International Journal of Surgery Case Reports 95 (2022) 107257

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Femtosecond laser assisted superficial lamellar keratectomy as a successful treatment of corneal opacity in a patient with Thiel Behnke corneal dystrophy

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ARTICLE INFO

Keywords:
Thiel Behnke corneal dystrophy (TBCD)
Histopathology
Bowman's layer
Anterior segment-optical coherence tomography (AS-OCT)
Femtosecond laser (FSL)
Lamellar keratectomy

ABSTRACT

Introduction and importance: Thiel Behnke corneal dystrophy (TBCD) and Reis Buckler corneal dystrophy (RBCD) are Bowman's layer dystrophies with overlapping clinical features causing diagnostic confusion. However, each entity has typical histopathological features. We describe in this case the successful use of Femtosecond laser (FSL) in the treatment of TBCD-related corneal opacity.

Case presentation: We present a 54-year-old male with bilateral superficial corneal opacities consistent with TBCD based on clinical appearance, anterior segment optical coherence tomography (AS-OCT), and In vivo confocal microscopy. Management options were discussed with the patient before proceeding with Femtosecond Laser Assisted Superficial Lamellar Keratectomy (FSLASLK). The histopathological findings of the excised left anterior lamellar corneal flap were typical of TBCD and the patient had a satisfactory outcome.

Clinical discussion: TBCD typically affects Bowman's layer centrally with progressive opacities involving the deeper layer of the corneal stroma and the periphery with advancing age. Histopathology typically shows subepithelial fibrosis with interrupted basement membrane and totally replaced Bowman's layer by uneven fibrous tissue forming the characteristic saw tooth pattern. The treatment of such cases is challenging with variable success and recurrence rates. Our case was managed successfully using FSL.

Conclusion: TBCD, even though a rare type of dystrophy, should be suspected based on the appearance of the corneal opacities clinically. It can be diagnosed by typical AS-OCT findings supported by histopathological confirmation and can be successfully treated by FSASLK.

1. Introduction

Bowman's layer is an acellular non-regenerating layer, 8–10 μm in thickness, and lies between the corneal epithelium and stroma. Two autosomal dominant corneal dystrophies mainly involve Bowman's layer: Reis Bückler corneal dystrophy (RBCD) and Thiel Behnke corneal dystrophy (TBCD) with overlapping clinical features but different AS-OCT, and In vivo confocal microscopic findings [1–3]. TBCD is an autosomal dominant well-defined corneal dystrophy based on the international classification of corneal dystrophies, in which the gene has been mapped and identified as a heterozygous missense mutation (Arg555Gln) in human transforming growth factor beta-induced gene (TGFBI) located in the long arm of chromosome 5 [1,3]. The disease starts early in childhood and progress with age. It causes corneal recurrent epithelial erosions (REE) in the first 20 years of life. However, gradual reduction of vision develops later due to corneal scarring and the REE diminishes. This epithelial-stromal dystrophy typically affects Bowman's layer centrally, starting with irregularly shaped scattered opacities followed by symmetrical subepithelial honeycomb opacities. However, with aging, opacities can progress to involve the deeper layer of the stroma and the periphery [3]. OCT, in vivo confocal microscopy, and histopathology all aid in differentiating between TBCD and RBCD. Multiple treatment modalities were described to treat TBCD related corneal scarring including blade superficial keratectomy, excimer laser and corneal transplants with different success and recurrence rates [4–6]. We describe the use of femtosecond laser-assisted superficial

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https://doi.org/10.1016/j.ijscr.2022.107257
Received 10 May 2022; Received in revised form 26 May 2022; Accepted 28 May 2022
Available online 31 May 2022

lamellar keratectomy (FSASLK) for corneal opacity in a case of TBCD in an aphakic patient and report the functional and morphological early outcome of this procedure. This case report has been prepared and reported in accordance with the SCARE criteria [7].

