Scleral rupture during retinal detachment repair with primary scleral buckle and cryoretinopexy in a patient with microspherophakia

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ABSTRACT

Purpose: The purpose of this report is to describe a case of a patient with microspherophakia (MSP) who had a scleral rupture during a retinal detachment (RD) repair with primary scleral buckle and cryoretinopexy.

Observations: A 48-year-old woman with MSP presented with six days of expanding loss of vision and photopsias. Examination revealed a superior retinal detachment involving the macula associated with two superior retinal tears. The patient underwent successful placement of a segmental buckle. During cryoretinopexy treatment of the tears, a 4 mm full-thickness scleral rupture occurred. The sclera was immediately closed with interrupted 8-0 nylon sutures and reinforced with a processed pericardium allograft. Subsequent combined phacoemulsification with capsulectomy, zonulectomy, and pars plana vitrectomy with retinal reattachment was performed nine days post buckle placement.

Conclusions and importance: This case illustrates that a patient with MSP, even observed in the absence of a genetic syndrome or familial condition, may be at increased risk of scleral rupture during RD repair. Though future investigations are necessary to confirm this association, surgeons should take a conservative approach by having a high clinical suspicion for compromised scleral integrity in patients with MSP and proceeding with caution in procedures that may pose a risk of scleral rupture. A pericardium allograft can be an effective adjunct for scleral rupture repair.

1. Introduction

Scleral rupture during retinal detachment (RD) repair is a rare occurrence. Risk factors for developing a scleral rupture during RD surgery include reoperation following a previously failed RD repair and pre-existing scleral pathology including high axial myopia.1 Scleral rupture is a serious event which can result in further intraoperative complications and poor anatomic and visual outcomes.1 Microspherophakia (MSP) is a rare congenital condition defined by a small, spherical crystalline lens with increased antero-posterior thickness and decreased equatorial diameter.2,3 It typically presents bilaterally with visualization of the lens equator after pupillary dilation and antero-posterior motion of the lens with positional changes due to laxity of the lens zonular fibers.2 High refractive myopia due to the spherical lens, impaired accommodation, lens subluxation, and angle closure secondary to pupillary block are frequently observed in MSP.2-4 MSP can be found in isolation, as a familial condition with no other systemic abnormalities, or in association with genetic syndromes.2,3,4 It most frequently occurs in Weill-Marchesani syndrome and Marfan syndrome, but has also been seen in other inherited conditions such as Alport syndrome and Klinefelter syndrome.2,3,4

To our knowledge, this is the first report of a patient with microspherophakia who experienced scleral rupture during RD repair.

2. Case report

A 48-year-old woman with ocular history significant for bilateral microspherophakia, associated angle closure glaucoma status post bilateral peripheral laser iridotomies, and cataract extraction with anterior chamber intraocular lens implantation of the right eye presented to the emergency department with a 6-day history of expanding inferior hemifield vision loss and photopsias in her left eye. Visual acuity was 20/25 in the right eye and 20/50 in the left eye with normal intraocular pressures in both eyes. Anterior segment exam was notable for an anterior chamber intraocular lens implant in the right eye and cataractous microspherophakic lens with phakodonesis in the left eye. The

Abbreviations: MSP, microspherophakia; RD, retinal detachment; OCT, optical coherence tomography; PPV, pars plana vitrectomy.

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microspherophakic lens is illustrated in Fig. 1, which was obtained at a prior visit. Dilated fundus examination of the right eye revealed an atrophic hole in the inferonasal periphery. Examination of the left fundus demonstrated a superior retinal detachment extending clockwise from 9 to 2 o’clock, involving the macula extending up to the fovea (Fig. 2). There were two superior horseshoe tears at 11 and 1 o’clock. Optical coherence tomography (OCT) demonstrated partial posterior vitreous detachment with attached posterior hyaloid at the optic nerve and separation over the macula (Fig. 3). The axial length of the left eye was measured to be 24.71 mm.

Options for retinal detachment repair included pneumatic retinopexy, pars plana vitrectomy (PPV), and/or scleral buckle repair. Due to concerns regarding glaucoma and microspherophakia, and the risks of pupillary block with a gas bubble, the decision was made to proceed with a primary scleral buckle with cryoretinopexy.

Following 360-degree conjunctival peritomy and isolation of the rectus muscles, the quadrants were inspected and showed a small area of scleral thinning in the superotemporal quadrant. Cryoretinopexy was performed under visualization via indirect ophthalmoscopy with care to instill balanced salt solution after treatment to release the cryoprobe from the sclera. During treatment of the 1 o’clock tear, the globe became soft. Inspection of this area revealed a 4-mm full-thickness scleral defect. This was closed with five interrupted 8-0 nylon sutures. Due to the irregular nature of the scleral defect and continued wound leak, a processed pericardium patch graft (Tutoplast Pericardium, Katena Products, Inc, Parsippany, NJ) was secured over the scleral defect with 8-0 nylon sutures. An anterior chamber maintainer was placed to reform the globe and a segmental 510 sponge was secured spanning from the superonasal to superotemporal quadrants with 5-0 polyester fiber sutures in a horizontal mattress fashion. Indirect ophthalmoscopy of the posterior segment showed good scleral buckle effect with cryopexy re-action over the tears, subretinal hemorrhage originating super-temporally extending to the superotemporal arcade, and persistent subretinal fluid. The conjunctiva was closed with 6-0 plain gut suture.

