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Penetrating Keratoplasty in a Child with Kerato iridolenticular Dysgenesis: A Case Report

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Abstract: Corneal opacities at birth present a challenging scenario to the treating physician in terms of diagnosis and management. A week-old term child delivered via unassisted vaginal delivery was referred for bilateral corneal opacities that were noted since birth. Examination under anaesthesia revealed anterior staphyloma of the corneas with normal intraocular pressures. Ultrasound biomicroscopy revealed kerato iridolenticular adhesion with mal development of the lens, while B-scan showed a flat retina and clear vitreous. Systemically, there were no other congenital abnormalities detected. A diagnosis of bilateral keratoirido lenticular dysgenesis with anterior staphyloma was made. Initial treatment was mainly topical lubricants, but medial tarsorrhaphy was performed on both eyes due to corneal thinning secondary to exposure keratopathy. After counselling her parents, the child underwent right eye penetrating keratoplasty at 6 months old. Although initially showing slight improvement in vision and also better cosmesis, the corneal graft eventually became fully vascularized again after one year post-operatively. It is therefore imperative that parents of children with this condition be counseled thoroughly and their expectations be handled delicately to ensure the best possible outcome.
Keywords: Cornea, keratoirido lenticular dysgenesis, penetrating keratoplasty, congenital abnormality, tarsorrhaphy

INTRODUCTION

Neonatal corneal opacities (NCO) manifest themselves as loss of transparency of the cornea and are evident at or soon after birth (<4 weeks) [1]. The prevalence of NCO is about 3 to 6 in 100,000 newborns [1, 2]. The aetiology of NCO may be genetic, metabolic, infectious, traumatic, toxic or a combination of these influences. All these factors affect development between the 6th and 16th weeks of gestation when differentiation of the anterior segment occurs [2].NCO can either be primary, affecting the cornea alone, or they can be secondary, being associated with anterior segment abnormalities [1]. Kerato irido lenticular dysgenesis (KILD) is placed in the latter category. We report this rare case of a child who presented with bilateral congenital corneal opacities at birth without any history of antenatal infection or drugs toxicity and no other systemic abnormalities.

CASE REPORT

A one-week-old term baby girl was referred to us for assessment of bilateral corneal opacities noted since birth. She was the first child of a non-consanguineous marriage. Antenatally, there was no history of maternal infection. Antenatal follow-up of the foetus showed the growth was corresponding to age. Postnatally, the patient was delivered via spontaneous vaginal delivery without complication. Birth weight was 2.9kg. The child was active and was not on oxygen supplementation. At birth, it was noted that there was bilateral corneal opacity. There was no other gross ocular abnormality detected by the mother or attending doctor in the labour room. The mother did not have any underlying medical illness and was not on any medication during pregnancy other than folic acid 5mg OD, vitamin C and vitamin B complex. There was no family history of congenital abnormalities in both sides of the parents' families and no history of consanguinity.

On examination, the child was active on handling and did not appear to have any syndromic features. Both the corneas were globularin nature and there was generalised corneal conjunctivalization bilaterally. Anterior chamber and fundus examinationswere not possible due to media opacity. Examination under general anaesthesia (EUA) was done. Both the corneas were bulging 5mm forward from the normal limbal plane. The corneal diameter was 10mm horizontally both eyes. There was no buphthalmos and the intraocular pressure was 13mmHg in both eyes. There was no epithelial defect.

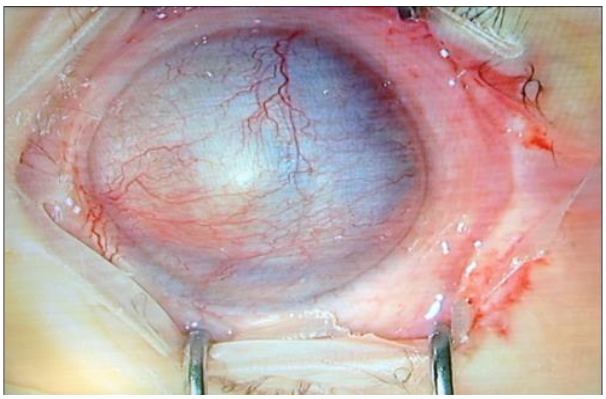

Fig-1: Intraoperative image of the left eye, showing complete conjunctivalization of the cornea.

