Surgical treatment of peripapillary choroidal neovascularisation

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Surgical treatment of peripapillary choroidal neovascularisation is a valuable therapeutic option which should not be undertaken too late

Peripapillary choroidal neovascularisation (PPCNV) comprises about 10% of all cases of choroidal neovascularisation.[1] Starting at the nasal margin of the disc the condition does not become symptomatic until fluid, exudate, blood, or the membrane itself have extended from the disc toward the macula, threatening central vision. Very large PPCNVs are defined as more than 3.5 disc areas or greater in size and involve 180° or more of the disc circumference.[2] Although less common than smaller PPCNVs, the very large ones may lead to severe visual loss.[3] Over time, scar contraction at the edge of the PPCNV causes breaks in Bruch's membrane, and the associate haemorrhage leads to a new circle of ''reparative'' fibrovascular ingrowth that manifests as progression or extension of the PPCNV complex.[1]

PPCNVs can be idiopathic or secondary to various conditions. In a recent survey,[4] Browning and Fraser reported that PPCNV was associated with age related macular degeneration (AMD) in 45% of cases, while 39% were idiopathic, so at least 84% of the patients will be over the age of 55 years. Although the diagnosis of ''idiopathic'' PPCNV[5] is unsatisfactory, the presence of these lesions in clinically normal eyes has been demonstrated in pathological studies.[6 7] The remaining cases of PPCNV occur secondary to multifocal choroiditis, angioid streaks, histoplasmosis, choroidal osteoma, optic disc drusen, congenital disc anomaly, pattern dystrophy, and peripapillary pseudopodal pigment epithelium and choroidal atrophy.[4]

In patients over 70, involvement of the second eye in PPCNV can be expected in 20–62% of all cases.[4 8] At this age, 75% of untreated cases have lost visual acuity (VA) to a level of 3/60 or less. The time between the involvement of the first and the second eye varies from simultaneous to seven years.[8] On fluorescein angiography, PPCNV may contain a significant occult component, leading to slow and unpredictable growth; in fact more than half the AMD related and idiopathic cases are entirely or mainly occult.[9] This makes PPCNVs difficult to treat by laser, which requires well defined lesion margins.

Results of laser photocoagulation for PPCNV vary. In 1988 Kies and Bird recommended that a large margin of normal tissue should be treated, and that there should be laser ablation of any angiographic abnormality around the lesion.[3] In their series of 55 cases, only 13 (23.6%) received laser treatment, and recurrences were observed in three quarters of these. Once the centre was affected by fluid, bleeding, or choroidal neovascularisation, VA did nor recover or improve spontaneously. Flaxel *et al*[2] reported 1996 on their results with laser treatment for very large (massive) PPCNV, measuring 3.5 disc diameters or more, and with treatment limited to the temporal portion of the neovascular complexes. Six of 10 treated cases showed stabilisation while four progressed to severe visual loss. In PPCNV related to histoplasmosis, Turcotte *et al*[10] reported stable vision in about 75% of cases after laser treatment. On the other hand, no statistical difference in final VA following laser treatment for AMD-PPCNV was reported by Ruben *et al* in 1994.[11] In Browning and Fraser's survey,[4] 73 of 115 eyes with PPCNV of various origins underwent laser treatment; in 14 (19.2%) a recurrence was noted, and multiple recurrences occurred in four (5.5%). Comparable results with recurrence rates of 20% and 28% were reported by Annesley *et al*[12] and Cialdini *et al*.[13] Finally, in the Macular Photocoagulation Study PPCNV subgroup, there was no improvement in visual outcome with laser ablation over three years of untreated follow up; furthermore, there was no significant difference in the rates of severe vision loss between treated and untreated eyes.[14]

WHAT ABOUT SURGICAL EXCISION FOR PPCNV?

Since Thomas and Kaplan introduced subretinal surgery for foveal choroidal neovascular membranes in 1991,[15] several groups have reported on membrane excision for PPCNV.

Successful surgery with improvement in vision was described in two single case reports of the surgical removal of AMD related extrafoveal PPCNV, where fluid accumulation had caused visual loss.[16 17]

In 2003, Sullu *et al*[18] presented a case report of a nine year old girl with binocular PPCNV related to papillary drusen, who already had submacular involvement in her left eye. Wrongly diagnosed as having papilloedema, this child had undergone extensive neurological examination. After surgery the VA improved in the left eye from 0.05 to 0.3, and no recurrence was observed. In 1998, Atebara *et al* reported on 17 young patients with extensive PPCNV related to histoplasmosis.[19] In the majority (82%, 14/17), the PPCNV had already reached the fovea. While all cases with a preoperative extrafoveal location of the PPCNV reached a VA of 20/20 postoperatively, half the remaining 14 eyes achieved a final VA of 20/40 or better. After 32 months recurrences were observed in 24% (4/17). No surgical complications occurred. In 2004, Kertes described three patients aged 25–30 years with histoplasmosis and PPCNV, all of whom underwent surgery.[20] The location of the PPCNV extended extrafoveally in two and was juxtafoveal in the third. VA improved after surgery in all three eyes, two reaching 20/20 and the third, 20/50. Postoperatively, one peripheral tear needed laser treatment.

