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Case Report / Приказ болесника

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Urrets-Zavalia syndrome following posterior segment surgery – case report and review of literature

Синдром Уретс-Завалије после хирургије задњег сегмента ока – приказ случаја и преглед литературе

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SUMMARY

Introduction Urrets-Zavalia syndrome (UZS) has been defined as a fixed and dilated pupil accompanied with iris atrophy and occasionally secondary glaucoma. The precise cause of the syndrome is uncertain. Most often it has been described following anterior segment surgery.The objective of this article is to present how to successfully handle patients with Urrets-Zavalia Syndrome (UZS) after posterior segment surgery.

Case outline This is case presentation of patient with UZS following scleral buckle procedure .The presented case is the first to our knowledge case of UZS following this type of posterior segment surgery. We are presenting all dilemmas and difficulties we encountered during diagnostic process. Delay in treatment was mostly due to lack of knowledge about linkage of this syndrome with posterior segment surgery. Once the diagnosis was confirmed parasympathicomymetic drops were administred. Patient responded well to the therapy and partial reduction of mydriasis and restoration of pupillary kinetics was observed.

Conclusion Two months after surgery, treatment of UZS was resulted in slight residual anisocoria with signs of iris atrophy. This could indicate reversible mechanism of UZS after posterior segment surgery with iris atrophy as only permanent consequence

Keywords: Urrets-Zavalia syndrome; retinal detachment; scleral buckling; pars planavitrectomy

САЖЕТАК

Увод Синдром Уретс-Завалије (*UZS*) дефинише се као фиксирана и дилатирана пупила праћена атрофијом ириса и повремено са секундарним глаукомом.Тачан узрок овог синдрома је још увек непознат.Најћешће је описан у вези са операцијама на предњем сегменту ока. Циљ овог рада је да представи како је могуће успешно решити случај *UZS* после хирургије на задњем сегменту ока.

Приказ случаја Ово је приказ случаја пацијента са *UZS* који је настао после операције аблације ретине методом *scleral buckling*-а. Приказани случај је први *UZS* после овог типа операције на задњем сегменту ока .Приказали смо све потешкоће и дилеме које смо имали у постављању ове дијагнозе.Каснији почетак лечења је био због недовољног знања о овом синдрому везаног за операције на задњем сегменту ока. Након постављања дијагнозе *UZS* аплицирали смо парасимпатикомиметске капи и добили парцијално смањење мидријазе и опоравак пупиларне кинетике.

Закључак Два месеца послео перације лечење UZS се заврсило благом резидуалном анизокоријом са знацима атрофије ириса. Ово указује на реверзибилни механизам UZS после операција аблације ретине са ирис атрофијом као трајном последицом.

Кључнеречи: *Urrets-Zavalia* синдром; аблацијаретине, класична операција аблацијеретине; витректомија

INTRODUCTION

Urets-Zavalia Syndrome (UZS) has been identified as a fixed and dilated pupil accompanied with iris atrophy and, occasionally, secondary glaucoma. It has been often described following penetrating keratoplasty (PKP) for keratoconus in patients who havemydriatics in therapy. [1] The precise cause of the syndrome is uncertain. There have been several reported cases of UZS after DALK for keratoconus[2], DSEK for Fuch's endothelial dystrophy[3], argon laser peripheral iridoplasty[4], surgical trabeculectomy and phacic ACIOL implantation[5]. The precise cause of the syndrome is uncertain.

We report a case of UZS following scleral buckling surgery which is, by the best of our knowledge, the first such reported case in the available literature .

CASE REPORT

A56 years old male patient was referred to our clinic because he noticed a "black shadow" in the lower nasal part of the left eye visual field three days prior to the examination. Best corrected visual acuity (BCVA) on the left eye was 0.3 on Snellen chart and 0.9 on the right. From teenage years our patient has been myopic -3.5 Dsph on the right, and -6.5 Dsph on the left eye. Other than high blood pressure, the patient had no health issues.

The examination of the left eye showed retinal tear on the one o'clock with retinal detachment in the upper temporal quadrant. The macula was attached.

The patient was operated day after. We performed scleral buckling with equatorial encircling band. On the first postoperative day retina was reattached, there was some corneal oedema and visual acuity was 3/60. Patient was discharged with standard corticosteroid and antibiotic therapy, but on the following day he returned to our emergency centre with extreme pain in the operated eye. His eye lids were swollen, he had corneal oedema, slightly wider left eye pupil and inflammatory reaction in the anterior chamber. Intraocular pressure was 30 mm Hg and responded well to the administrated local anti-glaucomatous drugs.

