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Urrets-Zavalia Syndrome following cataract surgery in a case of anterior megalophthalmos

Síndrome de Urrets-Zavalia após cirurgia de catarata em um caso de megaloftalmo anterior

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ABSTRACT | Intraoperative and postoperative cataract surgery in eyes with anterior megalophthalmos are challenging procedures. Herein we describe the case of a 53-year-old male with anterior megalophthalmos who developed unilateral Urrets-Zavalia Syndrome following cataract surgery.

Keywords: Cataract extraction/adverse effects; Córnea/surgery; Anterior chamber; Mydriasis; Syndrome; Humans; Case reports

RESUMO | O intraoperatório e o pós-operatório de cirurgia de catarata em olhos com megaloftalmo anterior é desafiador. Descrevemos o caso de um homem de 53 anos com megaloftalmo anterior que desenvolveu a Síndrome de Urrets-Zavalia unilateral após cirurgia de catarata.

Descritores: Extração de catarata/efeitos adverso; Córnea/cirurgia; Câmara anterior; Midríase; Síndrome; Humanos; Relatos de casos

INTRODUCTION

Urrets-Zavalia Syndrome (UZS) was first described following a penetrating keratoplasty and has been associated with other ocular surgical procedures[1]. This condition often develops with the use of mydriatic drops provided during the postoperative period, a short period of high intraocular pressure or surgical trauma. Anterior megalophthalmos (AM) is a rare disorder characterized by megalocornea (horizontal corneal diameter greater than 13 mm in adults), a very deep anterior chamber, stromal atrophy, subluxation of the lens, myopia and early development of cataracts[2,3]. Herein we report a case of a 53-year-old male with AM who developed unilateral UZS following cataract surgery. After a long follow-up period, the pupil remained dilated; however, there was satisfactory vision acuity.

CASE REPORT

Preoperative measurement

A 53-year-old man presented with cataracts, megalocornea, iridodonesis and mild lens subluxation in both eyes at the slim-lamp, with no ocular trauma, systemic diseases or iris atrophy. It was not possible to measure the depth of the anterior chamber in the optical coherence tomography, since this depth was more than the program could measure (Figure 1). His corrected distance visual acuity (CDVA) was 0.6 logMAR in both eyes (-6.50 -2.00 x 35 OD; -4.50 -3.50 x 110 OS). The intraocular pressures (IOP), measured using an applanation tonometer, were OD 14 and OS 12 mmHg. Gonioscopy revealed moderate pigmentation on the trabecular meshwork and open angle. The fundus examination was normal.

Corneal horizontal diameter (OD:15 mm; OS:14.50 mm); central corneal thickness using the Pentacam (software version 1.17r24, Oculus, Wetzlar, Germany (OD: 0.406 mm; OS:0.425 mm); white-to-white method (OD:13.4 mm; OS:13.7 mm); axial length using an IOLMaster (Carl Zeiss Meditec AG, Jena, Germany) (OD:26.31 mm; OS:26.26 mm) and the anterior chamber depth (OD:5.00; OS:5.20 mm) were measured. These findings ruled out keratoglobus and isolate megalocornea.

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Surgical procedure

A standard phacoemulsification surgical technique was performed in both eyes after dilating the pupils with phenylephrine and tropicamide. To avoid intraoperative miosis, epinephrine was added in the balanced salt solution (BSS) with the same amount in both surgeries. Briefly, a topical anesthesia with 0.75% bupivacaine hydrochloride was used, and a main clear cornea incision of 2.75 mm was performed. An ophthalmic viscosurgical device (OVD, OFTVisc 2%®) was injected into the anterior chamber, and a manual central continuous curvilinear capsulotomy with *Utrata* forceps was made. Hydro-dissection, hydro-delineation and phacoemulsification of lens were performed. The same parameters of phacoemulsification (Infiniti® Vision System, Alcon Laboratories Inc.) were used in both eyes: height of the bottle of BSS 110 cm H_2O aspiration rate of 35 ml/min, a vacuum level of 330 mmHg, burst mode and the "Faco-chop" technique were used. A foldable, single-piece intraocular lens (IOL) was implanted in the capsule bag through the main incision. Then, the OVD was carefully removed. No complications or mioses were observed during the surgeries. After 30 days, the same surgery was performed in the right eye. However, for this procedure we decided to implant a capsular tension ring to stabilize the capsular bag better.

Postoperative outcomes

In the first postoperative day in the OS, IOP was 24 mmHg, and a fixed and dilated pupil was noted. Short-term use of timolol maleate 0.5% drop remained IOP under control. There was no intraocular inflammation observed. Daily pilocarpine 1% drops were prescribed for 30 days. No rise in IOP was reported in the OD. The final CDVA was OD: 0.0 logMAR (Spherical equivalent -1.00); OS: 0.2 logMAR (Spherical equivalent -1.25). Figure 2 shows postoperative photos. The patient complained of seeing a circle in the OS during the first months following the surgery. We believe it was the IOL edge. The neuroadaptation improved vision quality in an intermediate length of time. At a three-year follow-up, he was satisfied, and no IOL dislocation was noted. However, a non-pharmacological paralytic mydriasis was maintained in the OS.

