

KERATOGLOBUS AND KERATOCONUS*

A CONTRIBUTION TO THE NOSOLOGICAL INTERPRETATION OF KERATOGLOBUS

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AN interesting work by Verrey (1947) discussing the nature of that very rare deformation of the cornea known as keratoglobus has led me to write about a case I observed many years ago, since I feel that the arguments I was able to deduce therefrom give significant confirmation of the conclusions reached by Verrey.

Previous Opinions

Ophthalmological literature does not agree as to the correct interpretation of the nature of keratoglobus. Less recent authors are generally inclined to identify it with megalocornea and to look upon it as a symptom of arrested hydrophthalmos.

"Keratoglobus, also known as megalocornea, is usually accompanied by infantile hydrophthalmos or glaucoma" (Bietti, 1925).

"There is a form of ectasia of the bulb known as hydrophthalmos, megalophthalmos, or also, because the enlargement of the cornea particularly attracts our attention, as megalocornea or keratoglobus: the affection is congenital or appears shortly after birth without any apparent cause" (Fuchs, 1889).

"Keratoglobus or megalocornea is a partial manifestation of megalophthalmos; usually both keratoglobus and megalophthalmos do not result from an abnormal development though still within physiological limits, but from an increase in tension which, moreover, remains latent, namely a form of hydrophthalmos that has become stationary" (Elschnig, 1923).

Among more recent authors, Prélat (1939) distinguishes keratoglobus from megalocornea proper, identifying the former with buphthalmos:

"When the increase in the diameter of the cornea is regular, it is given the name of megalocornea. This affection may be of two quite distinct types: sometimes it is what is known as essential megalocornea, whereas in other cases the increase in size is apparent in the whole of the eyeball which may be considerably dilated. This deformation, known as keratomegalia, keratoglobus or buphthalmos, is the consequence of iridociliary lesions developing during the intra-uterine period or during the first months of life and is very similar to infantile glaucoma, which is frequently one of the complications of hereditary syphilis".

Duke-Elder (1938), on the other hand, attributes keratoglobus and megalocornea to the same group of deformations, dealing with congenital hydrophthalmos as something apart:

"Megalocornea (keratoglobus, keratomegalia, cornea globosa, anterior megalophthalmos, etc.) is a bilateral developmental anomaly wherein, in the absence of a raised intra-ocular pressure, the anterior segment of the eye is larger than normal".

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Schieck (1931), however, considers keratoglobus as a separate affection erroneously confused with megalocornea :

"Megalocornea is characterized by an increase in the diameter of the corneal disk and is the opposite of microcornea, whereas keratoglobus is characterized by a lessening of the radius of the curvature of the cornea and consequently by an increase in the curving of its surface; its opposite is cornea plana".

According to Schieck, this distinction is not mentioned in the literature, because what has always struck oculists as the essential feature of the foregoing affections is the deepening of the anterior chamber, which is common to both. "For this reason", he concludes, "we find the names megalocornea and keratoglobus used indifferently, side by side, although this use is certainly erroneous".

Verrey's Case

I have reported at some length the contrasting opinions of ancient and modern classical authors regarding the nature of keratoglobus, because these quotations show so clearly that the nosological classification of keratoglobus is still uncertain and confused. In my opinion a good step towards the solution of this question has been made by Verrey, who cites a case of keratoglobus observed in a 23-year-old girl.

Case Report.—The two corneas were quite transparent, very globous and enlarged, with a basic diameter of about 13 mm.; they were also very thin, being about one-third the thickness of a normal cornea. The anterior chamber was very deep. A closer examination of the thickness of the cornea showed that the thinnest part corresponded to an area not far from the limbus, so that the all-round picture was of a pellucid ectasia, representing, so to speak, the reverse of keratoconus in so far as the least resistant part was marginal instead of in the central area of the cornea.

From these traits Verrey concluded that he was dealing in this case with an affection similar to keratoconus, and this opinion was borne out by the fact that he was able at a certain moment to observe in his patient's left eye the appearance of an acute affection of the type described by Pflüger (1876) as acute keratoconus, and consisting, as is known, in a sudden clouding of the cornea due to rupture of Descemet's membrane.

In fact, without any kind of premonitory symptom, the eye in question became irritated and began to water, and there was intense photophobia, the cornea being clouded and very much thickened by the rapid absorption of aqueous humour by the corneal parenchyma. The slit-lamp showed that at the points where the cornea was most seriously affected, the epithelium was detached, Descemet's membrane had lost its normal adherence to the front layers, and the parenchyma showed fissures resembling those to be found in the vitreous in a tumefied cataract. The symptoms came to a climax a few days later and then began to recede, but Verrey was unable to follow the later stages up to the final result.

