Retinal Detachment With Macular Holes in Infants With Retinopathy of Prematurity

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Objective: To describe retinal detachment with macular holes in infants with retinopathy of prematurity and the methods of surgical repair with their outcomes.

Methods: A retrospective review of 4 cases of retinopathy of prematurity and 1 case of retinopathy in a full-term infant that resembled retinopathy of prematurity, in which a macular hole and associated retinal detachment developed and surgical repair was attempted.

Results: The average gestational age of the 4 infants with retinopathy of prematurity was 26 weeks. All 5 patients had a history of vitreoretinal surgery prior to discovery of the macular hole. All 4 who underwent further surgical correction have partially or completely attached retinas and ambulatory vision.

Conclusions: Vitrectomy combined with fluid-air exchange failed to close a macular hole associated with retinal detachment in infants with retinopathy of prematurity. Athermal scleral buckling of the macula can close macular holes and reattach the retina. Scleral buckling appears to be the method of choice for repair.

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The patient is a female born at a gestational age (GA) of 28 weeks and with a birth weight (BW) of 1200 g. At 7 months of age, she had leukocoria and stage 5 ROP in both eyes. The detachment had an open-open configuration in the left eye, an open-sky vitrectomy was performed. When the patient was 10 months of age, the retina was attached on the nasal side of the disc, but there was a suspicious macular hole with detachment of the temporal retina. At this time, a circumferential scleral buckle was placed with subtretinal fluid drainage. The macula was left untouched, and by 11 months of age, the temporal retina was still detached and a closed vitrectomy with fluid-air exchange was performed. The subtretinal fluid was aspirated with a low suction through the macular hole by placing a tip of the back-flush needle in front of the macular hole. As the air exerted a posterior force on the retina during the fluid-air exchange, the macular hole enlarged to almost the size of the entire temporal retina (Figure 1). The edge of the hole remained elevated, but the retina nasal to the disc stayed attached. Endolaser was then applied at the nasal edge of the tear to prevent the detachment from extending to the attached nasal retina. Currently, the patient is 3 years old with the nasal retina staying attached in the left eye, a large macular hole, and 20/3270 visual acuity (VHD) in the left eye. The vision in her right eye, which was not treated surgically, has no light perception (NLP).

CASE 2

The only surviving twin, a female, was born at 25.5 weeks GA and with a BW of 750 g. She was diagnosed as having stage 4A ROP in the right eye, stage 3 in the left eye, and moderate-plus disease in both eyes at 8 weeks of age. Both eyes had ROP in zone 1. The window for treatment of threshold disease had passed to the right eye. Cryotherapy was performed in the right eye and laser in the left to reduce the neovascular activity. However, the disease progressed to stage 5 bilaterally; the right eye remains detached despite open-sky vitrectomy and currently has NLP visual acuity. Ultrasonography at 23 weeks of age revealed a funnel-shaped total RD with an open-closed configuration in the left eye, at which time open-sky vitrectomy was performed. The retina reattached in the left eye and remained attached until 33 months of age when a shallow RD with preretinal organization and subretinal bands developed. One month later, a total RD developed in the left eye, and a circumferential scleral buckle was placed. The retina was temporarily reattached, but at 38 months of age, the retina was totally detached again with a macular hole and extensive subretinal organization. At this time, a pars plana vitrectomy, membrane peel, and air-fluid exchange were performed to remove the fluid in front of the detached retina. With the retina still detached and subretinal fluid remaining, the macular hole was plugged with cyanoacrylate glue applied directly to the hole in the air-filled vitreous cavity. The retina did not re-attach even with drainage of subretinal fluid 1 month later. At 42 months of age, a radial scleral buckle with a sponge No. 506 was used to close the macular hole. No treatment with cryotherapy or laser was applied to the hole (athermal buckle). The retina reattached and the visual acuity was 20/760 (VHD) 1 year and 6 months after the last surgery. The family has since moved, and 9 years after surgery, the local ophthalmologist, who has observed this patient, reports that the cyanoacrylate glue persists and the retina remains attached (Susan G. Elner, MD, written communication, September 2003).