2. Presentation of case

A 54-year-old healthy male with no significant family or medical history presented to the ophthalmology clinic with left aphakia following pars plana vitrectomy (PPV) and lensectomy for retinal detachment (RD) 3 years before (in another institute). When he

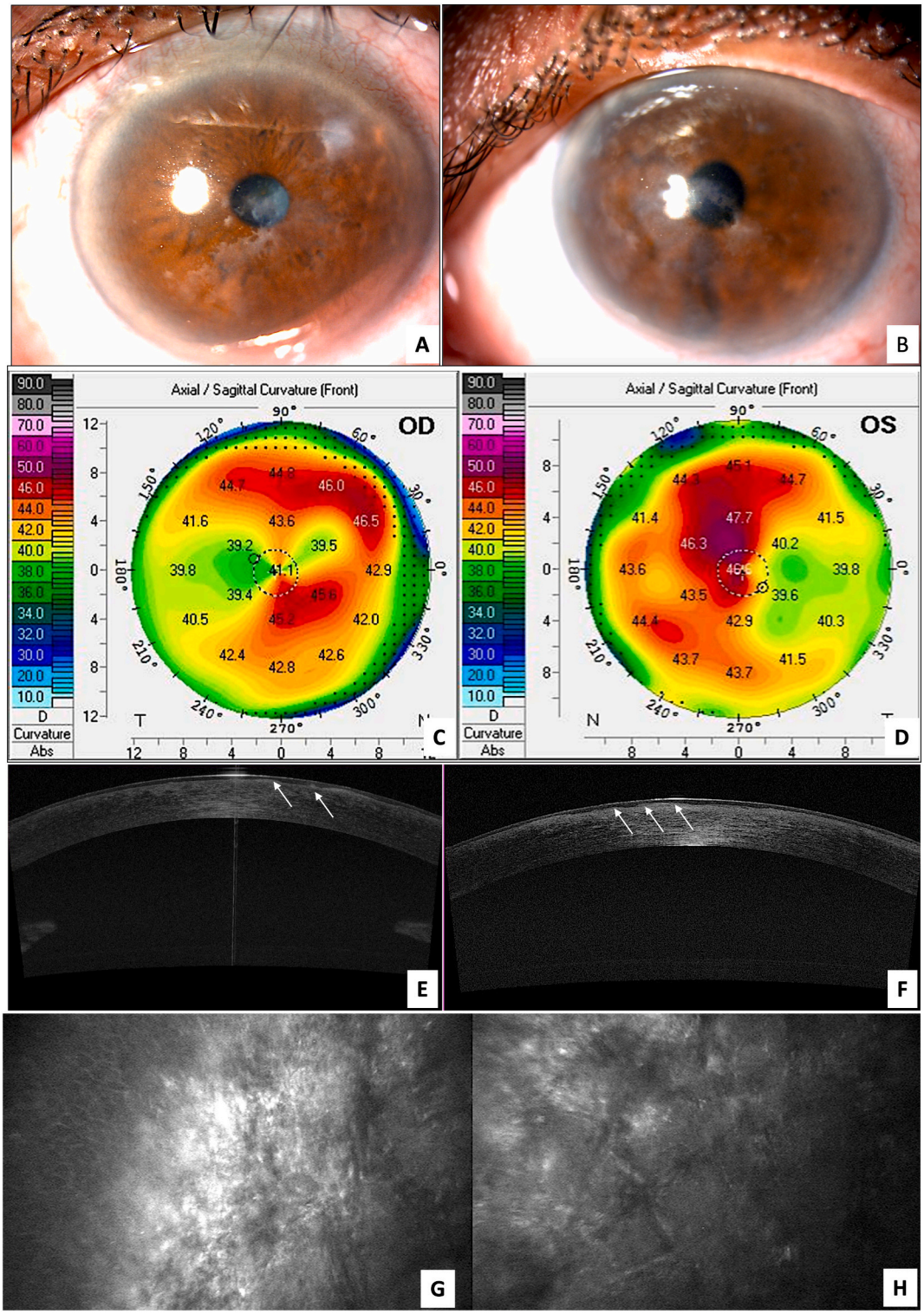

Fig. 1. A & B: Clinical appearance of the classical honeycomb corneal opacities in the right eye (A) and left eye (B). C & D: Corneal topography of the right (in C) and left (in D) eyes. E & F: Anterior segment optical coherence tomography (AS-OCT) showing subepithelial, and anterior stromal hyper reflective band with a saw tooth pattern (white arrows). G & H: Confocal microscopy of the right (in G) and left (in H) eyes showing replacement of Bowman's layer by hyper-reflective deposits at the basal epithelial layer.

