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Pre-Descemet's endothelial keratoplasty with glued intraocular lens implantation with pinhole pupilloplasty in a case of ocular comorbidity in achromatopsia

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Abstract:

Ocular comorbidities can happen as congenital defective gene associations. We present a 37-year-old female patient who was mentally challenged and had coexisting achromatopsia gene abnormality on genetic analysis. She was operated in childhood for congenital cataract, and posterior chamber intraocular lens (IOL) was implanted at 10 years of age elsewhere. The patient presented 27 years later with luxated IOL with endothelial decompensation. There was a coexisting steep and thin cornea noted on corneal topography. She was managed with pre-Descemet's endothelial keratoplasty with transpositioning of posterior chamber IOL to glued IOL with single-pass four-throw pupilloplasty. Postoperatively, the cornea was clear with centered glued IOL. The lesser postanesthetic challenges and faster rehabilitation are obtained in combination procedures with reduced complications in such rare scenarios.

Keywords:

Aphakia with corneal decompensation and keratoconus, glued intraocular lens implantation, pinhole pupilloplasty, pre-Descemet's endothelial keratoplasty, single-pass four-throw pupilloplasty, triple procedure

Introduction

Congenital cataracts are often treated by intraocular lens (IOL) implantation in primary or secondary sitting in the modern era.[1-3] The common complications in childhood cataract extraction include glaucoma, corneal decompensation, and amblyopia. Management of childhood cataract in a mentally challenged patient is even more difficult due to multiple comorbidities. We present a case of a mentally challenged young patient diagnosed with genetic disorder achromatopsia who was operated in childhood and had undergone corneal decompensation, and incidentally, there was an early keratoconus change on topography. As the IOL was luxated and there was secondary corneal decompensation, we wanted to manage the patient in a single sitting to avoid anesthetic and postoperative surgical complications. The patient was managed by triple procedure of pre-Descemet's endothelial keratoplasty (PDEK), glued IOL, and single-pass four-throw (SFT) pupilloplasty for faster rehabilitation.

Case Report

A 37-year-old female presented with the chief complaint of defective vision in the right eye since birth. She has underwent

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cataract surgery with IOL implantation at 10 years of age. She was a known case of mental instability since birth. The patient's visual acuity could not be accurately assessed due to her poor cooperation resulting from her mental disorder. On ocular slit-lamp examination, the right eye exhibited superior corneal bullae with diffuse edema with endothelial guttate changes suggestive of bullous keratopathy [Figure 1]. There was preexisting peripheral surgical iridectomy, subluxated posterior chamber IOL. The left eye showed a Grade 2 nuclear sclerosis cataract. Fundus examination was suggestive of bilateral retinitis pigmentosa characterized by peripheral "bone spicule" pigmentation, arteriolar narrowing, and optic disc pallor [Figure 2]. Specular microscopy showed low endothelial cell count with polymegathism and pleomorphism in the right eye and guttate changes in the left eye [Figure 2]. Scheimpflug Pentacam imaging revealed a steep cornea with high astigmatism and increased thickness due to corneal edema, which can mask the underlying thin cornea [Figure 3].

She was advised for IOL repositioning by glued IOL with PDEK in the right eye, followed by cataract surgery in the left eye. Genetic testing was done in view of multiple ocular manifestations associated with mental disorder, and it confirmed autosomal recessive achromatopsia with a mutation in the cyclic nucleotide-gated (CNG) B3 gene. The patient underwent IOL repositioning by glued IOL with PDEK and SFT in the right eye in a single sitting under general anesthesia.

Surgical technique

Informed consent was obtained from the guardian. Under sterile precautions with general anesthesia in the operating room, the anterior chamber entry was made with a side port knife and ocular viscoelastic device was injected. A 25-gauge transconjunctival trocar anterior chamber maintainer was placed at the inferotemporal conjunctiva. Superior limbal incision was made, and the anterior chamber was filled with viscoelastic device. Subsequently, anterior vitrectomy of the pupillary plane was performed. After making 2 partial thickness scleral flaps diagonally apart (180°), 2 sclerotomies were made 1.5 mm from the limbus below the flaps. Glued IOL was then performed using the conventional method [Figure 4a-c] by handshake technique to transpose the existing posterior chamber IOL into glued IOL.[4-6] The haptics were then externalized using the end-grasping forceps and placed into the intrastromal scleral tunnels. The pinhole pupilloplasty using SFT was then performed under direct visualization of pupil center and Purkinje image I intraoperatively with microscope [Figure 4d and e].[7,8] Once the pupil was reduced to 1.5mm size after SFT, it was confirmed by measuring (confirming the size) using an intraoperative pupil gauge (which has multiple holes of sizes from 0.5 to 3mm). The centration was adjusted under the operating microscope with respect to Purkinje image I.

