Clinical findings and management of rhegmatogenous retinal detachment

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Abstract

Rhegmatogenous retinal detachment (RRD) is a potentially blinding ocular condition that should be treated promptly. Depending on the initial clinical presentation of the detachment, patients can either have excellent vision or significantly reduced visual acuity as a result of the maculae involvement. In general, a macula-on retinal detachment that is threatening the posterior pole requires more prompt treatment compared to macula-off retinal detachment due to the impending risk of progression to macula-off. This article reviews the mechanism, risk factors, clinical findings and treatment options for RRD.

Key words: Rhegmatogenous retinal detachment (RRD), steamroller, pneumatic retinopexy (PR), scleral buckle, retinal tear, Schwartz-Matsuo Syndrome

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S Introduction

There are three types of retinal detachment: Rhegmatogenous retinal detachment (RDD), exudative retinal detachment and tractional retinal detachment. Unlike serous and tractional retinal detachments, a tear or break on the retina is necessary for RRD to occur. Subretinal fluid gains access to the outer layer of the retina via the tear and detaches the sensory retina from the retinal pigment epithelium (RPE) leading to RRD. The prevalence of retinal break is approximately 5 to 7 percent in the general population, with most of these breaks composed of atrophic holes and some horseshoe tears.¹ The incidence of RRD ranges from 6.3 to 17.9/100,000 people annually.²

A higher prevalence of RRD was found in 20-30 year-olds and in 60 year-olds, possibly correlated with high myopia in the younger age group and posterior vitreal detachment associated with aging in the older population.² This bimodal distribution was found in several research studies.¹⁻² Similarly, men have a higher incidence rate for RRD compared to women by a ratio of 1.3:1 to 2.3:1, but more young males have been diagnosed with traumatic retinal detachment than females.² The incidence of bilateral RRD has been found to range from 3-33 percent, but most studies suggested the incidence to be approximately 5-15 percent.¹⁻²

Other risk factors for RRD can include peripheral retinal degeneration, higher refractive error and status post cataract surgery.² Approximately 0.3 to 0.5 percent of patients with lattice degeneration develop RRD.² In addition, 25 to 50 percent of the RRD cases are associated with lattice degeneration.² Patients with a spherical equivalent refractive error of -1.00 to -3.00 diopters have a four times greater risk of developing RRD compared to emmetropic patients.² Patients with a spherical equivalent refractive error greater than -3.00 diopters have a 10 times greater risk for developing RRD compared to emmetropic patients.² High myopes have an increased risk for RRD due to anomalous vitreous interaction with the retina and thin peripheral retina from axial length elongation. Although the prevalence of myopia in Asian countries is approximately 1.5 to 2 folds higher than the age-matched norm population in Caucasians, studies had not found a consistent higher incidence of RRD in Asian countries.²

Patients with RRD often complain of flashes, floaters, curtain vision, shadows or reduced vision. However, patients with RRD can also be completely asymptomatic, which was estimated to be in 10 percent of the patients with RRD.³

Case Report

Patient R.K., a 38-year-old Asian male, complained of a sudden onset "black-grey shadow" at the right lower corner of his vision. He reported this shadow took over one third of his peripheral vision and had remained at the same location since the onset of the symptom approximately two days prior. R.K.'s ocular history was significant for positive HLA-B27 anterior uveitis secondary to

ankylosing spondylitis. This condition was diagnosed approximately 10 years ago without any ocular involvement at the time. R.K. denied photophobia and reported that he was not taking any medication for ankylosing spondylitis due to the limited symptoms of stiffness. His last physical evaluation with his internist approximately one year ago was unremarkable with stable, mild ankylosing spondylitis. No medical treatment was recommended, except for daily exercise.

At the patient's last annual eye examination, he was diagnosed with his first episode of anterior uveitis OS secondary to ankylosing spondylitis. R.K. was monitored and treated with pred-forte 1 percent and homatropine ophthalmic eye drops. Uveitis resolved with treatment.

R.K.'s entering visual acuity at distance was 20/20 OD and 20/20-1 OS with his habitual spectacles correction of -7.00 -2.00 x170 OD and -6.75 -2.00 x170 OS. Anterior segment evaluation was unremarkable without ciliary injection, keratic precipitates, or posterior synchiae OU. Mild anterior chamber reaction of three cells per view without flare was found in the right eye. Anterior chamber was deep and quiet in the left eye.



