TERRIEN'S MARGINAL DEGENERATION OF THE CORNEA: A case report
Terrien'in marjinal kornea dejenerasyonu: Vaka takdimi

Sarper Karaküçük[1] , G Ertuğrul Mirza [2], Nurullah Çağıl[1] , A Kerim Durukan[1]

Summary: *A rare case of Terrien's marginal degeneration is reported. The case had the characteristic features of the disease including progressive astigmatism caused by ectasia and thinning of the peripheral cornea. The literature relevant to this disorder is reviewed.*

Key Words: *Corneal diseases, Astigmatism, Corneal opacity, Corneal stroma*

Özet: *Terrien' in marjinal dejenerasyonu tanısı konulan ve ender rastlanan bir vaka takdim edilmiştir. Vakada hastalığın karakteristik özellikleri arasında bulunan periferik korneanın incelmesi, ektazisi ve buna bağlı ilerleyici astigmatizma bulunmaktadır. Bu hastalıkla ilgili literatür gözden geçirilmiştir.*

Anahtar Kelimeler: *Korneal hastalıklar, Astigmatizma, Korneal opasite, Korneal stroma*

Terrien's marginal degeneration is an uncommon disease of the peripheral cornea, characterized by gradual thinning of the stroma which in turn leads to corneal ectasia and gutter formation. It is usually bilateral, although it may be asymmetric. One eye may be involved many years before the other. About 75% of patients are males. The disease can be seen at any age between 10 and 75 years with the highest incidence in the 20 to 30 age group. We report a patient with Terrien's marginal degeneration with the classical features of the disease.

CASE REPORT

A 20 year-old-female was referred to the Ophthalmology Clinic of Erciyes University Faculty of Medicine complaining of burning sensation on both eyes in September, 1992. The refraction showed -0.75 -1.00x150° RE and -1.00-0.50x70° LE; her corrected vision was 9/10 on both eyes. The fundus examination was bilaterally normal .

Biomicroscopy of the right eye showed arcuate thinning at the inferior cornea near the limbus. On the left, areas of marginal degeneration with gradual and arcuate thinning were seen at the superior limbus, along with stromal deposits having some vascularization approaching from the peripheral conjunctiva (Figure).No inflammatory signs were accompanying the picture.

History revealed that the condition was present since 1981, the astigmatism showed progression and topical steroids were occasionally prescribed. No systemic abnormalities were found on examination. A diagnosis of Terrien's marginal degeneration was made; she was informed about the condition and instructed to come should any symptoms persist.

DISCUSSION

Terrien's marginal degeneration has been called with many other names including peripheral furrow keratitis, ectasic marginal dystrophy, peripheral corneal ectasia, and senile marginal atrophy. Fine, yellow-white punctate peripheral opacities which resemble arcus senilis are among the early clinical features. Pseudopterygia may occasionally develop, occurring at unusual or oblique meridians(8, 5).

The corneal thinning process in Terrien's marginal degeneration begins by an indentation in the clear cornea between marginal opacification and the upper limbus. Gradually a gutter-like furrow forms here. As the guttering progresses, its central wall makes a sharp inclination while the peripheral side

Erciyes University Medical Faculty 38039 Kayseri, TÜRKİYE
Department of Ophthalmology. Resident[1], Associate Prof.[2].

shelves gradually. The sharp central edge is demarcated by a grayish-white line. Irregular deposits of lipid can be seen within the deep layers of the furrow. The floor of the gutter gradually becomes thin and vascularized. Since the epithelial layer remains intact , it does not stain with fluorescein. Interpalpebral periphery is usually spared. As a result of continued thinning, a furrow in the lower periphery is seen along with an ectasia in the upper cornea (8, 10). Clinically progressive thinning and ectasia causes high astigmatism since the cornea flattens in the vertical meridian; gradual deterioration of vision occurs. The ectatic areas may eventually perforate, either spontaneously or with mild trauma. Patients are usually asymptomatic unless there is severe irregular astigmatism. Our patient also had high astigmatism gradually increasing since 1981. Occasionally, intermittent, mild ocular irritation may be seen , however, Austin and Brown (2), and İrkeç (7) have described recurrent episodes of painful ocular inflammation in Terrien's marginal degeneration, which they termed as inflammatory Terrien's Disease. Binder and associates described a similar case with moderately severe ocular inflammation (3).

