Terrien's marginal degeneration: case reports and literature review

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Terrien's marginal degeneration (TMD) is a rare, bilateral, asymmetric disease of unknown aetiology. The peripheral cornea, predominantly superiorly, undergoes lipid deposition, vascularisation, opacification and stromal thinning leading to 'gutter' formation, ectasia and eventual corneal perforation. Two cases are presented which demonstrate the typical clinical features of the various stages of this disease. The disease process and its spectrum of presentation are reviewed. Differential diagnosis and management of TMD are discussed with particular reference to computerised corneal topographical analysis, which has a limited role for diagnosis but is valuable for monitoring disease progression. (*Clin Exp Optom* 1994; 77: 3: 97–104)

Key words: astigmatism, computerised corneal topographical analysis, ectasia, pseudopterygium, Terrien's marginal degeneration

First described by Trumpy[1] in 1881 and later by others,[2] including Terrien,[3] Terrien's marginal degeneration is a rare condition, with approximately 200 cases having been reported.[4-25] Historically, various names have been given to the disease including peripheral furrow keratitis, ectatic marginal dystrophy,[3] peripheral sclerosis and atrophy, peripheral corneal ectasia, senile marginal atrophy, sulcus marginalis and keratoleptynis.[2] However, Terrien's marginal degeneration (TMD) is now the accepted term for a distinct but somewhat variable clinical entity.

CASE REPORT ONE

A 70-year-old male presented with symptoms of gradually reducing distance vision, epiphora and photophobia. There was no history of ocular pain or inflammation. The general medical history was unremarkable as was the immediate family history.

The patient had been examined 18 months earlier and bifocals were prescribed. Vision with the bifocals was R 6/12 and L 6/15. This improved to R 6/7.5 and L 6/7.5 with pinhole, but best corrected spectacle acuity was only 6/9 R and L. The patient's prescription now required R 2.00 D and L 2.50 D of cylinder, axis against-the-rule whereas 18 months earlier he required 1.00 D of cylinder against-the-rule for each eye. At an examination seven years ago, no astigmatic correction was necessary and vision was correctable to 6/6 in each eye.

Keratometry results showed R 2.00 D and L 2.50 D of against-the- rule astigmatism, with the left mire appearing slightly ovoid with greater steepening superiorly. Retinoscopy also indicated irregular astigmatism. Pupils and binocular vision findings were normal and intra-ocular pressures were within normal limits. The anterior chambers and fundi were normal. Minimal age-related lens changes were apparent.

Slit-lamp examination revealed peripheral corneal thinning with gutter formation, peripheral corneal opacification and vascularisation (Figure 1). These changes were predominantly along the superior corneal margin, but appeared to be distributed randomly around the cornea, affecting half the corneal circumference in total. These changes were bilateral, asymmetric and did not stain with fluorescein. Pterygium-like growths (pseudopterygia) were present at the seven o'clock position in the right eye and at the five o'clock and eight o'clock positions in the left eye. These growths were broad and flat at the leading edge rather than pointed as in a true pterygium. Tear break-up times in both eyes were of the order of three seconds with all the puncta open and in apposition to the globe. Klein keratoscopy showed distortion of the peripheral cornea of both eyes and a slight superior ectasia of the left cornea.

The fitting of rigid gas permeable (RGP) contact lenses to correct the irregular astigmatism and maximise VA was discussed. However, the patient declined this option as he felt his vision was satisfactory with an updated spectacle correction. New bifocals were prescribed and clip-on sunglasses were recommended to provide relief from glare. Tear supplements were recommended to reduce epiphora by preventing dryness, but these were of only limited benefit.

The patient was advised to present for review in 12 months, or sooner if vision became unacceptable or other symptoms such as pain or redness developed.

