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Congenital blinding corneal opacification in a female baby

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Abstract: Corneal opacification is a sign that can relate with many eye conditions. They are a leading cause of blindness. Some of them are reversible; but in others cases they remain permanent.
Keywords: Congenital, blinding, corneal opacification, child

INTRODUCTION

Congenital corneal opacity is an emergency and requires management by a pediatric corneal specialist. If not treated early, these would lead to permanent visual deprivation amblyopia [1]. Congenital corneal opacification has many causes and classifications. It has been classified as being glaucomatous if associated with Haab's striae. It can be infectious commonly due to Herpes simplex keratitis in the developed world. Developmental anomalies of the cornea alone include congenital hereditary endothelial dystrophy (CHED) and or posterior polymorphous dystrophy (PPMD). Both can coexist with glaucoma [2]. Primary Congenital Glaucoma is a worldwide diagnostic and therapeutic challenge. It is responsible for 0.01 – 0.04% of total blindness and 5% of childhood blindness. Often, infants present with the classic triad of blepharospasm, epiphora, and photophobia. Characteristically, the patients are male (65%) with bilateral involvement (70%) and diagnosed within the first year of life (80%) [3,4]. Primary Congenital Glaucoma refers to a specific form of developmental glaucoma characterized by an isolated maldevelopment of the trabecular meshwork (isolated trabeculodysgenesis) not associated with other developmental ocular anomalies or ocular disease that can raise the IOP. Also called primary infantile glaucoma, it is the most common form of developmental glaucoma. The condition is typically bilateral, but 25–30% of the cases may be unilateral. Relating to the age of onset we have: congenital glaucoma which exists at birth, and usually before birth. Infantile glaucoma occurs from birth until 3 years of life and juvenile glaucoma which occurs after the age of 3 to teenage years [5].

CASE REPORT

A 9-month-old female baby brought by her mother for bilateral hazy cornea since birth. The delivery was vaginal and normal according to her. After birth the hazy cornea was noticed by the midwife who referred the child to a pediatrician but the mother rather took the child to a native doctor. When she noticed that despite the medication given by the native doctor the situation was worsening, she decided to attend the pediatric ward of the Institute of African Tropical Ophthalmology. The child was the first issue of her parents all illiterate. On external examination of both eyes, the child was not reacting to light (no fixation and pursuit). There was a complete hazy cornea along with buphthalmos and photophobia. In addition, the left eye was exotropic. We suspected a congenital glaucoma both eyes. So we planned to examine the child under general anesthesia with if necessary trabeculectomy both eyes after prior informed consent of the parents. Before that, a full evaluation performed by a pediatrician was unremarkable. Examination under general anesthesia, revealed a total opacification of the cornea, a megalocornea. In the right eye, the corneal diameter was 15 mm horizontally and 14 mm vertically. In the left eye the diameter was 15 mm horizontally and vertically. The anterior chamber as well as the fundus was not reachable because of the corneal opacification. The corrected intraocular pressure measured with the tonometer of Perkins found 35 mmHg in the right eye and 32 mmHg in the left eye. So the final diagnosis was congenital glaucoma both eyes. We performed at the same time bilateral trabeculectomy. Six months follow up showed intraocular pressure within normal limit (12 mmHg in the right eye and 13 mmHg in the left). But there was no improvement of the corneal opacification. As keratoplasty was not yet realizable in our Institute,

and the parents could not afford medical care abroad, a routine follow up was to be done in collaboration with the Department of low vision.

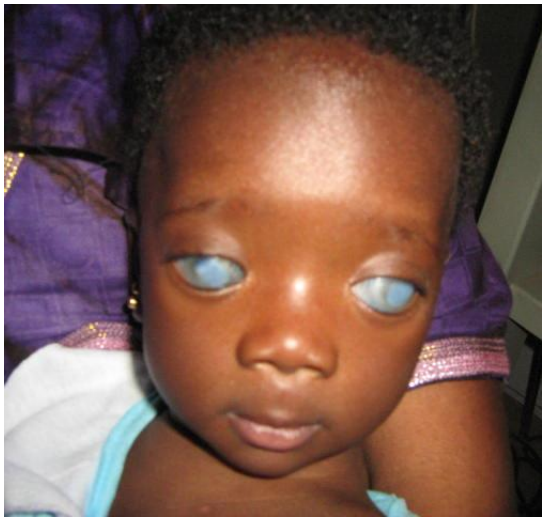

Fig.1: Photograph of the child with congenital corneal opacification both eyes

DISCUSSION

The case presented here, is a huge corneal opacification for the anterior chamber was not visible despite filtering surgery. One of the leading causes of congenital corneal opacification is Congenital Glaucoma.

Primary Congenital Glaucoma is diagnosed clinically in a neonate or infant by detecting the typical sign and symptoms of photophobia, epiphora, globe enlargement, corneal edema and opacification, and ruptures of Descemet's membrane (Haab's striae) [6, 7]. Clinically, our patient had globe enlargement along with the opacity and photophobia. Haab's striae were not visible because of the huge and total opacification of the cornea. In addition the IOP was very high.

A corneal diameter greater than 12 mm (normal 10.0 - 10.5 mm) in the first year of life is highly suggestive of Primary Congenital Glaucoma [8]. Other causes of cloudy cornea had to be ruled out.

The differential diagnoses for Primary Congenital Glaucoma is broad and would include congenital glaucoma associated with non acquired ocular anomalies or systemic disease/syndromes. Hence, careful surveillance of any systemic comorbidity is crucial in the well-being of the patient, as some systemic syndromes may carry life-threatening abnormalities that require prompt attention (e.g., cardiac outflow tract abnormalities in Axenfeld-Rieger syndrome) [9].

Sclerocornea is a congenital, non-progressive, non-inflammatory condition in which one or both corneas demonstrate some degree of opacification coupled with flattening of the normal corneal curvature. In all patients, opacification is more pronounced in the corneal periphery with varying involvement of the visual axis, in contrast with the primarily central opacification found in Peter anomaly. Sclerocornea is most commonly bilateral and asymmetric, although unilateral cases have been reported [10].

Peters' anomaly is characterized by central corneal opacity with defects in the corneal endothelium, Descemet's membrane, and posterior stroma. Variable degrees of iridolenticulocorneal adhesions are often also present [11]. Congenital hereditary endothelial dystrophy (CHED) is characterized by early-onset, bilateral diffuse corneal edema and clouding due to dysfunctional corneal endothelium Corneal clouding may be present at birth, but in many instances the signs are delayed or mild. In severe cases, deep amblyopia may develop [12].

The management of all these corneal opacities is mainly focused on corneal transplantation in addition to others options like filtering surgery in glaucoma surgery [13, 12].

CONCLUSION

Congenital corneal opacification is a challenging and sight threatening eye condition. A rapid diagnosis with the appropriate treatment is mandatory if we want to restore the sight of the fragile patients. Corneal transplantation is necessary in many cases.

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