Segmental Scleral Buckle Surgical Technique for Repair of a Rhegmatogenous Retinal Detachment in Retinoblastoma: A Case and Review of the Literature

Sydney P. Wendt^a Dianne A. Barrett^b Emmanuel Y. Chang^c Amy C. Scheffler^{b, d}

^aBaylor College of Medicine, Houston, TX, USA; ^bCizik Eye Clinic, University of Texas Health Science Center, Houston, TX, USA; ^cRetina and Vitreous of Texas, Houston, TX, USA; ^dRetina Consultants of Houston and Blanton Eye Institute, Houston Methodist Hospital, Houston, TX, USA

Established Facts

- Rhegmatogenous retinal detachments (RRD) are a known complication in patients with retinoblastoma who are treated with focal treatments such as cryotherapy or systemic treatments such as systemic or intra-arterial chemotherapy.
- Invasive surgery for the treatment of RRD in patients with retinoblastoma is known to increase the risk for extraocular tumor relapse.

Novel Insights

- We present a successful case of RRD repair in a patient with retinoblastoma via a posterior segmental scleral buckle without subretinal fluid drainage and without extraocular tumor relapse.

Keywords

Rhegmatogenous retinal detachment · Retinoblastoma · Segmental scleral buckle

Abstract

Purpose: The aim of this study was to describe the surgical repair of a rhegmatogenous retinal detachment (RRD) with a posterior break in a child with retinoblastoma (RB). **Methods:** Retrospective case report and review of the English language literature. Retrospective review of an 11-month-old male with bilateral retinoblastoma who developed a RRD

with a posterior retinal break in his better-seeing eye after treatment with cryotherapy. A review of all published cases to date of RRD in patients with RB is presented. **Results:** The patient underwent a posterior segmental scleral buckle without subretinal fluid drainage with successful reattachment of the retina and no extraocular extension of RB. **Conclusions:** RRDs in RB patients may be successfully repaired with anatomic success and no extraocular tumor extension. Even for patients with a posterior break, a segmental scleral buckle without drainage of subretinal fluid is a viable option and long-term excellent vision is a possible outcome.

© 2020 S. Karger AG, Basel

Introduction

Retinoblastoma (RB) is the most common primary intraocular malignancy in children [1]. Decades of clinical research have led to improved globe salvage rates and decreased mortality. To date, treatment options include chemotherapy (intra-arterial, systemic, intravitreal), cryotherapy, diode laser, plaque radiotherapy, and enucleation [2]. As with most treatment approaches, each modality has advantages, disadvantages, and risks. In eyes with retinoblastoma, rhegmatogenous retinal detachments (RRD) have been reported after focal treatment such as cryotherapy or after systemic treatments such as systemic or intra-arterial chemotherapy [2, 3]. Older publications have reported that the presence of an RRD was an automatic indication for enucleation [4]. However, more recent publications have detailed the treatment of RRDs with various techniques and good results. We report the surgical technique for a case of a successfully treated posterior RRD, which occurred after cryotherapy in a RB patient and review the literature on this rare complication.

Case Report

An 11-month-old boy was diagnosed with germline bilateral RB at 4 months of age. He was treated with systemic chemotherapy (carboplatin, vincristine, and etoposide) as well as focal laser therapy in both eyes at an outside institution for 6 months. Examination at presentation to our institution revealed grossly inactive disease with multiple types 3 and 4 regression patterns in both eyes. Over the next 6 months, he underwent 5 cycles of intravitreal chemotherapy (melphalan) in the right eye, one cycle of intravitreal chemotherapy in the left eye, transpupillary thermotherapy laser in both eyes, and one application of cryotherapy in the right eye, all for small recurrences. His disease was quiescent for the next 8 months. After this period, he had an additional small recurrence in the right eye, which was treated with one cycle of intravitreal melphalan and transpupillary thermotherapy laser. A persistent fish-flesh area in the left eye was also treated with laser. Three months later, he had 2 new, small recurrences in each eye, both of which were treated with cryotherapy. A month after treatment, he was found to have a macula-involving RRD in the better seeing right eye with a posterior slit-like retinal break at the edge of the cryotherapy scar 2.5 disc areas inferior to the optic nerve with hyaloidal contraction (Fig. 1). The contracture of the posterior hyaloid interface was evident because there appeared to be an early tractional component just anterior to the break. There were no changes in the vitreous base, and no posterior vitreous detachment.

