Surgical management of retinal detachment associated with the acute retinal necrosis syndrome

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Abstract

We operated on nine eyes in eight patients with retinal detachment associated with acute retinal necrosis (ARN) syndrome. The patients were treated with scleral buckling, vitreoretinal surgery, or a combination of these treatments. Vitrectomised eyes underwent combinations of lensectomy, membrane dissection, scleral buckling, air-fluid exchange, endolaser photocoagulation, cryotherapy, and retinal tamponade with C₃F₈ gas or SF₆ gas. Macular attachment was achieved in eight (89%) eyes. Vision improved in seven (78%) eyes, of which five (56%) achieved 20/200 or better vision. Three eyes that had received laser treatment posterior to areas of retinitis suffered retinal detachment despite this prophylactic treatment. Poor visual outcome resulted from viral infection of the optic nerve or macular involvement, macular hole formation, macular pucker, or hypotony.

Acute retinal necrosis (ARN) is a clinically defined syndrome characterised by confluent, peripheral necrotising vaso-occlusive retinitis, arteritis, and vitritis.¹⁻¹⁰ Although this syndrome was first described in healthy individuals, immunocompromised patients may also be affected.¹¹⁻¹³ Herpes viruses have been implicated as causal agents in ARN.¹²⁻²³ Retinal detachment is a frequent complication of the syndrome. The incidence of retinal detachment depends on the extent of peripheral retinal involvement and the degree of vitritis. It has been reported to be as high as 85%, despite the efficacy of acyclovir in hastening the resolution of the retinitis.^{1 24-29}

Measures for the prevention or treatment of retinal detachment in eyes with ARN have included prophylactic photocoagulation,^{9 24 25 30} prophylactic vitrectomy and scleral buckling with acyclovir infusate during the acute phase of the retinitis,^{29 31} scleral buckling alone,³² and pars plana vitrectomy, endolaser photocoagulation, and long-acting retinal tamponade.^{24 29 32-34}

We report here our surgical results for retinal detachment associated with the acute retinal necrosis syndrome.

Patients and methods

Between 1981 and 1988 we operated on nine eyes of eight patients with retinal detachments associated with the acute retinal necrosis syndrome. Five of the eight patients were females, three males; they ranged in age from 16 to 73 years, mean 40.1 years. Follow-up ranged from nine months to five years, mean 18 months.

Seven of the eight patients were treated with acyclovir either before or concurrently with

the development of retinal detachment. These patients were also treated with prednisone and aspirin during their time in hospital. In patients who were treated with prophylactic photocoagulation in an attempt to prevent the occurrence of posterior pole retinal detachment, three to six contiguous rows of laser photocoagulation burns were placed posterior to the confluent peripheral retinitis. Retinal detachment developed following large retinal breaks at the border of necrotic retina in seven eyes. One eye had a retinal break within an area of active retinitis (case 2), and one eye developed a break in the uninvolved retina (case 9). Three eyes underwent scleral buckling alone, while six eyes underwent vitreous surgery.

Those patients undergoing vitreoretinal surgery were operated on by the bimanual technique with a posterior infusion port and fibreoptic illumination. Meticulous anterior vitreous base vitrectomy, with 360° scleral depression, was part of each vitrectomy. Epiretinal membranes were dissected from the retinal surface and removed from each of these eyes.

Encircling scleral buckles were placed in all eyes that had vitreous surgery, not to support specific retinal breaks but to relieve peripheral anterior vitreous base contraction that might occur during the postoperative period. Vitrectomised eyes had air-fluid exchange with internal subretinal fluid drainage by means of pre-existing breaks or planned retinotomies. Confluent endolaser photocoagulation was used to treat the edge of attached uninfected retina posterior to the peripheral retinitis. Photocoagulation was also applied round all retinotomy sites and over the scleral buckle. Vitreous substitution with sulphur hexafluoride gas (25%), or perfluoropropane gas (20%) was then performed.

Results

Nine eyes of eight patients underwent surgery for retinal detachment associated with the acute retinal necrosis syndrome (Table 1). Macular reattachment was achieved in eight (89%) of them.

Two of the nine eyes had partial retinal reattachment. One eye with partial retinal attachment developed postoperative epiretinal membranes that detached a quadrant of extramacular retina and puckered the macula. The patient declined further surgery. The other such eye developed anterior retinal detachment due to anterior proliferative vitreoretinopathy, though the posterior retina remained attached.

Vision improved to the 20/200 level in five (56%) of nine eyes. Of the eight eyes in which the retina was reattached one had no light perception because of optic nerve involvement.