Following glued IOL and pupilloplasty, the donor graft was then harvested using the regular PDEK technique using pneumatic dissection with the aid of 0.06% trypan blue

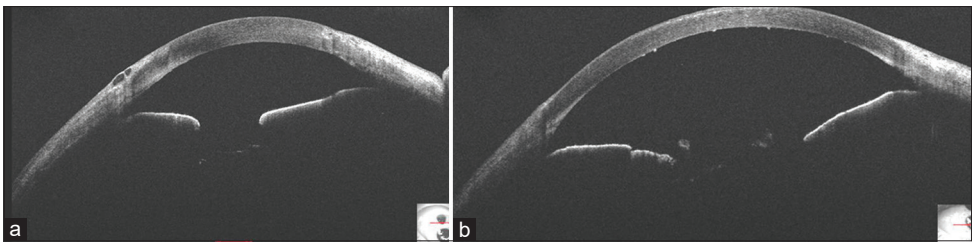

Figure 1: Anterior segment optical coherence tomography showing superior bullae (a) and thickened epithelium with luxated intraocular lens (b)

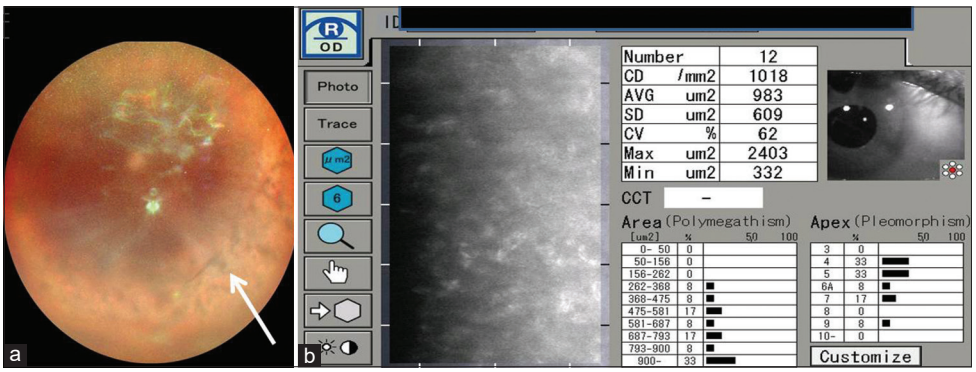

Figure 2: (a) Wide-angled fundus photograph of the right eye showing disc pallor with peripheral bony spicule (white arrow) and specular analysis showing endothelial polymegathism and low count (b)

