Clinical Case Report

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Spontaneous corneal perforation in Terrien's marginal degeneration in childhood

A case report

Junkyu Chung, MD[a], Kyung Hyun Jin, MD, PhD[b], Jaheon Kang, MD, PhD[a], Tae Gi Kim, MD[a,*]

Abstract

Rationale: Terrien's marginal corneal degeneration (TMD) is characterized by progressive peripheral corneal thinning. It appears primarily above the age of 40 years and is rare at younger ages. Spontaneous corneal perforation in TMD is a rare, but serious complication that may occur in childhood.

Patient concerns: This review presents the case of a 16-year-old girl presented with blurred vision in the right eye on awakening in the morning. Slit-lamp examination revealed superior corneal thinning with a corneal perforation.

Diagnoses: The best-corrected visual acuity (BCVA) was 20/50 in the right eye and 20/20 in the left eye. Intraocular pressures were 5 and 18 mm Hg in the right and left eyes, respectively, measured using a noncontact tonometer. Slit-lamp examination revealed superior corneal thinning with superficial pannus. A 1-mm corneal perforation was observed at the 11 o'clock position. The anterior chamber of the right eye was flat and the Seidel test result was positive. The left eye showed no apparent abnormality on slit-lamp examination. Corneal topography showed 4.3 D of against-the-rule astigmatism, and anterior segment optical coherence tomography revealed superior corneal thinning. We diagnosed it as childhood onset TMD.

Interventions: Multilayered amniotic membrane transplantation was performed over the perforation site and a bandage contact lens was placed.

Outcomes: At 1 week postoperatively, the BCVA of the right eye improved to 20/32, the amniotic membrane graft was well-attached, and the anterior chamber remained deep. At 2 months postoperatively, the BCVA was 20/25 and the anterior chamber depth was maintained.

Lessons: Spontaneous corneal perforation due to TMD is rare, but may occur in childhood. The possibility of corneal perforation should be considered even in childhood and good surgical results can be obtained with amniotic membrane transplantation.

Abbreviations: BCVA = best-corrected visual acuity, TMD = Terrien's marginal corneal degeneration.

Keywords: amniotic membrane transplantation, childhood, spontaneous corneal perforation, Terrien's marginal degeneration

1. Introduction

Terrien's marginal corneal degeneration (TMD) is characterized by thinning of the peripheral cornea with slow progression over several years.[1] It usually starts in the superonasal quadrant of the cornea and remains asymptomatic until the corneal thinning causes increased astigmatism and decreased visual acuity.[2] TMD occurs bilaterally and predominantly affects men, usually after 40 years of age. According to recent retrospective studies, 28% of patients with TMD have unilateral involvement and spontaneous corneal perforation occurs in 9% of patients with TMD.

Spontaneous perforation is rarely reported in TMD. Although the exact mechanism of corneal stromal loss is still not clear, it is presumed to be related to abnormalities in corneal homeostasis. Corneal homeostasis is the balance between collagen synthesis and catalysis. In TMD, this balance is lost in favor of catalytic processes, leading to a progressive thinning of the corneal stroma.[3] This process is time-consuming and eventually causes spontaneous perforation. However, it is known that most patients who have TMD do not progress to corneal perforation.[4] Based on a literature review, only 7 cases of spontaneous perforation in TMD have been reported, and to the best of our knowledge, there are no reports of spontaneous perforation in TMD in childhood.[2,5–7]

We report a rare case of unilateral TMD with spontaneous corneal perforation in a 16-year-old girl, who was treated with amniotic membrane transplantation.

Written informed consent was obtained from the patient for this case report. No ethical approval was obtained because this study is retrospective case report and did not involve a prospective evaluation.

2. Case report

A 16-year-old girl presented to the clinic complaining of blurred vision in her right eye that had started in the morning. There was

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[a] Department of Ophthalmology, Kyung Hee University Hospital at Gangdong, [b] Department of Ophthalmology, Kyung Hee University Medical Center, Kyung Hee University, Seoul, Republic of Korea.

