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Case Report

A Rare Case of Peter's Anomaly in New Born

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ABSTRACT

The authors report a rare case of congenital bilateral corneal opacities, corneal defects and iridocorneal dysgenesis. Clinical picture, radiological findings indicate this to be a case of type 1 Peter's anomaly.

Key words: Peter's anomaly, congenital corneal opacity, ocular congenital anomaly.

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INTRODUCTION

This congenital anomaly was first described by Albert Peters, a German ophthalmologist (1862-1938)[1].Peter's anomaly is rare and severe condition affecting both the eyes in more than half of the cases. It is defect in the endothelium and Descemet's membrane resulting in corneal clouding. Untreated case of Peter's anomaly can lead to amblyopia or congenital blindness. This condition is one of a group of disorders or differential diagnosis of congenital corneal opacities. Prevalence of Peter's anomaly is <1 / 1 000 000.[2]Previous observations points to a genetic etiology [3]

We report here a rare case of bilateral corneal opacity in day 1 old neonate with ocular features of Peter's anomaly.

Our case is of Peter's anomaly without any other associated systemic abnormalities in the child.

CASE

The case was referred to Ophthalmology OPD from Paediatric ward of DR D Y Patil Hospital & Research Institute, Kolhapur. Aneonate 1 day old male child (36weeks of gestation)born to parent residing in Karveer, Kolhapur. The birthweight of child was 1.8kgs. Baby was delivered by normal vaginal delivery in our hospital

No history of non-consanguineous marriage.

The child presented to us with bilateral corneal opacity.

There is no one from the maternal and paternal side presenting similar complaint. No one in family showed any congenital abnormalities.

There was no history of the antenatal symptoms of fever or rash.

METHODS

A Case study was done in Opthalmology OPD of Dr. D.Y. Patil Hospital and Research Institute, Kolhapur. Examination was done under general anaesthesia. Diffuse light examination, slit lamp microscopy, direct ophthalmoscopy, applanation tonometry and Bscan examinations were performed.

A complete pediatric work up was done.

Parental informed consent to participate in this study was obtained.

RESULTS

Ocular examination of anterior segment showed normal lids and conjunctiva.

Corneal opacity was present, anterior chamber and pupillary reaction could not be assessed with diffuse illumination.

On slit lamp examination bilateral corneal leucomatous opacity was seen [Fig 1]. The opacity is not extending completely to the periphery. Opacity was vertically oval in shape. Right eye corneal opacity size being 5mmX6mm both eyes [Fig 2]. Left eye opacity 6mmX6mm and showed central non opacified area about 2mmX2mm [Fig 3]. There was irdocorneal adhesion seen in the slit lamp biomicroscopic examination.

Corneal diameter measured by callipers were 10mm in both eyes.

Direct ophthalmoscopy showed no glow due to central corneal opacity.
IOP by Perkins applanation tonometer 15mmHg in both eyes.
USG B-scan reporting showed no abnormalities in posterior segment. Axial length of right eye was 17mm and that of left eye 16.7mm.
Systemic examination revealed no abnormalities.