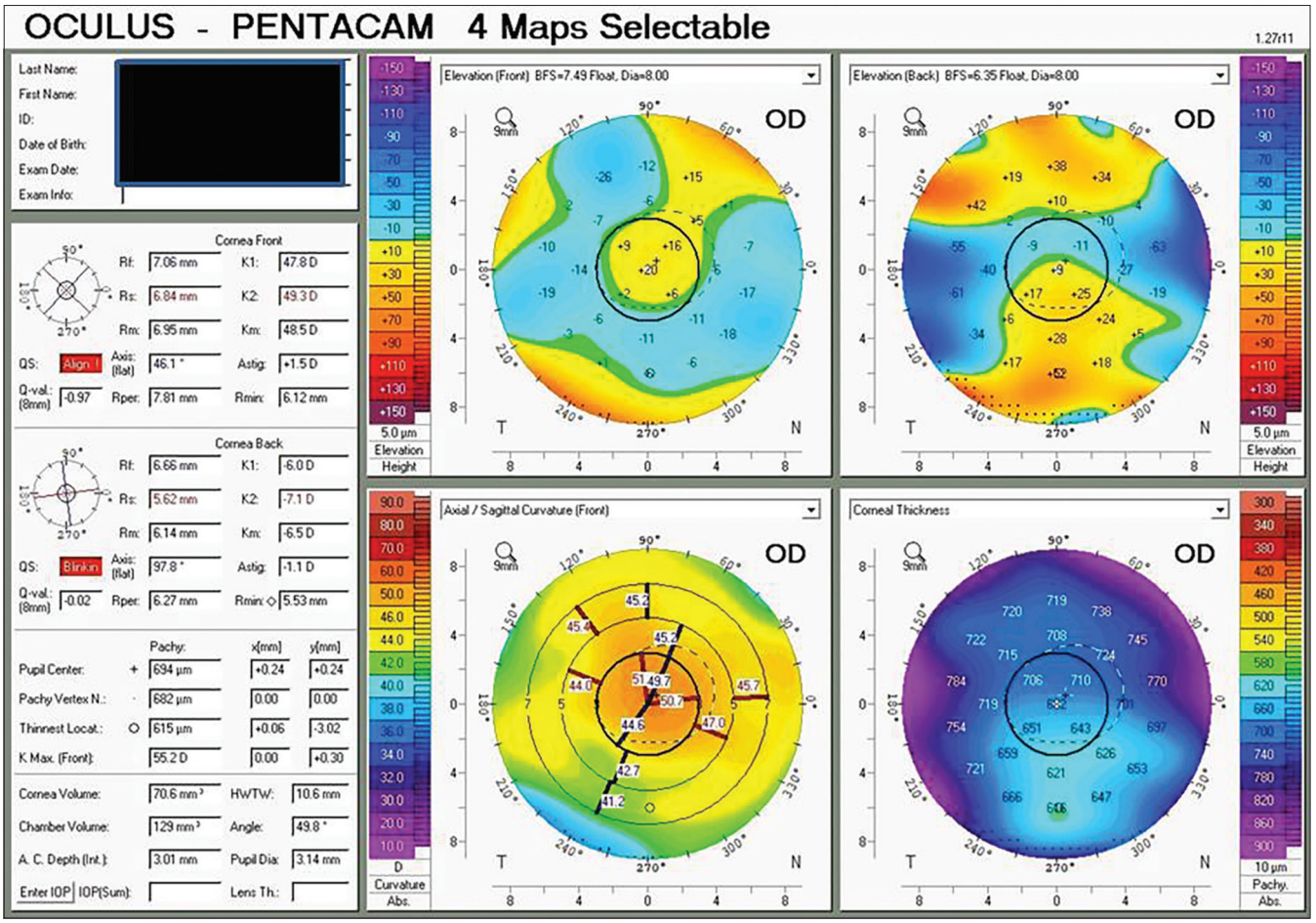

Figure 3: Preoperative pentacam imaging showing steep cornea with increased corneal thickness due to bullous changes

