A Case of Schwartz-Matsuo Syndrome with Electron Microscopic Findings

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ABSTRACT

A 45-years-old man presented with decreased vision in his left eye. His best corrected visual acuity was 0,7 OD and 0,4 OS. 1+ cells were detected in his left eye without keratic precipitate(kp), ciliary injection or iris synechiae and intraocular pressure (IOP) was 46 mmHg. Fundus examination showed a total rhegmatogenous retinal detachment in the lower quadrant of the left eye. Due to elevated IOP, 1+ cells in the anterior chamber, regmatogenous retinal detachment and absence of kp, ciliary injection the diagnosis was Schwartz-Matsuo Syndrome. Pars plana vitrectomy operation was performed for retinal detachment after the diagnosis. As predicted, a peripheral retinal tear was detected during pars plana vitrectomy and surgery resulted with normalization of the IOP. Antiglaucoma medication was stopped 2.5 months postoperatively. Electron microscopic examination of aqueous specimen demonstrated photoreceptor outer segments in varying stages of degeneration. We evaluated clinical and imaging findings of this Schwartz-Matsuo Syndrome which is rarely seen but important because the underlying treatable cause of elevated IOP may be overlooked.

Keywords: Schwartz-Matsuo Syndrome, retinal detachment, glaucoma, photoreceptor.

INTRODUCTION

Patients with rhegmatogenous retinal detachment generally have low intraocular pressures (IOP).[1] Elevated IOP secondary to rhegmatogenous retinal detachment was first described by Ariah Schwartz in 1973 an entity commonly known as Schwartz syndrome.[2] In a large retinal detachment case series, glaucoma was detected in 9.5% of patients, ocular hypertension in 6.5% and Schwartz-Matsuo syndrome in 2.1% on average.[3]

Schwartz-Matsuo Syndrome is defined as a condition characterized by uveitis developed without ciliary injection or kps, rhegmatogenous retinal detachment with oral dialysis or non-pigmented ciliary epithelial tears, presence of external segments of photoreceptor cells in aqueous humor and elevated IOP which normalizes after vitrectomy.[4,5] In this syndrome, tears are generally seen in the non pigmented epithelium of the pars plicata or pars plana of the ciliary body, or in around the vitreous base such as oral dialysis.[5] It is thought that the IOP increases with blockage of the aqueous humor outflow by obstruction of the trabecular meshwork with photoreceptor outer segments passing through the subretinal space after retinal detachment.[4] In this syndrome, the elevated IOP is resistant to corticosteroids and antiglaucoma treatment, but it usually returns to normal after a successful retinal detachment surgery. Previous intraocular surgery or ocular trauma can cause this syndrome by causing midperipheral tears.[3,5]

Case

A 45-years-old man presented to our clinic with decreased vision in his left eye. His best corrected visual acuity was 0,7 OD and 0,4 OS. His right anterior segment was normal. His left anterior chamber contained 1+ cells but no keratic precipitate (kp), ciliary injection or iris synechiae. The IOP at presentation was 12 mmHg OD and 46 mmHg OS. The patient had no history of trauma or surgery, but had myopic refractive error (−0.75 OD; −1.25 OS). Dilated fundus examination revealed a recent total macula-off

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retinal detachment and irregular optic disc margins in the left eye. Fundus examination was normal in the right eye. Posterior B-scan ultrasonography of the left eye revealed no pathology other than a total retinal detachment (Figure 1). In addition, cranial and orbital magnetic resonance imaging was performed in order to exclude the possibility of uveitis with ocular hypertension due to neoplastic etiologies in the differential diagnosis of our patient whose left optic disc margins were irregular. Imaging results were normal. At the begining medical treatment was as follows: 300 cc intravenous mannitol 20% and oral acetazolamide 250 mg three times a day, topical dexamethasone 0.1% four times a day, dorzolamide 2% + timolol 0.5% combination two times a day, brimonidine 0.2% two times a day, cyclopentolate hydrochloride %1 three times a day and oral potassium one time a day. Half an hour after the initiation of medical treatment, the IOP was reduced to 28 mmHg in the left eye.