On the check-up two days later intraocular pressure on the patient's left eye was 8 mm Hg, there was no corneal oedema or inflammatory reaction. We reduced anti-glaucomatous therapy and ten days later cancelled it completely because his intraocular pressure was 10 mm Hg. Left eye pupil was dilated but we thought it was the effect of the mydriatic eye drops.We were unable to immediately identify the cause of this reaction.

According to the data provided by our patient his visual acuity improved during the next period. However, in a two months he noticed a black shadow in the nasal half of the visual field and he had retinal re-detachment temporally with PVR. Upon the admission we noticed that the patient's left pupil was dilated, he had posterior synechiaes, iris atrophy particularly in the inferior part and incipient anterior subcapsular cataract. Left eye visual acuity was 0.1 on the Snellen chart with a shallow retinal detachment in the macular region.

We operated on the patient the next day. Phacoemulsification and pars planavitrectomywith internal silicon oil tamponade were performed. The patient was discharged the following day with a normal intraocular pressure and best corrected visual acuity 0.4 on the Snellen chart. On the check-up two weeks after the surgery we noticed an even more dilated pupil withwide posterior synechies and significant iris atrophy. [Figure 1]

It was only then that we suspected Urett'sZavalia syndrome so parasympathicmymethic eye drops were introduced 3 times daily. Three weeks later, the patient had noticed that the left eye pupil was narrower so he reported to the Clinic. We registered a significant narrowing of the left eye pupil [Figure 2].The best corrected visual acuity was 0.5 on the Snellen chart and the intraocular pressure was 12 mm Hg.

DISCUSSION

So far it was difficult to explain UZS etiology following keratoplasty. Past studies examined a number of possible causal factors including strong mydriasis further causing peripheral anterior synechiaes and glaucoma[6], direct iris trauma during surgery[7,8], iris ischemia following iris compression between the lens and the cornea during surgery[9], an abnormal immunological, neurological and iris in keratoconic eyes [10], IOP rise [11], preexisting anterior synechiae [12]. Furthermore, different studies reported UZS development following acomplicated DLK with intraoperative microperforation of Descemet membrane and air bubble in the anterior chamber. It was proposed that the air bubble could cause a pupil block, raised intraocular pressure and secondary iris ischemia with a dilated, fixed pupil [13].

Raised IOP and low ocular rigidity of the eye with keratoconus may cause occlusion of the vessels at the root of iris within the sclera resulting in iris ischemia, while preserving ciliary body function [10]. This may also be the reason for UZS developement in the presented case since scleral buckle may cause transient rise of the IOP as well as compromise scleral rigidity.

Pathophysiological mechanism of sympatethic spasm with parasympathetic inhibition was sugested since early resolution of UZS was described with an association of sympatholytic and parasympathomimetic drops [13].

Since it is the first case of the kind it was diagnosed late so the usage of the parasympaticomimethic drops started two months after primary surgery. We do not know what the permanent consequence of UZS following posterior segment surgery are and when is the deadline for the introduction of topical therapy to avoid more serious complications? We started with parasympaticomimethic drops two months after primary surgery. Resolution of UZS was succeeded after three weeks of therapy. Therapeutic effects were determined according to significant narrowing of the eye pupil and stabilisation of IOP.What is best time to start with therapy and how long we should to use it? Those questions need answers.

Resolution of UZS was succeeded after three weeks of parasympathicomymethic therapy. This could indicate reversible mechanism of UZS after posterior segment surgery with iris atrophy as only permanent consequence. The presented case is the first to our knowledge case of UZS following posterior segment surgery. Further investigation of this rare disease is warranted.

