American Journal of Ophthalmology Case Reports 17 (2020) 100610

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

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

Case report

Placoid choroidopathy after bilateral uncomplicated descemet's membrane endothelial keratoplasty

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ARTICLE INFO

Keywords:
DMEK
Endothelial keratoplasty
Placoid choroidopathy
Chorioretinitis

ABSTRACT

Purpose: To describe a case of bilateral, sequential placoid choroidopathy following uncomplicated Descemet's membrane endothelial keratoplasty (DMEK).

Observations: A 49-year old woman presented with flashing lights and central visual field scotomas after undergoing uncomplicated DMEK combined with cataract surgery for Fuch's endothelial dystrophy in the right eye. She was found to have placoid choroidopathy responsive to systemic steroids and for which a comprehensive work-up was unrevealing. Three and a half months later, she underwent DMEK surgery in the fellow eye and again developed placoid choroidopathy in the operated eye. Work-up was again unrevealing and the lesion followed a similar course to the first eye on systemic steroids. Over the course of seven (right eye) and three and a half months (left eye) of follow-up, the uncorrected visual acuity was 20/20 bilaterally and the retinal lesions had modestly improved.

Conclusions and Importance: We report a case of placoid choroidopathy following uncomplicated DMEK combined with cataract surgery in both eyes of a single patient. This case expands upon the reported complications following DMEK surgery and suggests a need to remain aware of posterior segment complications following endothelial keratoplasty.

1. Introduction

Endothelial keratoplasty has revolutionized our ability to treat conditions of corneal endothelial dysfunction. First described in the 1950's by Charles Tillet,[1] endothelial keratoplasty entered a renaissance period around the year 2000 with the introduction of deep lamellar endothelial keratoplasty, a procedure that ultimately evolved into Descemet's stripping automated endothelial keratoplasty (DSAEK). DSAEK, which involves transplantation of an allograft comprised of a partial thickness layer of posterior stroma *in addition to* the endothelium and Descemet's membrane, has since remained the standard of care for treating endothelial dysfunction in the United States.[2,3]

More recently, Descemet's membrane endothelial keratoplasty (DMEK), a procedure that replaces dysfunctional endothelium with an allograft comprised of *only* endothelium and Descemet's membrane, has gained popularity among corneal surgeons.[4] In multiple studies, DMEK has proven to have better and faster visual outcomes as compared to DSAEK, as well as a lower risk or allograft rejection.[5,6] In a recent AAO report, DMEK was also reported to have a safe complication profile, with the most common complication being partial graft detachment. To date, the only posterior segment complication described after DMEK surgery has been the development of cystoid macular edema (CME), which has been reported to occur in 7–14% of patients in two series.[7,8]

Here, we present the case of a patient who developed placoid choroidopathy following uncomplicated DMEK surgery combined with cataract extraction and lens implantation in both eyes. To the best of our knowledge, this is the first report to describe chorioretinitis/choroidopathy following any endothelial keratoplasty surgery.

2. Case report

A 49-year old Caucasian woman of Portuguese descent presented to clinic with a four-year history of progressive, bilateral blurred vision and significant glare from oncoming headlights. Her past medical history was remarkable for Fuch's endothelial dystrophy and narrow angles for which she had previously undergone bilateral peripheral iridotomies. Her family medical history was notable for mild glaucoma in her father and symptomatic Fuch's endothelial dystrophy in her sister.

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https://doi.org/10.1016/j.ajoc.2020.100610
Received 15 August 2018; Received in revised form 21 February 2019; Accepted 27 January 2020
Available online 01 February 2020

Her social history was unremarkable. Her only medications were daily calcium, lutein (20 mg daily) and astaxanthin (12 mg daily) supplements. Her best-corrected visual acuity was 20/40 in both eyes and slit-lamp examination was notable for bilateral confluent central corneal endothelial guttata, mild corneal stromal edema in the absence of any anterior stromal haze or microcystic epithelial changes, patent superior peripheral iridotomies measuring < 1mm x < 1mm in each eye and trace nuclear sclerotic cataracts. Examination of the posterior segment was unremarkable, including normal vitreous, retinal vasculature, optic nerve, macula and peripheral retina. The patient underwent combined extra-capsular cataract extraction, one-piece acrylic intraocular lens implantation, and Descemet's membrane endothelial keratoplasty in the right eye according to a previously published standardized technique, with the only exception being the inferior peripheral iridotomy, which had been performed 12 days preoperatively with argon and Nd:YAG lasers to a size of approximately 500 μm in diameter.[9] There were no intra-operative complications.