[*] Correspondence: Tae Gi Kim, Department of Ophthalmology, Kyung Hee University Hospital at Gangdong, # 892 Dongnam-ro, Gangdong-gu, Seoul 05278, Republic of Korea (e-mail: tk1213@hanmail.net).

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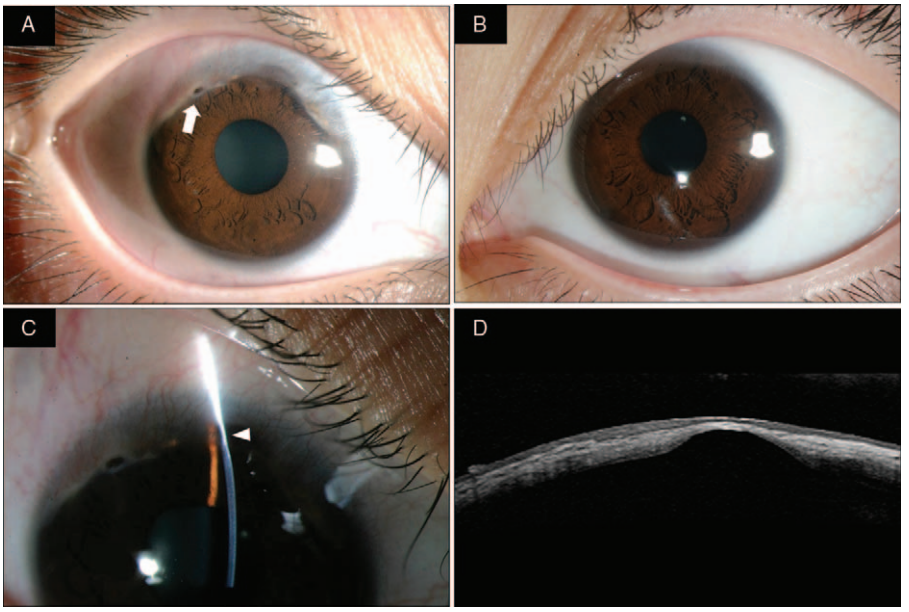

Figure 1. (A) Slit lamp photograph of the right eye showing superficial vascularization with corneal perforation (arrow). (B) Slit lamp photograph of the left eye showing a normal cornea. (C) Slit lamp photograph of the right eye displaying superior stromal thinning (arrowhead) with intact epithelium and a shallow anterior chamber. (D) Severe corneal stromal thinning with intact overlying epithelium as seen on anterior segment optical coherence tomography.

no previous history of ocular or systemic disease, trauma, ocular surgery, collagen vascular disease, or contact lens wear.

On examination, the best-corrected distance visual acuities (BCVAs) were 20/50 in the right eye and 20/20 in the left. Her intraocular pressure (IOP) was 5 mm Hg in the right eye and 18 x0200A;mm Hg in the left, measured using a noncontact tonometer. Slit-lamp examination showed superior peripheral corneal thinning in the right eye; the corneal epithelium was intact and superficial vascularization was present (Fig. 1A, C). The thinned cornea showed tissue loss of >50% of the stroma; anterior segment optical coherence tomography revealed corneal stromal thinning (Fig. 1D) A 1-mm corneal perforation was noted at the 11 o'clock position. Seidel test using fluorescein dye was positive and the anterior chamber was shallow. On corneal topography, 4.3 D of against-the-rule astigmatism was observed (Fig. 2). The left eye showed no apparent corneal abnormality on slit-lamp examination; the anterior chamber was deep, and corneal topography showed a 1.1 D with-the-rule astigmatism (Figs. 1B and 2). Serological tests for rheumatic diseases did not show any abnormalities.

A diagnosis of unilateral TMD with spontaneous corneal perforation of the right eye was made; multilayered amniotic membrane transplantation was performed on the right eye under local anesthesia on the day of admission. We used dried amniotic membrane (AmbioDry2; IOP Ophthalmics) in 3 layers. Two layers of amniotic membrane with the basement membrane side facing up were attached at the perforation site using Greenplast fibrin glue (Green Cross, Seoul, Korea). Finally, a larger piece of membrane was trimmed to cover the corneal perforation with the basement membrane side facing down. Subsequently, a purse-string suture was placed using 10-0 nylon to keep the amniotic membrane layers in place. At the end of the operation, the eye was covered with a soft bandage contact lens (Biomedics, Cooper Vision) (Fig. 3).

