Conjunctivo-limbal Autograft for Climatic Droplet Keratopathy with Ulcerative Keratitis: A Case Report

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Climatic droplet keratopathy (CDK) is one of the most frequent degenerative corneal diseases with high prevalence rates, especially in equatorial and polar regions. The profound hypesthesia observed in some cases with advanced CDK may contribute to the occurrence of trophic–corneal ulceration, rapid bacterial over infection, difficulty of healing and relentless evolution to perforation. Superficial keratectomy, lamellar or penetrating keratoplasty and excimer laser phototherapeutic keratectomy are various surgical options that have been tried in the management of CDK involving the visual axis, impairing the vision. We attempted conjunctivo-limbal autograft (CLAG) in a patient of advanced CDK with corneal ulceration and studied the outcome at the end of 6 weeks and found a better corneal epithelial regeneration post CLAG.

Keywords: Autograft, Climatic droplet keratopathy, Hypesthesia, Phototherapeutic-keratectomy

June 2014 with the chief complaints of pain and watering in the left eye. He had a history of diminution of vision in left eye since 1 year for which he underwent cataract surgery of the left eye in a camp about 3 months prior to presentation; with inadequate gain of vision post-operative in the left eye.

On examination of right eye, vision was 6/36 with pinhole improvement to 6/12. On the slit lamp bio microscopy upper lid showed multiple concretions in the palpebral conjunctiva. Cornea showed typical yellowish, elevated nodular lesions suggestive of CDK extending from corneal mid-periphery to limbus, from 4 o’clock to 8 o’clock position inferiorly. Lens showed early cataractous changes. Corneal sensation was normal.

On examination of the left eye, vision was finger counting at 1 m with no pinhole improvement. On slit lamp bio microscopy upper lid showed multiple concretions. Cornea showed typical yellowish, elevated nodular lesions suggestive of CDK extending from corneal mid-periphery to limbus, from 4 o’clock to 8 o’clock position inferiorly, there was an ulcer with dense fibrous scar at the base with vascularization, measuring about 2 mm × 2 mm × 1 mm depth extending from 6 o’clock to 4 o’clock position. The limbal conjunctiva adjoining the plaque showed pigmentary and papillomatous changes (Figure 1). Corneal sensation was normal. Pseudophakia with posterior chamber intraocular lens in the bag observed. Pre-operative keratometry showed extremely

INTRODUCTION

Spheroidal degeneration of the cornea is a degenerative condition whose exact etiology remains unclear. It is known by a number of other names, including bietti corneal degeneration, labrador keratopathy, climatic droplet keratopathy (CDK), droplet degeneration of the cornea, fisherman’s keratopathy, Eskimo corneal degeneration and elastotic degeneration of the cornea and is especially common in parts of the world with high levels of exposure to ultraviolet (UV) radiation, either from environmental or vocational predilection or both, plays a causative role in the etiology of spheroidal degeneration.1,2 Chronic exposure to UV radiation, especially when reflected from snow or sand, is widely regarded as the primary causative factor in spheroidal degeneration. High levels of reflected UV radiation have been demonstrated in the geographic regions where the condition is most prevalent. Temperature extremes, repeated corneal epithelial micro trauma and inadequate corneal hydration are currently thought to the pathogenesis of spheroidal degeneration.3

We are reporting a case of CDK with a non-infective ulcer, wherein we attempted conjunctivo-limbal auto grafting (CLAG) with superficial keratectomy for the non-infective ulcer.

CASE REPORT

A 65-year-old male patient presented to the Eye Department at Sri Siddhartha Medical College, Tumkur in the month of

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irregular broken mires due to the ulcerative plaque and spheroidal degeneration.

Schirmer’s and tear film break-up time (TBUT) revealed moderate dry eye in both eyes. Rest of the examination was essentially normal in both eyes.

Infective keratitis was ruled out. Swab was taken from the base and the margins of the ulcer, which was reported negative for any bacterial organisms and fungal elements.