Verrey concludes that this acute affection, similar to that sometimes observed in keratoconus, is a valuable argument in favour of the thesis that keratoglobus is more similar to keratoconus than to megalocornea. He consequently distinguishes two groups of deformations of the cornea which are apparently similar :

- (1) those in which there is simply an increase in the diameter of the cornea and consequently in the front segment of the eye;
- (2) those in which there is a change and a lessening in thickness of the corneal parenchyma of mesodermic origin with all the similarities that this suggests from the standpoint of general pathology."

Personal Observations

Many years ago, in the Eye Clinic of Naples University, I was able to observe a typical case of keratoglobus which, in its clinical aspect and the manner of its development, was almost identical with that described by Verrey. In this case also, the marked enlargement of the two corneas was accompanied by a considerable thinning, and an acute affection appeared in the right eye similar to that described above. Further it is interesting to remark that our patient had a son suffering from a typical keratoconus, yet another point in favour of Verrey's opinion concerning the nosological interpretation of keratoglobus.

Case of Keratoconus.—In September, 1932, an 18-year-old student came to the infirmary of the Naples Eye Clinic; he complained of visual failure in both eyes, but particularly in the left; this had started some years ago, progressing gradually until he was now unable to read ordinary print. As a consequence of this weakening of his sight he had been obliged to interrupt his studies a year ago.

The patient was suffering from a typical form of keratoconus in both eyes—but more advanced in the left eye—which had the characteristics of the second type of keratoconus described by Salzmann (1908). The cornea differed from that of the commoner cases in that the cone started from the marginal area near the limbus. The corneal membrane was transparent and very thin. There was no other change in the eye. Vision in the right eye was 1/10 with sph. $-6 = \text{cyl.} -5$ at 60° ; in the left eye it was less than 1/10. There was nothing the matter with the patient's general health. Since it was found impossible to improve his sight with spectacle lenses, contact lenses were prescribed, which were made by Zeiss. Thanks to these contact lenses, which the patient was able to tolerate for many hours a day, he was able to resume his studies.

In 1936 the condition had deteriorated so that the patient no longer benefited from the contact lenses which he had used so far, and as the apex of the cone of the left eye was becoming opaque, we decided to operate. This operation was performed in the Eye Clinic of Rome University, first on the left eye and then on the right, by means of galvanocautery puncture of the most salient portion of the cornea. The patient benefited so much from this operation that he was again able to resume his studies and other activities. The vision in the right eye had risen to 5/10 with sph. $-8 = \text{cyl.} -3.50$ at 75° , and the left vision to 2/10 with sph. $-6 = \text{cyl.} -4.50$ at 65° .

Subsequently the keratoconus remained stationary in the right eye, the sight of which remains as it was 13 years ago. In the left eye, however, the corneal deformation increased again, and some opaque spots formed around the small operation-leukoma. Keratoplasty was performed this year with excellent results.

Case of Keratoglobus.—Shortly after I first visited this patient, I saw his father who had been suffering from eye trouble since childhood. This man was 54 years old at that time, and a milkman by trade. There was nothing of particular interest in his

past history. He had always enjoyed good health; he had married a healthy woman who had borne him eight children, six boys and two girls; two had died in infancy, but the others were alive and healthy. Apart from the son who was suffering from keratoconus, none of the children appeared to suffer from eye trouble. The wife had never had any miscarriages and had died some years earlier of an acute disease, the exact nature of which had not been ascertained. The patient told me that he had suffered from a deformation of the eyes since childhood, which had never, however, interfered with his occupation. What was worrying him at the time when I saw him was an irritating burning sensation in both eyes. In fact, the patient's eyes were reddened by a flare-up of chronic catarrhal conjunctivitis, but what first attracted notice was the singular appearance of the corneae, which were typical of an advanced keratoglobus.

The corneae were extraordinarily large and prominent: the horizontal diameter at the base was 18 mm.; the anterior chamber was enormously deep. The corneal membrane was quite transparent and very thin, as could be seen from an examination with the slit-lamp, which showed that it was less than half the normal thickness: there did not appear to be any marked differences in thickness between one region of the cornea and another. Near the limbus there were neither opacities nor vascular loops. The iris presented a normal aspect and the refractive media were transparent. There was nothing particular about the fundus: the optic disk was pink and not cupped. The intra-ocular pressure was normal, vision could not be ascertained because of the patient's backward mental development but was approximately 4/10 in both eyes, with the aid of correcting-lenses for slight myopic astigmatism. The visual field was full.

An argyrol collyrium was prescribed, and the patient was not seen again for a number of years. In May, 1935, he returned, complaining that two days before, for no particular reason, the sight of his right eye had suddenly failed.