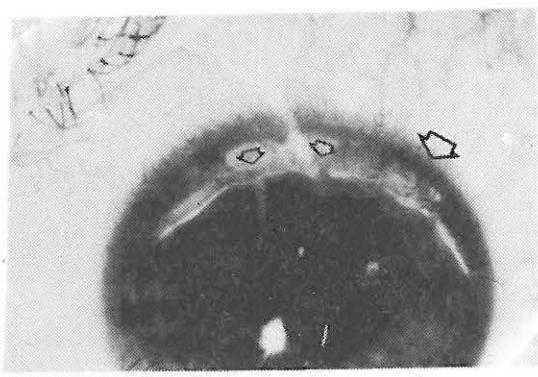

Figure. Arcuate thinning at the superior limbus (small arrows) and stromal vascularization approaching from the peripheral conjunctiva (large arrow)

Terrien's marginal degeneration affects primarily the corneal stroma.Pathological examination may show that the majority of stroma undergoes fibrillary degeneration along with lipid infiltration of collagen fibrils. Stromal thinning is always present, In advanced cases, the stroma may be extremely thin. In the epithelium overlying ectatic areas, multiple basal mitoses and hypercellularity is noted. Local defects in Bowman's membrane, and in some cases healed ruptures of Descemet's membrane can be seen.

The etiology of Terrien's marginal degeneration remains unknown. Various histopathologic and ultrastructural studies of affected corneas have demonstrated evidence of degeneration, particularly of the basal epithelium- basement membrane -anterior stroma complex (6, 11).

Differential diagnosis must be made between Mooren's ulcer, which is usually painful, accompanied with inflammatory signs and can cause severe melting (3). However, epithelium in Mooren's ulcer is non-intact, the lesion spreads centrally and circumferentially; rapid progression is sometimes seen and no lipid is associated with the lesions (1).

Disease progression in Terrien's marginal degeneration is classically slow. Usually the patient does not present until progressive irregular astigmatism deteriorates visual acuity. At this stage, spectacle correction of astigmatic error is sufficient. Rigid contact lenses or piggy-back (combined rigid-soft) contact lenses can be prescribed at advanced stages. In advanced cases, corneal irregularity and astigmatic error increases too much as to make contact lens fitting impossible. Surgical repair is then indicated both to decrease high astigmatism and to prevent perforation of thinned ectatic corneal regions . Excision of the ectatic tissue followed by suturing of the walls of the furrow, annular full thickness or crescentic lamellar keratoplasty or eccentric penetrating graft fitting to the shape of the defect is sufficient to solve the problem (4, 9).

Terrien's marginal degeneration must be

contemplated in any patient with progressive astigmatism. Once the diagnosis is made, the patients with Terrien's marginal degeneration must be informed about their disease and carefully followed-up. This can help better supporting them and solve disease oriented problems.

REFERENCES

1. *Arffa RC: Degenerations. In Arffa RC (ed): **Grayson's Diseases of the Cornea.** Mosby Year Book Inc. , St. Louis 1991 pp 333-363.*
2. *Austin P, Brown SI: Inflammatory Terrien's marginal corneal disease. **Am J Ophthalmol** 92:189-192, 1981.*
3. *Binder PS, Zavala EY, Stainer GA: Noninfectious peripheral corneal ulceration: Mooren's ulcer or Terrien's marginal degeneration? **Ann Ophthalmol** 14:425-435, 1982.*
4. *Caldwell DR, Insler MS, Boutros G, Hawk T: Primary surgical repair of severe peripheral marginal ectasia in Terrien's marginal degeneration. **Am J Ophthalmol** 97:332-336, 1984.*
5. *Goldman KN, Kaufman HE: Atypical pterygium A clinical feature of Terrien's marginal degeneration. **Arch Ophthalmol** 96:1027-1029, 1978.*
6. *Ivamato T: Electron microscopy in cases of marginal degeneration of the cornea. **Invest Ophthalmol Vis Sci** 11:241-258, 1972.*
7. *İrkeç M, Atabay Ç: Terrien' in marjinal kornea dejenerasyonu. **Türk Oftalmoloji Gazetesi** 13: 129-132, 1983.*
8. *Kanski JJ: Terrien's marginal degeneration. In Kanski JJ (ed): **Clinical Ophthalmology. A Systemic Approach**. Butterworths, London 1989. pp. 108-109.*
9. *Mannis MJ: Terrien's marginal degeneration. In Fraunfelder FT, Roy T (eds): **Current Ocular Therapy**. W.B. Saunders, Philadelphia 1990. p 426.*
10. *Robin JB, Schanzlin DJ, Verity SM, et al: Peripheral corneal disorders. **Surv Ophthalmol** 31: 1-36, 1986.*
11. *Suveges I, Levai G, Alberth B: Pathology of Terrien's disease: histochemical and electron microscopic study. **Am J Ophthalmol** 74: 1191-1200, 1972.*