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Case Report

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Glaucoma Associated With Bilateral Congenital Sclerocornea: Case Report

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Introduction:

Neonatal corneal opacification (NCO) encompasses a broad spectrum of disorders that have different aetiologies, with a prevalence of 6/100.000 newborns in Europe. Associated with other anterior segment developmental anomalies and glaucoma, it frecuently causes blindness from visual deprivation. The phenotypic characteristics are similar in the different clinical pictures of NCO and the terminology used is commonly not specific enough to describe separate entities, such ussclerocornea and Peters´ anomaly[1].

Case Report:

We report the case of a black male full term newborn, with bilateral total corneal opacification**(figure 1)** with vascularization at birth. Intraocular pressure (IOP) OD 55mmHg, OS 45mmHg (Tonopen and Schiötz tonometer).Corneal irregularity thickness oscillates 583 -906 microns.Ultrasoundbiomicroscopy (UBM) showed iridocorneal adhesions **(figure 2)**

We started treatment with oral acetazolamide and topic latanoprost, dorzolamide and timolol.One month later IOP was OD 25mmHg and OS 18mmHg.Trabeculotomia was the surgical technique as glaucoma surgery. Subconjunctivalbevacizumabwas administered in OS in order to perform a PK.Bilateralpenetratingkeratoplastywas performed at 4 month old. Unilateral corneal transplant in OS was performed at 5 months old.Corneal button histology was reported as sclerocornea with a markedly attenuated Bowman's layer, absence of Descemet's membrane and endothelial cells **(figure 3)**, and the presence of more densely and irregularly arranged collagen fibers in the anterior stroma with neovascularization

Discussion:

Sclerocornea is a rare form of NCO. It ´s an abnormality of the second mesodermal wave that forms scleral tissue instead of corneal stroma.Fiftypercent of the cases are sporadic and the remainder either autosomal dominant or recesive.A study of 2011 reports dominant mutations in PAX6, PITX2, FOXC1 and FOXE3 genes, and recessive mutations in B3GALTL and KERA genes.The decision to intervene surgically, and the procedures that will produce the best outcome, can best be determined with a clear understanding of the cause of the corneal opacity. Anterior segment imaging using UBM or OCTas can show us the presence or absence of iridocorneal or keratolenticular adhesions. Trabeculotomia is the first surgical option when glaucoma is associated. It is important to consider the need for a penetrating keretoplasty in one eye, when bilateral sclerocornea exist, within the first year of life to prevent sensory deprivation[3].

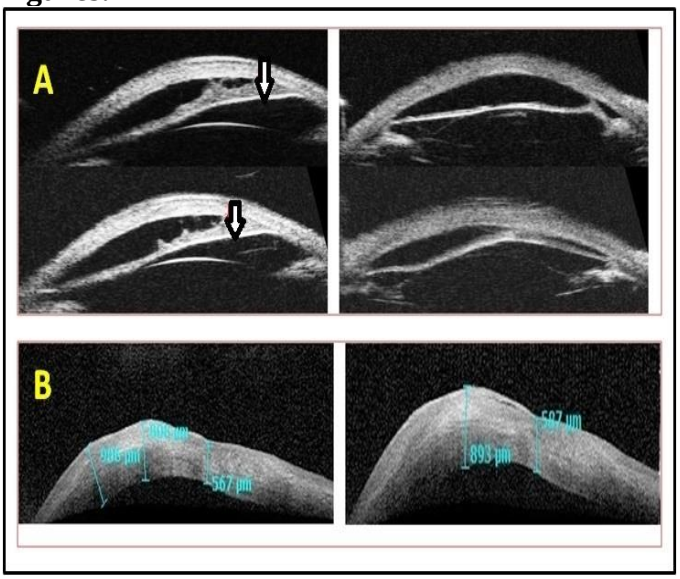
References:

1. Rezende RA, Uchoa UB, Uchoa R, et al. Congenital corneal opacities in a cornea referral practice. Cornea.2004; 23:565-570.

2. Mataftsi A, Islam L, Kelberman D, Sowden JC, Nischal KK. Chromosome abnormalities and the genetics of congenital corneal Opacification.Molecular Vision. 2011; 17:1624-1640

3.Medsinge A, Speedwell L, Nischal KK. Defining success in infant penetrating keratoplasty for developmental corneal opacities.AmOrthopt J. 2014; 64:81-8.

Figures:



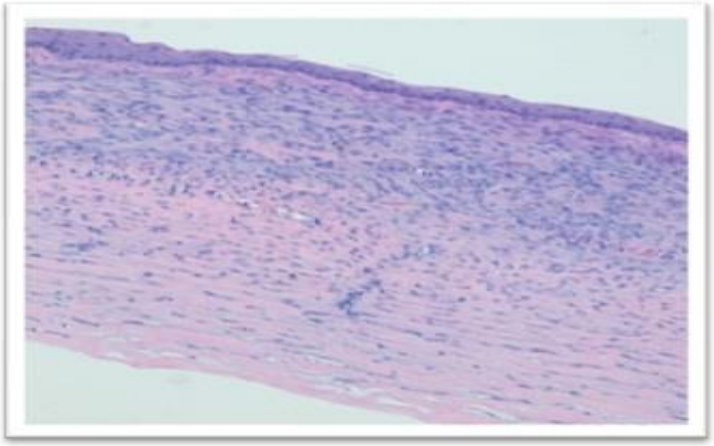


FIGURE 1
Bilateral corneal opacity

FIGURE 2
A. Ultrasound biomicroscopyshowsiridocorneal adhesions (white arrow). B Anterior segment optical coherence tomography (OCTas) shows corneal irregularity (measurements can be observed in light shade)

FIGURE 3
Histological section of corneal button, stained with hematoxylin-eosin where absence of Descemet's membrane is observed attenuation and Bowman layer with neovascularization