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**Case Report**

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# SCHWARTZ-MATSUO SYNDROME IN PATIENTS WITH RETINAL DETACHMENT FOLLOWING BLUNT TRAUMA: 2 CASES

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## Abstract

**Purpose**:We have reported two cases of Schwartz Matsuo syndrome secondary to blunt trauma that highlights the pathophysiology, diagnostic challenges and management considerations of this rare disease.

**Case Report**: A 74-year-old man with a history of frequent falls presented with pain and decreased vision in his left eye for 1.5 days. Best corrected visual accuity (BCVA) was 50/1250, intraocular pressure (IOP) was 56 mmHg, anterior chamber has plus 3 cells and near total retinal detachment (RD). The other patient was 64 years old, history of blunt trauma 10 months ago, BCVA 20/1250, IOP 40 mmHg, +2 anterior chamber reaction and total RD. IOP decreased after vitrectomy in both patients.

**Conclusion:** Schwartz Matsuo syndrome is characterized by high IOP, RD and anterior chamber reaction. The treatment is to control the IOP with antiglaucomatous drugs and repair the RD.

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## Introduction

The diagnosis of Schwartz-Matsuo syndrome is based on the evidence of rhegmatogenous retinal detachment (RRD), the presence of aqueous cells in the anterior chamber, and elevated intraocular pressure (IOP) (1). Electron microscopy studies have demonstrated the existence of photoreceptor outer segments in the aqueous and trabecular meshwork (2,3). After RD repair, IOP tends to decrease, and the prognosis is generally considered favorable (1). The objective of this study was to report two cases of Schwartz-Matsuo syndrome secondary to blunt trauma.

## Case presentation

Case 1:

A 74-year-old man presented to the emergency department of a tertiary referral hospital with blurred vision and blunt pain in his right eye of 1.5 days' duration. In the patient's medical history, it was reported that they experienced frequent falls resulting in head injuries due to the discrepancy in leg length. Best corrected visual acuities (BCVA) by Snellen's chart were 50/1250 and 20/20 in the right and left eyes, respectively.

Biomicroscopy revealed nuclear sclerosis in the phakic right eye with 3 or more pigmented flare in the anterior chamber and microcystic edema in the cornea. IOP values assessed by Goldmann applanation tonometry were 56 mmHg and 14 mmHg in the right and left eyes, respectively.

On dilated fundoscopic evaluation, the fundus had a blurred appearance due to corneal edema. There was a nearly total RRD with macular involvement (Image 1).The fellow eye examination was unmarkable.

Initial treatment initiated under emergency conditions was aimed at lowering IOP and included a treatment regimen of 300 cc intravenous mannitol 20% and oral acetazolamide 250 mg three times daily, topical dexamethasone 0.1% four times daily, a combination of dorzolamide 2% and timolol 0.5% twice daily, brimonidine 0.2% twice daily, cyclopentolate hydrochloride 1% three times daily and oral potassium once daily. IOP decreased to 34 mmHg in the first hour after treatment.