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Accepted for publication
8 March 1991

Table 1 Surgical results in acute retinal necrosis

Case	Age	Sex	Preop laser	Preop vision	Acyclovir	Surgery	Postop vision	Anatomical result	Follow-up	Comments
1	27	F	+	3/200	+	L, V, M, AFX, E, SB, C ₃ F ₆	5/200	Attached	24 mths	1. Developed peripheral RD, then PVR. Previous herpes zoster uveitis in other eye 2. Developed macular pucker postop, declined further surgery
2	35	F	—	HM	+	L, V, M, AFX, E, C ₃ F ₆ , SB	20/200	Attached	12 mths	1. Active retinitis and RD 2. Acyclovir (IV, and infusate 40 µg/ml) 3. Giant retinotomies created during surgery 4. Hypotony retinopathy, IOP normal 3 mths postop
3	36	M	+	7/200	+	V, M, AFX, E, C ₃ F ₆ , SB	20/160	Attached	12 mths	1. Developed posterior epiretinal membranes and macular hole 2. Increased nuclear sclerosis
4	73	F	—	20/400	—	SB (segmental)	NLP	Attached	12 mths	1. Optic nerve involvement caused NLP
5	51	M	—	20/300	+	V, C, AFX, SB, SF ₆	20/80	Attached	36 mths	1. Cataract developed, later removed 2. Treated before availability of endolaser
6	16	F	—	HM	—	1. V, SB, C, SF ₆ 2. L, V, M, AFX, E, SF ₆	6/200	Attached	24 mths	1. Developed postop ERM, and became redetached 2. Postop hypotony associated with anterior PVR
7	16	F	—	20/25	+	SB	20/20	Attached	18 mths	1. OD of case 6 2. Encircling scleral buckle for quadrantic RD
8	36	F	—	HM	—	L, V, M, AFX, C, SF ₆ , SB	NLP	Detached	60 mths	1. Prior to accepted use of acyclovir 2. Had VH due to neovascularisation 3. Developed proliferative vitreoretinopathy postoperatively and declined reoperation
9	63	M	+	HM	+	SB	20/60	Attached	9 mths	1. Break in uninfected retina, outside demarcated area

V: pars plana vitrectomy. M: membranectomy. L: lensectomy. AFX: air-fluid exchange. E: endolaser photocoagulation. C: cryotherapy. SB: scleral buckle. C₃F₆: perfluoropropane gas. SF₆: sulphur hexafluoride. S: silicone oil. RD: retinal detachment. PVR: proliferative vitreoretinopathy. PR: pneumatic retinopexy. ERM: epiretinal membranes. VH: vitreous haemorrhage. HM: hand movements. CF: counting fingers. LP: light perception. NLP: no light perception.

Three eyes had preoperative prophylactic laser posterior to the areas of retinitis. In two of these eyes the retina was treated for 360°. In one eye (case 9) the peripheral retinitis did not involve the entire periphery, and laser was placed round the infected retina, extending anteriorly to the ora serrata. Despite the prophylactic laser, the retina in these three eyes became detached, but was successfully reattached with surgery.

Discussion

Retinal detachment remains a serious complication of the acute retinal necrosis syndrome. As surgical techniques for dealing with complicated retinal detachment have become more refined, the reattachment rate has improved.^{1 24 29 31–34} The visual acuity results have not been as satisfactory, though recently Blumenkranz *et al* reported that five of six eyes achieved 20/200 or better vision after successful repair.³²

In our series the retina was reattached partially or totally in eight (89%) of nine eyes. Of the two eyes with partial attachment one (case 1) developed postoperative epiretinal membrane formation and macular ectopia and the patient refused further surgery. In the third patient (case 6, OS), the entire posterior pole was reattached. Nevertheless, anterior proliferative vitreoretinopathy redetached the periphery and probably played a part in the patient's postoperative hypotony.

Though vision improved in seven (78%) eyes, it did so to 20/200 or better in only five (56%). In three eyes the retina was reattached, yet vision failed to reach the 20/200 level. These eyes were thought to have associated complications that prevented significant visual improvement, including postoperative macular pucker (case 1), optic neuritis, presumed to be secondary to herpes virus (case 4), and hypotony following reoperation for proliferative vitreoretinopathy (case 6, OS).

Several authors have suggested that in eyes with clear enough vitreous to allow laser, prophylactic photocoagulation should be used to demarcate areas of active retinitis in an attempt to decrease the incidence of posterior retinal detachment.^{9 24 25 30} In our series three eyes had prophylactic laser treatment but developed posterior retinal detachment. In one of these eyes (case 9) the break developed outside the area of demarcated retina, presumably in uninfected retina. In case 1 proliferative vitreoretinopathy (PVR) developed and tractionally detached the retina away from the lasered area. Case 3 also developed PVR with an associated macular hole that resulted in posterior retinal detachment.

