



Rare and Interesting Case Report of Bilateral Congenital Corneal Dystrophy with Associated Left Eye Anterior Staphyloma



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Introduction

Corneal dystrophy is a very rare congenital entity with probable deposition of some unknown material in corneal stroma of an unknown etiology with strong heredo-familial tendency. A better modality of defining corneal dystrophy is as follows: lesions in the cornea of an unknown etiology which may manifest either at birth or at 1st or 2nd decade of life. May remain stationary or progressive. It has got a strong heredo-familial tendency. Clinically a bilateral central symmetrical corneal opacity with impaired corneal sensations and absence of deep vascularisation is corneal dystrophy. Unless proved otherwise, the other associated congenital anomalies with bilateral congenital dystrophy are very rare; they are in the form of

- Congenital glaucoma
- Keratoconus
- Anterior staphyloma
- Congenital absence of Descemet's membrane
- Cherry red spot
- Deafness known as congenital deafness syndrome
- Keratoconjunctivitis sicca
- Medullated optic nerve fibers
- Congenital ptosis
- Conjunctival xerosis

Keywords: Congenital glaucoma; Keratoconus; Anterior Staphyloma; Descemet's Membrane; Cherry red spot; Deafness; Congenital Deafness Syndrome; Keratoconjunctivitis Sicca; Medullated Optic Nerve Fibers; Congenital Ptosis; Conjunctival Xerosis; Microcephaly; Ichthyosis; Staphyloma; Ultrasound; Dystrophy; Keratoplasty; Spina Bifida; Retina; Congenital Anomalies; Corneal Stroma

Obstetric History

A first married cousin first pregnancy in the first trimester of pregnancy ultrasound revealed a male fetus with spina bifida and

microcephaly so the pregnancy was terminated. 2nd pregnancy: Female child delivered normal female child after 1st successful pregnancy. 9 years of age no associated congenital disorders. 3rd pregnancy: Male child delivered after 1st successful pregnancy. Died after 3 days due to generalized ichthyosis. 4th pregnancy: Female child delivered after 1st successful pregnancy. Had bilateral congenital corneal dystrophy with left eye associated anterior staphyloma with no other associated abnormalities seen at 24 hours of age. IOP measured with tonopen was normal. B-scan ultrasound showed attached retina [1].

Case Report

This baby at age of 3 months underwent right eye modified keratoplasty and at 3 years of age left eye cosmetic keratoplasty was done.

Discussion

It is very important to encourage parents of such patients who are very much depressed.

Conclusion

This female child is at present 5 years of age, very active and intelligent, goes to play school, sees well with her right eye, even is able to operate mobile phone and Tablet PC very efficiently. Bilateral congenital corneal dystrophy with associated congenital disorders are very rare; could only find [2,3] references in literature. One case of bilateral congenital corneal dystrophy with deafness was reported in King Khaled Eye Specialist Hospital, Riyadh, K.S.A. 2nd patient of bilateral congenital corneal dystrophy with right eye glaucoma has been reported in R.P. center for ophthalmic sciences, A.I.M.S., New Delhi, India. 3rd case of malignant melanoma of choroid has been reported in PGI Chandigarh. So my reported case is probably in literature [4-6].

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