Our patient underwent a successful 23 Gauge pars plana vitrectomy with the diagnosis of Schwartz-Matsuo Syndrome. A 3x4 mm full-thickness retinal tear was detected in the form of a 'L' shape at the superior temporal region near the pars plana. After the retina was reattached, 360 degree 4-line photocoagulation was performed around the retinal tear. 1000 cSt (centistokes) silicon oil was used as an internal tamponade agent in the surgery. No intraoperative complications were developed. The obtained aqueous sample during the surgery was fixed with gluteraldehyde and kept at 4°C after consulting with İstanbul Faculty of Medicine Department of Pathology. The sample was sent to İstanbul Faculty of Medicine Department of Histology to conduct an electron microscopic examination. In the postoperative period, topical moxifloxacin 0.5% eight times a day, prednisolone acetate 1% eight times a day and cyclopentolate hydrochloride 1% three times a day were initiated. On the first day postoperatively the retina was fully attached. The IOP was 37 mmHg OS. 300 cc intravenous 20% mannitol was administered and oral acetazolamide 250 mg three times a day was initiated. On the second postoperative day, his IOP decreased to 19 mmHg OS. The oral acetazolamide treatment was tapered off by reducing the dosing. His IOP was 12 mmHg OS in the control examination performed 1 week after the operation (with topical dorzolamide 2%, timolol 0.5% and brimonidine 0.2%). No subretinal fluid or cystic changes were detected on OCT performed 2 weeks after surgery. The topical antiglaucoma drugs were tapered off by decreasing dosage and the IOP improved to 16 mmHg without medication after 2.5 months of operation.

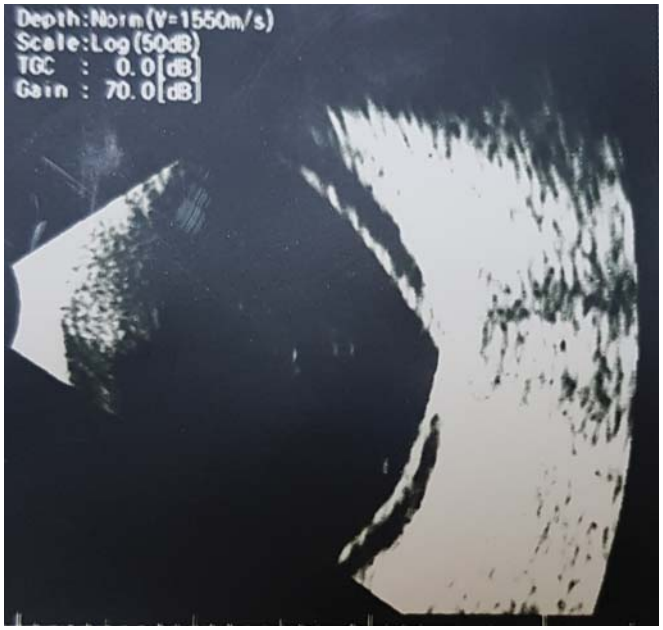

Figure 1: *Posterior B-scan ultrasonography of the left eye.*

The outer segments of photoreceptor cells which were partially or completely disrupted were observed in the electron microscopic examination of aqueous specimen of our patient. The outer segments were found to have intact or disrupted lamellar structures (Figures 2 and 3). Based on these findings the diagnosis was confirmed as Schwartz-Matsuo Syndrome.

DISCUSSION

Schwartz-Matsuo Syndrome is a very rare syndrome. Due to the fact that it can be confused with other diseases, the delay in diagnosis and treatment can result in severe vision loss. Unlike what is expected for most of the rhegmatogenous retinal detachments, in this sydrome the IOP is high and there is an anterior chamber reaction without kp and ciliary injection. Our patient had 1+ anterior chamber reaction and unilateral open-angle glaucoma associated with peripheric rhegmatogenous retinal detachment with a retinal tear in the temporal periphery. The risk factors for Schwartz-Matsuo Syndrome include young age, male sex, history of trauma and peripheral retinal tears.[2,5] Our patient was 45 years old and had no history of trauma.