DISCUSSION

This patient developed clinical findings compatible with UZS. It is postulated that the use of viscoelastic in cataract surgery could cause a toxic effect on the iris sphincter and iris vascularization, resulting in ischemia and, subsequently, fixed and dilated pupils[4]. It is possible that the depth of the anterior chamber observed in patients with AM contributes to the development of UZS, since a greater amount of viscoelastic (two tubes per eye) was required to fill the anterior chamber. Thus far, there have been no reports of UZS in patients with AM undergoing cataract surgery.

Cataract surgery in AM is challenging[2,5] and the choice of the intraocular lens IOL power and design is a difficult decision, due to high myopia and the deep anterior chamber. The *Haigis* formula was used to select IOL power[6]. The decision of a one-piece IOL was to minimize the mobility of the capsular bag during the

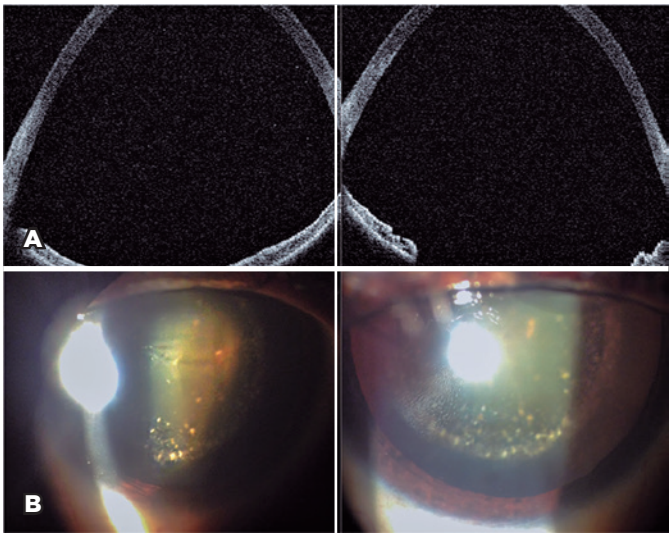

Figure 1. (A) Preoperative Visante anterior segment optical coherence tomography (OCT [Carl Zeiss Medited, Dublin, CA, USA]) showing a very deep anterior chamber in both eyes. (B) Preoperative slit-lamp photograph of the anterior segment of both eyes showing cataracts and subluxation of the lens.

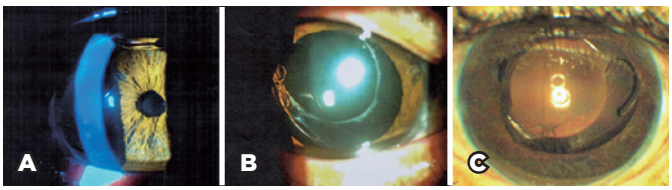

Figure 2. (A) Presence of megalocornea, sectorial iris atrophy and superior an air bubble in the anterior chamber of the right eye in the first postoperative. (B) Three years follow-up of the right eye showing the intracapsular ring and fibrosis of the border of the anterior capsule (pharmacological mydriasis). (C) Three years follow-up of the left eye following (no pharmacological mydriasis).

implantation[7]. Retropupillary iris-claw aphakic or a three-piece IOL implantation are described with satisfactory outcomes in these cases [2].

During the surgery, the capsulorhexis is difficult to perform due to the stiffening of the anterior capsule, and it is difficult to estimate its size. Since the enlarged capsular bag offers high risk of IOL displacement, this surgical step should be performed carefully. The depth of the anterior chamber required the surgeon's ability to adapt the positioning of the instruments and to maintain focus during the surgery. Unfortunately, we decided to implant an intracapsular ring in the second operated eye only. To prevent complex capsule IOL decentration, we recommend an intracapsular ring implantation in these cases. However, after a three-year follow-up, there was no complex capsule IOL decentration in either eye. We believe that the hardened consistency of the capsule bag in these eyes helped to avoid capsule contraction. An air bubble injection in the anterior chamber was cited as a risk factor for UZS in patients undergoing corneal transplant[8]. We used an anterior chamber air bubble in both eyes.

Most UZS cases do not respond well to the fixed and dilated pupil with topical pilocarpine. In addition, continuous use of pilocarpine drops could contribute to the occurrence of retinal detachment[9]. Like most patients, in this case did not respond well. Atrophy of the iris should also be expected at the postoperative period[10]. Glaucoma should be monitored in these cases due the high risk in AM[1]. This patient had high intraocular pressure in the immediate postoperative period better. More cases are needed to understand the pathophysiology and evolution of this syndrome. In conclusion, cataract surgery in eyes with AM should have a well-planned approach, and the UZS is a possible complication in these eyes.

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