CASE REPORT TWO

A 56-year-old, myopic male presented for a spectacle update with symptoms of reduced distance vision. There was a history of epiphora and recurrent chronic low-grade conjunctivitis. Repeated treatments with various antibiotics prescribed by his general medical practitioner over several years

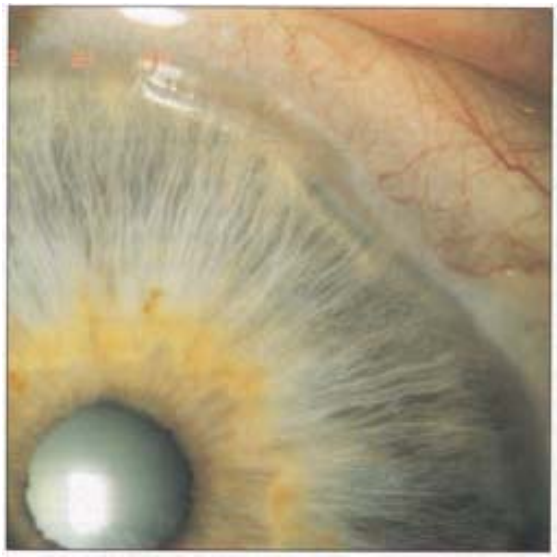


Figure 1. Case one, left eye showing superior temporal degenerative changes typical of TMD. Note the specular reflection from within the corneal gutter.

Figure 2. Case two, left eye showing TMD changes including obvious opacification and vascularisation in the inferior nasal cornea.

were unsuccessful. The general medical history was unremarkable but the immediate family history included conjunctival malignant melanoma.

The patient had been examined two years earlier and bifocals had been prescribed. Vision with the bifocals was R 6/7.5 and L 6/9. This improved to R 6/6 and L 6/6 with pinhole and spectacle correction. The axes of astigmatism rotated towards the vertical in both eyes with an increase in the strength of against-the-rule cylinder.

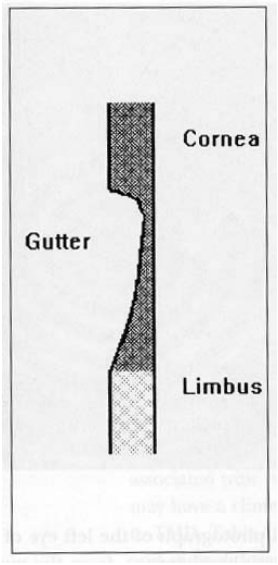
Keratometer mires and retinoscopy reflexes were both regular. Pupils and binocular vision findings were normal and intra-ocular pressures were within normal limits. The anterior chambers and lenses were normal. Both fundi showed old myopic degenerative changes.

Slit-lamp examination two years prior was unremarkable except for a corneal arcus which was unusually prominent for a 54-year-old without hyperlipoproteinaemia. At this visit, slit lamp examination revealed peripheral corneal thinning with gutter formation, peripheral corneal opacification and vascularisation (Figure 2). The gutter sloped gently from the limbal side, but rose sharply on the clear corneal side (Figure 3). These changes were along the inferior-nasal corneal margin of both eyes and the temporal corneal margin of the left eye, affecting one-quarter to one-third of the corneal circumference in total. This peripheral degeneration was bilateral, although much more extensive in the left eye and did not stain with fluorescein. A pseudopterygium was present at the eight o'clock position in the left eye (Figure 4). Tear break-up times in both eyes were of the order of three seconds with all the puneta open and in apposition to the globe.

Computerised comeal topographic analysis was performed using the Eyesis corneal analysis system (CAS). Both eyes demonstrated regular with-the-rule astigmatism on a flat cornea (Figure 5). Interestingly, the periphery of both corneae was two to three dioptres flatter than the central cornea. Unfortunately, the topographical changes occurring at the limbus are beyond the analytical powers of the Eyesis CAS. Careful observation of the polaroid Eyesis photograph of the left cornea (Figure 6), shows gross distortion at the limbus including flattening into the peripheral gutter and steepening over the pseudopterygium.

New multifocals were prescribed to alleviate the patient's visual problems. Tear supplements were recommended to reduce epiphora by preventing dryness, but these were of only limited benefit. The patient was advised of the presence of Terrien's marginal degeneration and counselled in the prognosis, including a future role for RGP contact lenses to correct irregular astigmatism and the possible need for surgical repair.

The patient was advised to present for review in 12 months, or sooner if vision became unacceptable or other symptoms such as pain or redness developed.



3a **3b**

Figure 3a. The peripheral corneal gutter from case two in section. Photograph of gutter showing the gentle slope from the limbus and the sharp rise to the central cornea.

Figure 3b. The peripheral corneal gutter from case two in section. Diagmmmatic representation of the photograph in 3a.

