Case Report

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Systemic treatment and surgical intervention in inflammatory Terrien disease

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Abstract:

This case report presents a unique instance of a 55-year-old male patient exhibiting features of both Terrien marginal degeneration (TMD) and Fuchs' superficial marginal keratitis. Characterized by peripheral corneal thinning vascularization, and a pseudopterygium, the patient experienced recurrent photophobia, redness, and tearing over 15 years. This case challenges the traditional distinction between TMD and Fuchs' superficial marginal keratitis, suggesting a potential common underlying disorder. Mycophenolate mofetil provided a partial response, while pseudopterygium removal led to sustained remission, emphasizing its therapeutic significance. This case highlights the first documented use of mycophenolate in TMD and supports the notion of shared vasculitic origins between TMD and Fuchs' keratitis, raising intriguing questions about targeted therapeutic interventions.

Keywords:

Fuchs superficial marginal keratitis, mycophenolate, peripheral thinning corneal diseases, superficial keratectomy, Terrien marginal degeneration

Introduction

It has been suggested that Terrien marginal degeneration (TMD) and Fuchs superficial marginal keratitis represent distinct expressions of a common disorder characterized by peripheral corneal thinning and having a vasculitic origin. In this context, we present a unique case involving a patient exhibiting features of both TMD and Fuchs' superficial marginal keratitis. While the patient showed a partial response to oral immunosuppression, notable symptomatic relief was achieved following the removal of pseudopterygium and conjunctival autografting. The patient consent form was obtained for publication purposes.

Case Report

A 55-year-old man presented with a 15-year history of recurrent episodes of photophobia, redness, pain, and tearing in each eye at different times. His medical history was unremarkable, with no history of ocular surgery or trauma. The ocular irritation typically responded well to topical steroids but would flare-up several times a year, as the steroids were tapered and discontinued. The patient could not tolerate topical 0.1% cyclosporine due to severe redness and tearing. Over the last couple of years, his vision had declined due to an increase in astigmatism.

During the initial examination, the best spectacle-corrected visual acuity was 20/25 in the right eye (RE: −1−2 at 175°) and 20/30 in the left eye (LE: −2−3 at 60°). Intraocular pressure measured 16 mmHg in the RE and 17 mmHg in the LE. Although the patient had recently started a new cycle of steroid drops, moderate conjunctival injection persisted in both eyes. The lacrimal meniscus was intact, and the lid margins showed no signs of inflammation. In the RE,

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a pseudopterygium at the nasal cornea invaded 2 mm of the cornea. In addition, there was peripheral corneal thinning from the 11-o'clock to 8-o'clock positions, accompanied by fine superficial vessels, linear lipid deposition near the thinning area, and an absence of epithelial defect [Figure 1a]. The LE exhibited similar findings, with peripheral thinning spanning 360°, fine superficial vessels, and a nasal pseudopterygium invading 3 mm of the cornea [Figure 1b]. The patient reported a significant growth in both pseudopterygiums over the last 2 years. Corneal tomography revealed superior steepening in both eyes, with with-the-rule astigmatism of 2.5 D in the RE and oblique astigmatism of 3.7 in the LE.

Laboratory investigations, including a comprehensive panel such as complete blood count, rheumatoid factor, antineutrophil cytoplasmic antibody, antinuclear antibodies, erythrocyte sedimentation rate, C-reactive protein, treponemal antibody (FTA-ABS), chest X-ray, purified protein derivative, and hepatitis C and B serology, all returned normal results. The patient initiated oral mycophenolate at 1 g every 12 h and was closely monitored with a presumptive diagnosis of inflammatory TMD. This condition, unlike Fuchs superficial marginal keratitis, manifests stromal lipid deposition in the thinning edge of the disease.

A year later, flare-ups decreased in both number and severity, but they still occasionally interfered with daily activities. Corneal tomography revealed superior steepening in both eyes, with with-the-rule astigmatism of 5.7 D in the RE and oblique astigmatism of 7.3 D in the LE. As the pseudopterygium had considerably grown in both eyes, the patient underwent a superficial keratectomy to remove the pseudopterygium in the LE. This procedure also involved removing the peripheral nasal superior pannus and placing a conjunctival autograft nasally, harvested from the superotemporal quadrant of the same eye [Figure 2b]. Six months later, the patient reported no flare-ups in the operated eye. In contrast, the RE, which had not undergone intervention, exhibited a marked increase in the pseudopterygium [Figure 2a] and corneal thinning close to the head of the pseudopteryigum [Figure 3]. Consequently, the same procedure was performed on the RE.

