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CHRONIC EDEMATOUS NEURITIS OF THE CORNEA (NEURITIS EDEMATOSA CR--ETC(U)  
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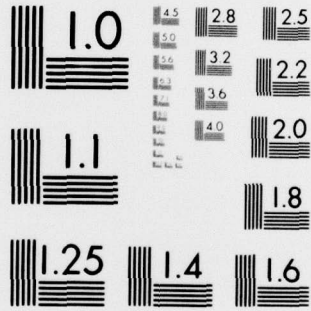
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AB

CHRONIC EDEMATOUS NEURITIS OF THE CORNEA

By Prof. Dr. Diego M. Argüello, Dr. Bruno Tosi and Dr. Carlos B. Gayoso

SUMMARY

The authors succinctly discuss the topic of familial corneal dystrophies, contributing their observations of a family exhibiting the symptoms of reticular keratitis.

They emphasize the significance of the striae, re-interpret the results of vital staining, and express their opinion that the striae are nothing other than profoundly altered corneal nerves.

\* \* \*

The still poorly defined topic of hereditary dystrophies of the cornea encompasses a wide variety of diseases which are characterized in common by the fact that they almost always arise after puberty, their chronic, progressive course, the symmetry and bilateralism of their lesions, the predominance of disturbances in the central portion of the cornea, the absence of vascularization, the lack of violent inflammatory signs, their unknown etiology and, particularly, their strong hereditary content.

These dystrophies may have their most significant manifestation in the corneal epithelium or the corneal endothelium, or they may extend through the entire thickness of the membrane (Berliner [2]). Three types of familial corneal dystrophy of the latter kind have been identified: the nodular type (dominant in nature), the macular type (which is recessive) and the reticular type (which is transmitted as a dominant trait). According to Bücklers [3], none of these forms assumes the characteristics of any of the others. Duke-Elder [6], on the other hand, cites cases in support of the identity of all

three forms, in which characteristics or aspects of each of the three types of dystrophy were observed in a single patient, a different form was present in each eye, or one type was seen to change into another.

The concept of reticular keratitis is still somewhat confused, both from the descriptive point of view and from that of how to approach and interpret the biomicroscopic picture at hand. Various corneal processes of different kinds and in different locations were described as forms of keratitis (reticular, lattice, net, grillagée) before the slit lamp came into use, but many of them were seen after the advent of biomicroscopy to actually consist of folds or tears in Bowman's membrane or folds in Descemet's membrane (López-Lacarrère [11]).

A review of cases reported by other authors suggests that the first biomicroscopic indication of true reticular keratitis is likely to be an improved visualization of the corneal corpuscles (Kleefeld [10]).

A second stage of evolution is marked by the initial appearance of lines or striae in the deeper region of the optic section, undoubtedly reflecting the presence of folds in Descemet's membrane; the endothelium loses its classic polygonal configuration, but no precipitates are visible.

The most characteristic element of the affection comes into the scene at this stage of the process, to such an extent that it suggested the name given to the disease. It consists of the presence of numerous radially arranged striae which originate near the limbus and dichotomize repeatedly as they extend toward the center of the cornea. These ribbon-like striae are waxy in appearance, have sharply defined, toothed edges, and are translucent when back-lit; their numerous branches may lie in different planes and usually intersect each other, but cases of true anastomosis have not been demonstrated (Gallemaerts [8], Kleefeld [10]).

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The content of the striae is finely granular, and they cannot be mistaken for blood vessels; they increase in number as they approach the center of the cornea and their details become more visible by back-light; they are almost always radial in direction, but a few occasionally extend concentrically with the center of the cornea for a small distance. Some authors describe them as optically empty tubes with a double outline, whose pointed extremities vanish at the periphery without reaching the limbus (Berliner [2]); for others, however, the striae do reach the limbus, where they become confused with the nerves of the cornea.

Numerous irregular opacities are observed in the central portion of the cornea. They may extend throughout the thickness of the cornea, so that the whole takes on a disciform, opaque appearance.

The epithelium exhibits areas of desquamation and, occasionally, true recurrent ulcers that may take on the characteristics of a herpetic keratitis (Adrogué [1]). Lloyd [12] believes that the nodules that form beneath the epithelium probably explain the development of the ulcers, since the eyelids would then rest more heavily and closely on these outermost points.

In the last stage, the striae become fragmented, the cornea scleroses, and deep blood vessels of the parenchymatous type have been seen to develop occasionally (Kleefeld [10]). As a general rule, however, the process is avascular throughout its evolution.

All the authors agree in noting serious disturbances of corneal sensitivity.

