“Contact lens” cornea in rheumatoid arthritis

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It has been noted that patients suffering from long-standing rheumatoid arthritis often display a characteristic corneal lesion.

This consists of a partial opacity of the periphery of the cornea involving part or all of its circumference. The cornea in this area is thinned, usually by about a third of its normal thickness and has a well-demarcated central edge. It is minimally vascularized by superficial vessels crossing the limbus. The appearance is quite striking even upon superficial examination and resembles that of an eye wearing a microcorneal contact lens, so much so that it could well be called “contact lens” cornea (Fig. 1).

FIG. 1 Drawing of slit-lamp appearance of cornea in the cases described (the lesion at 12 o'clock is a benign melanoma of the iris)

Five cases illustrating the clinical features referred to are summarized in the Table (opposite).

Case reports

Case 1, a woman aged 70 years, had experienced soreness of both eyes from time to time over the last 10 years, not severe enough to warrant seeking medical advice. She had suffered from rheumatoid arthritis for 30 years with severe joint deformity, and had been treated with salicylates only. She attended the clinic because the right eye had been sore for 5 weeks.

Examination

Both eyes showed an annular opacity extending 2 or 3 mm. inwards from the limbus. The right eye showed slight injection, but there was no staining after the instillation of fluorescein. There was a small area of scleral thinning at 12 o'clock on the right eye. The visual acuity was 6/6 in each eye, the media clear, and the fundi normal. There was no evidence of uveitis either past or present.
Table  Summary of five cases described

<table>
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<th>Case no.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Duration of RA (yrs)</th>
<th>Glaucoma</th>
<th>Eye</th>
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Eye lesions
coronal opacity
scleral thinning

Treatment
The symptoms resolved rapidly upon treatment with local mydriatics and steroids, and the appearance of the eyes was the same when the patient was reviewed 2 years later.

Full examination for glaucoma with applanation tonometry and Bjerrum screen testing showed no abnormality and the tear secretion was normal.

Case 2, a woman aged 64 years, was referred by a rheumatologist who had thought the appearance of the eyes was abnormal when she attended for treatment of arthritis. She had had minimal soreness of both eyes from time to time. Rheumatoid arthritis had been present for 20 years with severe joint deformity.

Examination
Both eyes had the characteristic ring of opacity involving the periphery of the cornea with slight superficial vascularization (Fig. 2, overleaf). The visual acuity of each eye was 6/9 after refraction. There was no evidence of glaucoma or uveitis, and tear secretion as tested by Schirmer's test papers was normal.

Result
The appearance of the eyes was unchanged when the patient was reviewed 6 months later.
Case 3, a woman aged 62 years, attended because of pain in the right eye for the previous month. She had had similar symptoms in a milder form from time to time which had resolved with treatment by drops prescribed by her doctor. She had suffered from rheumatoid arthritis for 13 years with multiple joint deformities.

Examination

The right eye showed the characteristic band of opacity on the cornea extending from 5 o’clock to 12 o’clock. The central edge of the opacity stained with fluorescein at 9 o’clock (Fig. 3).

The left eye showed the same type of opacity extending all round the peripheral cornea, but there was no staining point on this eye. The visual acuity was 6/18 in each eye, the decreased vision being due to lens opacities. There was no sign of uveitis or glaucoma and the tear secretion was normal.

Treatment

The staining area healed within 6 weeks after treatment with local steroids and mydriatics.
Case 4, a woman aged 62 years, had suffered from recurrent corneal ulceration of the left eye for about 4 years. The symptoms usually lasted for 3 months or so and resolved upon treatment with local mydriatics and steroids. There had never been any inflammation in the right eye. She had had rheumatoid arthritis for 7 years, and had been treated with salicylates only. The last and most severe attack had lasted for 3 months and showed no sign of resolving.

Examination

The visual acuity was 6/9 in each eye. The right eye showed a small area of opacity at 6 o'clock. The left eye displayed a ring of opacity on the cornea extending from 5 o'clock to 12 o'clock. It was 3 mm. or so in width and at a descemetocele was present at 9 o'clock (Fig. 4).

Treatment

It was felt that perforation of the left eye was likely in the weakened area, and an annular penetrating graft was therefore performed. The cornea healed without incident and corneal thickness has been maintained with no further episodes of ulceration in the 6 months follow up. Schirmer's test showed absence of tear secretion.

Case 5, a man aged 56 years, had experienced recurrent soreness of both eyes for several years, not severe enough to warrant seeking medical advice. He had rheumatoid arthritis of 17 years' duration and could walk only with the aid of crutches.

Examination

The visual acuity was 6/6 in each eye after refraction, with a moderate degree of myopia. Both eyes showed the typical annular opacity extending completely around each cornea, and there was also extensive scleral thinning all round in both eyes. Applanation tonometry showed that the ocular tension was 31 mm. Hg in the right eye and 58 mm. Hg in the left. The central fields on Bjerrum screen testing with a 2 mm. object showed no field loss and the discs did not appear to be cupped, although they were difficult to assess because of the myopic changes present. Gonioscopy showed open angles of medium width with no peripheral anterior synechiae.

