

Congenital Corneal Staphyloma

Hossein Salour,¹ MD; Nasser Owji,² MD; Alireza Sadeghipour,³ MD

¹Labbafinejad Medical Center, Shahid Beheshti University, MC, Tehran, Iran

²Khalili Hospital, Shiraz Medical University, Shiraz, Iran

³Rassoul Akram Hospital, Iran Medical University, Tehran, Iran

A 15-month-old boy was referred to the Oculoplastic Clinic at Labbafinejad Medical Center for treatment of a bulging and deformed blind left eye. He was the result of a normal full-term pregnancy and there had been no ocular discharge at birth. Examination of the left eye revealed a keratinized and ectatic cornea bulging 9 mm anteriorly (Fig. 1). The right eye had a chorioretinal coloboma. Due to severe deformity of the blind eye, we considered enucleation to be the most appropriate treatment.

The enucleated left globe measured 38×21×20 mm; horizontal and vertical corneal diameters were 21×20 mm respectively. The opaque cornea showed surface keratinization and a cystic forward projection measuring 16 mm. The cornea was edematous and up to 4 mm in thickness. The anterior chamber angle was occluded by iris adhesions. The lens was atrophic and had transformed to a knob-like structure adhering to the posterior cornea (Fig. 2). However, the posterior segment and optic nerve appeared normal.

On microscopic examination, the cornea demonstrated diffuse ectasia and the corneal epithelium showed hyperkeratosis, parakeratosis, acanthosis and irregular elongation of rete pegs. Bowman's layer was thin or absent in many areas. The corneal stroma was edematous, vascularized and diffusely infiltrated by acute and chronic inflammatory cells. Descemet's membrane and the endothelium were completely absent (Fig. 3). The posterior aspect of the cornea was entirely lined by atrophic iris tissue obliterating the anterior chamber angle. The optic disk, retina and sclera appeared normal.



Figure 1 Keratinized corneal ectasia and severe bulging of the left eye at the time of enucleation.

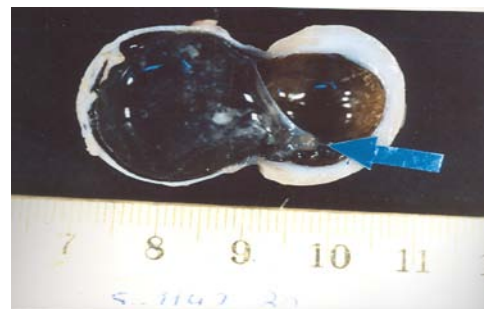


Figure 2 The cornea is edematous and up to 4 mm in thickness, the lens is transformed to a knob-like structure (arrow).

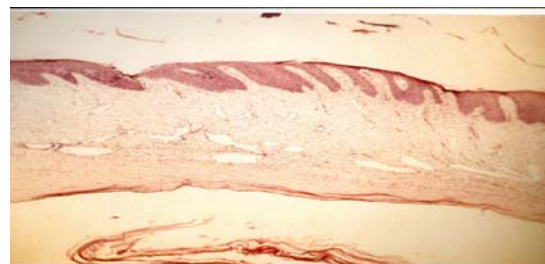


Figure 3 Diffuse corneal ectasia. The corneal epithelium is hyperkeratotic showing marked acanthosis. The stroma is edematous, vascularized and diffusely infiltrated by acute and chronic inflammatory cells.

DISCUSSION

Protrusion of an opaque cornea through the eyelids at birth is a very rare condition which may be due to keratectasia or congenital corneal staphyloma. These two conditions differ only in the presence of uveal lining of the cornea in congenital staphyloma. Keratectasia is probably not a developmental abnormality and seems to occur secondary to intrauterine keratitis or vitamin deficiency leading to corneal perforation.¹ The patient herein fulfills the clinical and histological definition of congenital corneal staphyloma.

Congenital corneal staphyloma is characterized by total corneal opacification, ectasia and forward protrusion between the eyelids.² The hereditary pattern of congenital corneal staphyloma is generally nonspecific and sporadic.³ The condition can be unilateral or bilateral;⁴ in unilateral cases, the unaffected eye frequently harbors another anomaly such as microcornea, keratoconus, keratoglobus, cornea plana, or corneal leukoma.² In the described case, the unaffected eye was colobomatous; an association not previously reported. The ectatic corneal protrusion is lined on its inner surface by uveal tissue causing the blue hue. Secondary angle closure glaucoma resulting from total anterior synechia leads to gradual bulging of the cornea. Corneal exposure can result in chronic keratitis, epithelial metaplasia and keratinization. Chronic corneal inflammation may also cause keloid formation in staphylomatous corneas⁵ and a dermis-like choristoma has also been reported.⁶

Histopathologic examination of congenital corneal staphyloma reveals severe corneal ectasia with the posterior aspect of the staphyloma lined by atrophic iris.^{7,8} The epithelium is intact but may show keratinization secondary to exposure.³ Bowman's zone is usually present, but may be fragmented,³ thin and atrophic.⁴ The corneal stroma is hypercellular, disorganized and vascularized.³ Inflammatory cells have been absent in the stroma of the staphylomatous cornea in many cases.⁴ Descemet's membrane and endothelium are typically absent.^{3,4} The posterior segment is normal, except for changes due to concomitant glau-

coma.³ Congenital corneal staphyloma is classified as a developmental abnormality and may resemble Peter's anomaly which is caused by abnormal migration of neural crest cells.⁴ On histopathologic examination of Peter's anomaly, Decrement's membrane and endothelium are absent at the site of the leukoma but corneal ectasia is an uncommon finding.⁹⁻¹¹ Considering the histopathologic similarities, congenital corneal staphyloma may represent a more extreme instance of mesenchymal dysgenesis as compared to Peter's anomaly.¹²

Congenital corneal staphyloma seems to entail a hopeless visual prognosis.⁶ The fate of most eyes has been enucleation, as has been the case with our patient, but some success has been reported with penetrating keratoplasty in milder forms of the disorder.³

Correspondence to: Hossein Salour, MD, Associate Professor of Ophthalmology, Ophthalmic Research Center, No. 5, Boostan 9 St., Amir Ebrahimi Ave., Pasdaran, Tehran 16666, Iran; Tel: +98 21 22585952, Fax: +98 21 22590607, e-mail: hosalour@hotmail.com

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