



## Case report

## Schwartz-Matsuo syndrome: An important cause of secondary glaucoma

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## ABSTRACT

**Purpose:** We report a case of Schwartz-Matsuo syndrome that highlights the pathophysiology, diagnostic challenges, and management considerations of this rare disease.

**Observations:** 31-year-old man with a history of left eye cataract presented with left eye photophobia and elevated intraocular pressure (IOP) of 64 mm Hg. Visual acuity 20/40. Open angles with an increased pigment of trabecular meshwork by gonioscopy, 2 + anterior chamber (AC) cell, superior retinal detachment, and 0.6 cup-to-disc ratio. Electron microscopy of AC fluid demonstrated outer segments of photoreceptors. IOP was lowered with oral and topical ophthalmic antihypertensives. Retinal detachment was treated with pars plana vitrectomy with endolaser, gas tamponade, and AC paracentesis. Follow-up VA 20/20 with normal IOP.

**Conclusions and importance:** Schwartz-Matsuo syndrome is characterized by elevated IOP with marked fluctuations, open angles, aqueous cells, and retinal detachment. Diagnosis is supported by electron microscopy of AC fluid with outer segments of photoreceptors. Treatment includes retinal detachment repair and antihypertensive therapy.

## 1. Introduction

Schwartz-Matsuo syndrome is characterized by elevated intraocular pressure (IOP) with marked fluctuation, cell in the anterior chamber, and rhegmatogenous retinal detachment.<sup>1-3</sup> Electron microscopy of aqueous fluid demonstrates photoreceptors outer segments. Following retinal detachment repair, IOP typically normalizes and prognosis is favorable. We report a case of Schwartz-Matsuo syndrome that highlights the pathophysiology, diagnostic challenges, and management considerations.

## 2. Case report

A 31-year-old man with a history of left eye cataract of unknown duration presented to glaucoma clinic as an urgent referral for significantly elevated left eye intraocular pressure (IOP) secondary to presumed uveitic glaucoma. Four months prior, the patient had an episode of left eye pain and photophobia. He was evaluated by an outside provider with an IOP of 40 mm Hg in the left eye, not on ophthalmic or systemic steroids. The patient was started on pressure lowering drops and treated for uveitis with improvement in his symptoms. One week prior to his presentation his symptoms recurred. Upon presentation, he was taking brimonidine tartrate twice daily and dexamethasone once daily in the left eye. He had a history of myopia and

astigmatism ( $-1.50 + 1.00 \times 013$  OD;  $-1.50 + 1.50 \times 163$  OS) but denied any prior ocular surgery or trauma. The patient was otherwise healthy, and there was no known family history of significant ocular disease.

On examination, visual acuity (VA) was 20/20 in the right eye 20/40 in the left eye. The left pupil was sluggish, and there was an afferent pupillary defect (APD). The right pupil was normal without APD. Visual fields by confrontation were full bilaterally. The IOP measured by applanation was 64 mm Hg in the left eye and 21 mm Hg in the right eye. IOP was rechecked and measured 70 mm Hg and 19 mm Hg, respectively. Gonioscopy revealed open angles bilaterally with slightly more pigment of the trabecular meshwork in the left eye compared to the right. Pachymetry was 596  $\mu$ m and 581  $\mu$ m. Anterior segment examination of the left eye showed 2 + anterior chamber (AC) cell, but no keratic precipitates and the eye appeared white and quiet. He was noted to have a posterior subcapsular cataract of the left eye, but otherwise, there were no transillumination defects or other anterior segment findings. On dilated fundoscopic examination, a superior retinal detachment and an increased cup-to-disc ratio of 0.6 was noted in the left eye (Fig. 1). The retinal detachment was previously diagnosed as retinoschisis; however, it lacked the usual characteristics, suggesting a chronic rhegmatogenous retinal detachment with a small unidentified hole. The right eye was normal.