There appears to be a spectrum of disease severity with the acute retinal necrosis syndrome. Some eyes have fulminant disease requiring advanced vitreous microsurgery, while others have only a mild manifestation of the disease.³⁵ Some eyes with retinal detachment and ARN can be successfully managed with scleral buckling; two were encircled, and one received only a segmental buckle. Scleral buckling, therefore, may be considered a surgical alternative in mild cases with small breaks, quadrantic involvement, and minimal vitritis.^{24 33 34} The majority of ARN cases, however, have multiple, large posterior breaks that are best treated with vitreoretinal surgery.

Good anatomical success rates and visual results have been reported with vitrectomy techniques without scleral buckling.³² In our series six eyes underwent vitrectomy in combination with various adjunctive procedures (Table 1). No eyes underwent vitrectomy without placement of a scleral buckle.

In most patients requiring vitrectomy for retinal detachment with ARN the lens needs to be removed. This allows easy access to the far anterior peripheral vitreous base. If this peripheral vitreous is allowed to remain, it may contract postoperatively and redetach the retina.³² Place-

ment of an encircling scleral buckle, combined with meticulous vitreous base dissection, may minimise this complication.

In one eye (case 6, OS) anterior retinal detachment and hypotony developed postoperatively, though the posterior retina was attached. In this case the lens was not removed at the time of the second operation. Another reason for removing the lens is the likelihood of developing a cataract with the use of long-acting gas substitutes or silicone oil. In our series two patients (cases 3 and 5) underwent vitrectomy without lensectomy. Both developed cataracts within several months, requiring cataract surgery.

Short-acting (SF_6) and long-acting (C_3F_8) gases were used as vitreous substitutes for retinal tamponade following retinal reattachment with air-fluid exchange (Table 1).

Hypotony was a postoperative complication in two patients with successful retinal reattachment (cases 2, 6, OS). In case 2 the retina was reattached and the eye had a normal intraocular pressure (IOP) for several weeks. Vision improved to the 20/128 level. The IOP fell to zero over several weeks and the fundus developed chorioretinal folds, retinal striae, and a boggy, thickened appearance. The vision dropped to 9/200. There was no significant intraocular inflammation at this time, nor was there any anterior proliferative response that might have created ciliary body detachment. After injections of retrobulbar steroids over a three-month period, the IOP rose to 10 mm Hg and vision improved to 20/200. The other patient who developed hypotony (case 6, OS) had traction on the ciliary body created by anterior PVR.³⁶

Retinal detachment usually occurs after the acute period of retinal necrosis has passed. The infected necrotic retina thins, so that only a fibroglial remnant remains over the disrupted retinal pigment epithelium in many areas. The vitreous develops various degrees of haziness and inflammatory opacification related to the severity and extent of retinitis, and also related to the immune system's ability to mount a response. The subsequent contraction of the vitreous creates traction on the retina, and large retinal breaks usually form at the borders of the involved and uninvolved retina. One of our cases (case 2) developed a retinal detachment during the acute phase of the retinitis. This eye had partial posterior vitreous separation and retinal breaks within the area of active retinitis. Surgery on such eyes is difficult because of the iatrogenic retinal breaks created by removing areas of attached vitreous from necrotic, detached retina.

Because retinal breaks usually occur when the contracting vitreous tears the necrotic, avascular retina, significant vitreous haemorrhage is not common in ARN. Vitreous haemorrhage can occur, however, in association with neovascularisation created by the inflammatory and ischaemic conditions prevalent in ARN.^{37,38} Pan-retinal photocoagulation to areas of non-necrotic and segmentally non-perfused retina has been shown to cause regression of optic nerve neovascular proliferation.³⁹ One eye in our series (case 8) had retinal detachment and vitreous haemorrhage caused by neovascularisation. At

the time of surgery the retina behind the vitreous haemorrhage was found to be totally necrotic. Postoperatively the retina became detached again and the eye was eventually enucleated. This patient's other eye also had ARN and disc neovascularisation that responded well to pan-retinal photocoagulation.

The acute retinal necrosis syndrome may be complicated by retinal detachment. For those eyes with small or single breaks and quadrant detachments, scleral buckling may be sufficient to reattach the retina. If complicated retinal detachment is present, including those cases with epiretinal membrane formation, marked vitritis, active retinitis, macular hole, or multiple large posterior tears, then pars plana vitrectomy, endolaser photocoagulation, and long acting retinal tamponade have a good chance of reattaching the retina. The visual recovery in these successfully reattached eyes depends on viral infection of the optic nerve, hypotony, macular hole formation, and postoperative epiretinal membrane formation.

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