

Chandler's Syndrome: A Rare Diagnosis With Unusual Presentation

Rashmi Ranjan^{1*}, Alok Sati¹, Pankaj Kumar², Parthasarathi Moulick¹, Sandeep Gupta¹

¹Department of Ophthalmology, Armed Forces Medical College, Pune; and ²Department of Pathology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

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Abstract

We report a case of Chandler's syndrome with unusual presentation and successful treatment with descemet stripping endothelial keratoplasty (DSEK). A 36-year-old healthy male presented with unilateral corneal edema following trauma with iron nail. On examination, there was microcornea with cornea having increased thickness due to diffuse microcystic and stromal edema, and descemet folds along with a few bullae seen inferiorly without any epithelial defect in the left eye. The anterior chamber was shallow with no inflammatory signs and the optic disc had a cupping of 0.7:1 with thinning of neuroretinal rim with digitally raised intraocular pressure. Endothelial morphology revealed bizarrely shaped endothelial cells with blackout areas, pleomorphism (minimum cell area 689/maximum cell area 1600), polymegathism (CV-19), and reduced cell counts (CD-834) with normal indices in the right eye. With the above findings of unilateral corneal decompensation, glaucoma and abnormal endothelial parameters, a diagnosis of Chandler's syndrome was made and the patient was taken up for DSEK after failure of medical therapy. With raised intraocular pressure (IOP) and shallow anterior chamber, DSEK was challenging and done under perioperative intravenous mannitol cover. At one-month follow up, the cornea was compact and well adhered to the host cornea, and the IOP was well controlled under anti-glaucoma medications. At 6-month follow up, the patient had a visual acuity of 20/60 with +1 dioptre sphere and -1 dioptre cylinder at 60° and the IOP was well controlled on tapering dose of topical steroid and antiglaucoma medications.

Keywords: Chandler's syndrome, descemet stripping endothelial keratoplasty, corneal decompensation, ocular trauma

Introduction

Iridocorneal endothelial syndrome (ICE) is a unilateral disorder with epithelial transformation of corneal endothelium (1). There are three subtypes of ICE depending on the extent of ocular involvement, including essential iris atrophy, Cogan Reese syndrome and Chandler's syndrome. The latter affects young to middle aged individuals, and primarily females, but is also

reported in males (2). Presenting with corneal edema, majority land with secondary glaucoma due to outflow obstruction. Here, we report an unusual case of corneal decompensation in a young male, secondary to ocular trauma, and its successful management by endothelial keratoplasty. To the best of our knowledge, this report is second to the existing report in literature (3).

Case Report

A 36-year-old healthy male, reported with diminished vision in his left eye for 5 days following trauma with the base of an iron nail. Ocular examination showed visual acuity of 20/20 in the right eye and counting fingers close to face with accurate projection of rays in the left eye. Right eye examination revealed no abnormality except for microcornea. The left eye cornea was normal in shape with horizontal diameter of 9 mm, but had increased thickness due to diffuse microcystic and stromal edema, and descemet folds along with a few bullae seen inferiorly without any epithelial defect. Anterior chamber (AC) appeared shallow uniformly with normal contents (Figure 1). Posterior segment was hazily seen with a vertical cup to disc ratio of 0.7:1 with thinning of neuroretinal rim more inferiorly. The intraocular pressure (IOP) was raised digitally as applanation could not be done due to corneal edema.



Fig. 1. Preoperative cornea showing diffuse corneal edema.

Central corneal thickness (Pachette 2, DGH, USA) was 651 μ m in the left eye and normal in the right eye. Glaucoma evaluation in form of gonioscopy, goldmann applanation, optic disc analysis by optical coherence tomography (Cirrus HD-OCT, Carl Zeiss Meditec, Dublin, CA), and visual field analysis (Humphrey Matrix, Carl Zeiss Meditec, Dublin, CA) was normal in right eye, but unrecordable in left eye due to corneal edema. Specular microscopy (SP 2000P, Topcon, USA) of the left eye revealed bizarrely shaped endothelial cells with blackout areas, pleomorphism (minimum cell area 689/maximum cell area 1600), polymegathism (CV-19), and reduced cell counts (CD-834) (Figure 2), whereas right eye was essentially normal.

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*Correspondence author: Dr. Rashmi Ranjan, Department of Ophthalmology, Armed Forces Medical College, Pune, India. Mobile: +91-8412847080; E-mail: rashminishi@gmail.com

In view of the salient findings, i.e. the unilateral corneal decompensation, glaucomatous disc and abnormal endothelial parameters, a provisional diagnosis of Chandler's syndrome was made and the patient was started on topical hyperosmolar (NaCl 5%) and antiglaucoma medications (Timolol 0.5% and Brimonidine 0.2%). However, conservative management failed and it was decided to attempt for descemet stripping endothelial keratoplasty (DSEK) as the pathology was confined to the endothelium.

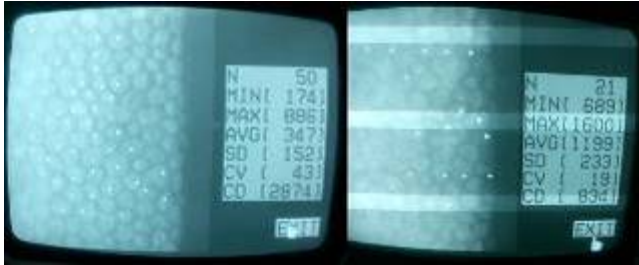


Fig. 2. Preoperative specular microscopy of the right and left eyes, respectively.