2%

Figure 1: Visual field defect OD found by Frequency Doubling Technique (FDT)



Figure 2: Tobacco dust or Shafer's sign OD

quadrants of the peripheral retina encroaching toward the posterior pole and inside the superior temporal arcade. Retinal detachment was present from approximately 11:00 to 4:00. A shallow, mildly undulated retinal detachment was also found inferior to the bullous retinal detachment encroaching toward the macula and optic nerve head. The shallow retinal detachment was approximately a half disc diameter (DD) away from the fovea and 1 DD away from the superior rim of the optic nerve head (Figure 3 a & b). Macula OD was threatened but intact. A small horseshoe-shaped retinal tear with adjacent retinal hemorrhage was found at 1:30 OD with no other tears or holes. Fundus evaluation of the left eye was unremarkable.



sign or scattered retinal pigment epithelium (RPE) pigments floating in the anterior vitreous OD (Figure 2). Right fundus evaluation revealed corrugated, bullous retinal

detachment extending from the superior to the superior nasal

Visual field testing by Frequency Doubling Technique (FDT) Screening C-20-1 found inferior temporal defects OD that corresponded with patient's visual complaint (Figure 1). Goldmann tonometry found symmetric IOP of 13 OD and 13 OS at 11:10 am. Dilated eye exam revealed positive Shafer's

Figure 3a and 3b: Bullous retinal detachment extending from superior nasal to superior temporal quadrants, combined with a shallow retinal detachment inferior to the bullous detachment OD, as indicated by white dashed line.

man

Referral

An urgent referral to the local ophthalmologist for macula threatened retinal detachment was made immediately after the diagnosis. R.K. was sent to the ophthalmologist for RRD repair in a yellow cab and he was treated on the same day. R.K. was advised not to drink or eat until he talked to the ophthalmologist due to the potential risk for food or fluid aspiration if general anesthesia and scleral buckle were indicated for his retinal detachment repair. Patient education on symptoms of retinal detachment and risk of developing retinal detachment in fellow eye due to high myopia was given to R.K. He was advised to see an eye doctor immediately with seeing flashes, shower of floaters or experiencing side vision loss. R.K's symptom only lasted for about two days prior to his treatment without macular involvement, so he had better visual prognosis compared to patients with macular off for longer than one week. With a successful retinal detachment repair, it was likely that he would continue to have 20/20 best corrected visual acuity in his right eye. However, he may encounter residual haze or visual disturbances at the periphery due to history of sensory retina detached from the retinal pigment epithelium.

A few hours after the referral, R.K's surgeon faxed his brief examination findings and treatment regimen to the office stating that "steamroller pneumatic retinopexy" was performed for "macula-threatened retinal detachment" on the same day. An exam summary soon followed the fax, which confirmed the fundus findings and the progressive nature of this particular rhegmatogenous retinal detachment. The ophthalmologist noted that he believed R.K. would have had a macula-off retinal detachment by the end of the day without the prompt referral. Steamroller pneumatic retinopexy was performed to push the fluid away from the fovea and prevent macula detachment during the surgery. It was then followed by laser photocoagulation at the location of the retinal tear OD one day after the initial procedure for pneumatic retinopexy. The initial surgery outcome was unremarkable and R.K.'s symptom of the black shadow had mostly gone away.

The seven-month follow-up with the surgeon revealed healthy and intact retina OD status post pneumatic retinopexy and laser photocoagulation treatment for the retinal detachment



Figure 4: Intact and flat retina OD status post steamroller pneumatic retinopexy.



Figure 5: Laser scar at 1:30 OD status post retinal tear repair.

(Figure 4 and 5). There was no change in R.K.'s pre-surgical best corrected visual acuity of 20/20 OD. Dilated fundus examination revealed no new tear or retinal detachment.

Discussion

Based on R.K's retinal presentation, the three differential diagnoses that were considered included rhegmatogenous, serous and exudative retinal detachments. Exudative or serous retinal detachment occurs when subretinal fluid accumulates underneath the sensory retina leading to retinal detachment. The etiology is often associated with inflammation or tumor. A tractional retinal detachment occurs when pre-retinal fibrovascular or fibrotic membrane pulls the retina anteriorly causing detachment. Some of the common etiologies for tractional retinal detachment include diabetes, connective tissue disease, sickle cells and trauma.⁴ Unlike rhegmatogenous retinal detachment, serous and tractional retinal detachments are not secondary to a retinal tear or break.⁴ Upon dilation, a horse-shoe shaped retinal tear was found at 1:30 of the right eye and the bullous retinal detachment was found to originate from that clock hour. This had concluded the final diagnosis for R.K: rhegmatogenous retinal detachment.



