Terriens Marginal Degeneration – A Varied Presentation

**Introduction**

Terriens marginal degeneration is an uncommon disease of the peripheral cornea, occurring at any age, with 75% being males. This condition may be bilateral or unilateral. Lesions begin usually superonasally rarely inferiorly with development of fine, white sub epithelial, peripheral opacities that spare the limbus. The opacities coalesce and this is followed by corneal thinning, typically with a sloping central edge and a fairly steep peripheral edge to the resultant furrow.

The epithelium is typically intact with yellowish white lipid deposits in the centre of the gutter with associated vascularisation. Etiology is unknown, although inflammatory, degenerative, and immune mediation have been proposed.

**Case Report**

A 22 year male patient presented to our hospital with an accidental detection of visual defect in the left eye while attending an eye camp. He had never used glasses earlier. His visual acuity was checked and was found to be 6/6 right eye and 6/12 left eye. He underwent a complete cycloplegic refraction and his final correction was found to be 6/6 with -1.00 Dioptra cylinder x 105° axis. The patient underwent a thorough slit lamp examination which revealed a unilateral, inferior circumlinear corneal thinning with intact epithelium, without neovascularisation, minimal lipid deposition and a clear area existing between the lesion and limbus. (Fig. 1a and b)

There was no epithelial defect noted by fluorescein staining. The patient underwent a corneal topography (Fig. 2) which clearly showed the area of inferior thinning with steepening 90° opposite to the central point of thinning. This is typical of Terriens. This has given rise to the high oblique astigmatism in this patient.

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Malabar Eye Hospital and Research Centre, Calicut

Fig. 1. (a) Classic presentation of Terriens marginal degeneration (b) Atypical presentation showing unilateral inferior circumlinear corneal thinning.
A diagnosis of unilateral Terriens marginal degeneration was made based on the presence of inferior lesion, absence of vascularisation and unilaterality. Our patient had subtle variation compared to a regular Terriens. The patient's fundus was normal.

**Discussion**

In Terriens degeneration when the thinning is restricted to the superior or inferior area of the peripheral cornea, there is relative steepening approximately 90° away from the midpoint of the thinned area resulting in astigmatism characteristic of this disorder. Histologically epithelium may be normal, thickened, and the thinned Bowmans layer and the lamellae may be split or fibrillated (1, 15) (Fig. 3).

Stroma is thinned. Whether or not there is vascularisation, the inflammation depends on the form of the disease. Lipid is found consistently. Though a lot of etiologies have been proposed, levels of circulating immune complexes are not elevated in patients with Terriens. In our patient the right eye was absolutely normal. Marginal corneal degeneration, dellen, collagen vascular diseases, sclerokeratitis, staphylococal marginal keratitis etc have been proposed as differential diagnosis. This patient was prescribed glasses and advised rigid contact lenses for his left eye, and advised regular followups to rule out early development of

![Fig. 2. Corneal Topographic map](image)
Lesion in the right eye. Usually pseudo pterygia occur in Terriens degeneration in 20% of cases. Very rarely extreme thinning occur when reconstructive surgery is indicated. A full thickness or lamellar corneo scleral graft (often hand fashioned to fit the defect) may be necessary. The progressive increase in against the rule astigmatism or oblique astigmatism with advanced disease can be arrested up to 20 yrs by grafting. Severe astigmatism can also be treated by a crescentric shaped excision of the gutter with suturing of the healthier margins. Corneal thinning may progress sometimes, despite intact epithelium, to the point at which a deep corneal break leads to hydrops or even frank perforation in about 15% of cases. In these cases keratoplasty either a full thickness or deep lamellar sector is necessary. So though an uncommon clinical entity we should keep Terriens degeneration in mind with its possible variations, and treat the patient accordingly with regular followups to see the progression of the disease.

References

15. Suyeges, M.D., Levai, G and Alberth,B. Pathology of Terriens degeneration.

Fig. 3. Histologic section shows limbus on the left (iris not present) and central cornea to the right. Not marked stromal thinning. (From Yanoff M,Fine BS. Ocular pathology, Ed 5.st. LOUIS: Mosby, 2002.)