Patient was posted for excision of the pathologic conjunctivo-limbal junction with superficial keratectomy of the ulcerative plaque area. The necessary pre-operative work up and consent for the procedures were done as per the hospital protocol. The operative procedure was done under local anesthesia. The palpebral conjunctiva was cleared off the concretions. Lamellar keratectomy was done along with removal of CDK spherules and the fibrotic plaque at the base was dissected (Figure 2a). The dissection was extended up to the adjoining limbal area; about 3 mm × 1 mm tissue was excised and sent for histopathological examination (HPE). CLAG was harvested from the superonasal area about 4 mm × 2 mm in size with deeper dissection at the actinically unexposed limbus (Figure 2b) and transplanted to the dissected area. Graft was secured with 10-0 nylon sutures, three interrupted sutures. Post-operatively patient was put on topical non-steroidal anti-inflammatory drug bromfenac 0.09% eye drops 4 times a day along with lubricant carboxy methyl cellulose 1% eye drops 4 times a day for 4 weeks.

The HPE report confirmed the presence of eosinophilic droplets in Bowman’s membrane and anterior stroma (AS) suggestive of CDK (Figure 3a and b).

Post-operative follow-up was done for 6 weeks.

Day 1 post-operative slit lamp biomicroscopy showed graft in situ with no sub graft edema (Figure 4a); 3 mm × 3 mm irregular area of fluorescein staining, extending from paralimbal to central cornea corresponding to the area of corneal epithelial defect (Figure 4b). Visual acuity was finger counting at 1 m.

In subsequent follow-up on day 15, the fluorescein positive area of the cornea was reduced in size with good healing of the cornea and graft margins regularly merged with the recipient conjunctiva (Figure 5a), suggesting a good take up of the graft. The keratometry values improved with regularized mires (Figure 5b). The visual acuity improved to 5/60 on snellen visual acuity chart.

At the end of 6 weeks, there was near total epithelial healing, leaving a small area of fibrous plaque of
about 1 mm × 1 mm in the 5 o’ clock mid-periphery (Figure 6a). The epithelium showed a recurrence of a small CDK spherule at the edge of the keratectomy area, in the periphery (arrow head in Figure 6a). Mottled fluorescein staining was observed (Figure 7a). On oblique illumination, the corneal epithelium showed near total healing with focal irregularities probably due to stromal thinning (Figure 6b). The best corrected visual acuity at the end 6 weeks was 6/60 improving to 6/24. The keratometry readings are as shown in Figure 7b. The comparative keratometry reading are shown in Table 1.

### DISCUSSION

CDK one of the most frequent degenerative corneal diseases with high prevalence rates especially in equatorial and polar regions.3

The photo disruptive effect of the UV component of the arc of light on corneal tissues depends on the length of time of exposure as well as distance from the arc of light.4 Repeated severe UV exposure over a limited period of time does initiate events resulting in extensive elastotic degeneration of the anterior corneal stroma in an interpalpebral distribution.5 CDK is characterized by the progressive opalescence of its most anterior corneal layers. Grading of the disease:

Grade I - There is appearance of multiple confluent, tiny, and translucent droplet-like sub epithelial deposits is observed primarily in the vicinity of the nasal and/or temporal comeoscleral limbus, leaving a clear fringe between the limbal conjunctiva and the affected area.

Grade II - The opalescence extends slowly throughout the interpalpebral area toward the center of the cornea in a band-shaped distribution, giving the cornea a misty or tarnished appearance. At this stage, visual acuity may be moderately or severely decreased. Small round, oval, or geographically well-demarcated “islands” of apparently normal clear cornea may be seen within the compromised areas.

Grade III - In more advanced stages of the disease, several large yellowish droplets or vesicles appear in the affected areas, some of them protruding beneath the corneal epithelium. They may be distributed in clusters of confluent vesicles over, or in the vicinity of, vascularized or non-vascularized plaques of AS opacity.6

Histopathologic examination of CDK tissue under a light microscope often demonstrates globular deposits of different sizes under the corneal epithelium, within Bowman’s membrane and the AS.7 The coalescence and increased volume of these spherules may cause disruption of Bowman’s membrane and elevation and thinning of the corneal epithelium.6,8 Histochemical analysis, the deposits stained lightly with eosin, minimally with periodic acid-Schiff, and demonstrates extracellular deposition of a complex of proteins including tryptophan, cystine, cysteine, and tyrosine. They stain positively for fibrin.1

The lesions are first seen at the limbus in the 3 and 9 o’clock positions.
And are restricted to the interpalpebral zone. They may be separated from the limbus by a clear zone, which is often lost as the condition progresses. The epithelium and its basement membrane remain unaffected unless advanced degeneration is present, whereas theBowman membrane is often disrupted or focally absent. No inflammatory cells are seen. Though the source of the material forming the spherules remains unknown, there is some evidence that the spherules form from an accumulation of plasma proteins, possibly immunoglobulins and albumins, which diffuse into the cornea - From the limbal circulation and are modified by UV radiation. This mechanism would explain the initial location of the lesions at the limbus as well as their proteinaceous composition.