It is worth noting that reticular keratitis patients claim better vision and less subjective discomfort than would be expected in view of the magnitude of the observable corneal lesions.

Anatomicopathologically, the fundamental change seems to consist of a

deposition of hyaline substance, causing disintegration of the corneal parenchyma and destruction of Bowman's membrane (Berliner [2]). For Fuchs [7], this seems to be a degeneration of the connective tissue, affecting primarily the elastic fibers.

The most characteristic element of the disease is precisely the one that is most discussed--namely, the significance of the striae.

Some observers (Kleefeld [10], Gallemaerts [8]), relying primarily on the biomicroscopic picture, identify the striae as corneal nerve endings that have undergone edematous degeneration; in fact, following a stria inward from the limbus, one observes that it begins as a nerve which after a short distance assumes the ribbon-like appearance we described above, with a diameter up to five times larger than a normal nerve. For this reason, Kleefeld [10] gives to reticular keratitis the name we adopted in this communication, chronic edematous neuritis of the cornea. Others consider the striae to result from breaks in the parenchyma or in Bowman's membrane, which are then filled with a light hyaline substance (Haas [9]). Stanka [13] states that it is only a dystrophic process of the corneal lamellae. Byers [4] believes that, in his case, the reticular keratitis resulted from endoocular hypotension, relating it to the folding of Bowman's membrane.

Those authors who deny any connection or linkage between the striae and the corneal nerves base their opinion especially on the results of vital staining. Methylene blue, considered specific for nerve endings, does not stain these elements, in contrast to brilliant cresyl blue, which appears to have some affinity for the corneal lamellae (Agrogué [1]).

The etiology of the disease also is not based on a sound foundation. Some authors suppose nutritional deficiency or endocrinal insufficiency, or point to tuberculosis as a causal factor. Adrogué [1] believes that the dif-

ferent forms of familial corneal dystrophy belong to the same clinical picture and follow attacks of herpetic keratitis.

Those authors who identify the striae with the corneal nerves maintain that reticular keratitis is due to an illness of the endings of the trigeminal nerve, that is, that it has a neurotrophic origin. The condition would then be an abiotrophy, a primitive dystrophy of the nerve fibers.

Treatment is symptomatic and has its widest application in the ulcerative episodes, which are sometimes recurrent. Kleefeld [10] recommends polyglandular extracts.

Our contribution to the casuistics is as follows:

A. G., 63 years old, smelter. Does not recall any hereditary background of interest; is the father of fifteen children, thirteen of whom are alive and healthy.

His vision was first affected only six years ago, because of a corneal ulcer in the right eye that required eight months to heal; a similar episode occurred two years ago, lasting two months. The patient attributed both processes to the penetration of a foreign body, but we were unable to verify this. He has always seen well with his right eye, since he was fond of hunting and used this eye for aiming.

The following interesting biomicroscopic picture was discovered upon examination with the slit lamp immediately after his first corneal ulcer:

O.D. — The presence of numerous striae is immediately apparent. Originating near the limbus, they extend radially toward the center of the cornea, bifurcating repeatedly and approaching the surface as they advance. The striae become tapered and thinner as toward the periphery and disappear from view before reaching the limbus, with the exception of four which continue directly as corneal nerves in the lower external sector (Figure 1, A). The striae have a slight waxy yellowish color when back-lit and are

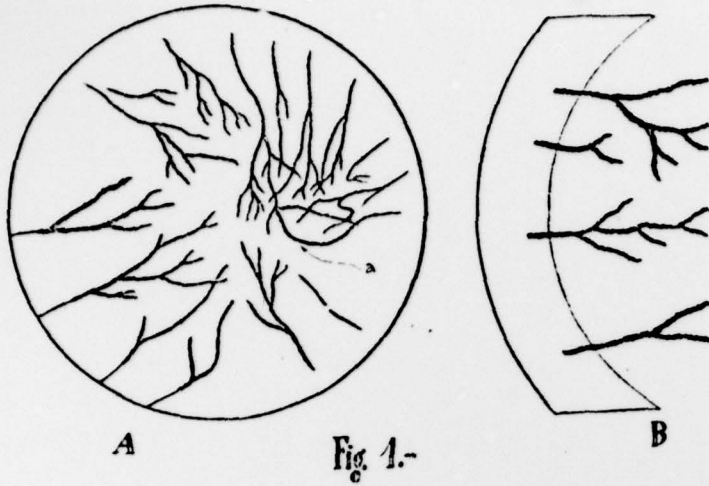


Figure 1. Chronic edematous neuritis of the cornea.