presented to us he was complaining of reduced vision in both eyes. There was no history of ocular inflammation or contact lens use. The visual acuity (VA) was 20/30 and counting fingers at five feet in the right and left eyes, respectively. Slit lamp exam showed bilateral diffuse sub-epithelial and anterior stromal corneal scarring involving the central and paracentral cornea (Fig. 1a & b). There were no signs of trachoma, and the pupils were normal. Dilated examination showed retained intact anterior capsular support in the left eye, and early cataract in the right, while retina was flat in both eyes with laser marks in the left eye. The visual acuity of the left eye improved to 20/200 with subjective refraction. The axial curvature map in the corneal topography showed high corneal astigmatism in both eyes. However, the astigmatism was more irregular in the left eye (Fig. 1c & d). Corneal thickness was 557 μm and 577 μm in the right and left eye respectively. A trial of hard contact lens (HCL) fitting of the left eye improved vision to 20/80-. Anterior segment optical coherence tomography (AS-OCT) showed thick sub-epithelial, Bowman's layer, and anterior stromal hyper reflective band with a saw tooth pattern reaching a depth of 120 to 140 μm (Fig. 1e & f). Confocal microscopy showed hyper-reflective deposits at the basal epithelium (Fig. 1g & h). Treatment options were discussed with the patient, and he agreed to proceed with FSASLK for the left eye, which had astigmatism and worse vision. An informed consent was obtained and complication including recurrence were explained. FSASLK was performed by an experienced corneal surgeon through creation of a free corneal cap of 9 mm in diameter using 150 kHz iFS femtosecond laser (Abbot Medical Optics, Santa Ana, California) with a bed energy of 1.5 u J. The corneal cap thickness was selected at 120 μm. The thickness was chosen based on corneal assessment by AS-OCT and previously reported FSASLK excision depth for treatment of RBCD.[8] After cap removal, a bandage contact lens was placed. No complications were noted, and the patient was put on preservative free lubricating eye drops.

The excised corneal tissue was sent for histopathological examination, which showed an irregular epithelium with areas of acanthosis and thinning. The basement membrane of the epithelium was interrupted with wavy sub-epithelial fibrosis corresponding to the interrupted areas. Bowman's layer was absent and replaced by a thick uneven fibrous layer, which formed a saw-tooth pattern under the epithelium and stained deep blue with Masson's trichrome stain (Fig. 2a & b). The histopathological findings were consistent with TBCD.

Most of the opacities in the left eye were removed post FSASLK (Fig. 2c & d) and the AS-OCT post-operatively showed evidence of resolution of the previously seen irregularities due to the superficial opacities (Fig. 2e & f). On last follow up 6 months following the left eye procedure, the patient reported better vision and was happy with the