Before a diagnosis of open-angle glaucoma secondary to retinal detachment is made, three conditions should be differentiated.[2] The first and most common group is that of retinal detachment occurring in an eye with preexisting chronic open angle glaucoma. The clues to this diagnosis are the bilaterality of the glaucoma and the presence of typical glaucomatous changes of the optic disks; so glaucomatous changes are seen in the other eye which is not detached. We excluded first group because our patient didn't have a

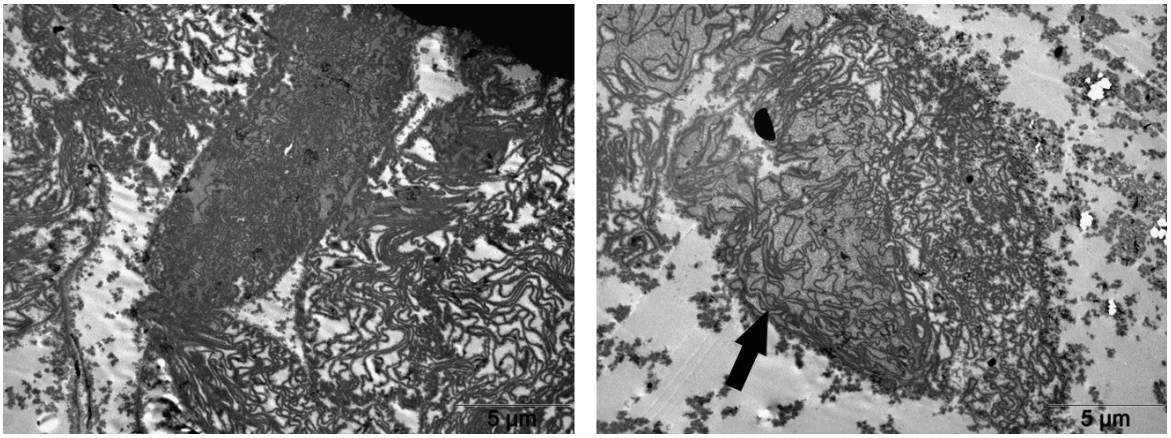

Figure 2: *Transmission electron microscopic examination of aqueous specimen showing outer segments of photoreceptor cells (indicated by the arrow) (5µm).*

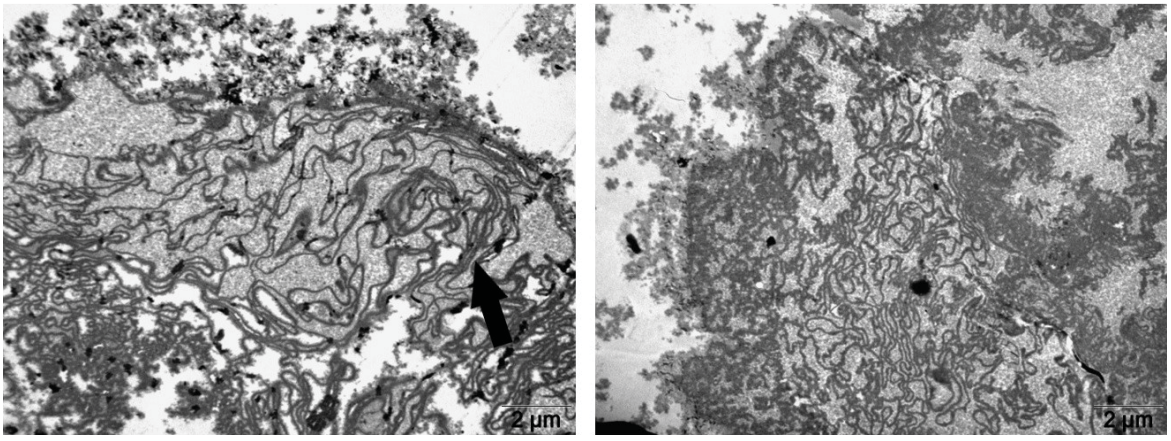

Figure 3: *Transmission electron microscopic examination of aqueous specimen showing outer segments of photoreceptor cells (indicated by the arrow) (2µm).*

