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CASE REPORT

Peter's anomaly type 2 with cataract

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Purpose: This study aims to report a case of Peter's anomaly.

Case report: An 18-year-old male came with complaints of white reflexes in the right eye, which had been slowly enlarging since birth, and no vision in the left eye. On examination, the visual acuity of his right eye was 6/36 with no light perception in his left eye. Slit lamp examination in his right eye revealed nystagmus, micro-cornea, corneal opacification, cataract with cornea lenticular adhesion, and no cornea developed in his left eye. The patient was diagnosed with Peter's anomaly type 2. The patient underwent cataract surgery with intraocular lens implantation and his postoperative best corrected visual acuity was 6/24.

Conclusion: Peter's anomaly is a rare disorder with anterior segment malformation where the visual outcome is poor.

Keywords: corneal opacification, cornea lenticular adhesion, Peter's anomaly

Introduction

Peter's includes anterior segment malformation and congenital corneal opacification. The actual cause of this disorder is not clear. The most common causes are associated with genetics, infections, toxic, and traumatic factors (1). It is a rare form of anterior segment malformation with abnormal anterior segment cleavage. This condition is associated with PAX6 gene mutations (2). It was first described by Dr. Alfred Peter's about 100 years ago (3). There are two types of Peter's anomaly based on the lens: type 1 cataract may or may not be present but the cornea does not adhere to the lens and type 2 includes cataract and the lens adheres to the cornea (4).

Case report

An 18-year-old male presented with complaints of white reflexes in the right eye, which had been slowly enlarging since birth, and there was no vision in the left eye. On examination, the visual acuity in his right eye was 6/36, and no light perception in his left eye. Slit lamp

examination of the eye revealed nystagmus, micro-cornea, corneal opacification, and cataract with cornea lenticular adhesion in the right eye, and the cornea was not developed in the left eye. In the right eye, intraocular pressure was 12 mmHg. No abnormality was found in the B-scan ultrasound of his right eye. A systemic examination revealed no abnormality. The above clinical findings supported the diagnosis of Peter's anomaly type 2. The patient was treated by cataract extraction with removal of cornea lenticular adhesion and intraocular lens (IOL) implantation. During the operation, the anterior capsule was stained with 0.1% trypan blue and excised cornea lenticular adhesion, which was very difficult and challenging. Performed anterior capsulorhexis and IOL implantation in the bag were also very challenging.

Discussion

Peter's anomaly is mainly sporadic, but sometimes it can be autosomal recessive or autosomal dominant mode. It is associated with chromosome (4) abnormalities (5). Many genes have been identified as potential causes of this disorder, including PAX6, FOXC1, PITX2, CYP1B1, MAF,



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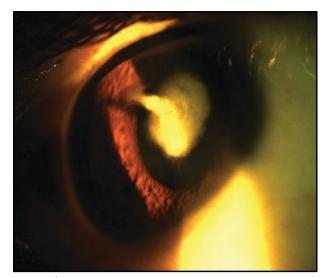


FIGURE 1 | Cornea lenticular adhesion.

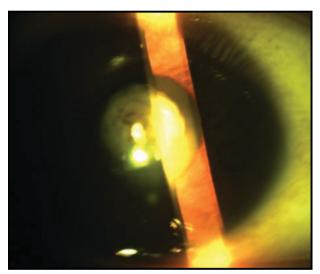


FIGURE 2 | Corneal opacification since birth.

and MYOC. The cause of Peter's plus syndrome is a beta-1,3-galactosyltransferase gene mutation. Cytomegalovirus infections rarely involve it, which causes many difficulties like microcornea, optic atrophy, coloboma, anophthalmia, Peter's anomaly, and chorioretinitis (6, 7). Systemic abnormality including hydrocephalus, congenital heart defects, and renal dysgenesis was associated with it (8). The patient may also have cleft lip and palate, genitourinary disorder, sacral hypoplasia, spina bifida, anal vesicocolonic fistula. Genitourinary anomalies are associated with multicystic dysplastic kidney, renal, and ureteral duplication, hydronephrosis, glomerulocystic kidney, and renal hypoplasia (9, 10).

Peter's anomaly with cornea plana, sclerocornea, cataract, glaucoma, and microphthalmos is associated with abnormal development of the anterior segment and

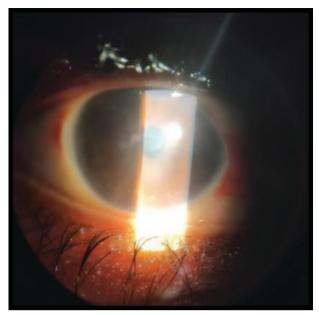


FIGURE 3 | 7th postoperative day.

mesodermal layer of the iris and within the anterior chamber angle (11).

Management of this disorder is not satisfactory and is actually very challenging. Postoperative visual outcome is not so good in the case of cornea transplantation, and some complications such as graft rejection, cataracts, glaucoma, and retinal detachment may occur (12). Whole corneal involvements and micro-ophthalmic globe are included with poor visual outcome. Amblyopia is a risk factor in cases of one-eye involvement.

Patients with cataracts need to undergo cataract surgery. To improve vision, peripheral iridectomy and pupil dilatation, in case of small corneal opacification, can be beneficial. A multidisciplinary management is required in such cases.

Conclusion

Early detection and appropriate treatment will help to preserve the residual vision and improve the quality of life. A multidisciplinary treatment is needed for the patient with systemic anomalies.

Author contributions

SN: article writing. SB: proof correction and photography. Both authors contributed to the article and approved the submitted version.

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