Case Report

A rare case of congenital corneal clouding with anterior staphyloma of the eye

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Abstract

There are only few case reports of congenital anterior staphyloma, an extremely rare condition. We report a case of congenital anterior staphyloma presenting as corneal clouding. A preterm girl baby was noted at birth to have bilateral corneal opacity with left sided microcornea. Ophthalmological evaluation revealed anterior staphyloma in right eye &total leucomatous corneal opacity in left eye. Despite antiglaucoma treatment, right sided bupthalmos & corneal thinning worsened. Corneal transplant was done at 7 months of age & baby improved symptomatically.Congenital anterior staphyloma occurs due to developmental aberration or secondary to inflammation. There are various genetic, metabolic, developmental & idiopathic causes of congenital corneal clouding like congenital glaucoma, birth trauma, peters anomaly, dermoid tumors, sclerocornea, infectious/inflammatory processes, metabolic causes & excess prenatal maternal consumption of alcohol. Complete evaluation of congenital corneal clouding include slit lamp biomicroscopy, funduscopy, tonometry, gonioscopy, ultrasonography, photo screening, ultrasound biomicroscopy & CT scanning in selected cases. Treatment is primarily surgical followed by management of amblyopia & optical therapy. Early penetrating keratoplasty, primary combined trabeculotomy-trabeculectomy & corneal grafting are associated with a favorable visual outcome. Early identification of ophthalmological problems in infancy & prompt intervention is mandatory for conditions like congenital corneal clouding.

Keywords: Congenital corneal clouding, Anterior staphyloma, Peter's anomoly

Introduction

Congenital anterior staphyloma is an extremely rare condition [1]. Parsons collected 15 cases over a period of 100 years [2]. Duke-Elder published 17 case reports [3]. Some cases have also been reported from India [4]. We are presenting a case of congenital anterior staphyloma presenting as corneal clouding.

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A 31 week preterm girl was delivered to a primi mother by LSCS due to decreased foetal movements. Immediate newborn examination revealed bilateral corneal opacity with left sided micro cornea. No history of fever, trauma, nutritional deficiencies or X-ray exposure during pregnancy was noted in the mother.

There was no family history of similar defect. General & systemic examination didn't reveal anything significant. Ophthalmological evaluation was suggestive of anterior staphyloma in right eye & total leucomatous corneal opacity in left eye. B-scan showed that both eyes were structurally normal inside. Despite treatment to reduce intraocular pressure, right eye bulged & cornea was thinned out. Corneal transplant (penetrating keratoplasty with extra capsular cataract extraction) was done at 7 months of age. Examination under general anaesthesia (post transplant) revealed that optic nerve was healthy & the Intraocular pressure was normalised. The progress of the eye is being closely & constantly reviewed.

Etiopathogenesis of congenital anterior staphyloma is controversial. Many attribute this condition to be inflammatory in origin [2,5], but some feel that staphyloma occurs due to developmental aberration- failure of mesodermal differentiation resulting in adhesion of cornea to iris[6]. Peters' anomaly causes corneal opacity due to developmental dysgenesis of anterior segment.

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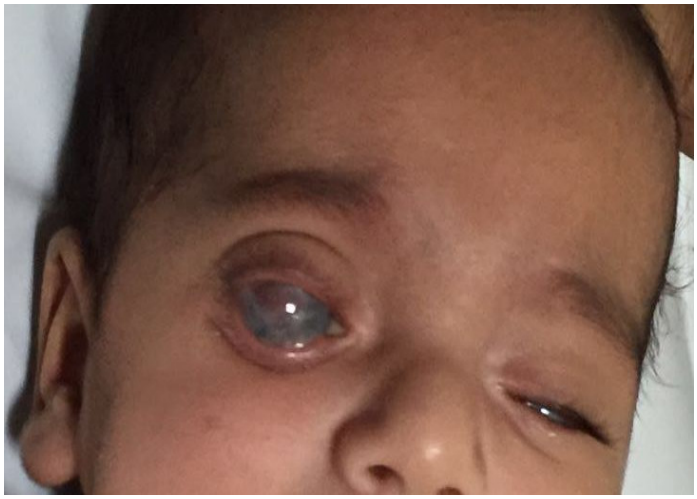

Various genetic, metabolic, developmental& idiopathic causes can result in congenital corneal clouding like congenital glaucoma, Birth trauma, Peters anomaly, Dermoidtumors, Sclerocornea, Congenital hereditary endothelial dystrophy, Infectious/ inflammatory processes, Ulcers due to Viral keratitis (Herpetic keratitis, Rubella keratitis), Tears in descemet membrane secondary to birth trauma or congenital glaucoma, Metabolic causes like Mucopolysaccharidoses (Hurler, Scheie, Hurler-Scheie, Morquio, Maroteaux-Lamy), Sphingolipidoses (Fabry disease), Mucolipidoses (GM gangliosidosis type 1, mucolipidoses types I & III), cornea plana, corneal keloids, oculoauriculovertebral dysplasia (Goldenhar-Gorlin syndrome), congenital corneal ectasia, congenital hereditary stromal dystrophy, posterior polymorphous dystrophy, Harboyansyndrome, Fryns syndrome, Sanjad-Sakati syndrome (hypoparathyroidism-retardation-dysmorphism syndrome), Encephalo-craniocutaneouslipomatosis & excess prenatal maternal consumption of alcohol. A new syndrome of hereditary congenital corneal opacities, cornea guttata, and corectopia is reported [7].

The evaluation of congenital corneal clouding should include a complete eye examination, including anterior segment evaluation, with slit lamp biomicroscopy, funduscopy, tonometry & gonioscopy. A-scan ultrasonography can reveal an enlarged globe. Tonometry is an essential component of the examination. Optic nerve head may be examined with a direct or indirect ophthalmoscope. Photoscreening can detect abnormal refractive errors & congenital glaucoma. Ocular ultrasonography may be useful in assessing other ocular abnormalities [8]. Ultrasound biomicroscopy is useful in evaluation of anterior segment structures that cannot be observed clearly because of the corneal opacity [9]. B-scan ultrasonography is used to evaluate posterior segment. CT scanning helps to diagnose protuberant congenital corneal opacities.

Treatment of corneal clouding is primarily surgical. After surgery, treatment of amblyopia & optical therapy can be helpful. Recently novel treatments for corneal clouding like transplantation of umbilical stem cells of mesenchymal type are being considered [10]. For those with bilateral corneal opacity, early penetrating keratoplasty prevents amblyopia. Poor prognostic indicators include bilateral disease, concomitant infantile glaucoma, lensectomy & vitrectomy at time of surgery, previous graft failure, extensive goniosynechiae & extensive corneal vascularisation [11]. Primary combined trabeculotomy-trabeculectomy is associated with a favourable visual outcome in infants with cloudy cornea associated with congenital glaucoma [12]. Corneal grafting for congenital opacities in infants has an excellent potential for long-term survival &should be performed as early as possible for unilateral or bilateral disease.

Conclusion

Early identification of opthalmological problems in infancy & prompt intervention is mandatory for conditions like congenital corneal clouding. Children with corneal opacities require special educational assistance based on their visual outcome. Parental education & support help in quick recovery.

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