CASE 3

The only surviving triplet, a female, born at 24 weeks GA and with a BW of 622 g, was diagnosed as having stage 3, zone 2 ROP in both eyes and was treated with cryotherapy bilaterally at 11 weeks of age. The treatment failed in both eyes, and the disease progressed to total RD and phthisis bulb in the right eye. The retina in the left eye detached at 15 weeks of age and was reattached with circumferential scleral buckle at 19 weeks of age. The retina remained attached until there was temporal dragging and RD with macular hole formation at 15 months of age. The RD was limited to a pie-shaped area with the tip at the disc temporally (Figure 2). This did not resolve, and at 33 months of age, a radial sponge was used to buckle the macular hole. There was minimal buckling effect, and the retina remained detached with an open macular hole. The sponge was replaced by another one with closure of the hole and reattachment of the retina. The retina has remained attached at 13 years of age, and the visual acuity is counting fingers at 2 feet.

CASE 4

The patient is a female, born at 27 weeks GA and with a BW of 1170 g; at 11 months of age, she had stage 5 ROP in the right eye and stage 4A in the left eye. The retina in the left eye remains attached in the macular area with inferotemporal detachment and nasal dragging at the present time (10 years of age). Ultrasonography of the right eye showed a total RD with a tight-tight funnel configuration. An open-sky vitrectomy was performed at 12 months of age. By 14 months of age, the retina was partially attached, but a macular hole developed. The retina subsequently totally detached with NLP visual acuity. No fur-
ther intervention was attempted considering the stable condition of the fellow eye that had useful visual acuity (20/70).

CASE 5

The patient is a male, born at 41 weeks GA and with a BW of 3600 g; he was diagnosed as having hypoplastic left heart syndrome at birth. Before being examined at our clinic, the patient was diagnosed as having total RD in the right eye, and a closed vitrectomy and lensectomy were performed in that eye at 6 months of age. At this time, the avascular retina was also ablated with diode laser. The retina redetached in the right eye, and another vitrectomy was performed in that eye, which was unsuccessful. The child was suspected to have familial exudative vitreoretinopathy. However, there was no family history of retinal pathologic features, and the patient’s parents and 5 siblings were examined and found to have no signs of any retinopathy. On our initial examination, the retina was totally detached and visible in the pupil, and the patient had a visual acuity of NLP in the right eye. There was a dense vitreous hemorrhage in the left eye, and a closed vitrectomy with endolaser to avascular peripheral retina was performed. The retina later detached in the left eye with vitreous hemorrhage, and another closed vitrectomy with endolaser was performed at 12 months of age, with attachment of the retina. At 14 months of age, the retina was once again detached, and a circumferential scleral buckle with subretinal fluid drainage was performed. The retina remained reattached until 24 months of age when the buckle was removed to allow growth of the eye. At the time of surgery, a macular hole was noted with detachment of the macular area. Indirect laser was performed to demarcate the area of detachment and limit the spread. At 26 months of age, the retina was totally detached, and a radial sponge was placed to support the macular hole. The retina reattached with some preretinal fibrosis. At 4 years of age, the retina redetached, and a closed vitrectomy with membrane peel and silicone oil injection was performed. The patient is currently 4 years old with attached retina in the left eye and 20/360 visual acuity (PL).

RESULTS

Three of 4 of the patients who underwent attempted repair of RD with macular hole have at least ambulatory vision in the repaired eye even years after the surgery (Table). The fellow eye in each case has NLP vision. The average GA and BW of the premature infants were 26 weeks and 936 g. Two patients (cases 2 and 3) had ultimate closure of the macular hole and retinal reattachment with a macular buckle. A third patient (case 5) also had reattachment with a macular buckle, but the retina redetached with opening of the macular hole. This patient required another closed vitrectomy and silicone oil tamponade to reattach the retina and close the macular hole. All 3 patients who had macular buckles placed (cases 2, 3, and 5) also had circumferential buckles placed in a previous surgery. One patient (case 2) with a macular buckle also had a prior attempt at closure of the hole with cyanoacrylate glue that failed. The 1 patient (case 1) who underwent closed vitrectomy with fluid-air exchange had enlargement of the tear and required endolaser to preserve the attached nasal retina. The 1 patient (case 4) in whom there was no intervention for the macular hole still has a totally detached retina in that eye.

