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Membranous cataract – a case report

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ABSTRACT

Presentation of a rare case of an 8-week-old infant with unilateral congenital cataract and microphtalmia. The child underwent an ophthalmic examination under general anesthesia and an ultrasound assessment in A- and B-scan modes. The patient met the eligibility criteria for cataract phacotomy and phacoaspiration surgery. Most of the resorbed nuclear cortex of the lens and the fibrous membrane with blood vessels in the posterior lens capsule were found intraoperatively. They were closed by endodiathermy and an opening was made in the membrane to keep the visual axis clear. The procedure was complicated by bleeding from the pathological blood vessels in the iris during peripheral iridectomy.

KEY WORDS: congenital cataract, membranous cataract, phacotomy.

INTRODUCTION

Congenital membranous cataract is a rare condition, and it is typically observed soon after birth. It develops following the degeneration and liquefaction of the nucleus and cortex of the lens which undergo spontaneous resorption. The anterior and posterior lens capsules become opacified and fuse into a characteristic fibrous plaque. The developing membrane may be accompanied by pathological vascularization. The first case of membranous cataract was reported by von Ammon in 1833 [1]. Since then, only a few dozen cases have been reported in the literature, both in adults [2, 3]



Figure 1. Anterior segment of the eye prior to cataract surgery - right eye

and in children [4]. The etiology of membranous cataract is unclear, but *Rubella* virus infection during the early part of pregnancy may be a contributing factor [5]. The presented case is the first report of congenital membranous cataract in the Polish literature.

CASE REPORT

An 8-week-old male infant was admitted to the Pediatric Ophthalmology Department with Strabismus Treatment Center at the University Clinical Hospital in Bialystok with a suspicion of congenital cataract of the right eye. The boy was born in week 38 of pregnancy, with a birth weight of 3,400 g. The pregnancy was complicated by gestational diabetes and maternal infections of the genital tract. The child was born through natural birth. The delivery, which took place in Iceland, was uneventful. Based on the information collected from history, the child did not open the right eye from birth until three weeks of age. The parents also noticed that the child's right eye appeared smaller than the left eye, but did not seek medical attention. In the third week of life, they observed leukocoria in the boy's right eye. A consulting ophthalmologist urgently referred the child to hospital. The patient had no remarkable family history of ophthalmic diseases.

Examination performed under general anesthesia revealed a white, completely opaque lens in the right eye, with blood vessels in the inferior iris visibly growing into the lens in a fan-shaped pattern. Further ocular structures

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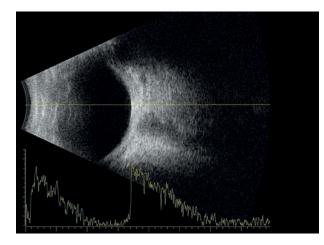


Figure 2. Ultrasound findings - right eye

could not be visualized; red fundal reflex was not seen (Figure 1). B-mode ultrasound showed retinal attachment in all sectors and normal echogenic appearance of the vitreous. In the right eye, a hyperechoic signal consistent with lens opacification was evident in the anterior segment (Figure 2). A-mode ultrasound revealed a difference in the size of both eyes - OD: 15.01 mm; OS: 18.69 mm (Figure 3). Ophthalmic examination of the anterior segment and fundus of the left eye found no abnormalities. The intraocular pressure in both eyes was 12 mmHg (measured by Tono-Pen). During wakefulness, the eyes were positioned in parallel, with no visible nystagmus. The child was scheduled for right eye cataract surgery, and preparations were undertaken for the procedure. To dilate the pupil, 1% tropicamide and 10% phenylephrine were applied topically; 0.5% levofloxacin was used for preoperative prophylaxis. To inhibit pupillary constriction during surgery and prevent anterior segment inflammation, 0.1% nepafenac was administered. In addition, 0.25% betaxolol was used to lower intraocular pressure, and an osmotic diuretic (20% mannitol) was given intravenously one hour prior to the procedure.

The surgery was performed under general anesthesia. After establishing corneal ports to maintain the depth of the anterior chamber, a viscoelastic was injected. Following phacotomy and phacoaspiration (a very small amount of lenticular masses), a hard fibrous membrane with ingrown blood vessels was visualized. The vessels were closed by endodiathermy and the membrane was excised using vitrectomy scissors. Subsequently, during parabasal iridectomy performed to prevent secondary glaucoma, bleeding from the abnormal iris vessels occurred. At the end of the procedure, following the removal of the viscoelastic, 0.5 mg of cefuroxime in 0.1 ml of 0.9% sodium chloride was administered by subconjunctival injection. The corneal ports were closed with 10-0 nylon non-absorbable sutures (Figure 4).