In 2003, Bains *et al* presented the surgical results in 17 patients over 55 years of age with extensive PPCNV, mainly AMD related or idiopathic.[21] Preoperatively the PPCNV was located extrafoveally in seven cases (41%) but 11 eyes (59%) already showed foveal extension. Visual acuity was stable or improved in six eyes (35.2%) and worsened in 11 (63.8%). After an observation period of 30 months the investigators concluded that surgical excision yielded improvement or stabilisation of VA in about one third of their elderly patients. Complications such as retinal detachment, macular oedema, and preretinal membrane formation were observed in five eyes (29%).

Eleven AMD patients with massive PPCNV not eligible for laser treatment or refusing it were included in a study by Blinder *et al* in 2005.[22] Cases were the PPCNV extended into the fovea were excluded, and the mean size of the membrane was 5 o'clock hours. After 23 months follow up seven cases (64%) had stable or improved VA, with a mean change of one line improvement. In three cases (27%), a recurrent membrane developed. In the same year, Kokame and

Yamaoka described the outcome of surgery in six elderly patients with extrafoveal PPCNV, where vision was threatened or affected by subretinal fluid, haemorrhage, exudate, or neovascular membrane growth.[23] After three years of follow up, VA was stable or had improved in five cases (83%) with a range of VA between 20/25 and 20/80. In three eyes there was early or late recurrence.

In this issue, Aisenbrey and coworkers report on the two-year functional and morphological outcome of subretinal membrane excision in eight patients with AMD related PPCNV (***see page 1027***).[24] Preoperatively, mean VA was logMAR 0.5; this improved to mean logMAR 0.3. Six of the eight cases gained vision. Although recent progression of the disease was the indication for surgery, in no case had the membrane extended into the fovea. Two years after surgery one recurrence was observed and was successfully removed surgically. The authors discuss newer treatment options, including photodynamic therapy, where a safety distance of 200 μm from the margin of the optic disc is recommended, and treatment with antiangiogenic agents. However, there is only one small case series on the successful treatment of PPCNV with photodynamic therapy, and none with antiangiogenic agents so far. Although small PPCNVs can be treated successfully with laser coagulation, the authors state correctly that large membranes may be ineligible for surgery because of the damage to the retinal pigment epithelium and the neurosensory retina that is caused by adhesions to the coagulated tissues. In agreement with the three recent reports cited above on the surgical excision of large PPCNVs, the authors recommend surgical intervention in older patients before the membrane has reached the centre of the fovea in order to maximise any improvement in vision.

Overall, younger patients with ocular histoplasmosis and other rare indications, with large, growing PPCNVs, have an excellent visual prognosis after subretinal surgery if the membrane is still extrafoveal. If the macula is already involved, there is a 50% chance of stabilisation or improvement, because the membrane in these cases tends to be located in the prepigment epithelium (type II),[25] and because the patients are younger and so some regrowth of pigment epithelium can be expected. Clearly, the decision to undertake surgery is easier if the fovea is threatened or involved by fluid or exudates, and if the patient's vision is already compromised. In elderly patients, subretinal surgery to remove extrafoveal PPCNVs might also be a promising therapeutic option leading to visual improvement. However, little or no chance of regaining vision can be expected if the macula is already involved in the neovascularisation process.

Possible complications related to surgery include endophthalmitis, retinal detachment, and haemorrhage, but these are rare. Cataracts will develop in most of the elderly patients if the lens is not removed in combination with vitreous surgery. Over the past 15 years subretinal surgery has developed technically, and we have learned to keep the retinotomies small, to prevent haemorrhages, and remove subretinal tissue with minimal trauma,[26] making subretinal surgery for PPCNV a valuable therapeutic option which should not be performed too late!

There are, however, limitations to what we can conclude from the current studies because the numbers of cases are small and there have been no randomised comparisons with alternative treatments. Such studies are needed to define the place of surgery in the current therapeutic armentarium. Other therapeutic options such as photodynamic therapy and antiangiogenic agents are being assessed, and there may be a place for combination therapies.

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