The patient's post-operative course was unremarkable and she reached an uncorrected visual acuity of 20/25 with complete graft adherence by the second postoperative week. She was treated with a post-operative regimen of moxifloxacin 0.5% QID, ketorolac 0.5% TID and a prednisolone acetate 1% QID.

Four weeks post-operatively the patient reported intermittent flashing lights and two distinct dark shapes in her central vision in the right eye. On further questioning she reported the two scotomas might have been present immediately after surgery. Her best-corrected visual acuity at this time was 20/20. Fundus examination of the right eye revealed hypo-pigmented deep retinal/choroidal lesions along the superior and inferior temporal arcades (Fig. 1A). The patient was referred to the retina service. Fluorescein angiography (FA) showed early autofluorescence followed by blocking of the choroidal lesions and late staining of the surrounding retinal pigment epithelium (Fig. 2A and C). Optical coherence tomography (OCT) revealed placoid sub-retinal deposits and attenuation of the RPE signal with distortion of the outer retinal architecture in conjunction with vitreous cell and debris consistent with a mild overlying vitritis (Fig. 3A and C).

A comprehensive systemic investigation, including complete blood count, rapid plasma reagin, fluorescent treponemal antibody absorption, quantiferon gold, angiotensin converting enzyme, lysozyme, Lyme immunoglobulins, blood cultures and chest x-ray were unremarkable. Systemic prednisone (40 mg daily) was initiated. Over the course of three months of follow-up, the patient's subjective symptoms improved but the chorioretinal lesions improved only minimally and in light of this systemic steroids were tapered off over six weeks. Although the patient had good visual acuity in the operated eye (20/30 uncorrected, 20/20 best-corrected), she was anisometropic and suffering from asthenopia. She was scheduled for combined cataract and DMEK surgery in her dominant left eye.

The patient underwent uncomplicated extracapsular cataract extraction, acrylic one-piece intraocular lens implantation, and Descemet's membrane endothelial keratoplasty in the left eye three months after the first eye. As with the right eye, an inferior peripheral iridotomy approximately 500 μm in diameter was performed 5 days preoperatively. Immediately following surgery of the left eye, she denied any visual phenomena similar to what she remembered seeing after the first procedure on her right eye. The DMEK graft was fully attached and visual acuity was 20/20 uncorrected one week post-operatively. Ten days post-operatively the patient reported distortion of vision with patchy central scotomas in the left eye; her visual acuity was 20/40 and fundus examination showed a triangle-shaped deep retinal/choroidal hypo-pigmented lesion along the superotemporal arcade (Fig. 1B), similar in appearance to the lesions that had presented in her contralateral eye. Fluorescein angiography (Fig. 2B and D) and OCT imaging showed a placoid chorioretinal lesion with sub-retinal fluid along the superotemporal arcade and a sub-foveal serous pigment epithelial detachment (Fig. 3B and D). A comprehensive laboratory investigation was repeated and was again unrevealing and she was started on 40 mg of oral prednisone which was tapered over the course of six weeks.

Over the course of seven and three months of follow-up for the right and left eyes, respectively, the patient's symptoms of flashing lights and scotomas resolved and visual acuity recovered to 20/20 uncorrected in