On post-operative day 3, fundus exam demonstrated persistent superior retinal detachment with subretinal fluid extending up to the fovea. Localized subretinal hemorrhage was present in the area of the detachment near the superotemporal arcade. Though the buckle provided imbrication over the area of the superotemporal break, persistent vitreous traction prevented adequate support and closure. The presence of an open retinal break, subretinal hemorrhage, and detachment in close proximity to the fovea were indications to explore further surgical options. After discussing the options with the patient, the decision was made for a combined phacoemulsification with capsulectomy, zonulectomy and PPV, perfluorocarbon liquid exchange and 14% C3F8 gas injection nine days after the initial surgery. The retina was successfully reattached at the end of the case and the patient was left aphakic. The subretinal hemorrhage was not evacuated, remained localized during the immediate post-operative period, and spontaneously resorbed.

At post-operative-month four, the patient’s aphakic vision was 20/400 which improved to 20/60 with pinhole. She then underwent secondary anterior chamber intraocular lens placement at 4.5 months post vitrectomy with postoperative uncorrected visual acuity improving to 20/25.

3. Discussion

Scleral rupture during RD repair and MSP are both rare occurrences and have not been previously reported together in this setting. The pathogenesis of MSP is unknown. However, it is thought to result from insufficient nutritional support to the tunica vasculosa lenticis during the 5th and 6th months of embryogenesis when the lens is spherical, leading to arrested development or abnormal insertion of secondary lens fibers. As mentioned previously, MSP can occur as an isolated defect or as a familial condition, either without systemic involvement or as part of a known systemic syndrome. MSP has been associated with multiple connective tissue disorders including Weill-Marchesani and Marfan syndromes. In Marfan syndrome, there is a mutation in the FBN1 gene encoding for fibrillin-1, a protein which is a key component of many ocular structures, including the lens zonules and sclera. Numerous genes have been implicated in Weill-Marchesani syndrome, including FBN1; ADAMTS10, which is involved in connective tissue remodeling; ADAMST17, thought to be involved in extracellular matrix organization; and LTBP2, a protein with expression in the trabecular network, ciliary body, lens capsule, and lens epithelium layer which has been identified as a cause of isolated familial microspherophakia.

Given the critical role that these genes play in ocular structure and maintenance, it is reasonable to postulate that defects in these pathways could compromise scleral integrity and predispose to scleral rupture. Indeed, connective tissue disorders have been linked with scleral rupture in multiple case reports. There has been a reported case of recurrent spontaneous scleral rupture in a patient with Marfan syndrome and MSP and other cases of scleral rupture in Marfan syndrome and Ehlers Danlos syndrome that can be explained by the scleral...
thinning that is known to occur in these conditions.5-8,11 Interestingly, there have been two case reports of spontaneous filtering blebs with scleral thinning in patients with MSP and mild craniofacial dysmorphism; neither of these patients had clinical features of a known genetic syndrome.12,13

Following the scleral rupture, our patient did consult a geneticist who noted that she had short stature and ocular abnormalities that could be consistent with Weill-Marchesani syndrome, but she did not exhibit the brachydactyly and cardiovascular defects associated with this condition. Ultimately, the patient did not end up pursuing genetic testing. Since our patient had no confirmed or highly suspected underlying systemic genetic disorder nor any family history of MSP, we suggest this is likely an isolated case of MSP. Even in isolation, we hypothesize that the unknown defect underlying this case of MSP could be associated with compromised scleral integrity, resulting in the scleral rupture that occurred during RD repair. Surgical RD repair introduces conditions which can exacerbate underlying scleral pathology. Domínguez-Yates et al. reported a case of a patient with pathological myopia who developed a scleral rupture when silicone oil was injected during PPV.14 Pathological myopia can present with scleral thinning, which the authors suggest could have contributed to scleral rupture during a transient rise in intraocular pressure.14 A case of scleral rupture in a patient with no underlying risk factors has been reported during perfluorocarbon liquid injection.15 A previous study found that the highest percentage of scleral ruptures during RD surgery occurred secondary to scleral depression during cryotherapy or laser photoagulation, likely due to increased intraocular pressure.1 In our case, scleral rupture likely resulted from application of cryoretinopexy in the setting of underlying compromised scleral integrity.

The immediate intraoperative goals following an unexpected scleral rupture are preservation of intraocular contents and restoration of intraocular pressure.1 In our patient, nylon sutures were initially used to close the sclera followed by reinforcement with a processed pericardium allograft. Stunf et al. reported a case of scleral thinning which was identified during a scleral buckling procedure and successfully treated with a donor scleral graft, avoiding rupture.16 In the case of a patient with Ehlers Danlos syndrome and scleral rupture, a 360-degree scleral donor graft was utilized for scleral reinforcement, with the authors suggesting this as an option for patients with severe scleral ectasia who are not good candidates for a scleral buckle procedure.10

4. Conclusions

Scleral rupture is a rare but serious intraoperative complication during RD repair which threatens visual outcomes. Patients with MSP may be at increased risk of scleral rupture, whether or not they have a known underlying connective tissue disorder. Further studies are necessary to support and elucidate the potential association between isolated MSP and compromised scleral integrity. For patients with MSP, surgeons should be aware of the potential for compromised scleral integrity and proceed with caution during the steps of a surgical procedure that may raise intraocular pressure or pose a threat for scleral rupture. In cases where unexpected scleral rupture occurs, use of a processed pericardium allograft can be an effective adjunctive method for use in the surgical repair strategy.

Patient consent

Written consent to publish the case report and associated images was obtained. This report does not contain any personal 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

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