He underwent a posterior segmental scleral buckle (SB) with the procedure performed in the following fashion. Preoperative acetazolamide was given to soften the eye to permit greater imbrication and buckle height. The recti muscles were isolated with 2–0

silk sutures after a conjunctival peritomy. The posterior break was then treated with retinal cryopexy located approximately 6–7 mm inferonasal to the optic nerve. Using thin small malleable retractors with 1/4 inch cottonoids, any fluid accumulation in the orbital space as well retracting orbital tissue was wicked away. This technique preserved visualization of the posterior sclera to the optic nerve. Once good posterior visualization was achieved, the posterior retinal break is marked externally with a scleral marking depressor. Preplacement of a 5–0 nylon suture in a horizontal mattress for a radial 506 sponge was then done. A 506 segmental sponge was then placed in a radial fashion and the nylon suture temporarily tied secure. A 30-gauge needle was then used to perform an anterior chamber paracentesis through clear cornea. This allowed further softening of the eye, which permitted greater imbrication of the horizontal mattress suture as it was further tightened and sutured in place. The bed of the sponge was then irrigated with antibiotic solution. An anterior anchoring suture was then placed on the 506 sponge as well as a layered closure of tenons over the sponge to prevent future anterior migration. The retina was then inspected to make sure it was perfused. Anesthesia was asked to raise the systemic blood pressure of the patient 10–20 mm Hg to facilitate retinal perfusion, which was confirmed. Oral diamox was then administered during the next several days after surgery to try to generate a greater indentation effect from imbricating sutures due to a softer globe.

Three years postoperatively, his retina has remained attached and both eyes have no active RB (Fig. 2). The patient's vision is 20/50 in the right eye and 20/200 in the left eye.

Discussion

RRDs were rare in the era of external beam radiation use for RB. Over the past 3 decades, as external beam radiation has been abandoned and newer modalities introduced, unsurprisingly, there has been a change in the types of vitreoretinal complications observed. Approximately 60 cases of RRDs have now been reported in RB patients after various newer treatment approaches [2, 5].

Previous studies have demonstrated that the mechanism of action of RRD in eyes with RB which received laser or cryotherapy and then developed a retinal break was an area of retinal thinning at the site of cryotherapy or laser photocoagulation [6]. Histopathology after cryotherapy has confirmed significant atrophy of the retina and choroid at the site of treatment with associated retinal pigment epithelium hyperplasia. Some studies have attributed poor RPE function due to chemotherapy exposure as the reason that microbreaks do not heal as well in these patients as they would in other patients [7]. RRDs have recently been reported after IAC in RB. In a retrospective series, Shields et al. [2] noted a 6% (10/167) risk for RRD in patients treated with IAC. Patients who developed a RRD tended to have a high tumor burden, and the

Fig. 1. Fundus photograph (**a**) showing healed recurrence at the posterior edge of a lesion at 4:00 with new slit-like break (arrow) and RRD demarked by blue lines. B-scan (**b**) showing tear at 5:00 posterior to the equator (red arrow) associated with RRD extending from 4:00 to 8:30.

Fig. 2. Fundus photograph (**a**) of the right eye taken 3 years after repair. The RRD has completely resolved. Widefield photograph (**b**) of the right eye 3 years after repair demonstrating complete resolution of the RRD and regressed RB.

RRD was attributed to atrophic holes that developed after full-thickness rapid tumor regression [7].