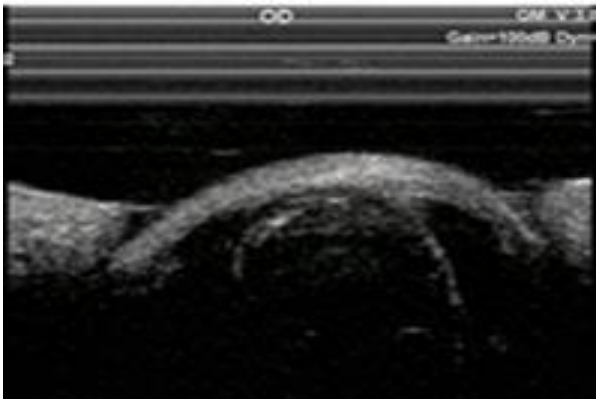

Fig- 2: UBM findings of keratoirido-lenticular adhesion and abnormal lens shape and development

Ultrasound biomicroscopy (UBM) showed kerato iridolenticular adhesions with maldevelopment of lens while B-scan showed flat retina and clear vitreous in both eyes. Systemic examination was normal; the child was moving all four limbs. There was no obvious neurological deficit. The lungs were clear. The liver or kidneys were not palpable. There were no cleft lips or palate and abnormal ears.

A diagnosis of keratoirido lenticular dysgenesis with anterior staphyloma of both eyes was made. Initially, management was aimed at preventing complications of exposure keratopathy. Topical lubricants were adequate at first, but at 2 months of age the child underwent medial tarsorrhaphy of both eyes under general anaesthesia due to thinning of the corneas.

The parents were then counselled regarding corneal transplant. Aims of restoring ambulatory vision and improvement of cosmesis were informed, although visual prognosis was guarded. At 6 months old, penetrating keratoplasty(PK) of the right eye was performed. Post-operatively, topical dexamethasone and ciprofloxacin were started 2-hourly and then gradually tapered off in 3 months.

At 6 months post-operatively, right eye examination showed a clear cornea graft at the centre (Figure 3) and fundus examination can be done. The right optic disc was normal in appearance (Figure 4). Right eye was cosmetically more acceptable post-operatively. The child was responsive to bright light.

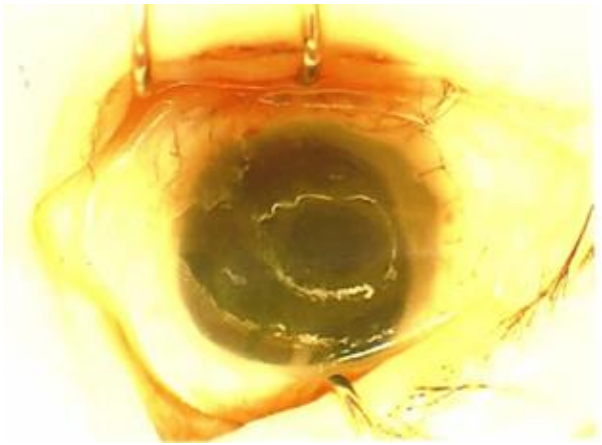

Fig-3: Appearance of a clear central corneal button, 6 months post-penetrating keratoplasty

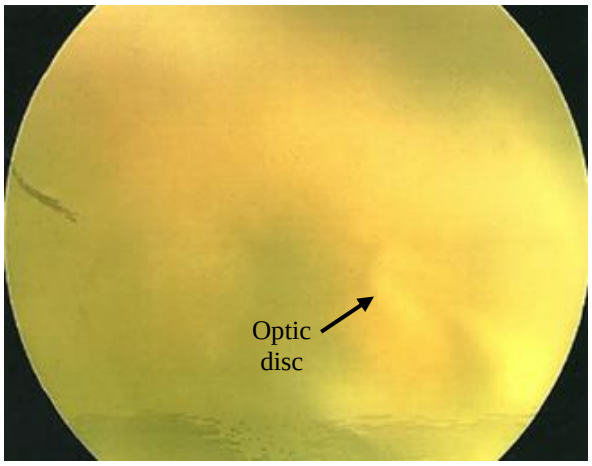

Fig-4: Fundus photograph showing a grossly normal appearance of the right optic disc.

Unfortunately, at one year post-operatively, there was recurrence of corneal conjunctivalization in the right eye. However, the child was still able to respond to bright lights and her parents felt that the eye was still acceptable cosmetically. After a lengthy discussion with the parents about the treatment options and prognosis, they decided for conservative management of the left eye.