The differential diagnosis for our patient with material, cells, and/or

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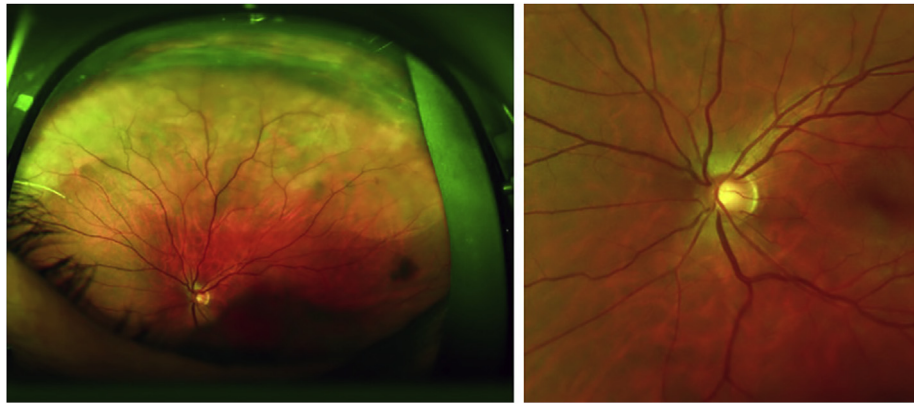
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**Fig. 1.** Left Eye Fundus Photography. A superior retinal detachment of the left eye and advanced cupping of the left optic nerve compared to the right eye.

debris in the AC and elevated IOP included uveitis, pigment granules associated with pigment dispersion syndrome, exfoliation material associated with pseudoexfoliation syndrome, erythrocytes secondary to hyphema, degenerated erythrocytes or ghost cells, lens-particle glaucoma, phacolytic glaucoma, and degenerated photoreceptors associated with Schwartz-Matsuo syndrome.<sup>1</sup> Uveitis and uveitic glaucoma are the most common, but our patient did not have a known autoimmune, infectious, or inflammatory condition. Additionally, the patient did not complain of photophobia, and there were no keratic precipitates, synechiae, or iris nodules on examination. Pigment dispersion syndrome classically occurs in myopic individuals, such as our patient, but he did not have the other classic findings of midperipheral transillumination defects, krukenberg spindles, or concave iris configuration and densely pigmented trabecular meshwork on gonioscopy. There was also no exfoliation material noted on the anterior lens capsule or peri-pupillary transillumination defects. Patients with erythrocytes or ghost cells in the AC typically report a recent history of ocular trauma, which our patient did not have. Likewise, trauma or surgery is common for lens-particle glaucoma. Phacolytic glaucoma usually occurs in elderly patients with a mature cataract, which is inconsistent with our patient.

Retinal nerve fiber layer imaging with optical coherence tomography (OCT-RNFL) was obtained and demonstrated cupping of the left optic nerve, but overall normal RNFL thickness suggesting a sub-acute process. A retina specialist was consulted, and an OCT confirmed the presence of a retinal detachment of the left eye instead of retinoschisis with adjacent loss of outer segments of photoreceptors (Fig. 2). A working diagnosis of Schwartz-Matsuo syndrome was made with suspected retinal detachment from prior ocular trauma.

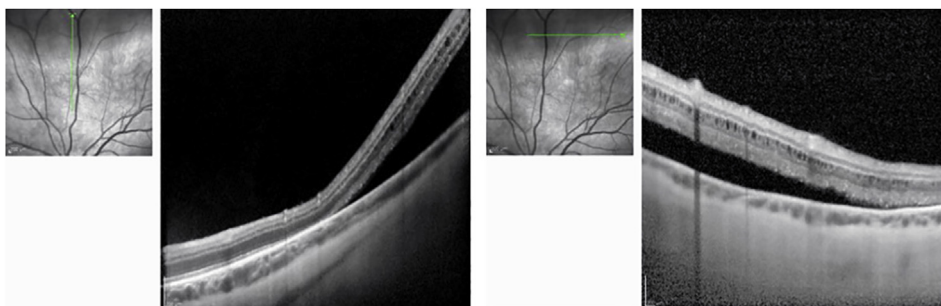
Left eye IOP was lowered to 48 mm Hg in the office with medical management and the patient was started on brinzolamide/brimonidine tartrate three times daily in the left eye, timolol twice daily in the left eye, bimatoprost at night in the left eye, and acetazolamide 250 mg orally four times daily. Follow-up two days later demonstrated stable VA at 20/40, IOP of 8 mm Hg, persistent mild AC cell, and optic nerve

cupping reversal from 0.6 to 0.3 in the left eye. The patient was continued on his current medical therapy and underwent an uncomplicated 23-gauge pars plana vitrectomy with endolaser, 20% sulfur hexafluoride gas tamponade, and AC paracentesis without simultaneous glaucoma incisional surgery one week later. The diagnosis of Schwartz-Matsuo syndrome was confirmed with electron microscopy of the AC fluid, which demonstrated pigmented epitheloid cells containing stacked membranous lamellar structures consistent with the outer segments of photoreceptors (Fig. 3).

