Boston keratoprosthesis and Ahmed glaucoma valve for visual rehabilitation in congenital anterior staphyloma

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Congenital anterior staphyloma entails grave visual prognosis. The majority of reported patients have undergone enucleation. We report a promising result of staphylectomy with implantation of a keratoprosthesis and a glaucoma drainage device in a seven-month-old child with a large, congenital anterior staphyloma.

Key words: Congenital anterior staphyloma, glaucoma drainage implant, keratoprosthesis

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Congenital anterior staphyloma is a rare anomaly. The fate of most eyes has been enucleation. When confronted with an advanced form of this condition in the only eye of a child, we aimed intervention towards possible visual rehabilitation.

Case Report

A seven-month-old male baby was brought for protrusion of the right eye since birth. His birth history was unremarkable. There was no history of ocular discharge in the immediate postnatal period. The right cornea was enlarged (horizontal diameter 15 mm), opaque and protruding between the lids [Fig. 1]. Other details of the anterior segment were not discernible. He was unable to fixate light. Digital tension was normal. Tonopen (Medtronic Solan, Jacksonville, FL) intraocular pressure (IOP) readings were 48, 50, 50 mm Hg (≤ 5% standard deviation). The posterior segment was normal on B-scan ultrasonography. Axial length was 20.2 mm. The left eye was phthisical.

We prescribed topical antiglaucoma medication for the right eye. He underwent contact transscleral diode cyclophotocoagulation (inferior 180 degrees) then. Post-procedure Tonopen IOP readings were 21, 24 mm Hg (≤ 5% standard deviation).

Four months later, he underwent penetrating keratoplasty using 14-mm diameter donor corneoscleral button along with extra-capsular lens extraction and anterior vitrectomy. Intraoperative indirect ophthalmoscopy showed healthy optic disc and retina. Simultaneously, pediatric Ahmed glaucoma valve (Model FP 8; New World Medical Inc., California, USA) was implanted in the superotemporal quadrant [Fig. 2].

Histopathology of the corneal button showed irregularly thickened epithelium with epidermidization. Corneal stroma was scarred and vascularized. Bowman’s membrane, Descemet’s membrane and endothelium were absent. Uveal tissue was adherent to the posterior surface of the cornea.

Post-surgery IOP measured 14 mm Hg by Perkins tonometer (Haag-Streit, Essex, UK). The child underwent amniotic membrane transplantation for central, persistent epithelial defect. However, this progressed to a sterile central melt necessitating cyanoacrylate glue and bandage contact lens application following which healing was complete with subsequent graft failure. After confirming adequate control of IOP and gradual graft failure over a period of six months, aphakic Boston keratoprosthesis (Massachusetts Eye and Ear Infirmary, Boston, MA) was implanted.

Six months later, the child was fixing and following light with the right eye. Retinoscopy value was +2.50 diopters. The keratoprosthesis was well-integrated [Fig. 3]. Ahmed glaucoma valve was in situ. An elevated conjunctival bleb was noted. Digital tension was normal.

Discussion

Congenital anterior staphyloma is a rare developmental anomaly. It is postulated to be due to lack of mesodermal differentiation or intrauterine keratitis; either blood-borne or trans-amniotic. It is characterized by ectatic opaque cornea lined by uveal tissue and can be unilateral or bilateral. The fellow eye in unilateral cases may have microcornea, keratoconus, keratoglobus or corneal leucoma. In our case, the fellow eye was phthisical.

Histopathologic examination of eyes with congenital corneal staphyloma shows intact epithelium that can be keratinized secondary to exposure. Bowman’s membrane may be absent. If present, it is usually fragmented and atrophic. Descemet’s membrane and endothelium are usually absent indicating early onset of the pathological process. Our patient fulfilled the clinical and histological criteria of congenital anterior staphyloma.

Initially, limited diode cyclophotocoagulation was done to reduce IOP and thereby reduce the possibility of expulsive choroidal hemorrhage during penetrating keratoplasty. The keratoplasty allowed assessment of the visual potential and later served as tectonic support for the Boston keratoprosthesis. Large size of the (corneoscleral) graft, disorganized anterior segment and young age of our patient were poor prognostic
indicators of graft survival. The graft did fail eventually. However, the eye was moist, fornices were well-formed, blink response was good and the fellow eye had nil visual potential. Additionally, successful visual rehabilitation with Boston keratoprosthesis in pediatric patients with congenital ocular anomalies or failed corneal graft was recently reported.\textsuperscript{[5]} Therefore, we offered Boston keratoprosthesis. Congenital corneal staphyloma has grave visual prognosis. Most of the earlier reported cases presented late after birth to the treating ophthalmologist; the involved eyes had been blind and underwent enucleation.\textsuperscript{[3,6,7]} Earlier only Schanzlin et al.,\textsuperscript{[1]} reported successful optical corneal transplantation, but in a milder form of congenital anterior staphyloma. The cornea was normal in diameter, had a protruding dumbbell-shaped anterior staphyloma extending 9 mm anterior to the corneoscleral limbus. IOP was 18 mmHg. Axial length was 15 mm. They sutured an 11-mm corneal button in place. The corneal graft was clear at one year.\textsuperscript{[1]} Our case was more advanced. Lunardelli et al.,\textsuperscript{[8]} did corneoscleral transplantation for a large congenital anterior staphyloma. The graft failed. But, their intention was cosmesis as the eye had no light perception.\textsuperscript{[8]} Furthermore, Jacob et al.,\textsuperscript{[9]} performed corneoscleral grafting in a similar case of large anterior staphyloma but eventually the graft failed.\textsuperscript{[9]} Large, corneoscleral grafts have poor success rates. In case of failed corneoscleral graft, keratoprosthesis offers promise for visual rehabilitation once the ocular surface and IOP are stabilized.\textsuperscript{[5]} This, to the best of our knowledge, is the first reported attempt to salvage an eye having congenital anterior staphyloma using a keratoprosthesis and a glaucoma drainage implant.

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