DISCUSSION

Kerato irido lenticular dysgenesis (KILD) is a spectrum of conditions which include iridocorneal adhesions, failure of the lens to separate from the cornea, lens separation but thereafter does not develop, lens separation and formation but later opposed to the cornea, and lastly, failure of lens to form at all [1]. In

our case, the lens was believed to have been formed and separated from the cornea but there was late corneal apposition as there was an intact anterior capsule reflectivity on ultrasound. Moreover, the lens was easily separated from the cornea during penetrating keratoplasty. Thus, lens aspiration was not done in this child. However, the cornea was vascularized instead of avascular as in most of the cases. It is believed that hypoxia, persistent fetal vasculature or aniridia might be the cause of lens apposition to the cornea in the later stages of gestation [1]. In this case, there was no evidence of persistent fetal vasculature or aniridia. The possible cause might be due to hypoxia during gestation.

KILD can be associated with congenital corneal staphyloma (CCS). CCS is presumed to be a developmental abnormality, characterized by an opacified, ectatic cornea that protrudes forward between the eyelids. Anterior segment of the involved eye is usually markedly abnormal [3]. Our patient had severe anterior staphyloma of both eyes which further progressed and resulted in corneal thinning despite intensive topical medications. Tarsorrhaphy was performed as a stopgap measure to prevent further complications to the compromised corneas.

Possible differential diagnoses for KILD include congenital hereditary endothelial dystrophy (CHED), intrapartum infections such as herpes simplex virus (HSV) infection, or even congenital glaucoma. However, there were key differentiating features in our patient which distinguished it from those conditions. There was no significant family history of inherited eye disorders (as in CHED) [4], negative corneal epithelial staining or associated conditions of herpetic infections such as pneumonitis, encephalitis or hepatitis, and there certainly was no classical signs of congenital glaucoma such as epiphora, blepharospasm and raised intraocular pressure.

Proper assessment of KILD is usually made difficult especially if there is conjunctivalization of the cornea such as in our patient. However, the introduction of ultrasound biomicroscopy (UBM) has provided clinicians with an invaluable tool for clinical diagnosis as well as to establish a strategy or surgical management. UBM is able to identify the pupil; it can also highlight the potential associated structural abnormalities such as kerato lenticular and iridocorneal adhesions, ectopialentis, aniridia and congenital aphakia [2]. Thus, UBM can facilitate the surgical planning and reduce the risk of complications of the surgery. As in this case, UBM was used to evaluate the anterior segment during examination under anaesthesia (EUA) prior to penetrating keratoplasty.

There are some successes in treating neonatal cornea opacities with penetrating keratoplasty. There are approximately 35% graft survivals for 7 years cornea grafts in Peter anomaly [5]. Bilateral cases of congenital corneal opacities warrant a surgical procedure to salvage some vision in at least one eye [2]. In this case, the child had bilateral corneal opacities and she had undergone right eye PK to salvage as much vision as possible. Recurrent conjunctivalization of the right cornea in this patient was most likely due to limbal stem cell deficiency. The child was still able to respond to light even with that recurrence. Cosmetically, her right eye was more acceptable.

Surgery in this young group of patients is primarily to minimize the risk of deprivation amblyopia. However, the data have not shown better visual outcome with early surgery [2]. Cornea graft in infant is more challenging as the sclera and cornea are not fully developed at birth until about age 2 [5]. Penetrating keratoplasty (PK) is the predominant technique because lamellar keratoplasty is technically more difficult to perform in children[7]. The poor prognostic factors for infant PK are bilateral disease, glaucoma, more than one procedure (eg lensectomy and vitrectomy), previous PK, corneal vascularization, kerato lenticular adhesions, goniosynechiae, absence of Bowman's layer and absence of Descemet's membrane

The decision to operate at a young age should be made with consideration of the risk of undergoing general anesthesia for initial surgery and multiple EUA for suture removal post operatively, as well as associated systemic abnormalities and the general health of the patient. Renata A et al suggested performing surgery at least at 4 to 6 months of age if possible when there is bilateral corneal opacification [2]. In this case, the infant underwent right eye PK at the age of 6 months old. Surgery should not be done too late as it will cause deprivation amblyopia. If the surgery was done too early in this child, the risk of general anesthesia was high

CONCLUSION:

Congenital cornea opacities are rare but important conditions to make an accurate diagnosis. This is in order to predict the natural history of the disorder and to look for associated ocular and systemic abnormalities to provide appropriate medical and or surgical therapy to the patient. Early detection of KILD and prompt treatment is important as appropriate medical and surgical therapy can reduce the risk of lifelong visual impairment and impact on the overall quality of life of the growing child.

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