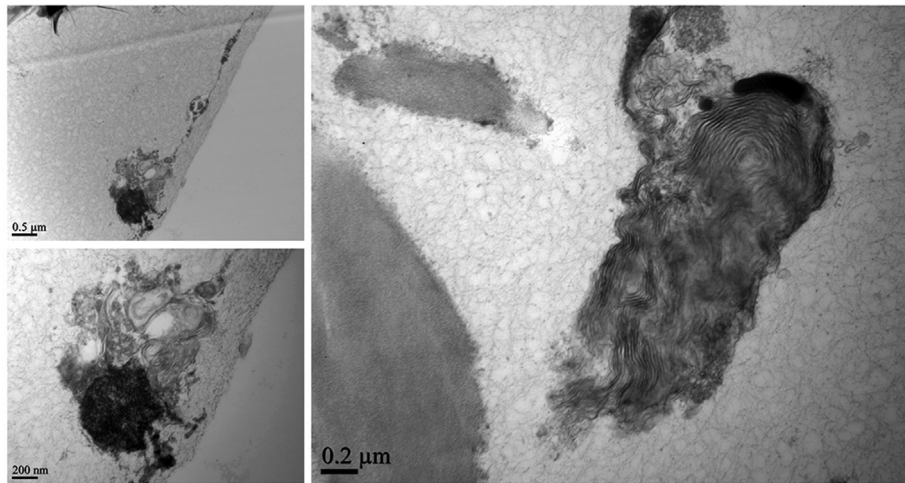
On post-op day one the patients left eye IOP was 10 mm Hg and acetazolamide and bimatoprost were stopped. The patient was started on typical post-op drops including Polytrim (polymyxin B sulfate/trimethoprim) and prednisolone acetate four times daily and atropine twice daily. One-week post-op the patients VA was 20/40 and IOP was 17 mm Hg in the left eye. A prednisone taper was started. One-month post-op the patients VA improved to 20/20 and IOP was stable at 18 mm Hg in the left eye. Brinzolamide/brimonidine tartrate twice daily and timolol twice daily was continued throughout the initial one-month post-op period. Post-op month two VA remained 20/20; however, an APD was present in the left eye. The patient had stopped brinzolamide/brimonidine tartrate twice daily and timolol twice daily two weeks prior due to ocular irritation. The IOP was 25 mm Hg, with a goal IOP at < 21 mm Hg. Brimonidine twice daily was started to lower the IOP. Humphrey visual fields were full in the right eye with an early superior arcuate deficit in the left eye and generalized depression, possibly due to the presence of cataract. The patients VA remained stable, IOP remained well controlled with topical antihypertensives, and the retina remained attached throughout subsequent follow-ups. Brimonidine was switched to dorzolamide hydrochloride-timolol maleate due to a medication allergy.

### 3. Discussion

Schwartz-Matsuo syndrome was first reported by Schwartz in



**Fig. 2.** Left Eye Optical Coherence Tomography (OCT). Subretinal fluid confirming the presence of retinal detachment with adjacent loss of outer segments of photoreceptors.



**Fig. 3.** Electron Microscopy of Anterior Chamber Fluid. Pigmented epitheloid cells containing stacked membranous lamellar structures consistent with outer segments of photoreceptors.

1973.<sup>1</sup> He described the classic findings of elevated IOP and iridocyclitis associated with rhegmatogenous retinal detachment and open AC angles. In 1986 Matsuo et al. discovered photoreceptor outer segments on electron microscopy of the aqueous humor and theorized that the free photoreceptor outer segments blocked the trabecular outflow leading to elevated IOP.<sup>2</sup> This theory was later supported by animal<sup>4</sup> and human experiments.<sup>5</sup> Since Schwartz and Matsuo described a series of eighteen cases, the syndrome has been reported eight times in the literature.<sup>5–10</sup> Risk factors include high myopia and history of ocular trauma, which increase the risk of retinal detachment. Retinal detachment is typically associated with low IOP secondary to increased outflow by active pumping of aqueous fluid through exposed retinal pigment epithelium. However, IOP in Schwartz-Matsuo syndrome is elevated. Theories for increased IOP include iridocyclitis causing reduced outflow and pigment granules released from the retinal pigment epithelium obstructing the trabecular meshwork.<sup>2,11</sup>