At 1 week postoperatively, the BCVA improved to 20/32 in the right eye. The grafted amniotic membrane was well attached to

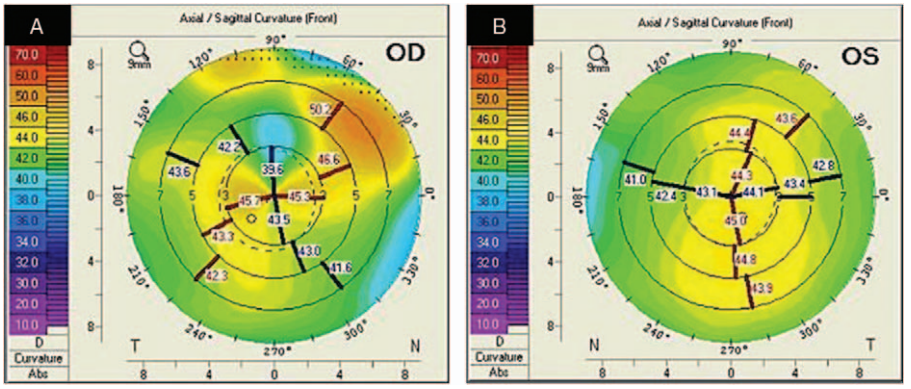

Figure 2. (A) Corneal topography of the right eye showing against-the-rule astigmatism with an irregular component. (B) Corneal topography of the left eye showing with-the-rule astigmatism and a normal corneal contour.

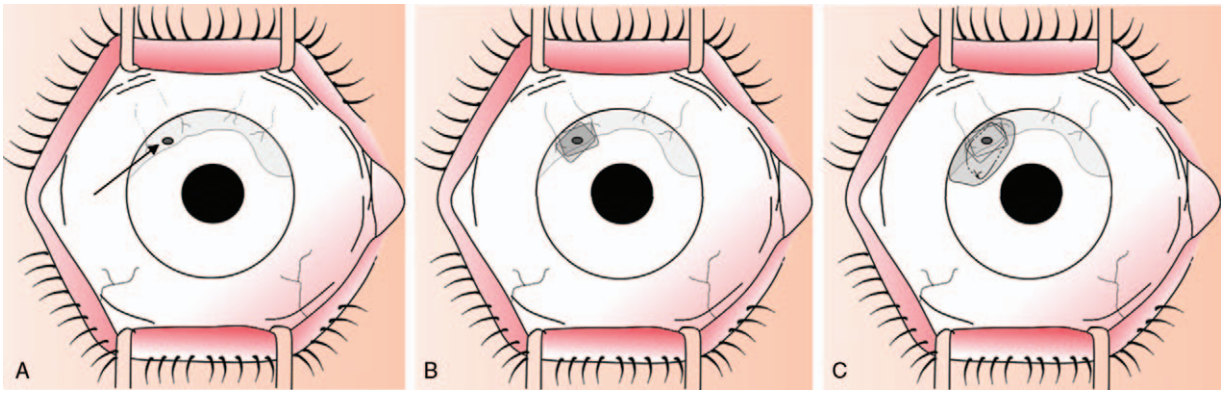

Figure 3. Schematic illustration of multilayered amniotic membrane transplantation. (A) Corneal perforation is observed at the 11 o'clock position (arrow). (B) Transplantation of 2 layers of amniotic membrane, with the side containing the basement membrane facing upward, was performed using tissue glue. (C) The third amniotic membrane was attached with the side containing the basement membrane facing down and fixed to the cornea with purse-string suture.

the corneal perforation site (Fig. 4A). No leakage was observed and the anterior chamber was deep (Fig. 4B). There was no recurrence of the perforation at 2 months postoperatively, and the BCVA improved to 20/25; moreover, the depth of the anterior chamber was maintained (Fig. 4C, D).