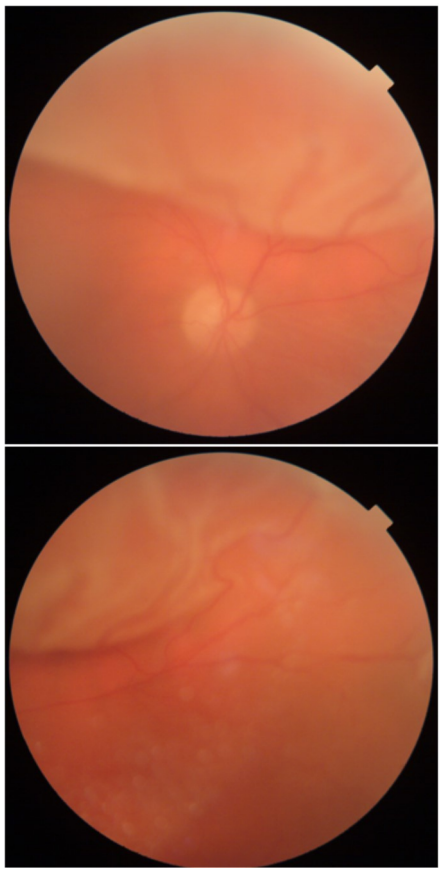

**Figure 1.** Retinal detachment with subretinal fluid extending to the macula in the left eye.

In addition to antiglaucomatous treatment, uncomplicated 23-gauge pars plana vitrectomy was performed for retinal repair. Using a 3-approach with 23 gauge trochars, a horse-shoe tear was observed at 11 o'clock, lower quadrant detached with subretinal fluid extending to the macula. The posterior hyaloid was peeled, and the vitreous base was cleaned by indentation. After retinal reattachment, 360-degree endolaser photocoagulation was performed around the tear and peripheral retina. 5000 cSt (centistokes) silicone oil was used as tamponade and the surgery was completed without complications.To ensure an effective tamponade effect of silicone oil through the tear, it is recommended to maintain an upright supine position.

Postoperative topical treatment consisted of moxifloxacin 0.5% and prednisolone acetate 1% administered eight times daily and cyclopentolate hydrochloride 1% administered three times daily. During the first day postoperative examination, the retina was reattached. BCVA improved to 20/400 and IOP decreased to 18 mmHg. In the control visit performed one week after the operation, the IOP was 12 mmHg. No subretinal fluid was detected on OCT performed one week after surgery. At 2.5 months postoperatively, the patient's BCVA had progressed to 20/50 and IOP remained at a level of 10 mmHg in the right eye without any antiglaucomatous medication.

Case 2:

A 64-year-old male patient with a history of blunt trauma to the right eye 10 months ago attended the outpatient department with blurry vision in the right eye for the preceding 2 months. BCVA were 20/1250 and 20/20 in the right and left eyes, respectively. Biomicroscopy showed moderate limbal hyperemia on the right, minimal corneal epithelial edema, and 2 flare in the anterior chamber. IOP was measured by applanation as 40 mmHg in right eye and 14 mmHg in left eye. Fundus evaluation indicated severe vitreous opacity,

pigmentation, and total RRD in the right eye.

Vitrectomy was performed on the patient. On postoperative day 1, IOP was 26 mmHg. After 15 days, the IOP was measured at 15 mmHg with the combination of dorzolamide and timolol. Two and a half months after the operation, the IOP remained stable at 12 mmHg without the use of any anti-glaucomatous medications.

## Discussion

Schwartz-Matsuo Syndrome was first proposed by Ariah Schwartz in 1973 in his article on 'Chronic Open-Angle Glaucoma Secondary to Rhegmatogenous Retinal Detachment'. Iridocyclitis caused by RD has been described as the cause of high IOP and it has been stated that open angle glaucoma will permanently improve after RD surgery. However, Schwartz could not clarify the exact location and cause of the change in the trabecular region that restricts aqueous outflow. In 1986, Matsuo et al. demonstrated free outer segments' of photoreceptors in the aqueous humor by electron microscopy and suggested that they may cause an increase in IOP by blocking trabecular meshwork outflow (2).

RD usually results in hypotonia due to the increased ease of outflow as aqueous humor is pumped from the exposed retinal pigment epithelium. Aqueous outflow through a subretinal pathway into the periocular connective tissue causes low IOP (4). In contrast, in Schwartz-Matsuo syndrome, obstruction of the trabecular meshwork by photoreceptor outer segments impairs aqueous outflow and causes RD with elevated IOP (1).

We report 2 cases of Schwartz Matsuo syndrome secondary to blunt trauma. In both cases, IOP decreased from the first postoperative day and returned to normal levels in the second week without the use of antiglaucomatous drops. In a case presented by Garlı et al, IOP decreased from 46 mmHg to 28 mmHg postoperatively, but the need for acetazolamide continued during the first week, and it took 2.5 months to discontinue all antiglaucomatous medications (5) .In both cases we presented, the IOP decreased to 18 and 26 mmHg on the first postoperative day, despite the use of 5000 silicone oil. Topical antiglaucomatous medications were only needed in the first postoperative week. Also, despite the presence of anterior chamber reaction in both cases, there was no evidence of any inflammatory pathology. The conjunctiva was white, with no ciliary injection. The cornea was transparent, and no keratic precipitate or synechiae were observed.