In the most recent appointment, 6 months after the last surgery, the patient continues with mycophenolate but has not required topical steroids. The best spectacle-corrected visual acuity was 20/20 in the RE (−1−2,5 at 160°) [Figure 4a] and 20/25 in the LE (−2−4 at 65°) [Figure 4b]. However, irregular astigmatism had only mildly reduced due to the thinning and scarred tissue [Figure 5].

Discussion

TMD stands out as an infrequent yet distinctive bilateral peripheral corneal thinning disorder, typically encountered in middle-aged individuals, predominantly males. The characteristic hallmarks of this condition

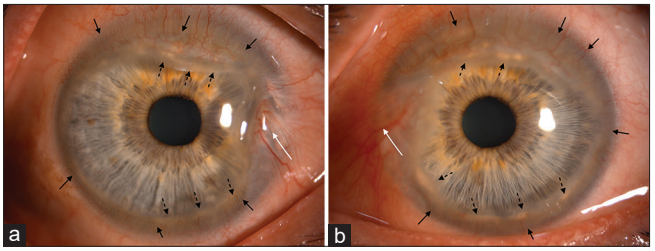

Figure 1: Slit-lamp photography of the right eye (a), left eye (b) at the time of the initial medical consultation. A progressive thinning and furrow formation with superficial vascularization are observed in the peripheral cornea 360° (small black arrows), particularly superiorly and nasally. Nasally a pseudopterygium is also visible (white arrow). In addition, lipid deposition occurs at the leading edge of the affected region (black dashed arrow). Both eyes exhibit also mild to moderate hiperemia

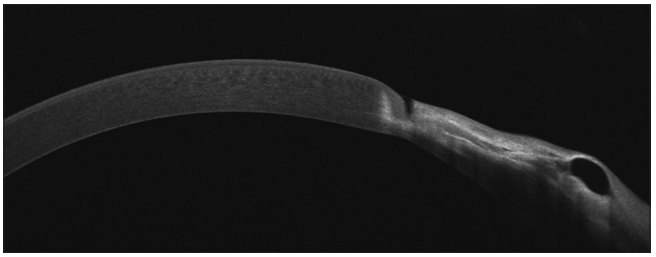

Figure 3: Anterior segment optical coherence tomography of the right eye showing the leading edge of thinning beneath the pseudopterygium

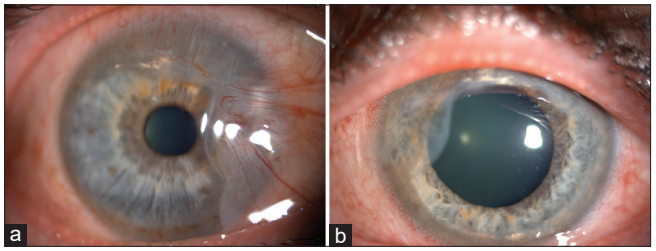

Figure 2: Slit lamp photography of the right eye (RE) (a), left eye (LE) (b) after superficial keratectomy and conjunctival autograft of the LE, performed elsewhere. There is a noticeable progression of the pseudopterygium in the RE, and a nasal thinned leucoma in the LE

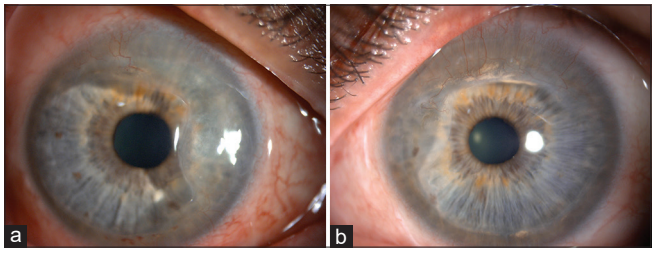

Figure 4: Slit-lamp photography of the right eye (RE) (a), left eye (b) after superficial keratectomy and conjunctival autograft of the RE. Notice the lipid deposition in the leading edge of the thinning bilaterally, which is a hallmark of Terrien marginal degeneration

