Peters’ anomaly: surgical considerations for cataract surgery. A case report

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ABSTRACT: We report the case of a young female carrier of type 2 Peters’ anomaly with microphthalmia and difficult-to-control glaucoma and vision loss in the left eye due to glaucomatous neuropathy. The cataract progressed in the right eye, reducing best spectacle-corrected visual acuity to 1/10. Treatment was performed solely on the cataract via the pars plana approach without placement of an intraocular lens. After surgery, best corrected visual acuity with a hard contact lens was 3/10 and intraocular pressure was 17 mmHg.

At 18 months of follow-up, improvement in vision and corneal stability was maintained and intraocular pressure was better controlled with hypotensive medication.


Peters’ anomaly was first described in 1906 as a rare congenital disease characterized by corneal dysgenesis involving the corneal stroma and Descemet’s membrane1. The disorder occurs bilaterally in 80% of cases2-4. A characteristic sign of the condition is corneal opacity, normally central, with a density that can vary from faint stromal opacity to dense central leukoma. The peripheral cornea is normally clear2-4. Corneal opacity can be associated with iridocorneal synechiae and other ocular malformations such as microphthalmia, cataracts, coloboma, sclerocornea, both angle and iris dysgenesis, ptosis, optic nerve and foveal hypoplasia.

Glaucoma develops in 50-70% of patients affected by this disease3-7. The cause of increased intraocular pressure (IOP) in these patients is thought to be incomplete development of the trabecular meshwork and Schlemm’s canal7,8.

Embryologically, Peters’ anomaly is classified as a mesenchymal dysgenesis of the anterior segment, and it is thought to be caused by incomplete migration to the corneal center of the mesenchymal cells that form the stromal keratocytes and corneal endothelium7. Homeotic genes control the differentiation of primordial cells, and would therefore be responsible for this abnormality. Mutations in the PAX6, PITX2 and FOXC1 genes have been observed. Most cases are sporadic, but cases of autosomal recessive and dominant inheritance have been found13. The crystalline lens may also be involved, which is why keratolenticular adhesions can be found, suggesting an incomplete separation of the lens vesicle or an anterior displacement of a normally developed crystalline lens4.

Peters’ anomaly is subdivided into type 1 and type 2, and a Peters’ plus syndrome has also been defined. Type 1 Peters’ anomaly is characterized by central corneal opacity with iridocorneal adhesions. In type 2 Peters’ anomaly there is a crystalline lens involvement and cataract or corneal lenticular adhesions, and is considered Peters’ plus syndrome if it is associated with systemic manifestations such as cleft lip or palate, short stature, abnormal external ears and mental retardation14.

CASE REPORT

A 30-year-old female patient carrier of bilateral type 2 Peters’ anomaly with glaucoma receiving triple therapy (timolol maleate every 12 hours, latanoprost every 24 hours and brinzolamide every 12 hours). The patient
reported progressive loss of visual acuity in the left eye over the last 6 years. Best spectacle-corrected visual acuity in the right eye was 2/10, and in the left eye the patient was only able to perceive light.

Biomicroscopic examination of the anterior segment revealed bilateral microphthalmia (axial length 17.77 mm), microcornea (white-to-white distance 9.0 mm), temporal paracentral corneal opacity in the right eye and central opacity in the left eye, stromal edema, corneal lenticular adhesions in area of corneal opacity, posterior synechiae and cataracts in both eyes (Figure 1A and 1B).

Figure 1. Leukomas in a patient with Peters' anomaly. A) Right eye. Temporal corneal leukoma with keratolenticular adhesions, posterior iridic synechiae and cataract. B) Left eye. Central corneal leukoma with keratolenticular adhesions and cataract.

IOP measured with a Goldmann applation tonometer was variable, ranging from 18 mmHg to 22 mmHg in both eyes. IOP measured with a TonoPen® (Reichert Ophthalmic Instruments) was 29 mmHg in both eyes.

Corneal pachymetry using optical coherence tomography of the anterior segment (OCT VisanteTM; Carl Zeiss Meditec AG) showed a mean corneal thickness of 870 µm in the right eye.