Exudative or serous retinal detachment occurs when subretinal fluid accumulates underneath the sensory retina leading to retinal detachment.

For rhegmatogenous retinal detachment to occur, there has to be a tear or break where the overlying liquefied vitreous has access to the underlying retina causing the detachment. When the sensory retina detaches from the underlying retinal pigment epithelium and the choroid, photoreceptors lose their nutrient and blood supply. This can lead to permanent damage if the duration of the detachment is significant. Unlike the rest of the retina, fovea has no dual blood supply from the retinal and choroidal vessels so macula-off retinal detachment would deprive the fovea of its sole blood supply from the



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choroid⁴ and often result in poor visual outcome. Only 28 percent of these patients achieve 20/40 with a macula-off retinal detachment.²

Pneumatic retinopexy is a surgical procedure where a gas bubble is injected into the vitreous to reattach the detached retina and to occlude the retinal break. This can either be performed with the traditional or the steamroller method. Laser photocoagulation is applied to the retinal tear one to two days after pneumatic retinopexy to seal the break. Alternatively, cryotherapy can be used instead of laser photocoagulation. Cryotherapy is usually applied before pneumatic retinopexy on the same day. However, cryotherapy could potentially release more retinal pigment epithelium pigments into the vitreous inducing proliferative vitreoretinopathy (PVR).

Pneumatic retinopexy mechanically reattaches the retinal layers and creates adhesion between the retinal pigment epithelium and the choroid. This would allow the retinal pigment epithelium to absorb the subretinal fluid subsequently.⁵ Pneumatic retinopexy is indicated for posterior, bullous or macula-involved retinal detachment, superior break and small break less than one to two clock hours.⁶ If there is macula involvement from RRD, pneumatic retinopathy often gives better visual outcome compared to scleral buckle.⁷ Some complications associated with pneumatic retinopexy includes new or missed breaks, reopening of break, cataract, cystoid macular edema, epiretinal membrane and proliferative vitreoretinopathy.⁵⁻⁷ Patients who have received pneumatic retinopexy are usually followed at one day, three days, one week, two weeks, four weeks, three months, six months, and then every six months.⁶ For R.K., he was examined postoperatively at one day, two days, one week, two weeks, four weeks, two months, seven months and then annually.

Unlike the traditional pneumatic retinopexy, steamroller technique requires the patient to face downward initially as the gas bubble is injected into the eye. Over 10-15 minutes, the surgeon would position the patient's head until the bubble rolls from the normal, non-detached retina to the detached area.⁶ This technique is indicated when there is retinal detachment within the arcades, bullous retinal detachment or a risk of iatrogenic macular detachment.⁵ However, by rolling the gas bubble toward the retinal tear, this could push more subretinal fluid and retinal pigment epithelium cells into the vitreous and induce proliferative vitreoretinopathy.⁷

Two types of gas that are most commonly used for pneumatic retinopexy are sulfur hexafluoride (SF6) and perfluoropropane (C3F8).⁵ About 0.3-0.5ml of the gas is injected into the vitreous. As C3F8 can expand larger than SF6 inside the vitreous, it can be used with large or extensive retinal breaks.⁵ However, C3F8 (30-45 days) takes much longer to dissipate compared to SF6 (10-14 days) so C3F8 may induce more complications, such as induced tear from bubble movement.⁵

In addition to the pneumatic retinopathy, scleral buckle can also be performed as a treatment for retinal detachment. Scleral buckle is often the treatment choice if retinal detachment is extensive or involving the inferior part of the retina. Scleral buckle repairs RRD by reducing the traction. It is the preferred procedure over pneumatic retinopexy when there are giant breaks, multiple breaks, no apparent break or dialysis. Complications associated with scleral buckle include new intraoperative retinal break, hemorrhage, choroidal detachment, proliferative vitreoretinopexy, ocular hypertension, glaucoma, diplopia and induced myopia by approximately one to two diopters.¹

With pneumatic retinopexy, the patient will need to maintain a certain head position 16 hours a day for 5 days since the bubble needs to be positioned to appose the detached retina for this technique to work properly.