COMMENT

There have been no reports in the literature regarding macular holes in infants and children with ROP. The success rates of surgical repair of idiopathic macular holes in adults have been reported to be as high as 80% to 90% and even 95% when considering reoperations. This has led to attempted surgical repair of macular holes of various etiologies. Success has been reported with traumatic macu-
lar holes both in adults and in the pediatric population. This success, however, has been in macular holes without any significant RD. All our cases had an RD. It is unusual for macular holes to be associated with RDs in adults, but there has been success with standard vitrectomy and gas fill in these patients. Our results show it is possible to successfully treat RDs from macular holes due to ROP with scleral buckling. Scleral buckling was the standard technique for repair of RD caused by macular hole in adults before the advent of vitrectomy.

Vitrectomy with fluid-air exchange, the standard technique for repair of RD with macular hole in adults, did not work for infants with ROP. Case 1 involved a vitrectomy with fluid-air exchange resulting in dramatic enlargement of the macular hole. The retina must have been under tension when it was pushed posteriorly by the incoming air in the vitreous cavity. Enlargement of the macular hole at the time of fluid-air exchange probably resulted from the existing traction not being completely removed by vitrectomy, from shortening of the retina by existing peripheral circular retinal folds, or from both. The traction must have persisted even with placement of a circumferential buckle prior to the vitrectomy. Despite success in closing posterior holes with cyanoacrylate glue, in case 2 this technique was not effective in permanently reattaching the retina. Potential toxicity of the glue to the retina is also possible and may limit long-term vision.

These cases, combined with the other 2 who also had vitreotomies prior to formation of the macular hole, suggest that the etiology of these holes is not entirely the same as that of adult idiopathic holes in which the anterior-posterior vitreous traction is considered an important factor in pathogenesis. Retinopathy of prematurity–associated macular holes are still found even after careful vitrectomy. The vitrectomy performed in an infant eye with stage 5 ROP probably does not completely relieve the traction on the retina from preexisting preretinal organization, even after shaving the membrane that is strongly adherent to the circular retinal ridge and removing the fibrous tissues bridging the circular trough, unless the surgery is done in a relatively early stage of the disease. Thus, foreshortening of the retina remained because of persistent circular retinal folds in our cases.

If macular holes in ROP are caused by tangential traction from the anterior circular membranes and retinal folds, this explains why our most successful cases were those in which a circumferential scleral buckle was first placed and then a macular buckle. All 3 of these patients had reattached retinas. The retina in 1 patient (case 5) subsequently redetached, and the patient underwent another vitrectomy and silicone oil tamponade. This case is atypical in the series, however, in that there was continued proliferation of retinal neovascularization due to retinal hypoxia resulting from a cyanotic heart condition. It is probable that the retina would have stayed attached if the underlying disease process had been controlled in this case. Macular holes are probably rare in RD with ROP, but the presence of a macular hole is a possibility in patients with ROP and could be the cause of failure in attempts to reattach the retina. All 5 patients described here had undergone prior vitreoretinal surgery, but we have also seen a case of ROP with macular hole–associated detachment in another infant without any prior history of vitreoretinal surgery. This patient was not operated on because the fellow eye had useful vision with no RD. A macular hole may be hidden by retinal folds or retrolental membrane and may therefore be undetectable in some cases with stage 5 ROP.

Surgical repair is difficult, but good anatomic reattachment with acceptable visual results is possible with athermal scleral buckling of the macula.

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REFERENCES