

Case report ● ● ● ●

Terrien's marginal degeneration: an unusual presentation in an Indian female

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Abstract

An interesting case of bilateral corneal ectasia resulting from its marginal degeneration in a young female is reported. Terrien's marginal degeneration is a rare disorder of unknown etiology. It is usually bilateral, although often asymmetric, and is seen mainly in young men. It generally starts superiorly as marginal opacification and stromal thinning. This case was unique in that, unlike routine presentation seen in this type of disorder, the gutter was located more anteriorly in the cornea and was not associated with any vascularization.

Keyword: ectasia, keratoconus, Terrien's marginal degeneration

Introduction

Terrien's marginal degeneration is an uncommon disease from the group of peripheral corneal degeneration (Golubovic, 1994). The condition is often bilateral and may occur at any age, although it typically occurs in middle-aged males. The disease first presents as a peripheral corneal haze. Over time, it exhibits a slowly-progressive peripheral corneal thinning with a sloping central edge that spares the limbus. We present a case of atypical Terrien's marginal degeneration with a more anterior, or mid peripheral, location of the gutter and an absence of any vascularization.

Case report

A 28-year-old female presented with complaints of diminution of vision in the right eye for the last one year. She was a house-wife and had never had any systemic or ocular illness or trauma to the eye in past.

Recorded visual acuity of her right eye was 3/60 and of the left eye 6/18. Inspection of the right eye showed bulging of superior cornea along with thinning and scarring of superior mid-peripheral cornea. The horizontal diameter of the cornea was 12 mm in both the eyes (normal 11.7 mm (Duke-Elder, 1964)). The vertical diameter was 11.5 mm (normal 10.6 mm) in both the eyes. Retinoscopy showed irregular shadows and it was difficult to conduct refraction. Slit-lamp examination of the cornea of the right eye showed intact epithelium with a 2-3 mm shallow groove with scarring at mid-periphery of the superior cornea about 4-5 mm from limbus (Fig.1). The thickness of the cornea on both sides of the groove was normal. There was no corneal vascularization. The anterior chamber appeared deep due to forward bulging of the inferior cornea, although this was not estimated quantitatively.



Fig. 1: (Slit lamp examination of right eye showing a 2-3 mm shallow groove with scarring at mid periphery of the superior cornea)

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Inspection of the left eye did not reveal any gross abnormality but the slit-lamp examination showed scarring and flattening of the superior cornea about 4-5 mm from limbus. This was also not associated with any vascularization. (Fig.2)



Fig. 2: (Slit lamp examination of the left eye showing scarring and flattening of the superior cornea about 4-5mm from limbus)

The iris and lens were normal in both the eyes. Gonioscopy revealed open angles. Fundus examination was normal. Corneal sensitivity as tested with a wisp of cotton wool was found to be normal.

Discussion

Duke- Elder (1964) has described Terrien's marginal degeneration of the cornea as a rare bilateral condition with opacification and vascularization of the periphery of the cornea, progressing by degeneration of the stroma to the formation of a gutter and finally resulting in ectasia. The evolution of the condition is extremely slow and the development of ectasia may take 10 to 20 years. The condition has been seen to exist three times more frequently in males than in females (Beachamp, 1982). The condition is bilateral in 86 % cases but may be asymmetric (Etzine & Friedmann, 1963). This case was of interest as it was seen in a female and had a more anterior (mid peripheral) location of the gutter with an absence of any corneal vascularization. Nirankari et al (1983) described a case of Terrien's degeneration with central corneal thinning and opacification in a 15-year-old female, features not having been reported in the literature of the last 30 years. We hereby report another case of similar nature in a 28-year-old female. No similar references except that of Nirankari et al (1983) were found in the literature.

This condition must be differentiated from keratoconus. In Keratoconus, there is thinning and forward bulging of the cornea but with no alteration of its transparency in the initial stages whereas in Terrien's degeneration, the disease begins with a yellowish-white deposition in the anterior stroma along with corneal flattening at the juncture of the furrow, as was seen in the present case.

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