RB patients with RRD in earlier reports were all managed with enucleation due to the belief that intraocular management of the RRD created an elevated risk for extraocular tumor relapse [4, 8]. Spaulding and Fuhs [4] and Stevenson et al. [8] reported a total of 4 cases of extraocular extension of RB after incisional surgery. Due to the reasonable concern of extraocular extension, there is a need to treat RDs in patients with RB without violating the globe. Including our case, other authors (Table 1) have reported successful RRD repairs utilizing SB without subretinal fluid drainage. None of these authors reported extra-ocular recurrences.

Anatomic reattachment rates and visual prognosis of RRD repair in RB are variable, but generally promising (Table 1). Forty-four of the 67 cases (67%) reported in the English language literature resulted in complete reattach-

ment of the retina. The most recent and largest series by Shields et al. [2] reported that in 10 cases of RRD after RB treatment, 4 patients underwent surgical repair, 3 of which remained attached after an average of 24 months of follow-up.

While our case documents a successful RRD repair using a posterior segmental SB without subretinal fluid drainage technique, there are some studies that report complications with a SB procedure (Table 1). A case series by Honavar et al. [9] reports complete retina attachment in 6 out of 8 patients who underwent a SB procedure. However, 5 patients developed RB recurrence, 4 patients needed enucleation, and 1 patient developed systemic metastasis [9]. Another complication that has been reported after a SB procedure is tractional RD. Stathopoulos et al. [10] suggest the use of pars plana vitrectomy with melphalan irrigation, with retinectomy if necessary, to manage tractional RD.

Table 1. Summary of visual and anatomic outcomes in patients with RRD following RB treatment

First author [ref.]	Year	Cases	Quiescent interval (last active RB to RD repair)	Surgical treatment	Final retina status	Visual outcome
Pruett [12]	1975	1	3.5 months	SB w/external drainage (1)	Complete reattachment (1)	Not reported
Buzney [13]	1984	3*	6 weeks–3.5 months	SB w/ external drainage (2), not reported (1)	Complete reattachment (3)	Not reported
Brooks [14]	1990	1	3.5 years	SB, vitrectomy (1)	Complete reattachment (1)	Not reported
Mullaney [3]	1997	1	11 months	SB w/o drainage (1)	Complete reattachment (1)	Not reported
Baumal [15]	1998	9	1 week–15 years	SB w/ external drainage (5), SB w/o drainage (2), PPV (1), Not repaired (1)	Complete reattachment (5), Persistent detachment then enucleation (4)	F + F (2), HM (1), CF at 1 foot (1), 20/200 (1)
Bovey [16]	1999	5	1–9 months	SB w/ drainage (2), No treatment (3)	Complete reattachment (2), Persistent detachment (2), RD reduced (1)	20/50 (1), 20/60 (1), Enucleated (1), Not reported (2)
Madreperla [17]	2000	4	9–15 years	SB w/o drainage, vitrectomy, and laser barricade (1) Vitrectomy, laser, and retinectomy (2), Vitrectomy and laser (1)	Complete reattachment (3), Inferior RD with preretinal fibrosis (1)	20/60 (2), HM (2)
Honavar [9]	2001	8	14 months	SB w/o drainage (5), SB w drainage (3)	Complete reattachment (6), Persistent detachment (2)	Not reported
Buerk [18]	2006	2	4.5–9 months	SB w/o drainage with indirect laser retinopexy post op (2)	Complete reattachment (2)	Not reported
Saunmya [19]	2010	9	1 month–15 years	SB w/o drainage (7), Vitrectomy with retinectomy using melphalan infusion and silicone oil tamponade (2)	Complete reattachment (9)	Not reported
Tanaka [5]	2014	3	1 month–3 years	SB and PPV (3)	Complete reattachment (3)	CF at 20 cm (1), 0.3 BCVA (1), F + F (1)
Yousef [1]	2016	3	4 weeks	SB w/o drainage (3)	Complete reattachment (2), enucleation (1)	6/9 (1), 6/12 (1)
Shields [2]	2017	10	1 weeks	SB w/o drainage (1), PPV (3), Laser barricade (1), No tx (5)	Complete reattachment (3), Persistent detachment (2)	Not reported
Elaraoud [20]	2018	5	2 months	SB (4), Observation (1)	Complete reattachment (3), Persistent detachment (2)	20/22 (1), 20/32 (1), 20/191 (1), 20/1383 (1), HM (1)
Stathopoulos [10]	2020	2	1–21 months	SB w/o drainage followed by PPV under melphalan irrigation (2)	Complete reattachment (2)	20/20 (1), 20/32 (1)
Current case	2020	1	1 month	Segmental SB w/o drainage (1)	Complete reattachment (1)	20/50 (1)