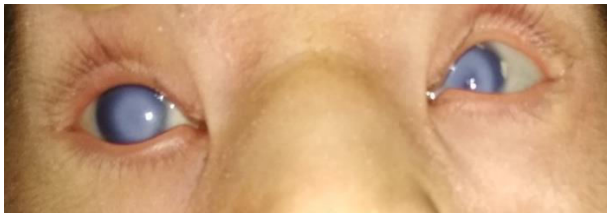

Figure 1:- Showing a new born with bilateral corneal opacity

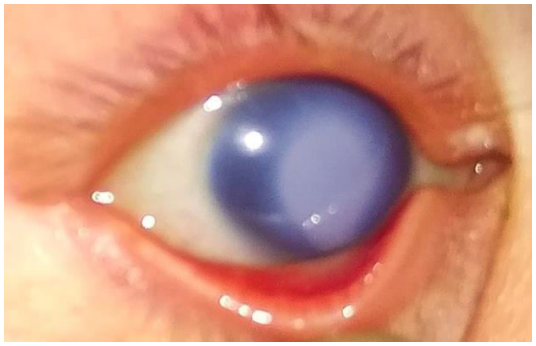

FIGURE 2:- Right eye corneal opacity size 5mmX6mm

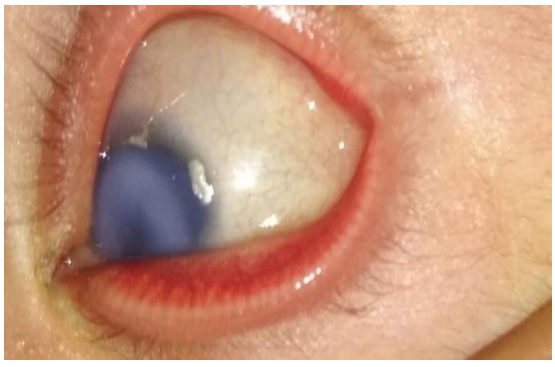

Figure 3:- Left eye corneal opacity size 6mmX6mm with central 2mmX2mm area of non-opacified area

DISCUSSION

Peter's anomaly is a rare condition in which there is opacification or clouding of the cornea leading to amblyogenic effect on a developing infant. Hallmark of Peter's anomaly is central defect in Descemet's membrane and corneal endothelium along with thinning & opacification of corresponding area of corneal stroma.[4,5,6,7]
Cause of Peter's anomaly is idiopathic. Genetic and environmental factors may play a role. Multiple genes loci have been isolated as a cause for Peters' anomaly including PAX6, PITX2, FOXC1, CYP1B1, MAF and MYOC. Abnormalities in chromosome 20, trisomy 13 and chromosome 11 have known to cause Peter's [11].Peter's can occur sporadically, but dominant and recessive inheritance has been reported.[10,11]
The critical event is 1st trimester of pregnancy, during the formation of the anterior chamber. Premature infants are at highest risk for development of Peters' anomaly. Studies have shown deficiency of heparan sulfate[12] and fetal alcohol syndrome[13] can lead to abnormal neural crest development in utero.
During embryogenesis structures of anterior segment are formed separately. If anterior segment development is abnormal then it can lead to attachment of cornea either to iris or to lens. This anterior segment dysgenesis causes Peter's anomaly. [9]
There are following subtypes of Peter's anomaly.[4.8]
A. Peter's anomaly I-the defect is in iris, corneal endothelium and Descemet's membrane
leading mild to moderate corneal opacity.
B. Peter's anomaly II-lens, though developed normally, is pushed forward against the cornea leading
to loss of descemet membrane causing severe corneal opacity that may involve the entire cornea.
C. Peter's anomaly can also be associated with Axenfeld Rieger syndrome.
Corneal opacification leads low vision early in life and causes amblyopia. Other conditions can be present such asglaucoma, cataract, and microphthalmia. Mostly Peter's anomaly is bilateral and such individuals may exhibit strabismus [9].
In Peter's anomaly anterior segment examination will reveal a central leucoma on the cornea with loss of endothelium and Descemet's membrane. Iris strands are attached to the opacified cornea. In type II Peter's lens is attached to the cornea.
Diagnosis can be done by genetic testing.
Differential diagnosis of congenital corneal opacifications are-

- Sclerocornea,
- Tears in Descemet's (eg. Congenital Glaucoma)
- Ulcers
- Metabolic
- Peter's
- Endothelial dystrophy
- Dermoid

Treatments for Peter's anomaly is to aim for visual maturation. Full thickness penetrating keratoplasty is the current standard indicated in Peter's for infants.
The success rate of penetrating keratoplasty was significantly higher in patients with Peter's anomaly type I (87.5%), as compared to patients with Peter's anomaly type II (14.2%)[3].
Combined surgery (cataract extraction and penetrating keratoplasty) can be done in presence of opacified lens and cornea lens adhesion (i.e. Triple Procedure).Peter's anomaly patient should be regularly monitored for raised intraocular pressure as glaucoma can be associated with it & if found raised appropriate medical or surgical management should be instituted.

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