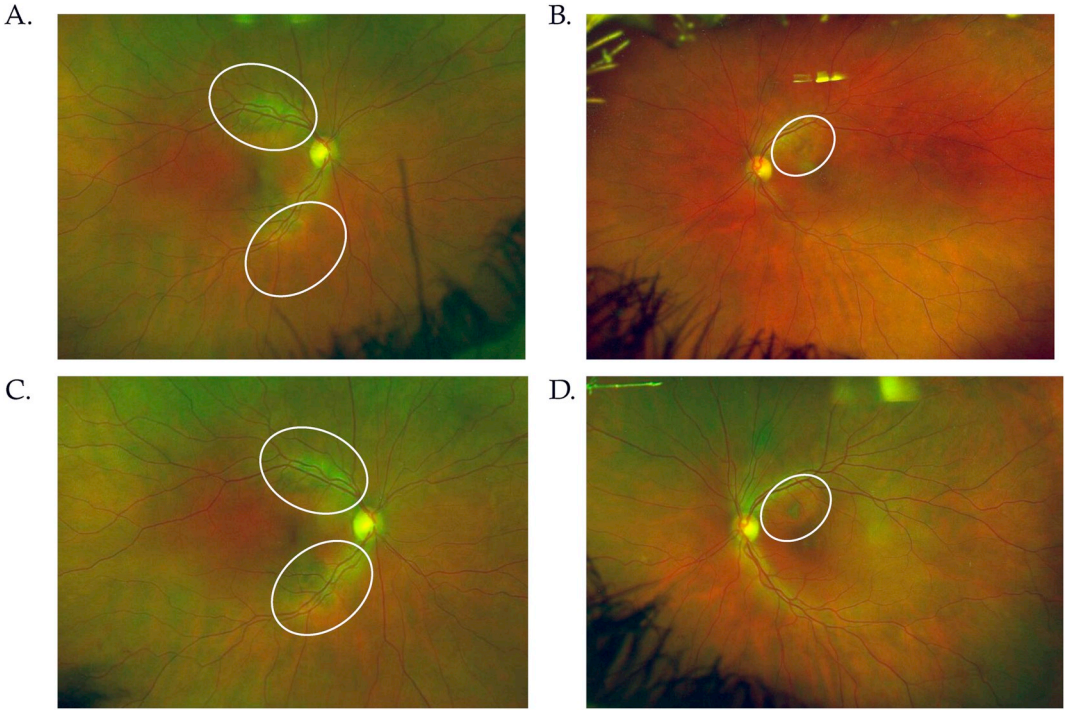

Fig. 1. Color fundus photos of the right eye 4 weeks (A) and left eye 10 days (B) after DMEK surgery demonstrating raised, hypopigmented deep retinal/choroidal lesions along the arcades (white circles). After 7 months of follow-up in the right eye (C) and 3.5 months of follow-up in the left eye (D) the retinal lesions appear more consolidated and hyperpigmented.