3. Discussion

TMD is a rare peripheral corneal ectactic disorder, typically known to occur bilaterally in men over 40 years of age.[1] Unlike the typical TMD, a variant of TMD occurs in younger patients, which is characterized by prominent inflammation, pain, and associated scleritis.[8] TMD is a slowly progressive degenerative disease, and spontaneous corneal perforation rarely occurs in young patients.[5] In this case, unilateral spontaneous corneal perforation occurred in a 16-year-old girl, which is extremely rare. To the best of our knowledge, this is the youngest of all spontaneous corneal perforation cases reported.

Complications of TMD are the result of corneal thinning, which include corneal astigmatism, trauma-related perforation,

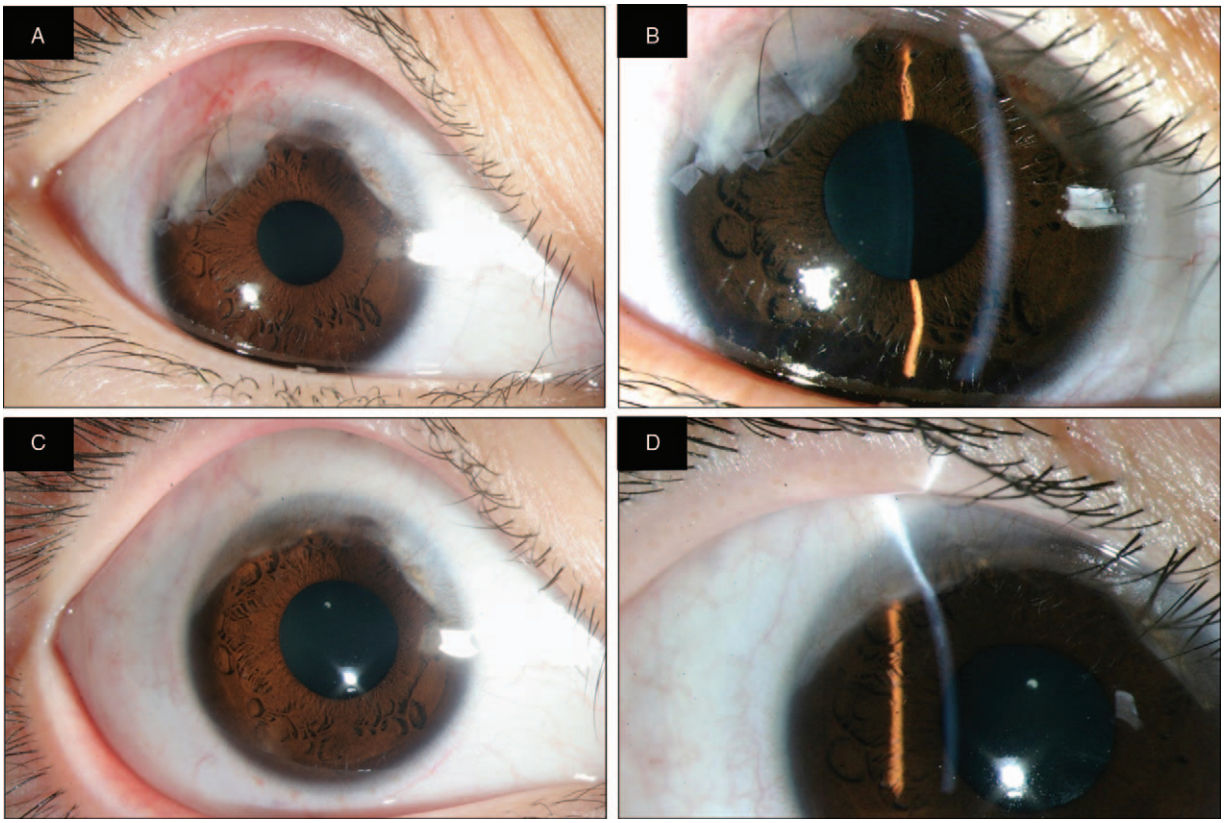

Figure 4. Slit-lamp photograph of the right eye at 1 week (A, B) and at 2 months (C, D) postoperatively. (A) Slit-lamp examination reveals well-attached corneal patch graft at the perforation site. (B) Anterior chamber is deep without any aqueous leakage. (C, D) The corneal perforation site was well covered with fibrovascular tissue and no additional aqueous leakage was observed and anterior chamber remained deep.