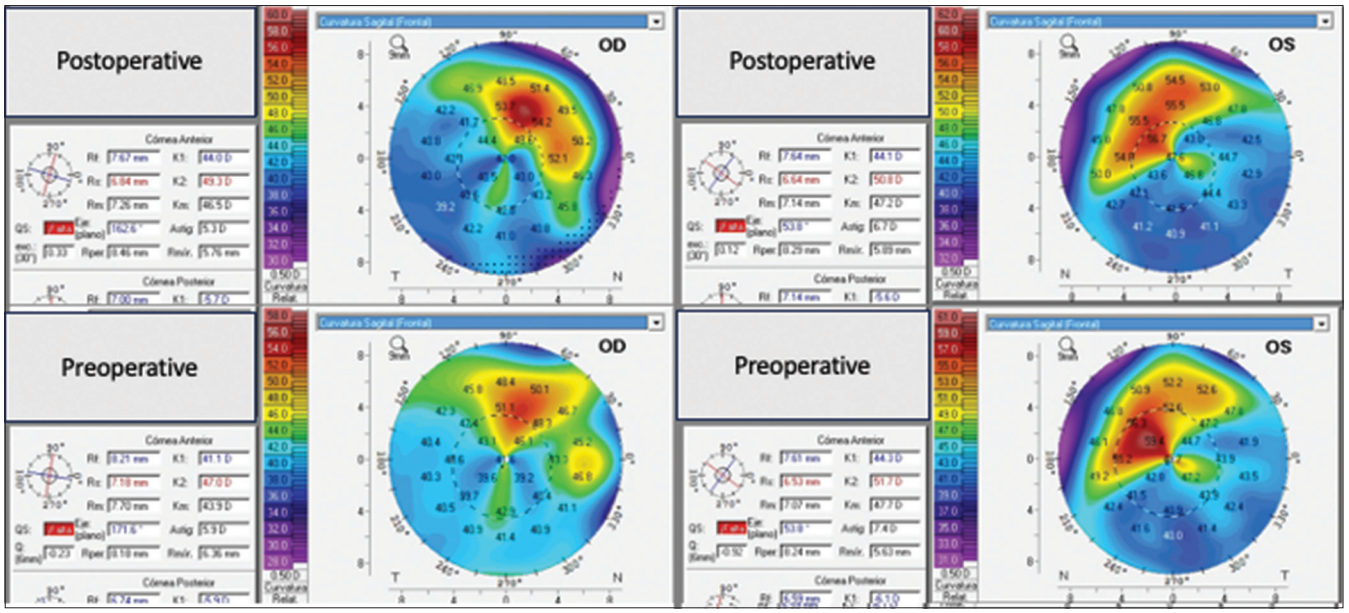

Figure 5: Corneal topography showing corneal curvature maps before and after the surgery in both the right eye and left eye

include the progressive thinning and furrow formation of the peripheral cornea, with a peculiar onset often occurring superiorly. A notable feature is the presence of superficial vascularization, which assumes a radial trajectory, occasionally displaying tortuosity and, at times, a circumferential appearance in the thinned area. In addition, lipid deposition occurs at the leading edge of the affected region. Intriguingly, TMD is recognized for its peculiar absence of epithelial defects or apparent inflammatory signs or symptoms.[1]

In the case of our patient, who exhibited all these classical features of TMD, a unique twist was observed – the presence of pseudopterygia and periodic ocular inflammation, more commonly associated with Fuchs superficial marginal keratitis. Despite the apparent clinical distinctions, there has been a historical consideration that both TMD and Fuchs superficial marginal keratitis may represent different manifestations of an underlying disease process.[2]

Within the broader spectrum of TMD, a specific and rare subset known as inflammatory TMD has been previously described.[3] This subtype is believed to be triggered by a hypersensitivity immune response directed against the basement membrane. While conventional management approaches have involved the use of topical anti-inflammatory treatments to control relapses, systemic treatment options have not been as extensively explored in poorly responsive patients, in contrast to other peripheral keratitis entities such as Mooren's ulcer.[4]

This case report makes a significant contribution by detailing the first published instance of TMD treated with mycophenolate mofetil, showcasing a partial response. There is no evidence in the literature regarding the effect of mycophenolate in treating this particular and rare disease; however, it has been used in corneal peripheral ulcers with an immune/inflammatory pathophysiology.[4] The pathophysiology of this disease is currently unknown, but several hypotheses suggest that it may be mediated by immune/inflammatory cells.[2,5]

What proved to be transformative in preventing relapses was the removal of the pseudopterygia. This intervention, consisting of a superficial keratectomy and conjunctival autograft, has previously demonstrated its efficacy in reducing flare-ups in patients with Fuchs superficial marginal keratitis.[6] Remarkably, this approach proved to be effective in the subset of inflammatory TMD characterized by the presence of pseudopterygia, thereby strengthening the hypothesis of a shared underlying pathophysiology between these seemingly distinct corneal disorders.

Moreover, there is a compelling suggestion that TMD and Fuchs superficial marginal keratitis might share a common vasculitic origin 1. This aligns with other peripheral ulcerative diseases such as Mooren's ulcer, where therapeutic strategies often involve systemic immunosuppression and surgical resection of the corneal lesion and adjacent conjunctiva.[7] Considering the similarities in treatment approaches for these conditions raises intriguing questions about the interconnectedness of their pathogenic mechanisms and potential avenues for more targeted therapeutic interventions in the future.

Data availability statement

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

The author declares that there are no conflicts of interests of this paper.

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