Visual Outcome After Surgical Removal of Choroidal Neovascularization in Pediatric Patients

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Objective: To assess the visual outcome after surgical removal of choroidal neovascularization (CNV) in pediatric patients.

Methods: A retrospective, noncomparative, consecutive case series of 17 eyes undergoing surgical removal of CNV of various causes in patients aged 18 years and younger.

Results: The cause of the CNV was presumed ocular histoplasmosis syndrome in 11 eyes, idiopathic in 3, and optic nerve coloboma, ocular toxoplasmosis, and trauma in 1 eye each. Two eyes had CNV within 100 µm of the center of the foveal avascular zone (juxtafoveal CNV) and 1 eye had peripapillary CNV, while 14 eyes had CNV beneath the geometric center of the foveal avascular zone (subfoveal CNV). In eyes with subfoveal CNV, median preoperative Snellen visual acuity was 20/200 (range, 20/80-3/200). With a median follow-up of 27 months (range, 6-45 months), median final visual acuity was 20/50 (range, 20/20-2/200); 10 (72%) had improvement of 2 or more Snellen lines after surgery, and 6 eyes (43%) had final visual acuity of 20/40 or better. In 3 eyes with juxtafoveal CNV or peripapillary CNV, all eyes had improvement of 4 or more Snellen lines. Postoperative recurrent CNV developed in 6 (35%) of 17 patients; 2 of the eyes underwent a second vitrectomy and 4 received laser treatment for the recurrences.

Conclusions: Pediatric patients may have good recovery of vision after surgical removal of CNV, and the removal of these membranes may be a viable alternative to laser photocoagulation in pediatric patients.


W h ile choroidal neovascularization (CNV) is a major cause of central visual loss in adults, it is also seen in pediatric patients and can cause permanent central visual loss.1-3

The Macular Photocoagulation Study (MPS) Group has demonstrated visual benefit of laser photocoagulation of non-subfoveal CNV associated with the presumed ocular histoplasmosis syndrome (POHS) or idiopathic membranes.4-10 However, because the eligibility criteria in the MPS included patients aged 18 years or older, there are no prospective, randomized, controlled data on laser treatment for pediatric patients with CNV. Sears et al11 recently reported the clinical experience at 3 centers with submacular surgery for subfoveal membranes in children. Twelve patients aged 7 to 17 years underwent surgical removal of neovascular complexes. With average follow-up of 20 months, median visual acuity improved from 20/300 preoperatively to 20/80 postoperatively.

The purpose of this study was to assess the visual outcome after surgical removal of CNV in pediatric patients.

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PATIENTS AND METHODS

We reviewed the charts of 19 eyes of 19 consecutive patients aged 18 years or younger who underwent surgical removal of CNV of various causes between February 1, 1993, and August 31, 1997. Of the 19 eyes, 2 were excluded because the follow-up periods were less than 6 months; therefore, this study group consisted of 17 eyes of 17 patients.

To determine the location of CNV, fluorescein angiograms were obtained preoperatively. In patients with juxtafoveal CNV, surgical extraction of the CNV was performed because, in the surgeon’s opinion, laser photocoagulation of the entire lesion would likely have damaged the center of the fovea. A patient with peripapillary CNV was judged to be a poor candidate for laser photocoagulation because of her young age (7 years) and because laser treatment would have ablated most of the temporal peripapillary retina. The initial size of the CNV was measured by means of a transparency prepared by the MPS with circles enclosing areas ranging from 1.0 to 16.0 MPS disc areas.

Preoperative and postoperative best-corrected visual acuities were determined by manifest refraction using the standard Snellen visual acuity chart, which was recorded in the following gradations: 20/20, 20/25, 20/30, 20/40, 20/50, 20/60, 20/70, 20/80, 20/100, 20/200, 20/300, 20/400, 6/200, 2/200, hand motions, light perception, and no light perception.

To analyze the visual acuity data, changes in visual acuity were categorized as improvement (increase of 2 or more Snellen visual acuity lines from baseline), stable (within 1 line), and worse (loss of 2 or more Snellen visual acuity lines from baseline).

Informed consent for submacular surgery was obtained from each patient and parent(s). Pars plana vitrectomy and removal of the CNV were performed by a single surgeon (M.A.T.) according to established techniques previously described. The posterior hyaloid is usually more adherent to the retinal surface in pediatric patients than adults. We developed the hyaloid lifter (Synergetics, Chesterfield, Mo) to detach the posterior hyaloid. This instrument, which was specially designed for disinserting the posterior hyaloid in all patients, is an angled 120° pick, approximately 2.5 mm in length and approximately 33 gauge at the tip. It was used in all 17 cases to engage the hyaloid near the optic nerve and mechanically detach the hyaloid at least beyond the major vascular arcades.

Multiple fluorescein angiograms were typically obtained for each patient during the postoperative period. Recurrent CNV was defined by angiographic criteria of new or persistent fluorescein leakage within or at the margins of the surgical scar.

