Bilateral Congenital Protruding Corneal Leukoma with Marked Edema: an Atypical Peters’ Anomaly

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Abstract

Purpose: Congenital corneal leukoma is rare with an incidence of 6/100000 and is one of the most important causes of amblyopia. Here we report an unusual case of bilateral congenital elevated corneal leukoma.

Methods: We encountered a 13-day-old full term male newborn. Ocular examination of both eyes revealed central elevated corneal leukomas with a narrow normal lucid interval to limbus. Corneal diameter of both eyes was 10.5 mm.

Results: After performing penetrating keratoplasty, histological examination reported that corneal thickness was about 2 mm and had central circular defect in Descemet’s membrane, endothelium, and some part of Bowman’s layer compatible with Peters’ anomaly.

Conclusion: Peters’ anomaly should be considered in the differential diagnosis of bilateral congenital bulged corneal leukoma with severe edema.

Keywords: Peters’ Anomaly, Congenital Corneal Leukoma, Corneal Edema


Introduction

Leukomas are discrete white opacified areas in the central or peripheral cornea. Congenital corneal leukoma is rare with incidence of 6/100000.1 It is one of the most important causes of amblyopia. Congenital leukomas may be associated with other anomalies of the anterior segment. These anomalies, including congenital glaucoma, are now known as neurocrisopathies. They are thought to result from defective terminal induction or migration of tissues derived from neural crest cells that form the cornea and chamber.2

Peters’ anomaly, one of causes of congenital corneal opacity, includes a central defect in Descemet’s membrane and absence of endothelium.3 Here we present an atypical presentation of Peters’ anomaly with bilateral involvement and severe increased corneal thickness.

Case Report

A 13-day-old male newborn was referred to Anterior Segment Services at Farabi Eye hospital, Tehran, Iran, due to corneal opacity. He was a full term baby, 2600 Grams weight. Ocular examinations revealed bilateral central elevated corneal leukomas with narrow lucid interval to limbus. Corneal diameter of both eyes was 10.5 mm. IOP was within normal limits respect to increased corneal thickness. Other intraocular structures were not evaluable. Ultrasonography showed normal posterior segment.

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The patient was evaluated for TORCH diseases (toxoplasmosis, others, rubella, cytomegalovirus, herpes) but we did not find any abnormal tests. Smear & culture of both eyes were negative. Thyroid function tests were normal, and tests for mucopolysaccharidosis were negative. Calcium level was also normal.

The patient underwent penetrating keratoplasty (PK) for both eyes with an interval of 2 weeks. After trephination we saw that there was no attachment between lens and cornea in both eyes. In both eyes after trephination clear lens was expelled spontaneously and anterior vitrectomy was performed. Vitreous of both eyes had some organization in central part. Temporal to the macula in both eyes a retinal scar could be seen. In the left eye, iris was normal and no peripheral anterior synechiae was seen, but in fellow eye, iris had abnormal vessels and diffused peripheral anterior synechiae (PAS) (Figures 1 and 2).

Histological examination showed increased thickness of corneas to about 2 mm. There was a central circular defect in Descemet's membrane and endothelium and some part of Bowman's layer compatible with Peters' anomaly. Stroma showed fibroblastic proliferation (Figures 4-7).

Examination 6 months after PK showed that both grafts were clear, but intraocular pressure of the left eye was increased (22 mm Hg) for which timolol drop was started.

Figure 1. Elevated corneal opacity during operation

Figure 2. Very thickened cornea during operation

Figure 3. Cornea after transplantation

Figure 4. Central dysgenesis of cornea (H & E stain, X100)
Discussion

Congenital corneal leukoma may result from traumatic, hereditary, developmental (dermoid tumors, sclerocornea, Peters’ anomaly) or inflammatory causes. They can be unilateral or bilateral. If congenital glaucoma is included its prevalence is approximately 6/100000. The most common primary cause of congenital corneal abnormalities is Peters anomaly (40.3%), followed by sclerocornea (18.1%), dermoid (15.3%), congenital glaucoma (6.9%), microphthalmia (4.2%), birth trauma, and metabolic disease (2.8%). Nearly 9.7% is classified as idiopathic. It may be associated with systemic abnormalities.3

History taking, including; obstetric (such as birth trauma), maternal, paternal, and family history, is the first step in approaching to a congenital corneal leukoma. Ruling out of the causes infectious and then developmental; the most important of them congenital glaucoma, is the next step. In the last step metabolic (mucopolysaccharidosis, corneal lipidosis, cystinosis, and von Glerke disease) and systemic disorders that affect the cornea (such as congenital ichthyosis and congenital dyskeratosis) should be evaluated.

In our case smear and culture were negative. Corneal thickness was increased with normal corneal diameter and normal IOP, so infective keratitis, congenital corneal ectasia-keratomalacia, and congenital glaucoma were ruled out. The patient was well nourished and without signs of vitamin A deficiency so that nutritional keratomalacia could be ruled out. Absence of endothelium, Descemet’s membrane and increased corneal thickness was compatible with Peters’ anomaly although iridocorneal adhesion was not present.

Peters’ anomaly includes a central defect in Descemet’s membrane and absence of endothelium. Often iris strands are attached to the edges of the defect. Clinically the defective central cornea is opacified. The lens may or may not be cataractous, and it may adhere to the defect in Descemet’s membrane by a keratolenticular stalk. The condition may be bilateral or unilateral and can be dominantly inherited. Isolated presentation often accompanied by chromosomal and other systemic anomalies, is common. Experimental studies have revealed that exposure of the
embryo to ethanol or isotretinoin can cause abnormalities suggestive of Peters’ anomaly.⁴

Conclusion

Here we describe an atypical presentation of Peter’s anomaly that to our knowledge has not been reported previously. Peters’ anomaly should be considered in the differential diagnosis of bilateral congenital bulged corneal Leukoma with severe edema.

References