Commentary: Clinical profile and demographic distribution of Terrien’s marginal degeneration in a multitier ophthalmology network in India

We read with great interest the article “Clinical Profile and Demographic Distribution of Terrien’s Marginal Degeneration in a Multitier Ophthalmology Network in India.”[1] We commend the authors for coming out with a relatively larger series on this not-so-common corneal degeneration. The major findings of the authors are similar to what is known in the literature. Here, we would like to share our experience on Terrien’s marginal corneal degeneration (TMCD).

Clinical diagnosis of TMCD is not difficult for an aware corneal surgeon. Most cases that we have seen till date at our referral center presented between 40 and 60 years. One case even presented at 80 years of age with cataract. The current article has reported associations with various diseases, such as vernal keratoconjunctivitis, but our cases were almost always idiopathic. The common presenting feature is poor vision, partly because of the irregular astigmatism and in some cases due to the associated cataract. Although corneal perforation following trivial trauma has been reported by several authors in the past, we have not come across such presentation till date. Lately, we have come across two cases referred to us with the diagnosis of corneal hydrops. On close examination of these two cases, we found a very interesting clinical sign that is “cavity formation in TMCD” in addition to the classic features of thinning and vascularization.[2]

Hattori et al.[3] have reported the interesting finding of cavity formation in TMD. They hypothesized that progressive stromal abnormality and thinning may lead to cavity formation with intact epithelial and endothelial layers. In later stages, the endothelial layer might get pushed or apposed to thinned out stroma or epithelium due to the intraocular pressure that can hide the cyst or cavity, giving the appearance of a localized thinning with posterior bowing. Out of our two cases, as mentioned above, the first case was a 45-year-old female with typical findings of an intrastromal cavity enclosed by Bowman layer and Descemet membrane with surrounding scarring in the right eye at 11–12 O clock position documented on slit lamp as well as on anterior segment optical coherence tomography (ASOCT). The second case was an 80-year-old female with an intrastromal cavity with bowing of posterior corneal surface at 9–11 O clock position in both eyes. ASOCT findings of both cases showed contiguous high reflectivity of the epithelial layer indicating a normal epithelium without defect, and the inner layer of the cavity was continuous with the endothelial layer of the intact central cornea. These findings suggest that the cavity forms within the stroma of the peripheral cornea with intact epithelium and endothelial layers. This peripheral thinning leading to cavity formation is probably a structural change during the pathogenesis of TMCD. The importance of highlighting this finding is that both our cases were diagnosed with corneal hydrops and could have been subjected to unnecessary intervention such as intracameral gas injection.

Managing TMCD is difficult since the thinning process involves the periphery.[4] Conservative management with spectacles or contact lenses is the most commonly used therapy. Indications of surgery (lamellar keratoplasty) include poor vision due to irregular astigmatism, severe peripheral thinning or hydrops, and corneal perforation. Several authors have described various techniques; we prefer tuck in lamellar keratoplasty (TILK) or crescentic patch graft in such cases.[5]
TILK is preferred when the central cornea is also thinned as in cases of advanced TMCD. If the thinning is involving only the peripheral part, a crescentic patch graft is a better option. The crescents can be customized depending on the extent of ectasis. However, it must be remembered that both the techniques are challenging and often require significant expertise.

A special challenge that may arise while dealing with such cases is handling the associated cataract. Selecting a site uninvolved in corneal ectasia for incision, preferring a scleral tunnel, appropriate use of viscoelastic, doing phacoemulsification with lowered settings, suturing the wound, and a careful intraocular lens power selection could result in excellent outcomes in such cases.

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**References**

1. Das AV, Pillutla NL, Chaurasia S. Clinical profile and demographic distribution of Terrien’s marginal degeneration in a multitier ophthalmology network in India. Indian J Ophthalmol 2021;69:3482-6.

2. Singhal D, Roop P, Maharana PK. Intrastromal cyst in Terrien’s marginal degeneration. Indian J Ophthalmol 2019;67:1475.

3. Hattori T, Kumakura S, Mori H, Goto H. Depiction of cavity formation in Terrien marginal degeneration by anterior segment optical coherence tomography. Cornea 2013;32:615-8.

4. Maharana PK, Dubey A, Jhanji V, Sharma N, Das S, Vajpayee RB. Management of advanced corneal ectasias. Br J Ophthalmol 2016;100:34-40.

5. Lohchab M, Prakash G, Arora T, Maharana P, Jhanji V, Sharma N, et al. Surgical management of peripheral corneal thinning disorders. Surv Ophthalmol 2019;64:67-78.