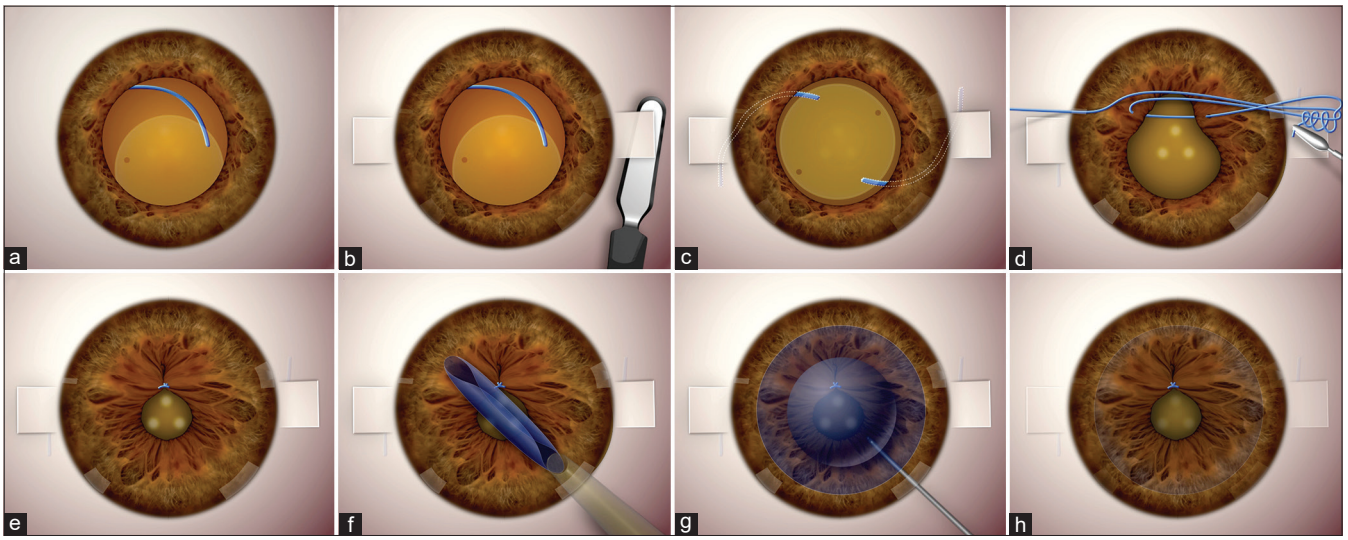

Figure 4: Surgical technique. (a) Preoperative luxated intraocular lens. (b) Two diagonal scleral flaps were made. (c) Intraocular lens transposed to glued intraocular lens by handshake method. (d) Single pass four throw pupilloplasty was performed using 10-0 prolene suture. (e) After pupilloplasty with centered pupil. (f) Donor pre-Descemet's endothelial keratoplasty graft injected into host anterior chamber. (g) Graft unrolled by pneumatic method. (h) Wound closed with air in place

dye.[9] The recipient's corneal Descemet's membrane was stripped by Sinskey's hook and removed. Subsequently, the donor PDEK graft was loaded into an IOL injector and inserted into the anterior chamber [Figure 4f]. The graft was centered under saline and unrolled by air [Figure 4g]. The corneal wound was sutured with 10-0 monofilament

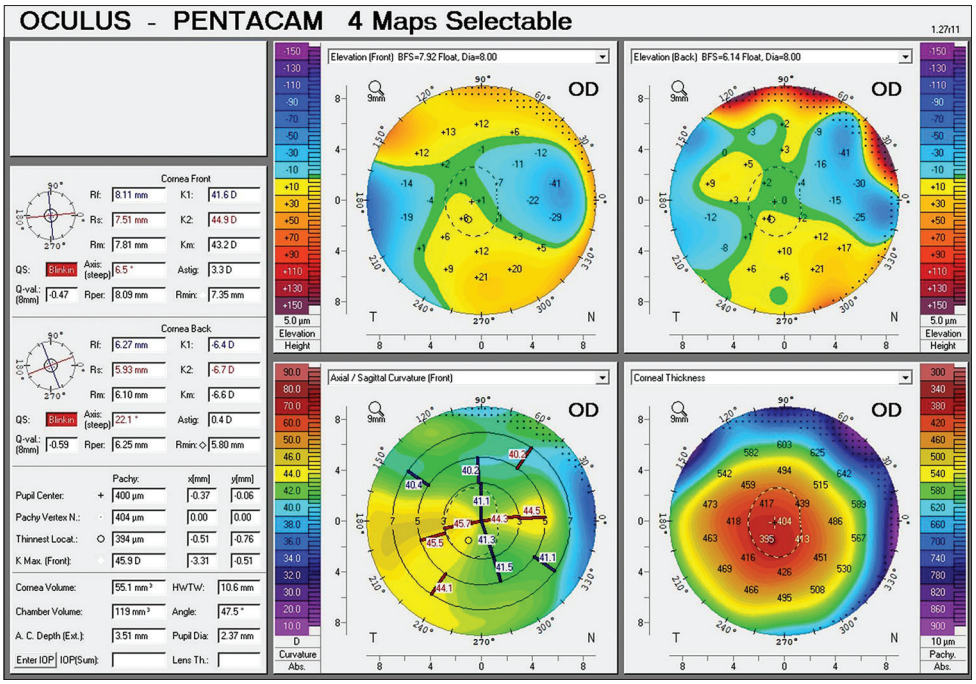

Figure 5: Postoperative Pentacam imaging showing a reduction in keratometry and corneal thickness