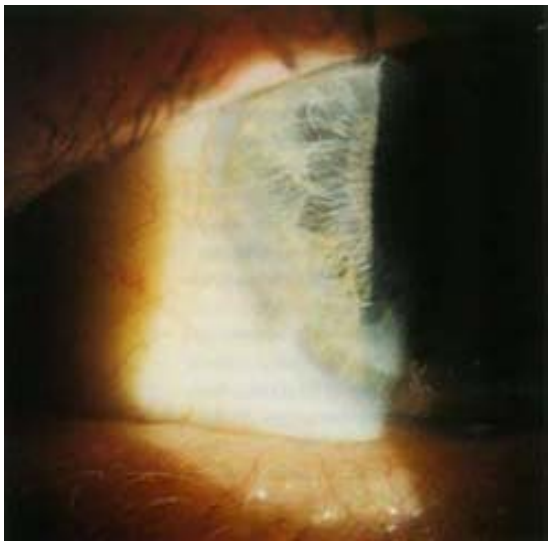


Figure 4. Pseudopterygium from the LE of case two. Note the broad, flat head and the oblique angle of insertion.

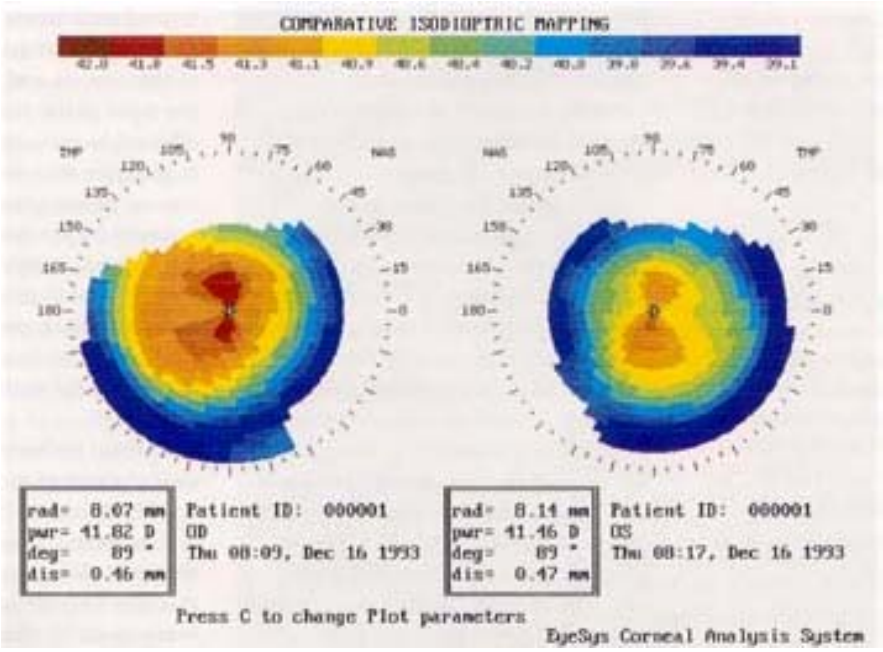


Figure 5. Computerised corneal topographical analysis (Eyesis) of both eyes of case two. Note the regularity of the astigmatism in the central corneae. Note also the excessive flattening in the far periphery.

DISCUSSION

These cases demonstrate the classical presentation of Terrien's marginal degeneration. Early reviews of this disease found that 75 per cent of reported cases occurred in males, the age of the patient varying from 10 to 70 years, with two-thirds being over age 40.[2] More recent reports have emphasised the presence of TMD in female patients and patients under the age of 40, including children as young as four years of age.[4,6,13,25-28]

The clinical picture of TMD can be considered as having five stages.[13,25,29-31]

Stage 1. Peripheral corneal opacification

Degeneration commences with a fine, white, punctate, peripheral opacity in a circumferential band; similar to arcus senilis. This opacity can appear around the entire cornea but is usually greatest superiorly and possibly also inferiorly. The opacification is generally thought to be due to lipid deposition.[32] As in arcus senilis, there is a zone of transparent cornea between the limbus and the opacity but in TMD vascularisation will cross this clear zone into the band opacity[2] and Figure 2. There are usually no symptoms during this stage but in one-third of cases a mild, unobtrusive irritation suggestive of mild conjunctivitis or dry eye may be present.[2]

Stage 2. Peripheral corneal thinning

A portion of the cornea affected by lipid opacity and vascularisation undergoes stromal thinning such that a gutter forms (Figure 1). The outer slope of the gutter descends gradually and the inner slope rises sharply (Figure 3). The epithelium always remains intact over the floor of the gutter and hence will not stain with fluorescein. The disease process is extremely slow but the gutter gradually spreads circumferentially as the disease enters stage three.

