Case report: Excimer laser phototherapeutic keratectomy (PTK) in gelatinous drop-like dystrophy.

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***Background**: Gelatinous drop-like corneal dystrophy (GDLD) is a rare corneal dystrophy, characterized by amyloid deposition in subepithelium and stroma of cornea, causing reduction in vision, photophobia and irritation because of irregularity in corneal surface. Excimer Laser Phototherapeutic Keratectomy (PTK) using Argon-fluoride laser is used to reshape, smoothen the corneal surface and removal of corneal opacity with minimal adverse reaction to the surrounding corneal tissue. A 19 year-old patient complained of irritation in both eyes for 3 months. She noticed whitish nodules on her cornea for about 5 years. After which she gradually suffered a progressive loss of her vision on both eyes.*

***Results**: right eye BCVA is 20/70, and counting fingers at 1 foot on left eye. Slit lamp examination revealed 2 grayish protruding subepithelial nodules at the central cornea of the right eye and multiple protruding subepithelial nodules that coalesce to a large lesion at the central cornea of the left eye. The patient was diagnosed as GDLD by clinical appearance. PKP was planned to restore vision, however the queue in Thailand for the procedure was 4-5 years. Thus Excimer laser PTK and fluid masking technique was performed for relieving the symptoms and temporary restoring the vision. Left eye laser setting was 6.8 mm zone of ablation with 45 microns in depth, performed after epithelium removal, the material deposit was reported as amyloid material.*

***Discussion**: The patient showed improving visual acuity and decreasing in eye symptoms following treatment. Duration of follow up was 2 months at time of writing with no clinical sign of recurrence of disease after laser operation.*

***Keyword:** Excimer Laser Phototherapeutic Keratectomy, PTK, Gelatinous drop-like corneal dystrophy, GDLD, PKP*

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Background

Gelatinous drop-like corneal dystrophy (GDLD) is a rare corneal dystrophy, characterized by deposition of amyloid in subepithelium and stroma of cornea. Clinical manifestation of GDLD usually present in first 2 decades of life with bilateral multiple white gelatinous subepithelial nodules (mulberry-like configuration) or band keratopathy-like lesion. With time, superficial vascularization has occurred and the lesion progresses to become larger and deeper which cause stromal nodules (kumquat-like lesion) and /or stromal opacifications

GDLD is classified as category I by IC3D Classification of Corneal Dystrophies [2], in which causative gene is Tumor-associated calcium signal transducer 2 (TACSTD2, previously M1S1), located in chromosome 1(1p32)[3]. Approximately about 25 mutations are reported on TACSTD2 gene. Loss of TASCSTD2 gene function resulting in loss of epithelial tight junction, allow the tear fluid influx into corneal tissue, and then lactoferrin in tear fluid turns to amyloid deposition in corneal tissue [4].

On Histopathology, GDLD is characterized by deposition of amyloid in subepithelial and stromal layer, area of epithelial hyperplasia and epithelial atrophy[5], destruction of Bowman's layer, stromal fibrosis and neovascularization. Descemet's membrane and endothelium are clearly visible. Transmitting electron microscopy reveals disruption of epithelial tight junction and amyloid deposit in basal epithelial layer.

GDLD can cause significant reduction in vision, photophobia and irritation because of irregularity in corneal surface. Wearing of soft contact lens in GDLD is proposed to relieve the ocular irritation. Surgical treatments are performed for visual restoration and rehabilitation including various type of keratoplasty. Limbal stem cell transplantation combined with keratoplasty or superficial keratectomy is an alternative option of tratment. However, recurrence after surgery is common within few years.

Boston type I keratoprosthesis[6] could be an option for advanced case of GDLD which required multiple keratoplasty.

Excimer Laser Phototherapeutic Keratectomy (PTK) using Argon-fluoride laser, emitting pulse at 193 nanometers is used to reshape, smoothen the corneal surface and remove corneal opacity with minimal adverse reaction to the surrounding corneal tissue. This technique has been used in many corneal disorders including recurrent corneal erosion, Reis-Buckler's dystrophy, anterior granular and lattice dystrophy. However it is not indicated in deep stromal lesions.

Recurrence after PTK is common in corneal dystrophy and may require repeating in PTK or corneal transplantation.

Case report

A 19 year-old patient presented to our eye clinic complaining of irritation on both sides of her eyes for 3 months. She denied any previous history of ocular trauma and surgery in addition to any underlying medical conditions. She also denied any family history of corneal

dystrophy and reported none of her relatives had the same symptoms. She has mild degree of eye irritation on both sides for about 10 years, and has noticed some whitish nodules on her cornea for about 5 years. After noticing the white nodules on the cornea, she reported a gradual progressive loss of vision on both eyes.