Examination showed moderate circumcorneal injection, together with loss of corneal transparency. Photophobia and lacrimation were also noted. The corneal membrane presented a cloudy and thickened appearance by reason of sudden spontaneous imbibition, such as often occurs in keratoconus: this condition was particularly evident axially and above, where the opacity was milky and deep with indistinct outlines. Examination with the slit-lamp showed that the thickness of the parenchyma in the most seriously affected region was very great, the epithelium was bedewed by oedema; no details of Descemet's membrane were to be perceived. The eyeball was slightly hypotonic, and the cornea somewhat flattened. Vision was reduced to finger-counting, while the visual field remained normal. No change was found in the other eye.

Atropine and dionin drops were prescribed as well as hot damp compresses. Eight days later there was already appreciable improvement, the cornea had grown clearer and the reactive conditions were making progress.

Recently I had news of this patient who is now 71 years old; as he cannot leave his home, which is very far away from Rome, owing to his age and the rheumatism that has been crippling him for some years, he was visited by a colleague. This visit in the patient's home gave the following results:

Senile spastic entropion of the lower lid and chronic conjunctivitis in both eyes.
Right eye: Enormously enlarged and bulging cornea, three-quarters opaque; folds in Descemet's membrane still evident. Horizontal diameter 17 mm. Depth

of anterior chamber greatly increased. Opacity of the crystalline lens. Vision = under 1/10. Ocular tonus and visual field normal.

Left eye: Enormously enlarged and bulging cornea of normal transparency. Diameter 18 mm. Depth of anterior chamber greatly increased. Central opacities of the crystalline lens. Normal fundus. Vision = 1/30. It was not possible to attempt a correction with spectacles.

This then is the case of a patient who has been suffering since childhood from an eye affection so typical of keratoglobus that as Verrey says, referring to his own case :

"If the expression keratoglobus did not exist, it would have to be invented, so suited is it to the corneal deformation of our patient. The clinical picture, in fact, was that of a pellucid ectasia of the two corneae, which were enormously enlarged and globous: they were less than half the normal thickness and the anterior chamber was exceptionally deep. The endocular tonus, the fundus of the eye and the visual field were normal".

In our case, as in Verrey's, there was an acute imbibition of the corneal parenchyma in one eye at a certain moment, due to rupture of Descemet's membrane, such as frequently occurs in keratoconus. This acute imbibition is often found in keratoconus and, though it is not frequent, we have observed it several times in our clinic and attention has recently been drawn to it by Amsler (1940).

The fact that this condition occurred in our patient and that there was a marked curvature and thinness of the cornea go to prove the affinity between keratoglobus and keratoconus.

Another point of considerable importance illustrated by our case, is the fact that one of our patient's sons suffered from keratoconus and had to undergo several operations. Unfortunately we were not able to carry out further genealogical researches which would undoubtedly have proved most interesting.

I feel that the foregoing observations help considerably to clear up the problem of the nosological interpretation of keratoglobus. This extremely rare deformation of the cornea deserves to be classified as a distinct affection with well-defined characteristics, and its name should only be used to indicate a pellucid ectasia of both corneas, with an exceptional enlargement and curvature of the corneal membrane, and a marked thinning of the parenchyma. These features of the affection and the possibility of an acute phase of clouding of the cornea due to rupture of Descemet's membrane during the course of the disease, as well as hereditary transmission in the form of keratoconus, prove that there is a close relationship between keratoglobus and keratoconus. Consequently keratoglobus must not be confused, as it has been so far by the vast majority of authors, either with megalocornea, which is characterized by an increase in the basic diameter of the cornea and consequently of the front segment of the eye (it is therefore a sign of anterior megalophthalmos),

or with arrested hydrophthalmos, since in keratoglobus we do not find a total increase in the size of the eye-ball, or increased tension, or cupping of the disk, or an advance of the limbus over the cornea, as has been pointed out by Schieck (1931).

Summary

(1) Having established that opinions still differ widely concerning the nature of the rare deformation of the cornea known as keratoglobus, a case is described that should help in establishing its exact nature.

(2) Both corneae of the patient in question were enormously enlarged and globous, and the anterior chamber was exceptionally deep, the corneae being less than half the normal thickness. At a certain moment an acute imbibition of the cornea occurred in one eye as a result of rupture of Descemet's membrane, such as sometimes occurs in keratoconus. One of the patient's sons suffered from keratoconus.

(3) The clinical features observed are very similar to those seen in Verrey's recent case.

(4) The conclusion is reached that keratoglobus is a distinct affection with well defined characteristics (great enlargement and curvature of the cornea, marked thinning of the parenchyma), and closely connected with keratoconus. Consequently it must not be confused with megalocornea or with arrested hydrophthalmos as it has been hitherto by the majority of authors.

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