The anatomical surgical success rate between scleral buckle and pneumatic retinopexy are similar.⁷ With a single operation, scleral buckle was found to have 75-85 percent success rate and pneumatic retinopexy was 84-90 percent.⁷ With multiple operations combining both scleral buckle and pneumatic retinopexy, success rate for retinal detachment repair was found to be 95-100 percent.⁷ Visual outcome with either procedures is good, but pneumatic retinopexy usually renders better outcome if macula was off for less than one to two weeks.⁶ The patient can return to normal posture after the scleral buckle procedure. With pneumatic retinopexy, the patient will need to maintain a certain head position 16 hours a day for 5 days since the bubble needs to be positioned to appose the detached retina for this technique to work properly.⁵ Scleral buckle is significantly more expensive by approximately four to 10 times compared to pneumatic retinopexy since general anesthesia and a surgery room are needed.⁵

Unlike bullous retinal detachment, shallow detachment can be very difficult to detect because there is no obvious loss of red fundus reflex or retinal elevation. However, one can look for an opaque, slightly out of focus, undulated, orange-peel like retina with folds or slight elevation. Having stereopsis will be critical for detecting subtle retinal changes. Scleral indentation and 3-mirror fundus contact lens can also be used to help clinicians to detect subtle retinal elevation. Visual field testing can also be useful in determining the location of the retinal detachment. Lastly, optical coherence tomography (OCT) and fundus photography are helpful (although not necessary) tools for detecting shallow retinal detachment.

Some patients with retinal detachment have slightly lower intraocular pressure in the affected eye due to decreased aqueous formation.⁸ However, some patients would also have elevated intraocular pressure due to preexisting glaucoma, ocular hypertension or Schwartz-Matsuo Syndrome.⁸ There are several papers written on an interesting syndrome called Schwartz-Matsuo Syndrome status post chronic untreated rhegmatogenous retinal detachment. These patients had tiny nonpigmented particles in the anterior chamber and elevated intraocular pressure with open anterior chamber angle. Pain, photophobia or ciliary injection were not present in these patients.⁹ The anterior chamber reaction can range from 1+ to 4+ cells and the intraocular pressure elevation was found to range from 21 to 56 mmHg.⁹⁻¹⁰

Transmission electron microscopy of the aqueous humor collected from patients with Schwartz-Matsuo Syndrome found rod photoreceptor outer segments, possibly released from the detached retinal tear, and no inflammatory cells were found.⁹ From cadaver experiment of the eyeball, these rod photoreceptors' outer segments blocked the trabecular



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meshwork outflow and led to intraocular pressure elevation.^{8-9, 11-12} Topical glaucoma and steroid medications could not lower the intraocular pressure.¹¹ Upon retinal detachment treatment, intraocular pressure went back to normal and the cells found in the anterior chamber resolved.¹¹ These findings suggested a non-inflammatory process that could potentially masquerade anterior uveitis due to the manifestation of the cells in the anterior chamber. However, the origin of these cells was from the RRD and its corresponding retinal tear. For Schwartz-Matsuo Syndrome to occur, the retinal break had to involve oral dialyses and a tear of the nonpigmented epithelium of the ciliary body.^{10, 12-13} Approximately 50 percent of the Schwartz-Matsuo cases had retinal tears secondary to blunt trauma.¹⁴

For R.K., the cells detected in the anterior chamber were most likely residual low grade anterior chamber reaction from his history of anterior uveitis secondary chronic ankylosing spondylitis. Schwartz-Matsuo Syndrome was ruled out in R.K.'s case because of the sudden onset of symptoms and negative history of blunt trauma. There was no scarring, fibrosis or a pigmented line that demarcated the border of the retinal detachment that would suggest the chronicity of the retinal detachment.⁴ R.K. also had low teens intraocular pressure in both eyes. Although R.K. denied increased photophobia, he reported stable low-grade photophobia for years. Nonetheless, it is important to include Schwartz-Matsuo Syndrome as a differential diagnosis for patients with anterior chamber reaction, elevated intraocular pressure and retinal detachment.

Conclusions

This case study discussed the clinical findings, diagnostic tools, risk factors, treatment options, epidemiology and the physiology of rhegmatogenous retinal detachment. In most cases, bullous retinal detachment can be easily detected and diagnosed. However, shallow retinal detachment can often be missed and the urgency of the referral could be mistaken. It is important for clinicians to look for subtle retinal changes and determine the most appropriate treatment and management plan. Schwartz-Matsuo Syndrome was considered as diagnosis due to positive findings of anterior chamber reaction and retinal detachment in this patient. This differential diagnosis was quickly ruled out due to nonconcurring clinical findings. However, it was of interest and educational value to review Schwartz-Matsuo Syndrome.

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