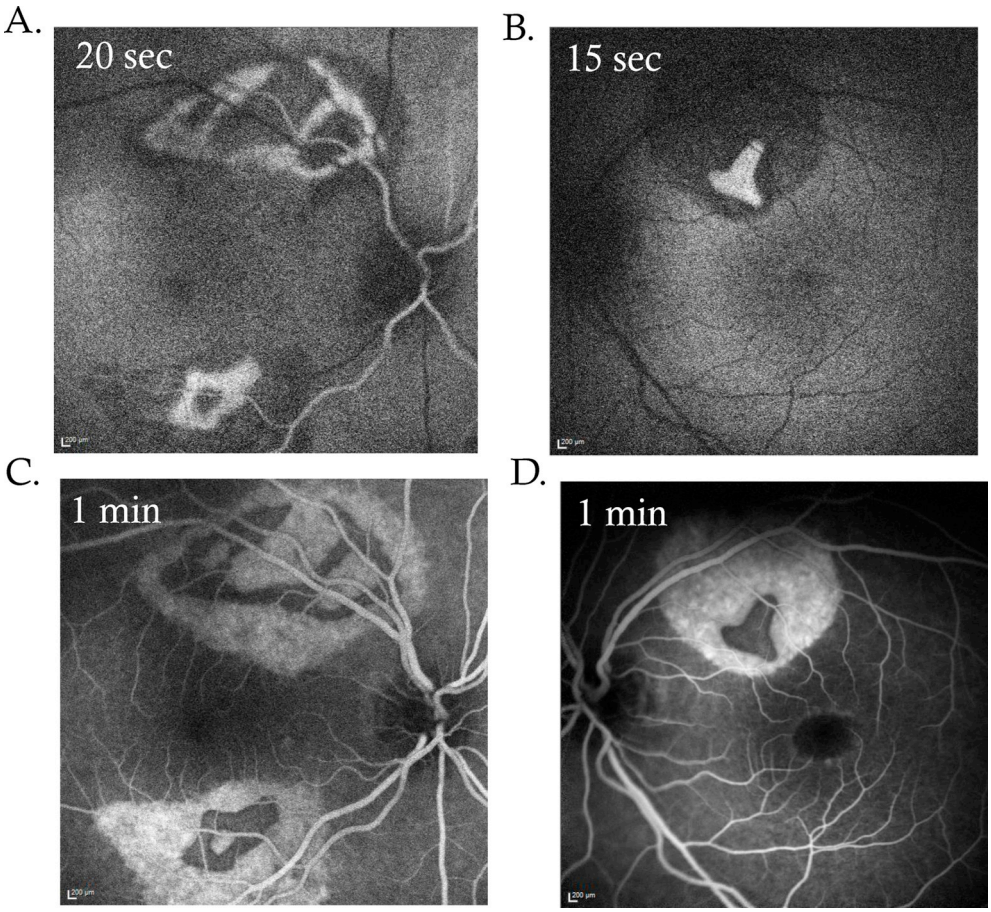

Fig. 2. Fluorescein angiography (FA) of the right eye 4 weeks after surgery demonstrating early autofluorescence (A) and late blocking with surrounding staining (C). FA of the left eye 10 days after surgery also demonstrating early autofluorescence (B) and later blocking with surrounding staining (D).

both eyes with mild distortion in the left eye due to the subfoveal lesions. The deep retinal/choroidal lesions in both eyes had improved as compared to the immediate post-operative period but were still evident on fundus examination (Fig. 1C and D) and OCT (Figs. 4 and 5). At the most recent follow-up (16 months post-operatively in the right eye and 12 months post-operatively in the left eye) the visual acuity continued to be 20/20 uncorrected in both eyes.

3. Discussion

Descemet's membrane endothelial keratoplasty (DMEK) is an increasingly popular corneal surgery for the anatomic replacement of diseased corneal endothelium that is both effective and safe.[10] In the present case report, we describe a patient with Fuch's endothelial dystrophy who developed placoid choroidopathy, RPE disruption, subretinal fluid and mild vitritis of unknown etiology after undergoing

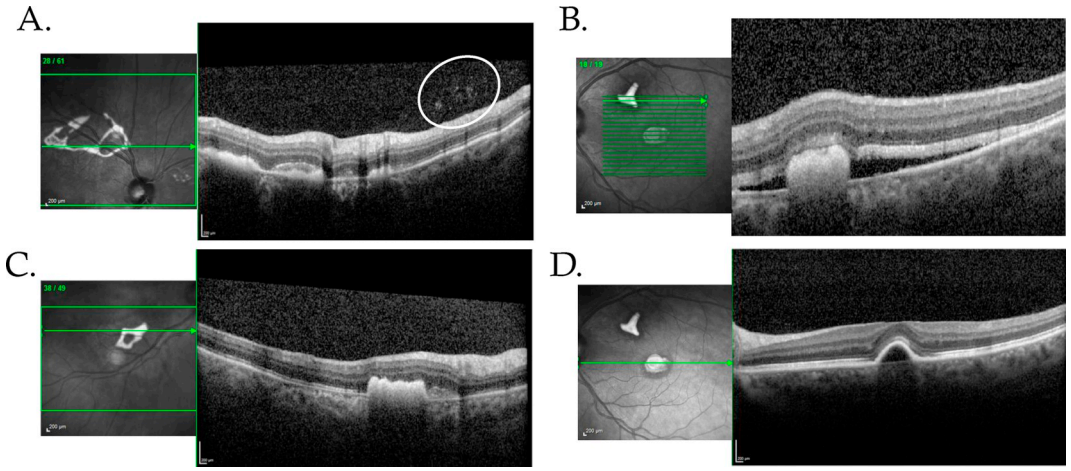

Fig. 3. Optical coherence tomography (OCT) of the right eye 4 weeks after surgery with slices through the superior (A) and inferior (C) arcades showing deep retinal/choroidal placoid lesions, retinal pigment epithelium attenuation (RPE) and overlying vitritis (white circle). Ten days after surgery in the left eye, OCT showed a similar deep retinal/placoid lesion with RPE attenuation and sub-retinal fluid (B) and a sub-foveal serous pigment epithelial detachment (D).