Performing DSEK was difficult due to raised IOP and shallow AC. Preoperatively, low IOP was achieved with intravenous mannitol and topical antiglaucoma agents. Surgery was performed under peribulbar anesthesia. Donor lenticule was prepared by manual dissection and trephined to 7 mm due to microcornea. Intracameral manipulation was difficult due to shallow AC, microcornea, and vitreous thrust. However, successful descemetorrhexis was achieved with help of cohesive viscoelastic (Hyopt 1%, Virchow Biotech, India) and donor lenticule was inserted using sheath glide. After repeated attempts, a partial air tamponade was managed with shallow AC peripherally.

On the first postoperative day, the AC was flat, but the lenticule was well adhered to the host cornea. Hence, AC reformation was attempted with air and fluid along with peripheral iridectomy and the patient continued on intravenous mannitol twice daily for a week. On the third postoperative day, AC was shallow and well formed (Figure 3). The patient was started on oral steroids (Prednisolone, 1mg/kg), topical steroids (Predacetate, 1%) in tapering doses and topical ocular hypotensives (Timolol, 0.5%; Brimonidine, 0.2%).

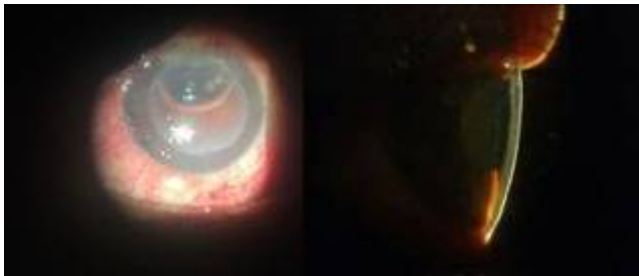


Fig. 3. Postoperative day 5 showing well adhered donor lenticule with formed but shallow anterior chamber.

At one-month follow up, the cornea was compact and the donor lenticule was well adhered to the host cornea with controlled IOP (Figure 4). At 6-month follow up, the patient had a visual acuity of 20/60 with +1 dioptre sphere and -1 dioptre cylinder at 60°. IOP was well controlled on tapering dosage of topical steroid along with anti-glaucoma medications.

Discussion

Unilateral persistent corneal edema without any epithelial defect raised our suspicion towards a list of causes like trauma, toxic endothelial insult, ocular surgery, inflammatory or infective process, and endothelial disorders like posterior polymorphous endothelial dystrophy or ICE syndrome.

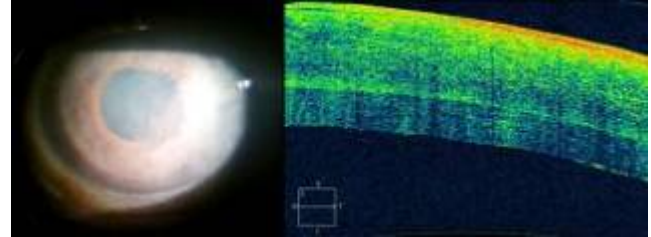


Fig. 4. Three months after operation showing compact cornea with formed anterior chamber.

Chandler's syndrome presents in young individuals with unilateral corneal edema in early stages and secondary glaucoma in late stages. Pathologically, metaplastic corneal endothelium proliferates over the AC angle leading to glaucomatous changes (4). The universal clinical sign is finely hammered silver appearance of all or part of the posterior corneal surface when viewed in specular reflection of slit-lamp. Specular microscopy of cornea reveals bizarre shaped endothelial cells and intracellular blackout areas with pleomorphism and polymegathism (5). We made the diagnosis of Chandler's syndrome primarily based on these findings and unilateral presentation of the disease.

Glaucoma is known to have an impact on corneal endothelium. Gangon *et al.* postulated that direct compression due to higher IOP damages corneal endothelium (6). Various literature supports that the patients of glaucoma with raised IOP have significantly lower endothelial cell counts (2370.5 cells/mm²) than normal matched population (2723.6 cells/mm²) (7). Considering the significant decrease in the endothelial count of 834 cells/mm², we concluded that there might have been additional damage to the metaplastic corneal endothelium with raised IOP.

Studies reveal that the corneal endothelium shows quantitative changes following blunt trauma with mean decrease in endothelial cell density of 6.4%. This gets compounded in the presence of angle recession by up to 12.2% (as against 1.2% in patients without angle recession) and may even decrease up to 21.2% if angle recession is greater than 180° (8). In the present patient, the impact of blunt trauma on corneal endothelium led to sudden corneal decompensation in an already compromised endothelium as evident by specular microscopic findings.

Usually these patients respond well conservatively, but for those with critical endothelial density of 800 cells/mm², curative treatment is to replace pathological endothelium with a healthy one. DSEK is favored as it is a sutureless surgery with lower graft rejection rate and most importantly, the endothelial disease being treated with a normal nonscarred cornea anteriorly. Even with high risk-profile for DSEK, the present patient was successfully managed and within a period of 6-month follow up, the patient showed significant improvement. This case report is unique in the sense that it highlights three factors i.e. metaplastic endothelium, raised IOP and corneal trauma that have led to corneal decompensation and its successful management by DSEK, a rare entity described in literature.

Conflicts of Interest: None

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