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 parasympathomimetic drops were administered. 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 пријатка атрофијом ириса и повремено са секундарним глаукомом. Тачан узрок овог синдрома је још увек непознат. Најчешће 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 пацијената са UZS који је настао после операције аблације ретине методом scleral buckling-a. Приказан 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. Касније 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 после операције лечење UZS се завршило благом резидуалном анизокоријом са знакима атрофијом ириса. Ово указује на нереверзибилни механизам UZS после операције аблације ретине са ирис атрофијом као трајном последицом.

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

**INTRODUCTION**

Urrets-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 have mydriatics 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**

A 56 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 D sph on the right, and -6.5 D sph 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 synechiae, 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 planavitrectomy with 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 with wide posterior synechiae and significant iris atrophy. [Figure 1]

It was only then that we suspected Urett’s Zavalia syndrome so parasympathic mimetic 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 synechiae 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 a complicated 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 the iris within the sclera resulting in iris ischemia, while preserving ciliary body function [10]. This may also be the reason for UZS development in the presented case since scleral buckle may cause transient rise of the IOP as well as compromise scleral rigidity.

Pathophysiologic mechanism of sympathetich spasm with parasympathetic inhibition was suggested 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 parasympaticomimetic 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 parasympaticomimetic 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 parasympatichimimethic 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
REFERENCES


Figure 1. Dilatated irregular pupil and iris subatrophy in the lower part two weeks after operation
Figure 2. Significantly narrower, still irregular pupil without posterior synechiae and significant opacification of the posterior capsule 4 weeks after operation.