Stage 3. Corneal ectasia

Stage three is reached when the corneal thinning spreads centrally, or becomes so circumferentially extensive that corneal structural integrity is compromised, resulting in corneal ectasia. Localised ectasias will form wherever thinning is greatest. If stage one and two changes predominantly occur in the superior cornea, it follows that corneal ectasia will usually begin superiorly. Classically, an ectasia in the inferior cornea is thought to be rare.[2] However, if the opacification, vascularisation and thinning occur in the inferior cornea then inferior corneal ectasia will follow.

Once gutter formation and ectasia occur, the patient will experience a slow, progressive decrease in vision due to the development of astigmatism. The corneal topographical changes leading to astigmatism begin with flattening over the areas of peripheral thinning,[21] as in any corneal tissue loss.[33] If thinning predominates in one corneal meridian then a relative steepening of the corneal surface is seen 90° from the centre of the thinned area.[21] This is precisely the same effect that is seen with a loose limbal suture after cataract surgery.[33] Therefore, if the thinning is centred on or near to the 90° axis, as is often the case, the astigmatism will be against-the-rule. As thinning involves a more extensive corneal area, this area can bulge forward creating a steep local ectasia. Often more ectasia occurs above the visual axis than below or ectatic areas are not symmetrical around the visual axis. In these cases there is irregular astigmatism which cannot be treated with spectacles.

Corneal sensitivity is lost in the ectatic cornea and may be impaired on the floor of the gutter but will not be affected in the central cornea.[2]

In stages one, two or three of the disease pseudopterygia may form. They occur in 20 per cent of cases and grow at an oblique angle into the cornea at locations other than three and nine o'clock.[4] Pseudopterygia have similar proportions to true pterygia but are broad and flat at the head rather than pointed.[32]

Corneal perforation occurs in 15 per cent of cases, either spontaneously or as a result of trauma.[4] Perforation may be asymptomatic or only mildly symptomatic and may seal with an iris prolapse and conjunctival overgrowth.[34,35] However, surgical repair may be required for hypotony if self-sealing does not occur.[15] Also, corneal hydrops may complicate perforation.[15,19,20] Perforation may lead

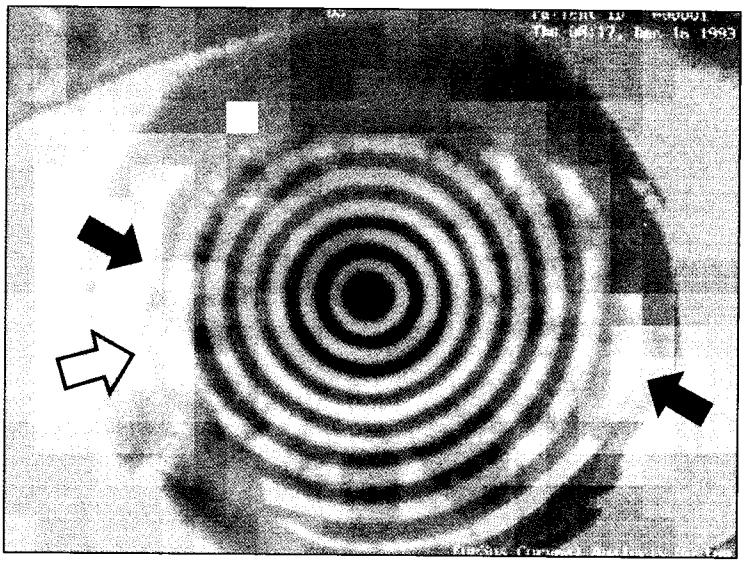


Figure 6. Eyesis polaroid photograph of the left eye of case two. Arrows denote gross distortion of the peripheral cornea, from the guttering (closed) and over the pseudopterygium (open).

to improved vision if scarring from the healing process causes corneal shape changes opposing those created by the disease.[36]

Stage 4. Total corneal ectasia

Stage four occurs when the thinning is so extensive that the gutter has encircled the cornea and total ectasia results. This is rare and results in corneal protrusion similar to that seen in keratoconus, but the central cornea will not be as steep as the corneal thinning is peripheral not central as in keratoconus. Gross astigmatism is typical in this stage although the central cornea may be remarkably symmetrical.[29]