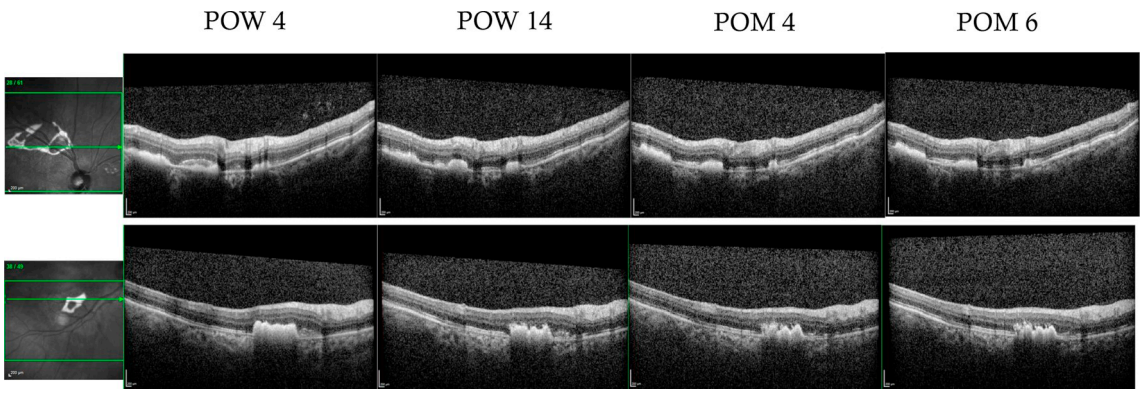

Fig. 4. Serial optical coherence tomography (OCT) images of the right eye showing changes over time of the superior (top row) and inferior (bottom row) lesions.

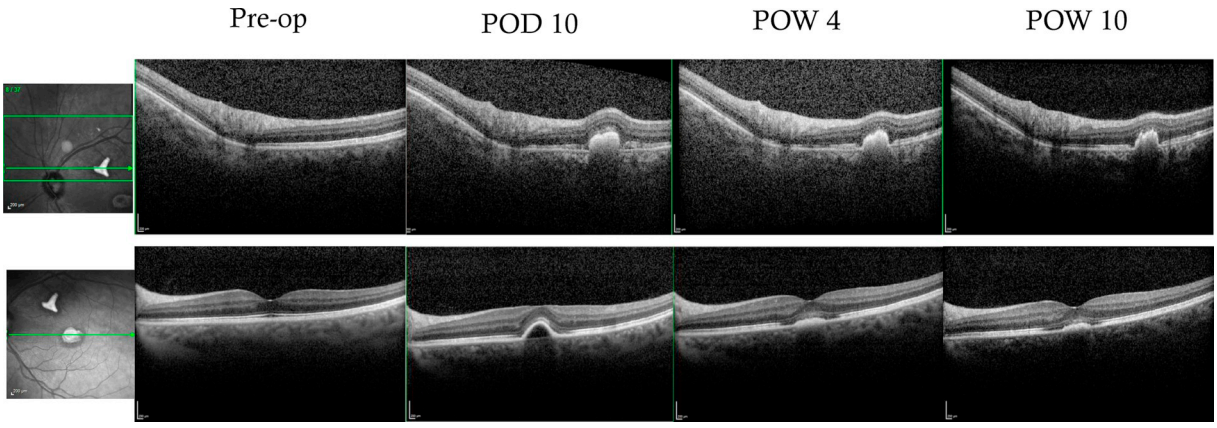

Fig. 5. Serial optical coherence tomography (OCT) images of the left eye showing changes over time of the superior placoid lesion (top row) and serous pigment epithelial detachment (bottom row).

DMEK combined with cataract surgery in both eyes. To our knowledge, this is the first reported case of choroidopathy/chorioretinitis after DMEK.