Conflict of interest: None declared

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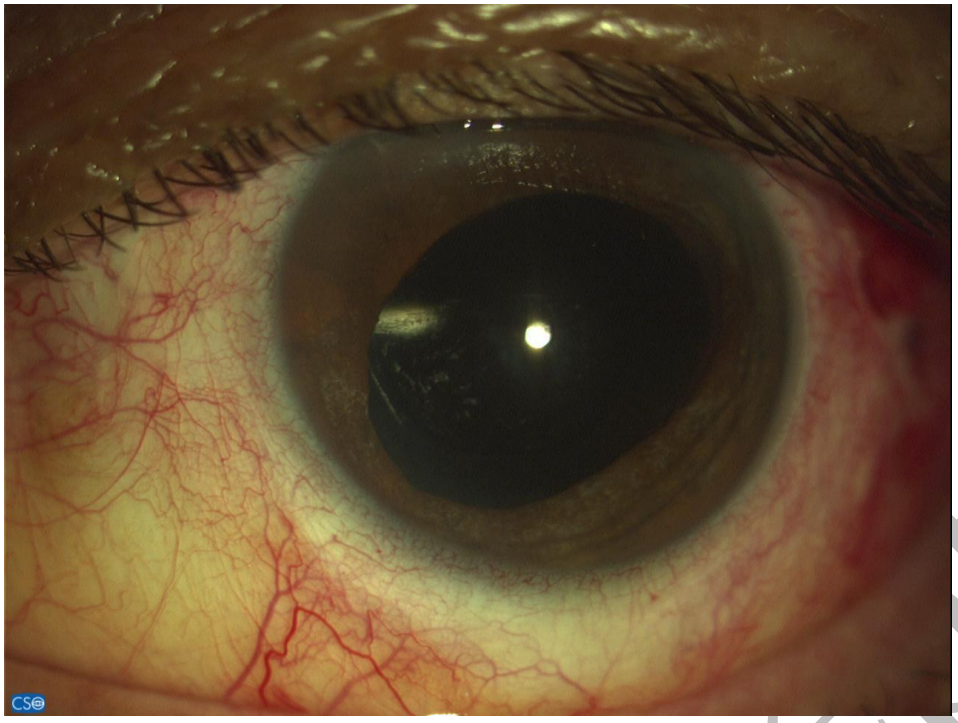

Figure 1. Dilatated irregular pupil and iris subatrophy in the lover part two weeks after operation

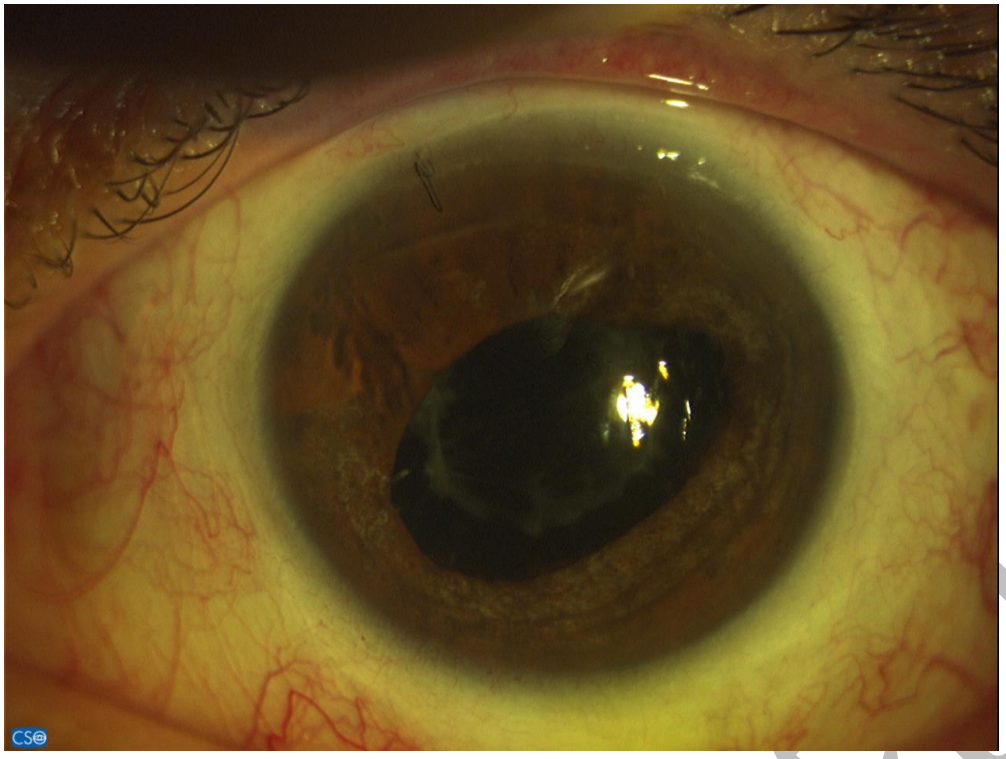

Figure 2. Significantly narrover, still irregular pupil without posterior synechiaes and significant oppacification of the posterior capsule 4 weeks after operation