The patients had a median follow-up time of 27 months (range, 6-63 months). Fourteen patients had more than 12 months’ follow-up. A healthy 16-year-old adolescent boy was examined because of a 5-month history of progressive central vision loss in the right eye. A retina specialist had previously diagnosed congenital toxoplasmosis, and at the time of surgery, 3 (21%) remained unchanged, and 1 (7%) had worsening of 2 lines. There were no patients whose vision worsened 3 or more Snellen lines. Six patients (43%) had visual acuity of 20/40 or better, and 5 eyes (29%) were 20/100 or worse at the last visit (Figure 1).

Intraoperative complications included a full-thickness small retinal break over the CNV during subretinal manipulation in 1 patient (patient 2). The break was successfully closed postoperatively and the patient’s visual acuity recovered to 20/30. The loss of retinal pigment epithelium (RPE) resulted in no improvement in postoperative visual acuity. There were no complications associated with general anesthesia, which was used in all cases.

Postoperative recurrent CNV developed in 5 patients (36%), with a median interval of 1 month (range, 1-6 months). Of the 4 eyes that had received previous laser photocoagulation, 2 (50%) developed recurrent CNV after surgery. Of the 10 eyes that had not undergone previous laser photocoagulation, 3 (30%) developed recurrent CNV. In these instances, laser photocoagulation was performed in 3 eyes with extrafoveal or juxtafoveal recurrent CNV, and a second operation was performed in 2 eyes with subfoveal recurrence. No other postoperative complications, such as retinal detachment, endophthalmitis, or visually significant cataract, were observed during the follow-up interval.

PATIENTS WITH NONSUBFOVEAL CNV

Two eyes of 2 patients had juxtafoveal CNV within 100 µm of the center of the foveal avascular zone, and 1 eye had peripapillary CNV (Table 1). Both eyes with juxtafoveal CNV achieved good visual recovery to 20/20, and the eye with peripapillary CNV had improvement of 4 Snellen lines (from 20/200 to 20/60). Choroidal neovascularization was noted in 1 eye (patient 15) 1 month after surgery, and the membrane was treated successfully with laser photocoagulation. In 1 patient, aged 7 years (patient 17), the retinotomy was open and elevated from the RPE and there was marked subretinal fluid in the macular area on the first postoperative day. As in all other cases, an intraoperative air-fluid exchange had been performed and the retina flattened. Balanced saline solution was then reinjected until approximately 30% of the vitreous cavity was filled with air and 70% filled with fluid. Given her young age, perhaps the patient was noncompliant with face-down positioning for 18 to 24 hours, as we advise in all cases. The patient underwent a second surgery with air-fluid exchange, endolaser photocoagulation to the retinotomy, and long-acting gas injection (15% sulfur hexafluoride). The retinotomy remained closed and the retina reattached thereafter.

SELECTED CASE REPORTS

Patient 7 (Subfoveal CNV)

A healthy 16-year-old adolescent boy was examined because of a 5-month history of progressive central vision loss in the right eye. A retina specialist had previously diagnosed congenital toxoplasmosis, and at the time of
referral a subfoveal CNV had arisen from the presumed toxoplasmosis scar. Preoperatively, the best-corrected visual acuity in the affected right eye was 20/100. On dilated fundus examination of the right eye, an old chorioretinal scar was observed just temporal from the fovea. No other inflammatory lesions were observed in the eye. Clinically, a small neovascular complex was noted just nasal to the scar. Fluorescein angiography disclosed classic choroidal neovascularization beneath the geometric center of the fovea (Figure 2). During vitrectomy, the membrane was reflected temporally, leaving the subfoveal RPE intact. The membrane and the scar were then extracted in 1 piece. Postoperatively, visual acuity returned to 20/20 (Figure 3). It has remained stable during 27 months’ follow-up without recurrent CNV.

Patient 15 (Juxtafoveal CNV)

A healthy 14-year-old adolescent girl was referred for surgery for juxtafoveal CNV of presumably idiopathic cause. Preoperatively, best-corrected visual acuity in the affected right eye was 20/200. Angiographically, a well-
defined neovascular membrane was seen that appeared to be anterior to the RPE. A thin layer of blood extended under the center of the foveal avascular zone (Figure 4). During vitrectomy, the membrane was peeled back, leaving the subfoveal RPE intact, and the membrane was extracted in 1 piece. Postoperatively, however, a recurrence of the neovascular complex developed at the choroidal ingrowth site. This lesion was treated successfully with laser photocoagulation. The final visual acuity returned to 20/20 and has remained stable during 63 months' follow-up without additional recurrence of CNV (Figure 5).