The child's overall postoperative health status was good. Systemic treatment consisted of an antibiotic (amoxicillin with clavulanic acid) and a glucocorticosteroid (dexamethasone) in view of the nature of cataract and the associated longer duration of surgery and high risk of postoperative in-

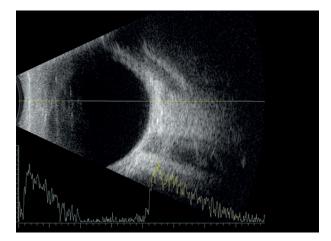


Figure 3. Ultrasound findings - left eye

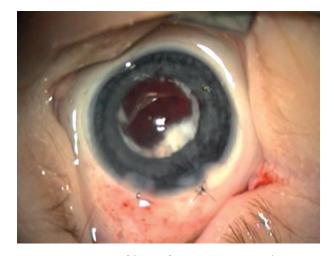


Figure 4. Anterior segment of the eye after cataract surgery – right eye

flammatory reaction presenting as inflammatory exudate in the anterior chamber. In addition, 1% tropicamide (to prevent adhesions), 0.5% levofloxacin, as well as 0.1% nepafenac and 0.5% loteprednol, were administered topically in order to reduce the risk of inflammation. On the first postoperative day, a blood clot and mild edema of Descemet's membrane were observed in the anterior chamber on the nasal side. The child was discharged on the third postoperative day with the recommendation to continue systemic antibiotic and topical therapy, and use spectacle correction (OD: +13 Dsph, OS: +2.00 Dsph), patch the healthy eye during the child's waking hours, and place a hard shield over the operated eye during sleep. The ophthalmic follow-up examination was conducted eight days after the patient's discharge. The eye was calm, and the limbal fixation sutures were secure. The cornea appeared clear and smooth. The integrity of the anterior chamber was preserved; the pupil was moderately dilated (following mydriatic administration). Superficial blood vessels were seen on the iris, with parabasal iridectomy visible from above. A small blood clot in the process of resorption was noted on the iris on the nasal side. The view of other ocular segments was slightly hazy; red fundal reflex was seen.

Table I. Classification of congenital membranous cataract

Classification of congenital membranous cataract based on slit lamp evaluation of membrane features	
1	Solid and compact opacity
II	Thin, fibrous, and sticky
III	Fibrotic and brittle
IV	Thin and compact
Classification of congenital membranous cataract based on operating microscope findings	
1	Partially resorbed lens in the cortical/nuclear layer
П	Mostly resorbed lens in the cortical/nuclear layer
III	Thickened solid lens with complete resorption of the cortical/nuclear layer
IV	Lens with few blood vessels, transparent peripheral lens area
۷	Lens with numerous blood vessels, no red fundal reflex

DISCUSSION

The incidence of congenital cataract worldwide ranges from 1.8 to 3.6 per 10,000 live births per year [6]. Membranous cataract accounts for a small proportion of congenital cataract cases, but there are no detailed statistics on the prevalence of the disease in the pediatric population. The patient described in this report is the third such case recorded in the Pediatric Ophthalmology Department in Białystok in 1995-2021.

The etiology of cataract may involve a variety of hereditary, environmental and traumatic factors. There have been reports of congenital membranous cataract coexisting with congenital rubella [7] and developing as part of various syndromes including Hallermann-Streiff-François syndrome, Lowe syndrome [8], and Marinesco-Sjögren syndrome [9]; however, the disease is not necessarily associated with any systemic disorder. The patient described in this case report has not, as yet, shown any features of the diseases and syndromes listed above, and his development is age-appropriate. Upon hospital admission, blood samples were collected as part of the diagnostic process for a congenital infection with a TORCH pathogen. The infection was ruled out.

Congenital membranous cataract can be classified into several types based on the clinical characteristics of the lens. Evaluation can be done during slit-lamp examination or under an operating microscope [4] (Table I).

Similarly to all other types of congenital cataract, membranous cataract can only be treated surgically. Surgery is indicated in cases where lenticular opacities are so prominent that they hinder normal development of the process of vision [10]. In order to prevent amblyopia of the operated eye, optical correction of aphakia should be implemented immediately after the procedure (spectacle lenses, contact lenses, IOL implantation after the child reaches an appropriate age). Longterm visual rehabilitation is also necessary. Reports of primary IOL implantation in membranous cataract are rare [2, 11], and mostly refer to a slightly older population. In the reported case, implantation was not performed in view of coexisting microphtalmia, vascular anomalies in the anterior segment of the eye, and the patient's challenging anatomy.

CONCLUSIONS

In children with congenital cataract, a specific type of the condition, i.e. membranous cataract, should be taken into account in the differential diagnosis prior to surgery. Membranous cataract may be associated with a complicated surgical course (e.g. bleeding from abnormal blood vessels), coexistence of other ocular anomalies and poorer postoperative outcomes. The presented case of an infant with congenital membranous cataract shows that the condition is a serious, though rare, problem in pediatric ophthalmology. A better understanding of the mechanisms underlying the development of the condition might significantly improve functional outcomes in the operated eyes.

DISCLOSURE

The authors declare no conflict of interest.

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