Case Report

Case Report – Bilateral Manifestation of Terrien's Marginal Degeneration

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Abstract

Purpose: To report our findings in a case of asymmetrically bilateral Terrien's marginal degeneration (TMD) involving the left eye more than the right eye.

Observations: A 35-yr old male visited our OPD with a complaint of diminution of vision in the left eye. He had a history of pterygium surgery in the left eye 20 days back elsewhere. There was no improvement in visual acuity of the left eye with best correction. Slit lamp examination showed bilateral peripheral corneal thinning with neovascularization and central macular grade corneal opacities in the left eye. Corneal topography showed ectatic areas on anterior and posterior elevation map, areas of high-risk corneal thinning on pachymetry in the left eye, high Kmax values in both eyes despite normal corneal pachymetry in the right eye, indicating progression of disease in the right eye as well. Based on slit lamp examination, results of corneal topography, AS-OCT and lipid profile, we diagnosed him as a case of bilateral Terrien's marginal degeneration involving the left eye more than the right eye. The patient is otherwise asymptomatic and was started on lubricants in both eyes with close follow-up.

Conclusion: We found a rare presentation of an early asymmetrically bilateral TMD associated with a history of pterygium. Corneal topography along with slit lamp examination and increased lipid profile levels helped in diagnosing the case as TMD while ruling out other close differentials of the disease like Pellucid marginal degeneration and Keratoconus.

Keywords: Terrien's marginal degeneration (TMD), Ectasia, Corneal topography

Introduction

Terrien's marginal degeneration(TMD) is an uncommon but slowly progressive non-inflammatory, unilateral or asymmetrically bilateral peripheral corneal thinning and is associated with corneal neovascularization, opacification and lipid deposition.

Our report presents an asymmetrically bilateral observation in a 35-year-old male. Informed consent was obtained from the patient for publication of this case report.

Case Report

We report a case of a 35-year-old male with a history of pterygium surgery elsewhere (left eye) 20 days back who presented in our OPD for diminution of vision (left eye). On clinical examination, unaided visual acuity was 20/20 in the right eye (OD) and 20/40 in the left eye (OS) with no further improvement in visual acuity of the left eye. On slit lamp biomicroscopy both eyes showed milky white peripheral corneal opacity involving stroma circumferentially with intact epithelium and sloping edges in superior, nasal and temporal quadrants. Also, an area of wide corneal thinning nasally with superior corneal vascularization and multiple macular-grade corneal opacities in the central cornea with clear cornea in between were additionally seen in the left eye.

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Investigations

The corneal topography(OCULUS Pentacam) in the right eye showed areas of peripheral flattening in all quadrants in the axial map. BAD (Berlin/ Ambrosio Enhanced Ectasia Display) MAP showed Kmax (maximum keratometry value) 48.7D, indicating moderate ectasia risk, with the corneal thickness spatial profile (CTSP) and percentage thickness increase(PTI) showing deviation from normal.

In the left eye, on the axial map, peripheral flattening was observed. BAD map showed high-risk nasal thinnest location(TL) and vertical displacement with high risk of ectasia nasally in anterior and in full periphery except inferiorly in posterior elevation map with Kmax 61.90D. The CTSP and PTI showed deviation from normal and quick sloping (slope before 6mm corneal diameter), denoting high risk of ectasia.

AS-OCT (Huvitz machine) showed characteristic thinning in the peripheral cornea with stromal opacification.

Rest of the general and ocular examination, including fundus, were within normal limits.

The lipid profile of the patient showed increased blood levels of total cholesterol (215 mg/dL), triglyceride level (204 mg/dL), LDL (131.27 mg/dL), VLDL (40.80 mg/dL) and cholesterol/HDL cholesterol/HDL cholesterol/HDL cholesterol / HDL-cholesterol ratio (5.01 mg/dL).

Differential diagnoses are as follows-

- Pellucid marginal degeneration (PMD) – Usually bilateral, clear, inferior (typically 4–8 o'clock) peripheral corneal thinning.[1] Butterfly pattern is seen on corneal topography.
- Keratoconus – Central or paracentral cornea undergoes thinning and steepening, most commonly involving the inferior cornea. This causes high irregular astigmatism and poor visual acuity.[2] On corneal topography, an asymmetrical bow tie pattern is seen.
- Mooren's ulcer – Associated with intense limbal inflammation, conjunctival congestion in one or both eyes. Usually of autoimmune etiology,[3] can be triggered by surgery or trauma.
- Arcus senilis – Age-related change at the limbus, starts at the superior and inferior cornea. Not associated with corneal vascularization.[4]

Treatment

As there was no improvement in visual acuity on best correction in the left eye, we started our patient on lubricant carboxymethyl cellulose 0.5% in both eyes and advised him to avoid rubbing of eyes, use protective eyewear and regular follow-up.

Discussion

TMD is an uncommon but distinct variety of marginal corneal thinning predominantly affecting young males.[5]

It causes non-inflammatory peripheral corneal thinning which may be unilateral, asymmetrically bilateral and is associated with corneal vascularization, opacification and lipid deposition.[5,6] as seen in our case.

Rarely, it can also be associated with pterygium, which was the history finding in this case and the patient can be otherwise asymptomatic.

Classically, there is peripheral flattening and high against-the-rule astigmatism.[7] We observed ectatic areas on the anterior and posterior elevation map, areas of high-risk corneal thinning on pachymetry in the left eye, high Kmax values in both eyes despite normal corneal pachymetry in the right eye, indicating progression of disease in the right eye as well. The spatial profile graphs in both eyes were seen to be deviating from normal in this case, showing quick sloping in the left eye indicating high risk of ectasia, but in the right eye the deviation attributes to the initial progression of disease.

AS-OCT (Huvitz machine) showed characteristic peripheral thinning with stromal opacification.

Biomicroscopy along with symptoms of the patient helps in differentiating TMD from other close differentials of the disease like PMD presenting with typical inferior corneal thinning, Keratoconus having central or paracentral steepening with bow tie pattern on corneal topography, and Mooren's ulcer being a painful condition with marginal ulceration and severe congestion.

Generally, no treatment is required unless perforation or impending perforation occurs. Conservative management for astigmatism includes spectacles or rigid gas permeable contact lenses with close monitoring of corneal thinning.[6]

Conclusion

We found a rare presentation of an early asymmetrically bilateral TMD. Corneal topography along with slit lamp examination and increased lipid profile levels helped in diagnosing the case as TMD. Protection of the ocular surface is the goal of treatment in TMD. Such patients should be educated about their disorder and the need for regular follow-up.

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