F + F, fix and follow; HM, hand motions; BCVA, best-corrected visual acuity; CF, count fingers. * One of the cases is from Pruett [12].

Our case presents a unique successful repair in a patient with a posterior retinal break. SB repairs of retinal detachments are typically reserved for peripheral retinal breaks while retinal detachments with very posterior breaks generally require vitrectomy to successfully repair [11]. Given our patient's recent history of retinoblastoma, concern for iatrogenic extraocular extension of RB precluded the use of vitrectomy for retinal detachment repair. Furthermore, since tamponade agents do not have to be utilized in SB, amblyopia risks are minimized in these young children with developing vision. We report the use of a carefully placed posterior segmental buckle without subretinal fluid drainage as an alternative means to successfully reattach the retina without vitrectomy in this setting. With the use of these safety techniques, we were able to safely and successfully perform the retinal reattachment surgery only 1 month after the patient's most recent active recurrence. This case is also unique in that we have long-term follow-up with an assessment of the child's visual outcome.

In conclusion, RRDs in RB patients are serious, but may be successfully repaired with good long-term visual and systemic outcomes. Careful surgical techniques should be considered when approaching RRD in RB eyes and attempts to salvage these eyes. We report herein the surgical technique for a successful case of RRD repair via posterior segmental SB without subretinal fluid drainage and without extraocular tumor relapse. With further innovations in RB treatment such as IAC, advanced disease can be successfully treated, and more eyes are being salvaged. However, we expect that the incidence of vitreo-retinal complications such as RRDs will likely increase over time as more eyes with more advanced disease undergo attempted salvage. Awareness of this complication

and its appropriate management is vital for ocular oncologists and retinal specialists who care for these patients.

Acknowledgement

There are no acknowledgments to declare.

Statement of Ethics

Our research complies with the guidelines for human studies and was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Written informed consent was obtained to publish details of case including the publication of images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding Sources

No funding or grant support.

Author Contributions

All authors attest that they meet the current ICMJE criteria for Authorship. A.C.S. and E.Y.C. contributed to the conception of the study, writing of the manuscript, and editing of the manuscript. D.A.B. and S.W. contributed to writing of the manuscript and editing of the manuscript. All authors approved the final version and agree to be accountable for all aspects of the work.

References

- 1 Yousef YA, Manna M, Khalil MB, Nawaiseh I. Surgical repair of rhegmatogenous retinal detachment in eyes harboring active retinoblastoma. *Ophthalmic Genet*. 2016 Sep;37(3):314–7.
- 2 Shields CL, Say EA, Pefkianaki M, Regillo CD, Caywood EH, Jabbour PM, et al. Rhegmatogenous Retinal Detachment after Intraarterial Chemotherapy for Retinoblastoma: The 2016 Founders Award Lecture. *Retina*. 2017 Aug;37(8):1441–50.
- 3 Mullaney PB, Abboud EB, Al-Mesfer SA. Retinal detachment associated with type III retinoblastoma regression after cryotherapy and external-beam radiotherapy. *Am J Ophthalmol*. 1997 Jan;123(1):140–2.
- 4 Spaulding AG, Fuhs JC. Retinoblastoma and retinal detachment. *Surv Ophthalmol*. 1968 Nov;13(3):152–6.
- 5 Tanaka M, Yokoi T, Ito M, Kobayashi Y, Noda E, Matsuoka K, et al. Three cases of rhegmatogenous retinal detachment associated with regressed retinoblastoma after conservative tumor therapy. *Retin Cases Brief Rep*. 2014;8(3):223–6.
- 6 Lim TH, Robertson DM. Presumed rhegmatogenous retinal detachment in patients with retinoblastoma. *Retina*. 2000;20(1):22–7.
- 7 Tawansy KA, Samuel MA, Shammas M, Murphree AL. Vitreoretinal complications of retinoblastoma treatment. *Retina*. 2006;26(7 Suppl):S47–52.
- 8 Stevenson KE, Hungerford J, Garner A. Local extraocular extension of retinoblastoma following intraocular surgery. *Br J Ophthalmol*. 1989 Sep;73(9):739–42.
- 9 Honavar SG, Shields CL, Shields JA, Demirci H, Naduvilath TJ. Intraocular surgery after treatment of retinoblastoma. *Arch Ophthalmol*. 2001 Nov;119(11):1613–21.