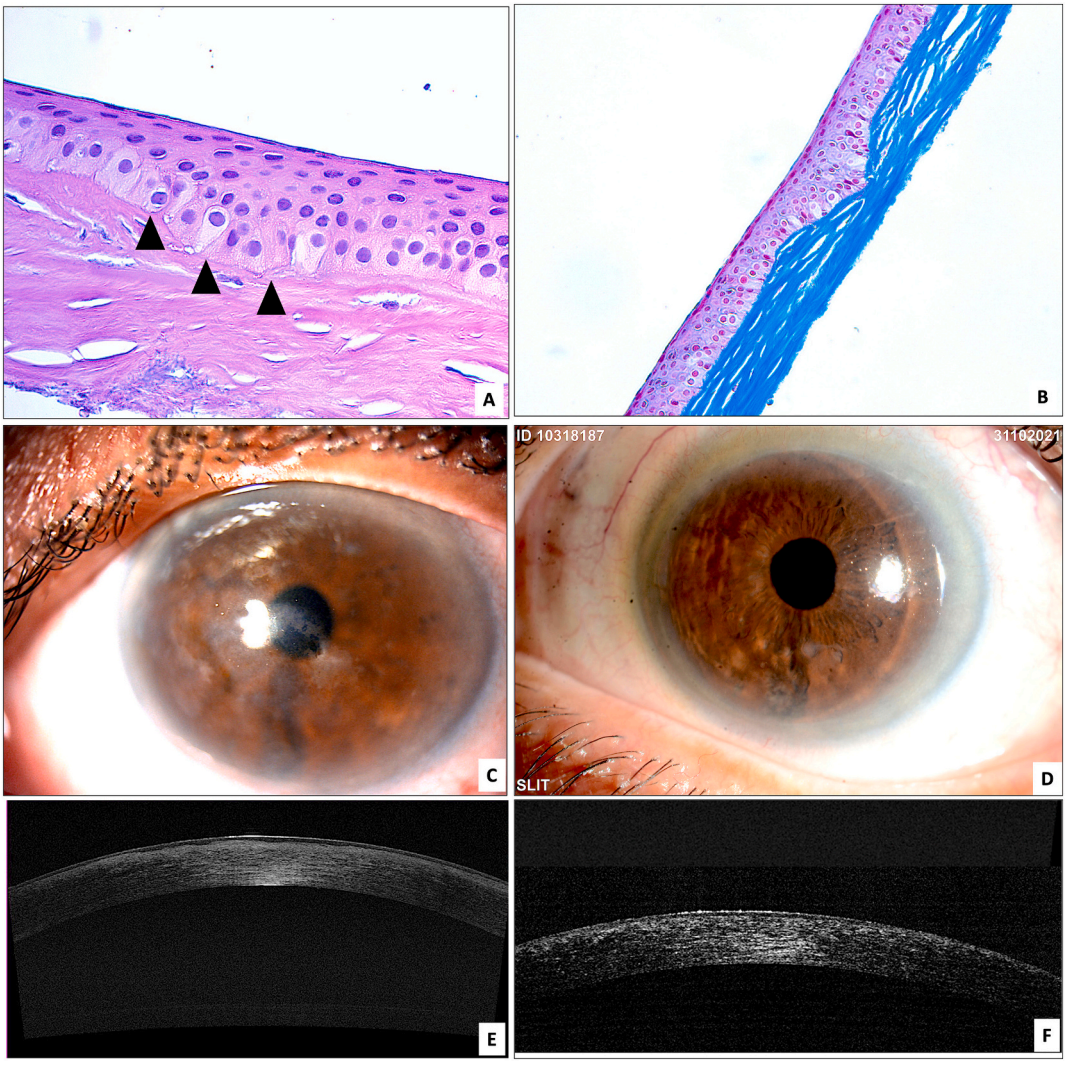

Fig. 2. A: Thick epithelium with saw-tooth pattern (Arrow heads), absent Bowman's layer, and subepithelial fibrous layer (Original magnification X 400 Hematoxylin & Eosin). B: The subepithelial fibrous layer deep staining (Original magnification X 200 Masson Trichrome). C: Slit lamp appearance of the left cornea at presentation. D: The left cornea, which is clear after treatment. E & F: Comparison of the AS-OCT before and after treatment showing resolved anterior corneal irregularities following FSASLK.

results. Visual acuity with subjective refraction improved to 20/100, and secondary intraocular lens implantation was planned in the future within few months.

3. Discussion

Corneal dystrophies affecting Bowman's layer (TBCD and RBCD) are uncommon and their clinical diagnosis might be challenging because of their similar clinical presentation. Since RBCD has a high rate of recurrence after treatment that occurs earlier and in a more severe degree compared to other TGFBI related dystrophies, confirming the diagnosis might guide the treatment plan and predict the outcome [3]. As shown in our Table (Table 1) genetic testing will aid in differentiating TBCD and RBCD and is also helpful in estimating the risk of recurrence after various treatment modalities to choose the optimal management [1–3]. Additionally, new treatment approaches for the prevention of corneal dystrophies and recurrences that involve genetic manipulations are currently being investigated [8]. The findings that were observed in our patient including slit lamp examination, AS-OCT, in vivo confocal microscopy, and histopathological features were all consistent with TBCD. The main goal of treating corneal opacities related to TBCD is to improve vision and to delay the need for keratoplasty. Compared to keratoplasty, PTK is a relatively safe, simple, repeatable (provided that not more than one third of the corneal thickness is removed, which is about 40–100 um), and has a short recovery time [4]. However, it might leave the patient with corneal irregularities, high order aberrations, haze and hyperopic shift [5,8]. Heida et al., reported ten eyes of five patients with TBCD that undergone PTK and reported a 2-line increase of BCVA in all patients with a recurrence in 5 eyes that didn't result in change of vision significantly [4]. Hsaio et al., described the use of combined wavefront-guided photorefractive keratectomy (PTK) to overcome the drawbacks in the refractive outcome that come along with PTK [5]. In their case the vision improved from 20/200 preoperatively in the right eye to 20/20 with a single recurrence that involved faint superficial opacities that didn't affect the vision and didn't require additional treatment [5]. Unfortunately, all excimer laser treatment will eliminate the chance of obtaining a sample to aid in the diagnosis of the disease. The use of Femtosecond laser (FSL) to treat anterior corneal dystrophy was investigated by Steger and his co-authors [8]. They reported FSASLK in 8 eyes with anterior corneal dystrophies that included RBCD, macular corneal dystrophy, lattice corneal dystrophy and granular corneal dystrophy, with good outcome in addition to the ability to provide histological diagnosis of the treated dystrophy [8]. In their study 5 of the 8 eyes had recurrence of the disease one of them was RBCD, and there were no cases of corneal ectasia within a 2 year follow up period [8]. FSL has an advantage over PTK in these cases since larger corneal zone can be treated with FSASLK (9.5 mm) compared to PTK (usually 6.0–6.5 mm), thus, avoiding having a transition zone [8]. Deeper corneal pathologies and patients with thin corneas that may require removal of more than 1/4th corneal thickness and might not be suitable for FSL. Visual improvement might be achieved with FSASLK despite the incomplete removal of corneal opacity due to the correction of most corneal irregularities. Additionally, FSL allows for a smooth regular planar corneal surface with precise control of flap diameter and thickness [9,10]. Corneal transplantation, including deep anterior lamellar keratoplasty and penetrating keratoplasty have good clinical outcome, however they carry risks of infection, rejection, failure, refractive challenges, and recurrence and shouldn't be considered as a first treatment option for TBCD [8].