previous diagnosis of glaucoma and the optic nerve head and the IOP was normal in the right. In addition, retinal detachment secondary to pilocarpine use in glaucoma cases due to vitreofoveal traction by displacement of the lens and vitreous to the anterior chamber should also be included in the differential diagnosis.[6] We have also ruled out this diagnosis since our patient didn't use topical pilocarpine.

In the second group, glaucoma is secondary to subacute or chronic uveitis. A non-rhegmatogenous retinal detachment may be caused by the uveitis. A retinal detachment may also be rhegmatogenous and unrelated to the uveitis. If uveitis is the primary process, kp, iris atrophy, ciliary injection, anterior and posterior synechiae are likely to be present. None of these findings were present in our patient. In the third group, elevated IOP and retinal detachment share common underlying etiology or association such as Marfan's syndrome, Stickler's syndrome, angle-recession glaucoma and traumatic retinal detachment.[7] Our patient's findings were incompatible with these diagnoses.

Occasionally diagnostic conclusions may have to await the findings in the postoperative course. If uveitis subsides promptly after retinal reattachment surgery and IOP readings normalize within a few weeks after retinal reattachment, the evidence would suggest that retinal detachment had been the primary etiological factor for both uveitis and glaucoma. If, on the contrary, the glaucoma and abnormal aqueous dynamics persist, it is more likely that the glaucoma is either a chronic open angle type or that it has occurred secondarily to a condition other than retinal detachment.

Matsuo and colleagues found a great number of photoreceptor outer segments and few cells in the electron microscopic examination of anterior chamber fluids of seven cases with Schwartz-Matsuo syndrome.[5] In all cases the cells in aqueous humor disappeared after retinal detachment surgery and the IOP returned to normal. They etiologically suggested that the outer segments of the photoreceptors occlude the trabecular meshwork passing from subretinal space to the aqueous humor. Lambrou and associates found decreased outflow facility in human

eye-bank eyes and cat eyes in vivo following injection of retinal rod outer segments into the anterior chamber and demonstrated that these accumulate in the trabecular region and the IOP rises.[4]

After retinal detachment surgery, our patient's IOP quickly returned to normal and the reaction in the anterior chamber disappeared. Despite the precise cause of IOP elevation in our patient is unclear, in accordance with the mechanism of the Schwartz-Matsuo syndrome, we have detected a large number of photoreceptor cell outer segments in electron microscopic examination of aqueous specimen of our patient.

Photoreceptor outer segments and glycosaminoglycans produced by visual photoreceptor cells have been identified in the aqueous of patients with retinal detachment who do not have elevated IOP.[7] Thus, the presence of photoreceptor outer segments in the aqueous does not necessarily produce Schwartz-Matsuo syndrome. It appears that Schwartz-Matsuo syndrome may occur following release of a sufficiently large quantity of outer segments into the aqueous, due perhaps to the size and position of the retinal break or the size of the detachment. Breakdown products of outer segments may also contribute to the reduction of outflow facility and elevation of IOP in Schwartz-Matsuo syndrome.

Yalvaç et al. reported two cases of Schwartz-Matsuo syndrome with post-traumatic retinal dialysis and peripheral retinal tears respectively. Their patients' IOP also normalized after retinal detachment surgey[8]. There are various reports of photoreceptor outer segments in the aqueous humor in cases of Schwartz-Matsuo syndrome in the literature.[9-12] In addition, Clark et al. described a case of photoreceptor outer segment glaucoma with electron microscopic evidence of photoreceptor outer segments in the trabecular meshwork recently.[13] However, contrary to the original description by Schwartz and Matsuo, the IOP in their patient did not normalize after the surgery.