The recent studies show that the matrix metalloproteinases (MMPs) have a role in the disruption of Bowman’s membrane in CDK. Under normal conditions, MMPs are expressed at relatively low levels, and their tight regulation gives way to up-regulation only when degradation is required. The authors recently reported that in human reflex tears collected from CDK patients, the levels of gelatinases were increased, while their tissue inhibitors MMPs were decreased.

Electron microscopy has shown that globules are round, electron-dense and sharply demarcated structures, always surrounded by basement membrane material and adjacent disorganized collagen fibrils. Kaji et al. found immunoreactivity against advanced glycation end products (AGE) in the cornea of CDK patients when compared with normal cornea and other corneal diseases such as bullous keratopathy or band keratopathy. This finding led them to postulate that pathogenesis of CDK might be an aggregation of AGE-modified proteins as the cause.

They demonstrated by immunoperoxidase studies that the droplets accumulate in the areas of the highest plasma-protein concentration. This, together with the fibrinoid tinctural properties and the strong evidence against a keratin or collagen derivation as shown by both Johnson and overall and also by Klintworth, suggests that the droplets are likely to be denatured plasma proteins.

As viewed in confocal microscopy, CDK is a corneal degenerative disease characterized by its progressive opacity because of accumulation of globular deposits in Bowman’s layer (BL) and AS, as well as abnormal corneal sensitivity. As per confocal microscopy Grade I CDK was characterized by incipient changes in the BL and AS but normal sub-basal and stromal nerve plexus. Grade II CDK showed hyper reflectivity and globular non-reflective deposits in the BL and AS. In Grade III, the AS showed fibrosis with increment of diffused hyper-reflective deposits and large non-reflective deposits. Concomitant to these changes, there were an increased number of dendritic cells at the peripheral cornea and limbus. Early changes in CDK did not affect the sub-basal and stromal nerves, but the progression of the disease lead to a significant density decrease of sub-basal nerves, and some structural changes in stromal nerves, such as uneven thickness and irregular configuration, that might be responsible for corneal hyposensitivity found in advanced stages of CDK.

The profound hyposensitivity observed in some cases with advanced CDK may contribute to the occurrence of corneal ulceration, rapid bacterial over infection, difficulty of healing and relentless evolution to perforation. CDK patients presented severe dry eye disease, as was shown by the Schirmer’s II test, the TBUT test, and vital staining compared with the control group.

Superficial keratectomy, lamellar or penetrating keratoplasty, and excimer laser phototherapeutic keratotomy are various surgical options that have been tried in the management of CDK involving the visual axis, impairing the vision.

We attempted a conjunctivo limbo-corneal resection with a limbal conjunctival autograft from the superonasal conjunctival area for CDK lesions, considering that abnormal CDK deposits are from the limbal circulation. Hence replenishing the limbal stem cell from the actinally unexposed superior limbus could lead to the growth of a better corneal stroma and epithelium probably devoid of the CDK.

In our patient, the superficial lamellar keratectomy healed with regular corneal tissue improving the post-operative keratometry values following CLAG. The fibrotic scar, which was present pre-operative, showed partial resolution. There was a correlatable improvement in visual acuity and patient’s symptoms. In our case, the pre-operative corneal sensations were preserved, and recurrence of only single CDK spherule was seen at the end of 6 week’s post-operative. We plan to observe the patient for recurrence of CDK and dry eye status.

Confocal microscopy can give more information about the regeneration of the nerves and corneal epithelium following CLAG.

**CONCLUSION**

CLAG can be a simple, effective technique in restoring the normal corneal architecture, with improvement in visual and subjective and objective parameters.
REFERENCES


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