

Terrien's Marginal Degeneration: An Uncommon Eye Disease and Treatment Modalities

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ABSTRACT

Outermost layer of the eye is cornea which is very clear and transparent. Many diseases can affect the cornea in that Terrien's marginal degeneration is also one of them, and their causes are unknown. But it is associated with other diseases like scleritis, episcleritis, and keratoconus, and this condition can be diagnosed by corneal topography, slit-lamp microscopy, ultrasound biomicroscopy, and scanning-slit topography. Medically, it can be treated by contact lenses and surgically by different types of keratoplasties.

Keywords: Cornea, Degeneration, Keratoconus, Keratoplasty, Slit lamp.

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INTRODUCTION

The cornea is the outermost and clear layer of the eyes. Disease of the cornea involves a variety of conditions that affect the vision.¹

Terrien's marginal degeneration is a corneal disease which is an uncommon but distinct variety of thinning of margin of cornea. It results in slowly progressive noninflammatory, asymmetrically bilateral, or sometimes unilateral corneal thinning at peripheral level of eye, and it is associated with neovascularization of cornea, lipid deposition, and sometimes opacification. Degeneration may result in high degree of oblique astigmatism.

ETIOLOGY

The causes of this disease are unknown/idiopathic, but it is associated with other conditions such as scleritis, keratoconus, episcleritis, anterior basement membrane dystrophy, and posterior polymorphous dystrophy.²

TYPES

- **Classic form:** This type most commonly affects patients after 40 years of age without condition, rheumatoid arthritis with slow progression.
- **Inflammatory type:** Inflammatory type affects youngsters which is characterized by recurrent inflammation and acute pain.³

PATHOPHYSIOLOGY

Corneal thinning, it can be localized or may involve extensive proportions of the cornea at peripheral area. Degeneration of corneal tissue starts from superiorly with very mild, punctuates below epithelial layer and anterior stromal opacities, and leaves a clear area in between the opacities and also the limbus. After opacification, the development of a superficial, peripheral, and fine vascular pannus will progress over the years to involve subepithelial opacity at the increasing edge.⁴ The thinning spreads all circumferentially but rarely involves the inferior limbus and also leaves the epithelium intact. Gradually, the central wall will steep, the peripheral wall slopes, and then a yellow line of lipid or fat deposits and appears at the central edge of the furrow called as

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leading edge of the pannus. Vessels transverse the furrow and will pass beyond it. In rare condition, spontaneous perforation may result. But perforation can result easily with minor trauma. If there is a rupture in Descemet's membrane, it will result in interlamellar fluid or even a corneal cyst.⁵

CLINICAL MANIFESTATIONS

With initial mild disease, patients are asymptomatic in nature, but in advanced disease, the patients exhibit common symptoms like

- Progressive blurred vision without pain
- Progressive change of glasses.

Less common symptoms are:

- Recurrent attacks of ocular irritation.
- Pseudo-ptyerygium
- Hydrops can occur in severe cases⁶

DIAGNOSTIC EVALUATIONS

- **History and assessment:** This disease is diagnosed with collection of related history and assessment of salient features like steep central edge, intact epithelium, sloping peripheral edge, leading edge of lipid, superficial vascularization.

- **Corneal topography:** This measures curvature of the cornea, elevation, and the amount of astigmatism of cornea.⁷
- **Scanning-slit topography:** This method uses optical slits that will scan along different points of the cornea. This allows corneal measurements of both the anterior and posterior cornea.
- **Ultrasound biomicroscopy:** Ultrasound biomicroscopy procedure quantifies the corneal thickness and majorly helps in detection of progression of disease.
- **Slit-lamp examination of eye**

MANAGEMENT

- **General therapy:** Terrien's marginal degeneration disease generally does not require treatment. It is necessary to treat the condition when a perforation occurs.
- **Medical therapy:** Most commonly, the medical therapy is used for contact lenses to manage the corneal astigmatism.
Rigid gas-permeable (RGP) contact lenses: The contact lens called as piggyback RGP lenses can be used. In piggyback, a soft contact lens is worn under the gas-permeable lens, acting as a cushion to reduce or eliminate any discomfort caused by the gas-permeable lens.⁸
- **Surgery:** Surgical correction is necessary when perforation is imminent because of progressive thinning of cornea.⁹
 - **Penetrating keratoplasty:** Patients with Terrien's marginal degeneration are usually poor candidates for this procedure because the thinning of cornea occurs near the limbus. Large eccentric grafts are necessary, and it should be positioned very near to the limbus so there is a chance of rejection of graft and complication related to sutures.
 - **Lamellar crescentic keratoplasty:** In this procedure, crescent-shaped, that is, partial-thickness donor lamellar cornea is used.
 - **Full-thickness keratoplasty:** In this surgery, thinned entire cornea is removed and the donor's Descemet's membrane is retained.
 - **Annular lamellar keratoplasty:** This type of graft may require in case of severe condition of Terrien's marginal degeneration, involving complete 360°.¹⁰

CONCLUSION

Terrien's marginal degeneration is uncommon corneal disease, and it does not require any medical treatment unless there is a perforation. With the help of contact lenses, we can treat this disease. In case of severe degeneration, surgery can eliminate the symptoms of this disease. Keratoplasty is a corneal surgery to treat the marginal degeneration of cornea.

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