Patients typically present with complaints of blurry vision, eye pain, and nausea associated with the elevated IOP, as well as floaters, photopsia, and scotoma secondary to the retinal detachment. A history of blunt ocular trauma, recent ocular surgery, and atopic dermatitis may be elicited.<sup>3,6</sup> On examination, IOP is elevated, with marked fluctuation in pressures.<sup>3</sup> Pigmented aqueous cells are present in varying number, but there are usually no other signs of uveitis. On gonioscopy, the anterior chamber angles are typically open; however, angle recession may indicate prior ocular trauma.<sup>7</sup> Finally, there is retinal detachment with tears most commonly located at the ora serrata or non-pigmented epithelium of the pars plana or pars plicata.<sup>3</sup> Identification of photoreceptor outer segments in the aqueous humor by electron microscopy assists in the diagnosis. OCT will demonstrate the presence of subretinal fluid indicative of retinal detachment.

The differential diagnosis of Schwartz-Matsuo syndrome includes iritis, open-angle glaucoma, and Posner-Schlossman syndrome. Anterior synechiae and keratic precipitates in iritis indicate an inflammatory condition, which is inconsistent with Schwartz-Matsuo syndrome. Additionally, the aqueous cells in Schwartz-Matsuo syndrome are unresponsive to corticosteroids.<sup>2</sup> Open-angle glaucoma should be considered. However, the presence of aqueous cells and retinal detachment are atypical. Finally, Posner-Schlossman syndrome, which is characterized by acute unilateral recurrent attacks of elevated IOP with mild AC inflammation, demonstrates few cells and little flare, fine keratic precipitates, and is responsive to corticosteroids.<sup>12</sup>

Schwartz-Matsuo syndrome should be managed as a secondary cause of glaucoma, with first line treatment including repair of the retinal detachment and washout of the AC cellular debris. Typically,

IOP returns to normal following retinal detachment repair.<sup>8</sup> Immediate glaucoma management includes maximizing medical therapy before retinal surgery with oral carbonic anhydrase inhibitors with or without pilocarpine.<sup>7</sup> Pilocarpine may help to open the trabecular meshwork pores but is associated with miosis and increased risk of retinal detachments. Our patient presented with significantly elevated IOP (64 mm Hg, recheck 70 mm Hg) managed with a combination of antihypertensives before retinal surgery. Following retinal detachment repair, IOP normalized, requiring antihypertensive medication adjustment. Only if IOP remains elevated after retinal surgery should surgical glaucoma treatment with laser trabeculoplasty, glaucoma drainage implant, or trabeculectomy be considered. Similar to the treatment of pigmented cells with pigment dispersion syndrome, laser trabeculoplasty should lower the IOP, but may not achieve satisfactory results. Additionally, laser trabeculoplasty may place patients at higher risk for post-operative IOP spikes given their compromised trabecular function. A tube shunt is preferred over trabeculectomy given the possible conjunctival scarring following retinal surgery and the risk of hypotony maculopathy with the use of antifibrotic agents in young, highly myopic patients. Tube shunts may induce diplopia via restrictive strabismus when combined with sclera buckle.<sup>13</sup> An additional consideration is the timing of the tube shunt implantation, whether at the time of retinal surgery or not. Visual prognosis in Schwartz-Matsuo syndrome is often good.<sup>8</sup>

#### 4. Conclusions

We presented a case of Schwartz-Matsuo syndrome, which characteristically presents with elevated IOP and marked fluctuations in pressures, open AC angles, aqueous cells, and retinal detachment. The diagnosis is supported by electron microscopy demonstrating outer segments of photoreceptors in the AC fluid. Surgical repair of the retinal detachment and medical management with antihypertensives are the mainstays of treatment. Prognosis is good, with typical resolution of elevated IOP after retinal detachment repair.

#### Patient consent

Consent to publish the case report was not obtained. This report does not contain any person information that could lead to the identification of the patient.

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## Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

## Declaration of competing interest

The following authors have no financial disclosures: T.E., J.C.L., T.M.N., A.C.M.

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