- 10 Stathopoulos C, Sergenti J, Gaillard MC, Munier FL, Daruich A. Pars plana vitrectomy under mephalan irrigation for recurrent retinal detachment in eyes treated for retinoblastoma: a case report. *BMC Ophthalmol*. 2020 Jan;20(1):34.
- 11 Heimann H, Bartz-Schmidt KU, Bornfeld N, Weiss C, Hilgers RD, Foerster MH; Scleral Buckling versus Primary Vitrectomy in Rhegmatogenous Retinal Detachment Study Group. Scleral buckling versus primary vitrectomy in rhegmatogenous retinal detachment: a prospective randomized multicenter clinical study. *Ophthalmology*. 2007 Dec; 114(12):2142–54.
- 12 Pruett RC. Scleral buckling in retinoblastoma: case report. *Ann Ophthalmol*. 1975 Aug;7(8): 1115–20.
- 13 Buzney SM, Pruett RC, Regan CD, Walton DS, Smith TR. Scleral buckling for retinal detachment in patients with retinoblastoma. *Am J Ophthalmol*. 1984 Oct;98(4):473–7.
- 14 Brooks HL Jr, Meyer D, Shields JA, Balas AG, Nelson LB, Fontanesi J. Removal of radiation-induced cataracts in patients treated for retinoblastoma. *Arch Ophthalmol*. 1990 Dec; 108(12):1701–8.
- 15 Bauman CR, Shields CL, Shields JA, Tasman WS. Surgical repair of rhegmatogenous retinal detachment after treatment for retinoblastoma. *Ophthalmology*. 1998 Nov;105(11): 2134–9.
- 16 Bovey EH, Fernandez-Ragaz A, Héon E, Balmer A, Munier FL. Rhegmatogenous retinal detachment after treatment of retinoblastoma. *Ophthalmic Genet*. 1999 Sep;20(3): 141–51.
- 17 Madreperla SA, Hungerford JL, Cooling RJ, Sullivan P, Gregor Z. Repair of late retinal detachment after successful treatment of retinoblastoma. *Retina*. 2000;20(1):28–32.
- 18 Buerk BM, Lai WW, Sharma MC, Shapiro MJ. Minimally invasive method for repair of rhegmatogenous retinal detachment following treatment for retinoblastoma. *Ophthalmic Surg Lasers Imaging*. 2006 Jan-Feb;37(1):82–5.
- 19 Saumya Pal S, Gopal L, Khetan V, Nagpal A, Sharma T. Rhegmatogenous retinal detachment following treatment for retinoblastoma. *J Pediatr Ophthalmol Strabismus*. 2010 Nov-Dec;47(6):349–55.
- 20 Elaraoud I, Ch'ng S, Karl D, Kalogeropoulos D, Chavan R, Sharma A. Management of retinal detachment in retinoblastoma with globe conserving treatment. *J Curr Ophthalmol*. 2018 Sep;31(1):43–8.