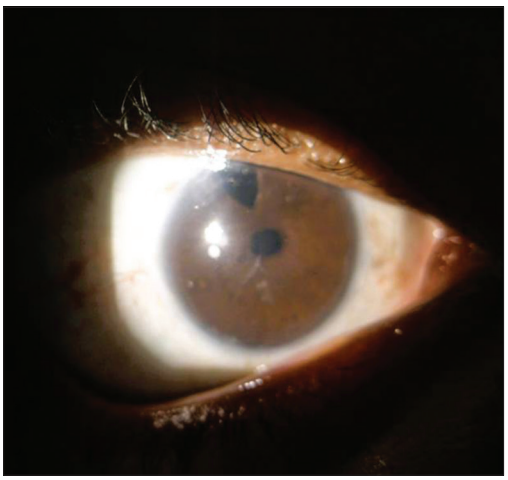

Figure 6: Postoperative slit-lamp photograph of the right eye showing clear cornea with pre-Descemet's endothelial keratoplasty graft *in situ* with surgical peripheral iridectomy and well-centered pupil and intraocular lens

nylon, and the patient was stabilized in supine position for an hour [Figure 4h]. Postoperatively, topical antibiotic and steroid combination drops were given and tapered over 4 weeks, while low-potent steroid drops were kept till 6 months. Postoperative topography [Figure 5], anterior segment optical coherence tomography, and higher-order aberrations were analyzed. Her postoperative period was uneventful, and she was able to perform her day-to-day activities independently [Figure 6].

Discussion

Achromatopsia is a rare, bilateral inherited retinal degeneration affecting all three types of cone photoreceptor cells, which results in reduced visual acuity, photophobia, hemeralopia, and severe loss of color discrimination. It is an autosomal recessive condition with the most common mutations affecting the genes that code for or regulate cone CNG channel subunits, including CNGB3 in 50% of cases and CNGA3 in 25% of cases, a smaller fraction of GNAT2, PDE6C, PDE6H, and ATF6.[10-12] Nystagmus is the first symptom often encountered within the 1st several weeks of life. They also have a high rate of hyperopia requiring spectacle correction. There are no systemic abnormalities associated with achromatopsia, and patients can expect a normal life expectancy. Electroretinogram is the gold standard tool in diagnosing achromatopsia showing diminished cone function and normal rod function. Currently, there is no approved treatment for achromatopsia. Subretinal adenoviral gene therapy targeting CNGA3 is under clinical trials.

This case report emphasizes the unique diagnostic and treatment challenges posed by the rare coexistence of keratoconus, endothelial decompensation, retinitis pigmentosa, and achromatopsia. The association of these conditions has not been extensively reported in the literature.[13] The coexistence of keratoconus and endothelial decompensation can mask the severity and progression of each condition due to their opposing effects on corneal thickness. Careful management is required, including genetic screening and counseling for patients in view of their guarded prognosis and long-term follow-up. Spectacle correction in milder forms is effective. However, in associated cataract and

corneal diseases, surgical intervention may be needed. Refractive surgery should be avoided, and cataract surgery should be performed with consideration of the stage and progression of the conditions.

Combined glued IOL and PDEK have been reported by us on complicated cases where deficient capsule and endothelial decompensation are coexistent.[14] However, here we combined IOL explantation due to luxated previous IOL and pinhole pupilloplasty to improve the quality of vision in the aberrated cornea. In pinhole optics, there have been prior reports on pinhole pupilloplasty showing satisfactory results in eyes with irregular astigmatism.[8] The technique of combining pinhole pupilloplasty helps filter the stray light from the periphery of the cornea in cases with higher-order corneal aberrations.[7,8] It is known that ocular aberrations are directly proportional to the pupil diameter. The other alternate options in keratoconus are toric IOL or contact lens. However, corneal intrastromal segments or contact lens may not be ideal for this patient due to poor mental stability and need for follow-up. Hence, near permanent procedure would be advisable in this scenario. The postoperative complications are less due to endothelial keratoplasty, and early rehabilitation is possible. In conclusion, early detection and appropriate intervention are crucial in achieving favorable functional outcomes in patients with this rare combination of eye disorders.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient guardian has given her consent for her images and other clinical information to be reported in the journal. The patient guardian understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Data availability statement

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

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Nil.

Conflicts of interest

The authors declare that there are no conflicts of interests of this paper.

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