The campimetric study was not performed due to poor visual acuity.

Examination of the fundus showed an intact retina, but the details and morphology of the optic nerve could not be seen due to the opacity of the media (corneal opacity and cataract) and lack of pharmacological mydriasis.

Over the course of the year, the vision in the right eye deteriorated to 1/10 due to cataract progression.

In view of the patient's age, history of progression in the contralateral eye, loss of visual acuity in the right eye, regular control of IOP and impediments to obtain a correct assessment of the optic nerve and retina, specialists of corneal retinal and glaucoma pathologies worked together to assess the patient and determine the therapeutic strategy.

It was decided to perform cataract extraction via pars plana and lensectomy, without implantation of an intraocular lens.

Limbal paracentesis was performed under retrobulbar anesthesia, the adhesions between the anterior capsule and corneal endothelium were separated with viscoelastic fluid (Healon®, Abbott Medical Optics Inc., Santa Ana, CA, USA). Following this, 23G trocar cannulas were inserted 1.5 mm from the corneal limbus to perform lensectomy, extraction of capsular residue with an intraocular forceps, vitrectomy via pars plana with aspiration of posterior hyaloid and to examine the peripheral retina. There were no intraoperative incidents, and the three sclerotomies were sutured with Vicryl® 7/0 (Ethicon, Somerville, NJ, USA).

There were no complications during the postoperative period, and the patient reported an improvement of vision.

At present, 18 months after the operation and with triple topical therapy (brinzolamide + timolol, Azarga®, Alcon: 1 drop every 12 hours combined with brimonidine; Alphagan®, Allergan: 1 drop every 12 hours), best spectacle-corrected visual acuity with hard contact lens is 3/10 (+20.00 −0.50 × 90), and IOP measured with an applation tonometer is 17 mmHg. Biomicroscopic assessment revealed stable corneal opacity with no endothelial decompensation or aphakia, and mydriasis that permits assessment of the posterior pole (Figure 2A and 2B). Due to visual improvement, campimetry can now be used for glaucoma follow-up.

DISCUSSION

In most cases of Peters' anomaly corneal opacity has to be treated with penetrating keratoplasty and glaucoma surgery due to angular dysgenesis and poor IOP control.

In cases such as this, with sectoral corneal opacity, visual acuity is usually better preserved and deterioration is usually due to cataract or glaucoma progression. It is sometimes difficult to determine the exact cause of visual decline due to limitations found during different examinations.
In most cases of type 2 Peters’ anomaly reported in the literature, cataract surgery was performed in combination with penetrating keratoplasty\(^\text{15,16}\). The multidisciplinary team from our hospital, after evaluating the case and the risks and benefits of each treatment option, decided to treat the cataract only. The lack of vision in the contralateral eye and ocular characteristics: temporary partial paracentral corneal opacity, microphthalmia with narrow anterior chamber and posterior synechiae, were determining factors in this decision. Among the risks was the possibility of corneal decompensation following release of corneal lenticular adhesions present throughout the area of opacity. The presence of healthy endothelial tissue in 70% of the cornea suggested that it might be capable of maintaining corneal stability after surgery.

Due to axial length, a high dioptric power lens (about 44 diopters) would be needed to compensate for crystalline lens extraction. This can be achieved with a piggyback or with monofocal lens CT Xtreme D (Zeiss) with a central dioptric measurement between +45 and +60 diopters due to diffractive power. We thought the implantation of two lenses in the capsular bag or sulcus in a microphthalmic patient with a narrow anterior chamber could aggravate glaucoma progression. This lead to the decision to choose postoperative aphakia and subsequent optometric correction.

Considering the aphakia option and the characteristics of the anterior segment, which could limit visualization and hamper access to the crystalline lens, the pars plana approach was chosen.

In conclusion, management of the cataract only in cases of type 2 Peters’ anomaly with partial opacity is a more conservative alternative to corneal transplantation; it can restore visual acuity and improve control of IOP and glaucomatous neuropathy.

REFERENCES


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