Our patient had no known history of prior retinal or choroidal disease and her DMEK surgery was uncomplicated in both eyes. The differential for her retinal findings included inflammatory (Vogt-Koyanagi-Harada syndrome, sympathetic ophthalmia, sarcoidosis, acute posterior multifocal placoid pigment epitheliopathy) and infectious (syphilis, tuberculosis, Lyme) etiologies, all of which were excluded by laboratory testing, examination/imaging or clinical course.

Although the etiology of our patient's findings is unknown, it is possible that she had an underlying choroidopathy that was somehow exacerbated by intraocular surgery, as routine as it was. However, because of the sequential nature of her surgeries and symptoms, it is clear that there were no sub-clinical retinal or choroidal findings present pre-operatively in the left eye, which underwent extensive pre-operative imaging due to the right eye's complicated post-operative course. It is also conceivable that an elevated post-operative intraocular pressure could have caused a choroidal blood flow disturbance leading to the described chorioretinal lesions. On review of the patient's post-operative course, we found that intraocular pressure ranged from 13 to 19 mmHg in the right eye and 15–16 mmHg in the left eye and there was a consistent absence of signs or symptoms of elevated intraocular pressure including shallow anterior chamber, retinal vascular changes, periocular pain/headache or nausea/vomiting.

Another possible etiology includes microscope light-induced retinal phototoxicity, which has been described after a variety of ophthalmic procedures including full-thickness penetrating keratoplasty combined with extracapsular cataract extraction and intraocular lens implantation.[11] Features of retinal phototoxicity include lesions at the level of the retinal pigment epithelium (RPE) with or without retinal edema, early leakage on fluorescein angiography and the development of RPE atrophy over time.[12] Although our patient had RPE changes and atrophy, both eyes also had features inconsistent with phototoxicity, including overlying vitritis, absence of leakage on FA and achievement of 20/20 visual acuity in both eyes at the most recent follow-up. Risk factors for retinal phototoxicity include length of surgery and use of photosensitizing medication, among others.[13] Although significant time can sometimes be required intraoperatively for graft/air bubble positioning,[14] both of our patient's surgeries were performed by an experienced, fellowship trained DMEK surgeon (CSS) in about 60 minutes. Nonetheless, retinal phototoxicity has been reported to occur with a duration of light exposure as short as 15 minutes.[15,16] In addition, although the operating microscope for both cases was inspected for eccentric light sources, it is possible that our patient's pre-existing superior and inferior peripheral iridotomies may have served as intraocular eccentric light sources leading to phototoxicity.[17] Interestingly, our patient was on lutein (20 mg daily) and astaxanthin (12 mg daily) as dietary supplements, both of which are carotenoids thought to prevent light-induced retinal degeneration through their anti-oxidant properties.[18–20]

To date, both DSAEK and DMEK have proven to be effective and safe options for the surgical management of corneal endothelial dysfunction. Although rare, posterior segment complications have been reported after these surgeries, including posterior DSAEK graft dislocation leading to proliferative vitreoretinopathy with retinal detachment[5] and cystoid macular edema after DMEK.[7,8] To the best of our knowledge, this is the first report of placoid choroidopathy and chorioretinitis following DMEK, or any form of endothelial keratoplasty surgery.

In the present case, we report the development of choroidopathy/chorioretinitis after sequential, bilateral uncomplicated DMEK surgery. Over the course of 16 months of follow-up in the right eye and 12

months of follow-up in the left eye, our patient has continued to show slow resolution of her retinal lesions and has maintained 20/20 uncorrected vision in both eyes. Our patient's course emphasizes the need to be aware of posterior segment complications following endothelial keratoplasty.

4. Conclusions

Placoid choroidopathy/chorioretinitis is a potential complication following DMEK surgery and should be maintained on the differential for patients reporting scotomas, photopsias or decreased visual acuity post-operatively.

Patient consent

Written patient consent to publish the described case was obtained.

Declaration of competing interest

The authors have no conflicts of interest to disclose.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2020.100610.

Funding

Research to Prevent Blindness Grant to the Weill Cornell Department of Ophthalmology.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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