On ophthalmic examination, her right eye visual acuity is 20/100 which improved to 20/70 after pinhole correction and counting fingers at 1 foot on left eye which is not improved with pinhole. Slit lamp examination revealed 2 grayish protruding subepithelial nodules at central cornea on right eye and multiple protruding subepithelial nodules that coalesce to be a large lesion at central cornea on left eye. No epithelial defects on both eyes. Normal anterior chamber depth, lens and pupillary light reaction.[3]

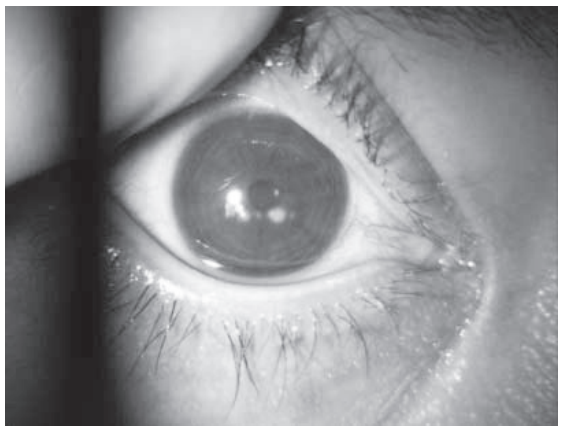

Fig 1: Right eye of patient demonstrates grayish subepithelial deposition at central cornea

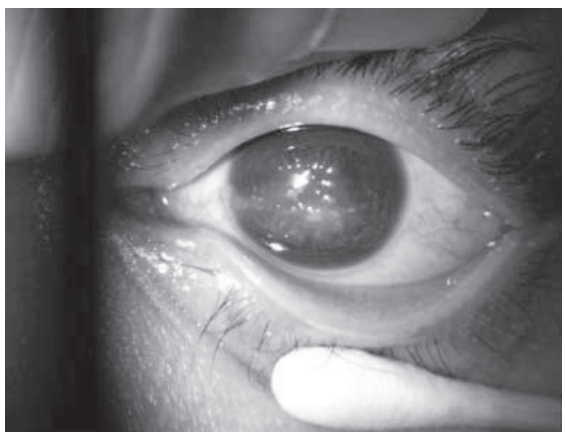

Fig 2: Left eye of patient demonstrates multiple grayish subepithelial deposition with haziness of central cornea

The patient was diagnosed as having gelatinous drop-like corneal dystrophy based upon clinical appearance. Plan of treatment is to perform penetrating keratoplasty to restore vision. But in Thailand, the waiting time is at least 4-5 years in surgical listing queue to receive the donor-cornea for transplantation. Thus Excimer laser PTK and the fluid masking technique were performed to temporarily restore vision and relieve the symptoms.

A decision was made to perform the laser operation on her left eye first and the laser setting was 6.8 mm zone of ablation with 45 microns in depth, performed after epithelium removal. Operation was successfully completed without complications. The material deposit on the cornea which we sent to identify was reported back as amyloid material.

One week after the laser operation, the corneal epithelial defect closed, left eye visual acuity has improved to 20/200 and 20/70 with pinhole correction. Her eye symptoms improved significantly, in particular, the symptom of photophobia has almost completely subsided.

Discussion

This is a case of a young female patient who presented with visual loss, eye irritation, photophobia and grayish subepithelial nodules on cornea of both eyes. A diagnosis of Gelatinous drop-like dystrophy of cornea was made. The management plan was to perform penetrating keratoplasty. Whilst waiting for the surgical listing queue, the patient received excimer laser PTK to relieve the eye symptoms and temporarily restore vision.

GDLD is a rare corneal dystrophy occurring in the young population. It still proves a challenge to today's ophthalmologists in diagnosis and providing treatment. Almost every patient will experience a recurrence within a few years following keratoplasty and superficial keratectomy techniques, thus patients may need multiple surgical intervention.

From literature review, there were few reports about excimer laser PTK for treatment of primary GDLD. T. Yamaguchi [8] reported direct ablation with excimer laser on the surface of cornea produces more surface irregularities. While Mouamen M. Seleet report lesion peeling and Phototherapeutic Keratectomy (PTK) and followed by Mitomycin C application appeared to be effective for GDLD for 6 months. Due to the rarity of disease, only reports in small case series with limited time of follow-up period are available.

Sustainability of treatment was also questioned, from case series reported by Shimazaki,[9] GDLD patients who received keratoplasty almost all developed subepithelial haziness on the graft's location within 1 year and amyloid deposition typically recurred within a few years.

This case demonstrated improving visual acuity and decreasing in eye symptoms following excimer laser PTK with fluid masking technique. Duration of follow up is currently about 2 months with no clinical sign of recurrence of disease after laser operation.

Conclusion

Excimer laser PTK appears to be effective for treatment of primary Gelatinous drop-like corneal dystrophy with good initial response in visual improvement and relieving of eye symptoms.

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