4. Conclusion

We describe a case of TBCD clinically manifesting with corneal opacities and successfully treated with femtosecond laser to remove the corneal opacities with no perioperative or postoperative complications. His treatment modality allowed for histopathological examination of the anterior corneal tissue with confirmation of his diagnosis along with the other typical appearance by AS-OCT and confocal microscopy. We demonstrated a high success rate of FSASLK in such a case by clearing the cornea, normalization of the corneal surface and the reduced irregular astigmatism resulting in an overall visual improvement.

Table 1
[a]Differentiating features between TBCD and RBCD.

Feature(s)	TBCD	RBCD
Onset and clinical course	Childhood to adolescence	Early childhood
Symptoms	Milder course of recurrent epithelial erosions relatively compared to RBCD, photophobia, and diminished vision	Recurrent epithelial erosions, photophobia, and diminished vision
Signs	Honeycomb pattern of opacities	Geographic opacities
AS-OCT	Moderate reflectivity with saw tooth pattern	Highly reflective and sharp borders
In vivo confocal microscopy	Deposits are round with dark shadows	Deposits are highly reflective without shadowing
Histopathology	Layer of connective tissue and hyaline material obliterating Bowman's layer	Absence of Bowman's layer, sub-epithelial deposits with saw-tooth pattern
Electron Microscopy	Presence of curly fibers	Absence of curly fibers
Gene (Mutation)	missense mutation (Arg555Gln)	missense mutation (Arg124Leu)

TBCD: Thiel Behnke corneal dystrophy; RBCD: Reis Buckler corneal dystrophy; AS-OCT: Anterior segment optical coherence tomography.
[a] References: [1–3].

Provenance and peer review

Not commissioned, externally peer reviewed.

Source of funding

This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

Ethical approval

IRB is not required for case reports. However, information was obtained and reported in a manner that was compliant with the standards set forth by the Health Insurance Portability and Accountability Act, and the Declaration of Helsinki as amended in 2013.

Consent

General informed written consent was obtained from the patient for publication of this case report and accompanying anonymous use of photos. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Research registration

Not applicable.

Guarantor

Hind M. Alkatan.

CRediT authorship contribution statement

- Mohammed M. Abusayf: Review of chart for data collection, literature review and first draft of the case report.
- Mohannad F. Tobaigy: Literature review and first draft of the case report.
- Abdullah Alfawaz: The primary treating ophthalmic surgeon providing clinical images.
- Hind M. Alkatan: Histopathological examination, final tissue diagnosis, and taking images. Critical overall review and revision of the manuscript for submission as Corresponding author.

Declaration of competing interest

None.

Acknowledgements

The authors would like to thank [3]King Saud University Medical City represented in the laboratories, materials, manpower, and use of infrastructure to support this case report.

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