Table 1

Summary of published treatment options for spontaneous corneal perforation in TMD.

Therapeutic modality	Number of patients	Advantages[14–18]	Disadvantages[14–18]
Close observation[2]	3	- Noninvasive No surgically induced astigmatism	- Restricted to relatively simple cases of corneal perforation
Bandage contact lens[5]	1	- Eliminates need for operation Less scarring No surgically induced astigmatism	- Restricted to relatively simple cases of corneal perforation
Conjunctival flap[2]	1	- Provides support for weakened cornea and anti-proteases and antibacterial substances	- Conjunctivalization of the cornea Extensive corneal vascularization
Lamellar keratoplasty[7]	1	- Reduces risk of endothelial graft rejection Retains structural integrity Efficient visual rehabilitation relative to penetrating keratoplasty	- Surgically induced astigmatism Technically difficult
Penetrating keratoplasty[6]	1	- Applicable to large area of corneal perforations	- Invasive Risk of graft rejection Surgically induced astigmatism

and may be associated with episcleritis or scleritis, which generally result in ocular discomfort. In atypical cases, corneal thinning has been known to cause a trabeculectomy-like filtering bleb.[6,9,10] In addition, spontaneous perforation, although rare, may occur as in this case.

TMD should be differentiated from other peripheral corneal disorders, such as Mooren's ulcer. TMD is generally easy to differentiate from Mooren's ulcer because TMD lacks a central undermined edge to the thinning trough and the epithelium remains intact; in addition, patients with TMD do not experience pain. Other differential diagnoses include pellucid marginal degeneration and furrow degeneration, which are characterized by the absence of lipid infiltrates associated with the central region of the thinning region, rendering them distinguishable from TMD. In addition to this, staphylococcal marginal keratitis, collagen vascular diseases, and infectious corneal ulcers should be excluded from TMD.[11]

TMD can be managed conservatively because patients are often asymptomatic and the disease progresses slowly. Conservative treatment includes spectacles and contact lenses to correct the corneal astigmatism. In case of episcleritis or scleritis, topical steroids may be used to control the inflammation. Surgical treatment is required when corneal perforation is imminent or present. Chan et al reported that corneal lamellar keratoplasty was the most frequent operation performed in 23.3% of patients with TMD requiring surgical treatment.[1] Conjunctival flaps and amniotic membrane transplantations are the other surgical options.[2,12] However, previous reports of corneal perforation mostly focus exclusively on corneal perforation due to iatrogenic causes or trauma; treatment methods for spontaneous perforation are rarely reported. Table 1 summarizes the treatment options for spontaneous corneal perforation in TMD reported in published cases. In this case, amniotic membrane transplantation and a bandage contact lens were used, which enabled rapid corneal recovery and maintenance of the anterior chamber. AMT can be used to restore the thickness of the cornea when used as a multilayer. The basement membrane reinforces the differentiation and migration of the corneal epithelial cells, and prevents apoptosis.[13] Previously, it was reported that spontaneous corneal perforation could be successfully treated using a simple bandage contact lens.[3] However, in this case, the size of the corneal perforation was >1 mm and the aqueous leak was severe; hence, we could not treat the perforation using a bandage contact lens alone.

In TMD, typically, superior corneal thinning results in against-the-rule astigmatism, which may be the presenting symptom. In this case, the patient had 4.3 D of the against-the-rule astigmatism; however, she did not complain of decreased vision because the astigmatism was corrected with glasses. She presented with blurred vision only after the corneal perforation that lead to leakage of aqueous humor occurred. As in this case, the condition may progress to corneal perforation without any subjective symptoms.

4. Conclusion

In conclusion, spontaneous corneal perforation in TMD may occur in childhood. Therefore, when monitoring younger TMD patients, patients should be cautioned about the possibility of spontaneous corneal perforation and be observed carefully. AMT is an effective treatment option for rapid restoration of corneal perforation.

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