BOHR

Peter's Anomaly Type 2 with Cataract

Sarmin Akter Nipa* and Sujit Kumar Biswas

Chittagong Eye Infirmary and Training Complex, Pahartali, Chittagong 4202, Bangladesh **Corresponding author: dr.sarmin.nipa@gmail.com*

Abstract.

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 anomaly 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, 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





Figure 1. Cornea lenticular adhesion.



Figure 2. Corneal opacification since birth.

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 mesodermal layer of the iris and within the anterior chamber angle [11].



Figure 3. 7th postoperative day.

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.

AUTHORS CONTRIBUTIONS

- First author: Article writing.
- Second author: Proof correction and photography.

REFERENCES

 Meyer I, Rolin H, Medeiros A, et al. Peters' Anomaly, Clinical and Therapeutic Aspects: Case Report. Arq Bras Oftalmol. 2010;73(4):367–369.

- [2] Trabaccli G, Piantanida A, Bandello F, et al. Congenital Aphakia in Peters' Anomaly Syndrome: A Case Report. Acta Ophthalmol Scand. 1997;75(5):595–597.
- [3] Peters A. UeberangeborneDefektbildung der DescemetschenMembran. KlinMblAugenheilk. 1906;44:105–119.
- [4] Ozeki H, Shirai S, Nozaki M, et al. Ocular and Systemic Features of Peters Anomaly. Graefes Arch ClinExpOphthalmol. 2000;238(10):833–839.
- [5] Hittner HM, Ferrel RE, Antoszyk JH, et al. Autosomal Dominant Anterior Dysgenesis with variable expressivity–probable linkage to MNS blood group on chromosome 4. Pediatr Res. 1981;15:56.
- [6] Almarzouki HS, Tayyib AA, Khayat HA, Alsulami RE, Alzahrani SM, Alkahtani AS, et al. Peters anomaly in twins: A case report of a rare incident with novel comorbidities. Case Rep Ophthalmol 2016;7:186–92.
- [7] Cheeran MC, Lokensgard JR, Schleiss MR. Neuropathogenesis of congenital cytomegalovirus infection: Disease mechanisms and prospects for intervention. ClinMicrobiol Rev 2009;22:99–126.

- [8] Townsend WM, Font RL, Zimmerman LE. Congenital corneal leukomas. Histopathological findings in 13 eyes with noncentral defect in Descements membrane. Am J Ophthalmol. 1974;77:400.
- [9] Kivlin JD, Fineman RM, Crandall AS, Olson RJ. Peters anomaly as a cause of genetic and nongenetic syndromes. Arch Ophthalmol. 1986;104:61–64.
- [10] Tuli N, Kumar S, Sood S. Peters plus syndrome and absence of kidney: a case report. Cases J. 2009 Jan 1;2(1):2. doi: 10.1186/1757-1626-2-2. PMID: 19118497; PMCID: PMC2635348.
- [11] Kenyon KR. Mesenchymal Dysgenesis in Peters' Anomaly, Sclerocornea and Congenital Endothelial Dystrophy. Ex Eye Res. 21(2):125– 142.
- [12] Gollamudi SR, Traboulsi EI, Chamon W, et al. Visual Outcome after Surgery for Peters Anomaly. Ophthalmic Genet. 1994;15(1):31–35.