Stage 5. Central corneal opacification

Small opacities appear in the ectatic central cornea remote from the marginal degeneration.[31] This is probably analogous to the cone scarring seen in keratoconus and may represent a breakdown of corneal transparency due to gross stretching. This stage is rarely seen, since perforation, radical treatment or mortality has usually intervened.[31]

Terrien's marginal degeneration is classically thought to occur without inflammation, but some reports have emphasised an inflammatory variant.[4] Inflammation can occur at any stage of the disease, usually effects younger patients and is recurrent. The signs and symptoms include conjunctival hyperaemia, lacrimation, photophobia and disabling pain.[4,32] The cornea appears unaffected in such cases, the inflammation being centred in adjacent deep episclera or superficial scleral tissue.[4] Austin and Brown speculated either that TMD could be considered as a disease with a spectrum of presentations, at one end of which is an inflammatory variant, or that all TMD cases involve inflammation but the memory of previous inflammation fades as the disease runs its protracted course.[4]

The aetiology of TMD remains unknown with only speculative theories proposed to date. Many conditions have been reported to be associated with TMD, but all associations are inconsistent and seemingly incidental.[18,23] Pathological investigations have reported stromal material in phagocytic cells and the suggestion of a hypersensitivity reaction, but these have been inconsistent and fouled by questionable diagnosis of the disease.[2,4,37] The significance of such errors has been highlighted in a recent report that finds strong evidence for an autoimmune reaction in Mooren's ulcer, but not for TMD.[24]

Differential diagnosis

Marginal corneal disease has been classically differentiated into a series of distinct clinical entities that may be associated with systemic disease and which may have a clinically similar appearance to TMD. Table 1 lists all such conditions and their differentiating features.

Unfortunately, the diagnosis of corneal disease is seldom simple. Significant overlap in presenting signs and symptoms often causes difficulties in disease differentiation.[7] In particular, the presence of inflammation in eyes with circumferential TMD changes has led to the diagnosis of inflammatory TMD, but if the TMD type changes are unilateral and confined to a smaller portion of the circumference, then the diagnosis may be considered to be Mooren's ulcer.[4,7,38] Such difficulties have led some to consider corneal disease as being not classifiable into distinct clinical entities but a continuous spectrum of disease.[7,38] Similar spectra exist for corneal stromal dystrophies, atopic eye disease and anterior segment mesodermal dysgenesis.[39-42] This approach recognises the limited importance of differential diagnosis, if patient management is based on the clinical signs and symptoms rather than disease nomenclature. This is a far more sensible approach that would allow diagnoses such as Terrien's type marginal degeneration (with

Condition	Differentiating features
Pellucid marginal degeneration	Inferior corneal thinning only; no pseudopterygia; inferior corneal ectasia only.
Marginal furrow degeneration	Asymptomatic, involves 360° of the cornea, no opacification, no vascularisation, no pseudopterygia, no ectasia, no change in astigmatism.
Dellen	Localised corneal depression only, not extending circumferentially, adjacent to raised lesion, unlikely to be bilateral.
Mooren's ulcer	Inflammatory, epithelium is disrupted, with consequent fluorescein staining, tends to be localised, rapidly progressive.
Rheumatoid disease	Associated systemic disease, usually inflammatory, can have non-inflammatory peripheral gutter formation with vascularisation, opacity, lipid deposition, but progresses more rapidly and leads to keratolysis rather than keratectasia.
Keratoconus	No gutter, gross central/inferior ectasia, dramatic reduction of vision, often in young people, no peripheral opacification, no pseudopterygia.
Fungal ulcer	Inflammatory, localised, rapidly progressive, history of trauma.
Acne rosecea	Associated systemic disease, inflammatory more extensive pannus formation, loss of epithelial integrity.
Arcus senilis	Possible systemic hyperlipidaemia, no gutter formation, no vascularisation, no ectasia.

Table 1. Differential diagnosis of Terrien's marginal degeneration

inflammation et cetera) or corneal arcus with early Terrien's type changes. This approach also facilitates assessment of changes close to the normal end of the spectrum, since TMD, like keratoconus may be common in early or abortive but clinically insignificant forms (for example, corneal arcus with early Terrien's type changes).