**COMMENT**

Choroidal neovascularization in the pediatric population is uncommon. Thus, little has been written in the ophthalmic literature concerning the management of CNV in pediatric patients. Wilson and Mazur³ reported 5 cases of CNV of various causes in children aged 18 years or younger. Of these patients, 3 with extrafoveal CNV had laser photocoagulation, and visual acuity improved in 2 patients. Two patients with presumed subfoveal CNV were not treated because of the location of CNV, and no improvement in visual acuity was seen. Goshorn et al⁵ described 27 eyes of 25 patients aged less than 21 years. In that report, 11 (58%) of 19 untreated cases of CNV spontaneously involuted, with 9 of these achieving a final visual acuity of 20/50 or better. However, only 6 of these 11 eyes had subfoveal CNV; the other 5 had juxtafoveal or peripapillary location. This report suggested that the natural course of CNV seemed to be more favorable in pediatric patients than in adults.

Surgical results in our series suggest that pediatric patients may have good recovery of vision after surgical removal of subfoveal CNV. Approximately 70% of the patients with subfoveal CNV had improvement of 2 or more Snellen lines, and nearly half of the patients regained visual acuity of 20/40 or better at the final examination regardless of the cause of CNV. The results are similar or better in terms of eyes with either improvement of vision or acuity of 20/40 or better when compared with previously reported surgical results in POHS in adults.¹²⁻¹⁶ In the series of Goshorn et al,³ approximately 30% of eyes achieved improvement of 2 or more Snellen lines, compared with 72% of eyes in the present series. Our results suggest a possible benefit of subretinal surgery, although it is hard to compare these dissimilar data (Table 2).

Of the 4 eyes that failed to improve in final visual acuity, 1 eye lost foveal RPE at the time of surgery, and 1 eye had postoperative subfoveal recurrence. The remaining 2 eyes had preoperative problems: severe loss of preoperative vision and congenital optic nerve abnormality. The relationship between corticosteroid use or previous photocoagulation and visual prognosis was unclear because of the small number of patients.

The MPS showed the benefits of laser treatment for extrafoveal or juxtafoveal CNV secondary to POHS or idiopathic origin.⁴⁻¹⁰ However, we have little information about the results of laser treatment for subfoveal CNV in POHS or idiopathic cause. In a pilot study of laser photocoagulation for subfoveal CNV in patients with POHS, no patients had improvement of vision after treatment.¹⁷ Olk et al¹⁸ reported the natural course of subfoveal CNV secondary to POHS. Only 11% of patients had
improvement in vision of 2 or more Snellen lines with a median follow-up of 42 months. However, it is impossible to compare our surgical results with these results of laser treatment or natural course because the patients’ preoperative findings were not identical in these series.

Of the 4 eyes that failed to improve in final visual acuity, 2 eyes experienced intraoperative or postoperative complications: 1 eye lost foveal RPE at the time of surgery, and 1 eye had postoperative subfoveal recurrence of CNV. Foveal RPE loss may occur in association with the removal of CNV and lead to dysfunction of the central retina. Because most of these membranes were considered preoperatively to be type II membranes (as proposed by Gass19), it was hoped that meticulous manipulation of the membrane would prevent the loss of subfoveal RPE if the ingrowth site was not under the fovea. In 1 patient with severe visual loss before surgery, visual acuity did not improve after surgery, although the membrane was removed successfully. We speculate from the angiogram that extensive RPE disturbance had existed before surgery despite the small size of the membrane.

Recurrent CNV was observed in 6 patients, and the frequency was the same as that reported in adult patients with POHS.11,15,16,20 Of 6 patients with recurrence, 4 with extrafoveal or juxtapfoveal recurrent CNV underwent laser photocoagulation and the 2 with recurrent subfoveal CNV underwent a second vitrectomy. Although 1 of 2 eyes that had a second surgical intervention recovered good visual acuity (20/30), careful and longer-term follow-up is necessary for evaluating the role of repeated surgery for subfoveal recurrence.

Vitrectomy carries inherent risks of ocular complications, although we did not encounter severe vision-threatening postoperative complications in this series. In 1 patient, aged 7 years, the retinotomy remained open with some subretinal fluid on the first postoperative day. On the second postoperative day, we performed a second air-fluid exchange and endolaser application around the retinotomy, and the eye was left with an approximate 30% air bubble. Given her young age, perhaps the patient was noncompliant with face-down positioning for 24 hours.

These results indicate that pediatric patients may experience improved visual function after surgical removal of CNV. However, a total of 7 patients in this series required either reoperation or postoperative laser treatment, for a total rate of additional intervention of...
41%. With these additional therapies, visual acuities were reasonably good. One must exercise caution in advocating vitrectomy surgery in pediatric patients with subretinal membranes. The limited available data on the natural course do suggest that some eyes will do well with observation alone. A randomized, prospective trial would be the only certain means to determine which management option is best. Given the infrequency with which CNV is seen in the pediatric age group, such a study would likely not be feasible. Longer follow-up of additional cases may offer some additional guidance.

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