Treatment

The patient was treated with miotics which controlled the raised ocular tension until his death a few months later.
**Differential diagnosis**

The relative lack of pain and acute inflammatory appearance differentiates this type of marginal ulceration from the necrotizing sclero-keratitis seen in association with the connective tissue diseases, most commonly with polyarteritis nodosa. This condition is acutely painful, causes rapid loss of corneal substance, and responds only to high doses of systemic steroids.

Terrien's marginal ulceration usually affects the superior cornea and usually has a more marked inflammatory element.

Senile marginal degeneration usually affects the interpalpebral area and does not often extend completely round the periphery of the cornea.

A very similar appearance on superficial examination may be seen in uveo-keratitis with sclerosing keratitis; this is illustrated by the two following patients:

**(A)** A woman aged 59 years had suffered from recurrent anterior uveitis for approximately 27 years and had herself noted a white ring surrounding both corneae for the past 5 years. Superficial examination showed an appearance very similar to that of Case 5, with the “contact lens” cornea surrounded by scleral thinning, but slit-lamp examination revealed that the opacity involved the posterior third of the cornea and was accompanied by deep vascularization. This patient had in addition numerous peripheral anterior synchiae in both eyes and had been treated for glaucoma for several years. She had had an iridectomy performed upon the right eye. Both discs were cupped and the visual fields considerably contracted.

Extensive investigations for rheumatoid arthritis, including serology and x-ray examination of the small bones of the hands and feet, were all negative.

**(B)** A woman aged 42 years had suffered from attacks of inflammation in both eyes lasting for several weeks at a time for many years. She had also been treated on one occasion for “acute” glaucoma. The visual acuity was 6/6 in each eye and both showed a white ring involving the periphery of the cornea. There was no scleral thinning but there was mild activity in both anterior chambers. Again, the slit lamp showed that the opacity involved the posterior third of the corneal thickness and that the superficial cornea was not affected. It is interesting to note that this patient also had numerous peripheral anterior synchiae causing raised intraocular pressure in each eye with accompanying visual field loss and cupping of the disc. She had had glaucoma surgery performed upon both eyes.

**Discussion**

Although rheumatoid arthritis is known to be accompanied by deficiency of lacrimal secretion and the corneal changes resulting therefrom and may also be accompanied by the various types of scleritis (Lyne and Pitkeathly, 1968), there are few accounts of corneal changes similar to those described above. Case reports of marginal furrows, which are almost certainly the same entity, include those of Brown and Grayson (1968), Frasca (1958), and Collier (1952).

Brown and Grayson described nine cases of marginal furrows of the cornea affecting patients suffering from rheumatoid arthritis of more than 5 years' duration. The furrows were seen most commonly at the inferior aspect of the limbus and were bilateral in four of the patients. Only one of their cases corresponded at all closely to those above in being circumcorneal. Inflammation and vascularization were minimal.

Four of the five patients in my series were women, and the average age was the middle sixties. This is to be expected in view of the association with rheumatoid arthritis, which is itself more common in women than men, and in which average age at onset is the middle thirties.
The mechanism of development of these lesions is not easy to explain. They may be due to the circumferential extension of a "dellen". These lesions consist of a shallow oval ulcer about 1.5 x 2 mm. in size accompanying a limbal swelling due to any cause.

Barraquer (1965) suggested that they were due to a discontinuity of the tear film caused by elevation of the lid by the limbal swelling and leading to desiccation of the cornea adjacent to it. From my own observations of these lesions, I am inclined to doubt the existence of the dry spot which I have been unable to demonstrate on the slit lamp. On the contrary there is usually a pool of fluid present which tends to stagnate as shown by the persistence of fluorescein in that situation when instilled, and it may be that lack of exchange of metabolites is the cause. It is likely that a pterygium results from the recurrent breakdown and healing of a "dellen", advancing a little further after each episode as Barraquer suggests. If, instead of progressing centrally, the "dellen" occurred at different points upon the limbus, an appearance similar to the "contact lens" cornea could be produced.

On balance, however, it seems more probable that the circumcorneal breakdown is due to a deficiency in the perilimbal vascular supply. It is known that the collagen of the sclera undergoes changes in scleritis associated with rheumatoid arthritis, accompanied by varying degrees of inflammation. It is known also that the arterioles are affected in rheumatoid arthritis, both generally throughout the body and in the eye. So far there has been no opportunity to examine an eye with this type of corneal lesion histologically.

Summary

A peripheral corneal opacity of a distinctive character involving the anterior third of the corneal thickness, was seen in five patients suffering from long-standing rheumatoid arthritis. The opacity was present over the whole of the circumference of the peripheral cornea in the majority of the eyes affected and the condition was bilateral to some degree in all five patients.

Inflammation and vascularization were minimal.

I wish to thank the consultants of the Manchester Royal Eye Hospital for allowing me to describe their case and Mr. R. Neave of the Department of Medical Art, University of Manchester, for the slit-lamp drawing.

References

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