Computerised corneal topographical analysis

Horner and colleagues[25] have shown that superior corneal ectasia in TMD is easily demonstrable with computerised corneal topographical analysis (CCTA) and they suggest that a corneal modelling picture with greater steepening in the superior quadrant compared with inferiorly may prove to be diagnostic of TMD. However, a superior corneal ectasia will occur only in TMD if the thinning is predominantly superior. This may often be the case but there is no doubt that thinning can be greatest inferiorly and then ectasia will form inferiorly and the CCTA picture may be indistinguishable from keratoconus.

Wilson and co-workers[21] examined the corneal topography of four patients with TMD and as in our case two (Figure 5), found flattening of the peripheral cornea over areas of peripheral thinning. They also enhanced the understanding of astigmatism development by showing a relative steepening 90 degrees away from the midpoint of the thinned area.[21]

CCTA in early TMD needs to be sensitive to peripheral corneal flattening and distortion. Unfortunately, all of the commercially available devices for CCTA are poor at measuring to the periphery of the cornea and are unable to cope with distorted mires as seen in case two (Figure 6). CCTA may be able to detect ectasia later in the disease, but by the time this occurs a slit lamp diagnosis could have been made 10 years earlier. Again, so-called corneal topographical 'signatures' could be easily mimicked by trauma to a normal eye. It is extremely difficult to support the assertion that CCTA has a role in the diagnosis of TMD.

Management

Despite a long course, the extremely slow progression of TMD ensures that serious complications such as gross visual disability or perforation are rare. In the early stages of the disease, treatment is required only to relieve symptoms. Mild irritation may be relieved with the use of ocular lubricants and refractive changes compensated for with spectacle or contact lenses. In cases with inflammation, corticosteroid therapy may be required, but this is often ineffective.[4]

Computerised corneal topographical analysis is valuable for assessment of corneal optical changes while monitoring the disease progress. Although visual acuity, contrast sensitivity and refraction are also adequate for this end. In addition, CCTA is useful for patient education in the mechanism of vision loss and good for prognosis determination as ectasia encroaches on the optically important part of the cornea.

As the condition progresses and the magnitude of the astigmatism increases, or as irregular astigmatism develops, visual correction with rigid gas permeable (RGP) lenses may be required. Such lenses may need to be of small diameter to prevent irritation to the degenerated peripheral cornea and of high oxygen transmissibility to minimise compromise to corneal physiology.[25]

Once corneal thinning has progressed such that spontaneous perforation becomes possible, protective eyewear should be prescribed to shield the eye from trauma. When perforation becomes imminent, surgical intervention may be advised. Many different procedures have been tried including conjunctival flap or scleral autograft.[30] However, a full thickness or lamellar corneo-scleral graft is the treatment of choice.[2,10,22,32] Hand-fashioning of the graft usually will be required to fit the irregularly shaped defect.[43] In advanced ectatic disease, astigmatism reduction may be achieved by lamellar or penetrating keratectomy of the ectatic cornea and suturing the normal thickness stroma together.[11,13] Damage to Desçemet's membrane may require the use of biological tissue cement, biosutures, monofilament sutures or permanent Prolene sutures to ensure wound closure.[11,13]

The patient should be educated about the nature of the eye condition and the likely prognosis. The possibility of inflammatory attacks should be discussed and the patient advised to return in the event of pain or a reduction in vision. The patient should be carefully and regularly reviewed; annually is appropriate early in the disease. However, if the patient wears contact lenses or if corneal thinning becomes precarious, more frequent reviews would be required.

CONCLUSION

Two examples of the asymmetric, bilateral corneal changes seen in Terrien's marginal degeneration are described. Both are in the earlier stages of the disease and exhibit peripheral corneal lipid deposition, vascularisation, gutter formation, corneal thinning and in one case corneal ectasia. The distinctive features of pseudopterygia and increasing against-the-rule astigmatism are also present. Although TMD may present across a varied spectrum, both of these cases fit into the classical picture of this distinct clinical entity. CCTA was performed for one case, but is of little value for the diagnosis of TMD, although it is useful for monitoring the progress of TMD and determining visual prognosis. Only treatment for refractive change and discomfort were required in these cases, although treatment of inflammatory episodes or fitting of RGP lenses to correct for irregular astigmatism is sometimes necessary. Surgical intervention is required if the condition progresses to imminent or actual corneal perforation, but this is unlikely in either of these cases, considering the patients' ages and the slow progression of the disease.

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