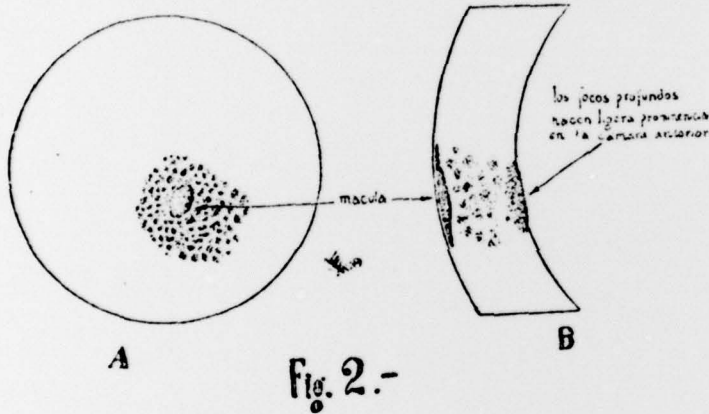


Figure 2.

The deep foci protrude slightly into the anterior chamber

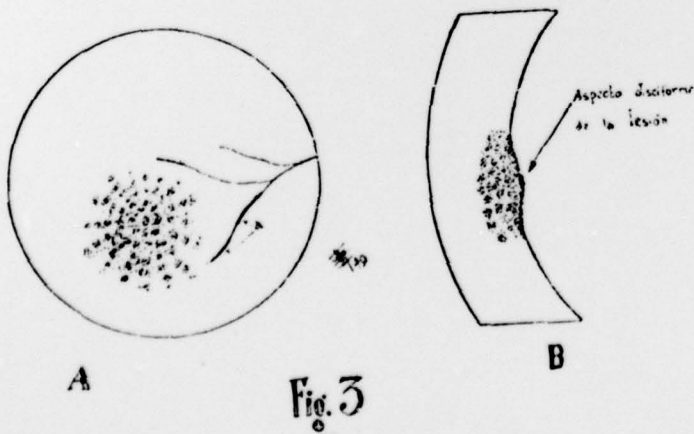


Figure 3.

Disciform appearance of the lesion

opalescent at the optic section; they are several times thicker than a first-order corneal nerve and have finely indented or wrinkled edges (Figure 1, B). They stand out clearly from the surrounding medium, crossing over each other with a few apparent anastomoses (Figure 1, A, a). We did not observe the continually changing, kaleidoscopic picture, varying according to the angle of incidence of the light or the depth of the biomicroscopic focus, that has been described by some authors, nor the appearance of optically empty tubes, giving rise to the production of small holes at the optic section, that the striae are said to have.

A multitude of small foci, nodules or opacities is observed intermixed with the striae in the central portion of the cornea (Figure 2, A, drawn separately for simplicity). They have the same waxy appearance as the striae, are irregularly and idiosyncratically shaped, occasionally touch each other, and have somewhat indefinite edges. They are scattered in all the corneal layers but predominantly in the deeper ones, where they pile up to form a disciform opacity which protrudes slightly into the anterior chamber (Figure 2, B). A corneal macula (Figure 2, A and B) is also observed, a result of the recurrent ulcer mentioned above.

It is appropriate to mention at this point that of the three known forms of familial corneal dystrophy, the only one reported to involve ulceration is precisely the reticular for we are describing.

Repeated instillation of a 0.5% solution of methylene blue did not stain these elements; on the other hand, 1% brilliant cresyl blue, instilled numerous times in alternation with 5% cocaine, did stain not only the striae but also the entire thickness of the cornea, including the opacities. The staining encompassed a large central area, leaving a peripheral ring unaffected by the coloring agent; although a few striae were somewhat attractive to the stain for some distance beyond the limit reached in the cornea, an appreciable portion of each of them was not stained.

The corneal sensitivity of this eye was clearly diminished, and vision was measured as 1/10.

O.S. — The most noteworthy aspect in this eye is the presence of the small opacities already described with reference to the right eye (Figure 3, A). Most of them are located in the deep layers of the cornea, forming also in this case a disciform opacity herniating toward the anterior chamber. The corneal nerves are visible and normal in appearance, except one which, after dichotomizing twice, appears thickened and wrinkled, resembling the striae described for the right eye but much thinner than them (Figure 3, A, a). It may represent an incipient process stage that attacks the corneal nerves. Sensitivity is not affected and vision is 6/10. Neither eye exhibits corneal blood vessels, and the patient does not claim any discomfort.

We were able to examine only twelve of this patient's thirteen children. Eleven had absolutely no suggestive signs in their corneas, except for one (a 29-year-old male) whose corneal nerves were somewhat more visible than is normally the case.

