Ophthalm. (Chicago)*, **30**, 732.
- STALLARD, P. L. (1930). *Trans. ophthalm. Soc. U.K.*, **50**, 556.
- WISE, G. (1944). *Amer. J. Ophthalm.*, **27**, 1406.