The first case presented a new RD, while the second case suggested chronic RD based on the duration of the patient's complaints and fundus examination. The fundus examination revealed the presence of large macrocysts, demarcation lines, and dense vitreous opacity due to scattered retinal pigment epithelium, indicating that the detachment had become chronic. While considering these cases, there may not be a direct relationship between chronicity of RD and increased IOP in this syndrome. Blunt trauma and high IOP are associated with angle retraction, which is a risk factor for RD. It has been reported that 60% of eyes with nonpenetrating or concussive trauma develop some degree of angle recession (6). Postoperative gonioscopic examination revealed that the angle elements were intact, and normalization of IOP allowed us to rule out angle recession. If the IOP had not decreased in the postoperative period, angle recession caused by trauma might have been suspected instead of Schwartz Matsuo.

Schwartz-Matsuo syndrome is triggered by retinal tears, particularly in the nonpigmented epithelium of the pars plana or pars plica close to the peripheral ora serrata, allowing fluid and photoreceptor outer segments to leak into the anterior chamber. In Case 1, the tear was located in the peripheral area at the edge of the 11 o'clock lattice degeneration. Wang et al reported a case in which the retinal tear originated from the pars plana nonpigmented epithelium and was

detected by ultrasound biomicroscopy (7).

Differential diagnoses of IOP elevation due to fine particles include lens-associated glaucoma, pigment dispersion syndrome, uveitis, ghost cell glaucoma, and Posner-Schlossman syndrome. Phacolytic glaucoma is characterized by corneal edema, anterior chamber reaction, disruption of lens capsule integrity and absence of keratic precipitates. Since both patients did not have hypermature cataracts, phacolytic glaucoma was excluded. Another possible diagnosis is pigmentary glaucoma, which is characterized by accumulation of pigment granules in the trabecular meshwork and decreased aqueous outflow. Pigmentary glaucoma increases the susceptibility to RD, both because it is more common in myopic males and because of its association with lattice degeneration (8). The presence of Krukenberg's spindles and iris retrolumination defects supports the diagnosis of pigmentary glaucoma. In pigmentary glaucoma, the anterior chamber reaction is generated by particles released from the pigment epithelium of the iris. These particles are typically dark in color and show a granular shape. In our cases, the reaction was seen as fine colorless granules in the anterior chamber. Another pathology that may be ruled out based on cell morphology and pigmentation in the anterior chamber is ghost cell glaucoma. After prolonged vitreous hemorrhage, red blood cells that have become depleted of their hemoglobin and infiltrated into the anterior chamber from the damaged vitreous appear khaki colored. Posner-Schlossman syndrome is typified by recurrent episodes of acute unilateral mild anterior chamber reaction. IOP is markedly elevated during attacks, usually above 40 mmHg. However, the anterior chamber reaction is responsive to corticosteroids. Our patient's anterior chamber reaction progressed to plus 4 cells despite topical dexamethasone treatment until surgery.

It should be kept in mind that the patient may have previously undiagnosed open-angle glaucoma. However, RD and aqueous flare do not explain this condition. Open-angle glaucoma typically involves both eyes. Normal IOP and cup-to-disc ratio and no family history of glaucoma rule away the diagnosis of open-angle glaucoma.

Exudative RD may develop secondary to uveitis or uveitic glaucoma. It is important to evaluate the patient for the presence of an intraocular inflammatory process. Inflammatory diseases such as sarcoidosis, systemic lupus erythematosus, psoriatic arthritis, ankylosing spondylitis, inflammatory bowel disease or infectious diseases such as tuberculosis, syphilis and HIV/AIDS can cause uveitis (9). The fact that our patients were 74 and 64 years old, did not describe uveitic attacks, and had no systemic complaints led us away from the diagnosis of uveitis. Findings such as keratic precipitates, pigment in the anterior lens capsule, iris nodules and posterior synechiae are evidence of inflammation in the anterior segment. These features are not generally seen in Schwartz-Matsuo syndrome. Some causes of uveitis, especially toxoplasmosis, herpes simplex virus, varicella-zoster virus, cytomegalovirus and syphilis, tend to present with elevated IOP. The difficulty in trabecular meshwork outflow in these infectious uveitis is due to trabeculitis caused by inflammatory debris. (10).

**Conclusion:**

In this article, we report two cases of Schwartz-Matsuo syndrome characterized by high IOP, aqueous cells, and RD. Retinal detachment developed secondary to blunt trauma in both cases. Management should include maximizing medical glaucoma treatment until surgery and repairing the RD as soon as possible. Early intervention in RD, preventing photoreceptor damage as well as controlling high IOP may be important in protecting the optic disc. Prognosis is generally favorable and IOP elevation typically resolves after RD repair.

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