One 39-year-old daughter, however, whose only recalled visual background is an inflammation of the right eye lasting some time two to three years ago, exhibits the following biomicroscopic picture (which was discovered accidentally, since she experiences no ocular disturbances whatsoever):

O.D. — The nerves are clearly visible and abundant. They can be seen as easily by back-lighting as by direct focal illumination (whereas they are normally visible only by the latter method). It can be stated without hesitation that some of the nerves first appear in the limbus with every characteristic of nerves, bifurcate once or twice, and then merge directly into the formations we have been calling striae and believe represent pathologically elongated corneal nerves; it is true, however, that these striae still have not reached the enormous thickness observed in this patient's father. The central portion of this eye also exhibits small waxy opacities of various shapes and sizes, distributed through all the corneal layers but concentrated in the deeper ones. Lastly, a superficial macula is observed in the lower parapupillary field, undoubtedly a result of a corneal ulcer, which explains the irrita-

tive process that occurred two years ago.

O.S. — The left eye reveals nerves having the cited characteristics, but no parenchymal opacities are evident. The nerves are more visible and more opalescent than in the normal state and are thickened in places to the point that they can be seen easily by retroillumination. Some of the striae are direct continuations of nerves, but for others such an origin could not be confidently established.

The sensitivity of the cornea seemed normal, although the great tolerance evidenced in the ability to bear biomicroscopic sessions without prolonged discomfort is noteworthy. Vision was 10/10 in both eyes.

We are thus in the presence of a chronic affection of the cornea, of a dystrophic and familial nature, transmitted as a dominant trait (to only one descendant for the time being).

We, also, have thought deeply about the significance of the lines or striae.

First of all, let us discard the hypotheses of folds and ruptures of Bowman's membrane and of folds of Descemet's membrane, since they present a completely different biomicroscopic picture and their location is also different in the optic section of the corneal prism (Tosi [14]).

From careful observation of our two patients, we reached the conclusion that in their case the striae should be identified with the corneal nerves, for the following reasons:

(1) the biomicroscopic picture: the origin, arrangement, location and dichotomization of the striae are consistent with our knowledge of the anatomy of the corneal nerves;

(2) from the study of the family we present, the earliest sign of this corneal dystrophy appears to be a greater visibility of the nerves, not of

the corneal corpuscles as maintained by some authors (Kleefeld [10]);

(3) the parallellism between the corneal anesthesia and the degree of visibility of the striae--the disturbed corneal sensitivity appears to be the clinical expression of its anatomic alteration;

(4) the appearance of herpetic corneal ulcers, which are known to develop only in the corneal dystrophy type which is accompanied by striae;

(5) our patient's right eye, with its exuberant development of striae, did not exhibit any nerves, while nerves were clearly visible in his left eye, which had but a single stria indicating an early stage of the disease;

(6) in this patient's daughter, whose nerves were not yet seriously altered, it could be clearly seen how the striae were direct continuations or prolongations of the nerves; and

(7) the fact that some striae do not appear to originate in the nerves can be explained in part by the opacity of the parenchyma and in part because the nerve endings are sometimes not visible unless the light beam incides on them at a particular angle.

The only argument against the nerve hypothesis is the results of vital staining, inasmuch as 0.50% methylene blue did not stain the striae but 1% brilliant cresyl blue did so.

We share the belief of Adrogué [1], that the vital staining is not decisive. In fact, we have used 1% methylene blue, instilled repeatedly in alternation with 5% cocaine, in a patient whose corneal nerves were highly visible, and the nerves were nevertheless not stained at all, so that the clinical value of methylene blue as a specific staining agent of corneal nerve fibers in vivo is very relative. As for brilliant cresyl blue, we have already noted that it stained only the striae in the central portion of the cornea, where the cornea's architecture--together with its chemical affini-

ties, upon which the staining effects depend--is undoubtedly profoundly altered. If one assumes that the striae represent corneal lamellae, as some authors maintain (and for which brilliant cresyl blue would be specific), they should become stained throughout their length, not partially as in the case of our patient. It can also be argued that, in reality, what the brilliant cresyl blue stains is the detritus of a profoundly altered nerve. Furthermore, against the specificity of brilliant cresyl blue, we note that mercurochrome instilled by the same technique is capable of intensely staining the entirety of the corneal parenchyma for several days without staining the nerves.

For all these reasons, we are inclined to believe that reticular keratitis has its most important effect at the level of the corneal nerves, thus justifying its designation as chronic edematous neuritis of the cornea, with which we headed this modest contribution to the national casuistics.

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