The prognosis for patients with Schwartz-Matsuo syndrome which is rarely seen is good. Vision remains stable following retinal reattachment surgery. Our patient's best corrected visual acuity was 0,4 OS and remained stable postoperatively. Microstructural ischemic irreversible retinal damage associated with macula-off total retinal detachment may be the reason why our patient's visual acuity did not improve postoperatively. The IOP may return to normal within days following surgical reattachment of the retina. However, as illustrated by our case, antiglaucoma medications may be required for several months postoperatively.

REFERENCES

1. Burton TC, Arafat NI, Phelps CD. Intraocular pressure in retinal detachment. Int Ophthalmol. 1979;1:147e152.
2. Schwartz A. Chronic open-angle glaucoma secondary to rhegmatogenous retinal detachment. Am J Ophthalmol. 1973 Feb;75(2):205-11. PubMed PMID: 4735339.
3. Phelps CD, Burton TC. Glaucoma and retinal detachment. Arch Ophthalmol. 1977;95:418e422.
4. Lambrou FH, Vela MA, Woods W. Obstruction of thetrabecular meshwork by retinal rod outersegments. ArchOphthalmol. 1989 May;107(5):742-5. PubMed PMID: 2719584.
5. Matsuo T. Photoreceptor outer segments in aqueous humor: key to understanding a new syndrome. SurvOphthalmol. 1994;39(3):211-233.
6. Puustjärvi T. Retinal detachment during glaucoma therapy. Review. A case report of an occurrence of retinal detachment after using membranous pilocarpine delivery system [Pilokarpin lameller (Ocusert) 11 mg]. Ophthalmologica.1985;190(1):40-4.
7. Netland PA, Mukai S, Covington HI. Elevated intraocular pressure secondary to rhegmatogenous retinal detachment. Surv Ophthalmol. 1994 Nov-Dec;39(3):234-40. PubMed PMID: 7878522.
8. Yalvaç I, Ünlü N, Karagöz Y, Akgün Ü, Acar MA, Kocaoğlan H, Kasım R. İki Olgu Nedeni ile Schwartz Sendromu. Retina-Vitreus/Journal of Retina-Vitreous. 1995; 3:94-6.
9. Gutiérrez C, Merayo J, Cuevas J, Lázaro M, Rebolleda G, Muñoz-Negrete FJ. Glaucoma asociado a segmentos externos de fotorreceptores en el humor acuoso (Síndrome de Schwartz-Matsuo) [Glaucoma related with photoreceptor outer segments in aqueous humor. Schwartz-Matsuo syndrome]. Arch Soc Esp Oftalmol. 2001 May;76(5):315-8. Spanish. PMID: 11373708.
10. Mitry D, Constable I, Singh J. Photoreceptor outer segment glaucoma in rhegmatogenous retinal detachment. Arch Ophthalmol. 2009 Aug;127(8):1053-4. doi: 10.1001/archophthalmol.2009.147. PMID: 19667346.
11. Fan W, Zheng X, Yuan S, Xie P, Liu Q. Angiogenic Cytokine Profile of a Patient With Schwartz-Matsuo Syndrome. Ophthalmic Surg Lasers Imaging Retina. 2018 Sep 1;49(9):721-725. doi: 10.3928/23258160-20180831-12. PMID: 30222809.
12. Etheridge T, Larson JC, Nork TM, Momont AC. Schwartz-Matsuo syndrome: An important cause of secondary glaucoma. Am J Ophthalmol Case Rep. 2020 Jan 3;17:100586. doi: 10.1016/j.ajoc.2020.100586. PMID: 31956728
13. Clark A, Alkhotani A, Yucel YH, Sylvester C, Kertes PJ, Birt CM. Electron Microscopic Evidence of Photoreceptor Outer-segments in the Trabecular Meshwork in a Case of Schwartz-Matsuo Syndrome. J Glaucoma. 2019 Sep;28(9):843-845