Surgical management of optic disc pit associated maculopathy

Y.K. GHOSH¹, S. BANERJEE¹, A. KONSTANTINIDIS², I. ATHANASIADIS², G.R. KIRKBY¹, A.K. TYAGI¹

¹Department of Vitreo-retinal Surgery, Birmingham and Midland Eye Centre, City Hospital NHS Trust, Birmingham
²Ophthalmology Department, Coventry and Warwickshire University Hospital - UK

INTRODUCTION

Optic disc pit and associated serous retinopathy is a rare anomaly occurring in less than 1 in 10,000 ophthalmic patients (1). Of the patients with optic disc pits seen in an ophthalmic department, up to 75% present with a sensory retinal detachment of the macula (2, 3). In the series published by Rubinstein and Ali (4), macular detachment was seen in over 93% of the cases. This condition is associated with a poor visual prognosis (3, 5) in spite of spontaneous retinal reattachment occurring in some patients (2, 4). Treatments tried in the past included bed rest (7), eye patching and mannitol (6), steroids (7, 8), and photocoagulation with Xenon arc or laser (3, 9, 10). Laser treatment in combination with scleral buckle, cryotherapy, drainage of subretinal fluid, and intraocular air injection have also been tried (11). In 1988, Schatz and McDonald (12) first suggested a systematic treatment plan for this condition, which has since then seen various modifications, including vitrectomy with fluid/gas exchange (13). Subsequently the success rate for reattachment of the retina has increased.

We present a series of seven eyes with optic disc pit and serous retinal detachment involving the macula. All these patients were treated by pars plana vitrectomy, intraocular tamponade, and laser photocoagulation. This group included two patients from the pediatric age group. All these cases had complete reattachment of the retina with good visual recovery.

MATERIALS AND METHODS

This was a retrospective case note analysis of nine patients who had been treated at Birmingham & Midland Eye Centre between 1999 and 2002 by two different vitre-
oretinal surgeons. The patients were identified from the databases of the two surgeons.

We examined the records relating to nine eyes of eight patients with optic disc pits. One of the fellow eyes had an asymptomatic optic disc pit, showing no serous detachment, and consequently no surgery was necessary. Complete postoperative follow-up records could not be traced in one patient. Seven eyes had a sensory retinal detachment and underwent surgery to reattach the sensory retina.

Preoperative data were collected for demography, laterality, presenting visual symptom, and visual acuity. The duration of visual symptoms and clinical findings were also noted. Postoperative data included type of surgery performed, complications, repeat surgery, follow-up, and stability of final visual acuity.

All patients had their best-corrected visual acuity (BCVA) recorded at every appointment, had their intraocular pressures measured with Goldmann tonometer, and underwent dilated slit lamp fundus examination with 90 D and 78 D lenses. Patients were reviewed in clinic 2 weeks, 6 weeks, and 3 months postoperatively. Patients 3 and 5 had short follow-up period as they came from other regions of the United Kingdom and were discharged back to the care of their local hospitals. The rest of the patients had a 9- to 14-month follow-up period as this was the time interval between their surgery and the time this article was prepared.

One interesting case is discussed in detail as it highlights the marked improvement in visual acuity and total flattening of the macula after a second procedure.

Case report

An 11-year-old boy presented with a history of blurred vision in the right eye following a scratch on the cornea about 7 days previously. On examination an optic disc pit with serous detachment involving the macula was diagnosed. His visual acuity was 6/18+3 unaided in the right eye improving to 6/12 part with pinhole. The visual acuity in the left eye was 6/3 unaided. He underwent vitrectomy with posterior hyaloid peel, SF₆ (sulfur hexafluoride) gas (25%) injection and argon laser to the temporal margin of the optic disc. He was advised to posture face down. The visual acuity was 6/24 postoperatively. A poor foveal reflex associated with residual fluid under the macula was noticed. He underwent repeat surgery including laser and 25% SF₆ gas injection in April that year and was advised to posture. He recovered significantly with postoperative visual acuity being 6/12 with refractive correction. On his last follow-up 9 months postoperatively, this best-corrected visual acuity had been retained and the macula was attached.

RESULTS

Out of the seven patients four were male and three were female. The age range was 8 to 63 years with a mean of 32.71 years. Six patients had the condition in their right eyes while one had it in the left eye. All of the patients had presented with blurred vision. The duration of their symptoms was between 2 weeks in a child to 5 months in an elderly man, before they presented to an eye clinic.

It was not possible to identify separation of the layers of the retina clinically as optical coherence tomography was not available at that time. They all underwent three-port pars plana vitrectomy and posterior hyaloid peel. In one case during the fluid/air exchange, viscous fluid was drained over the disc pit. Argon green laser photocoagulation, sufficient to cause slight blanching of the outer retinal layer, was then applied to the temporal margin of the optic disc across the papil-
lomacular bundle. Laser treatment was aimed at sealing off the entry site for the subretinal fluid originating from the disc pit. Internal tamponade with air was then inserted in all the cases. Four patients had intraocular air exchanged for 20–25% SF₆/air mixture and two had 14–16% C₃F₈/air mixture. One patient had silicone oil injection. All patients were asked to posture in a face-down position in order to maximize the tamponade of the gas. This posturing was required for 50 minutes every hour for a period of 2 weeks.

Two patients were found to have persistent serous fluid in the macula at first postoperative review. One of them had an intraocular C₃F₈ (14%) injection within 2 weeks, which successfully flattened the macula. The other patient improved spontaneously within 6 weeks without any further treatment. Two other patients initially had no fluid at first postoperative visit but then had a recurrence of the serous detachment at 6 weeks. They underwent repeat vitrectomy, laser, and intraocular 25% SF₆ gas injection with good anatomic and visual outcome. We treated one patient who had had four previous surgeries at another center with repeated recurrences. Silicone oil tamponade was inserted for 2 months with good anatomic result in terms of flattening the macula. This patient had a postoperative rise in intraocular pressure in the operated eye. This was controlled medically and subsequently he had a cataract surgery with intraocular lens implantation and removal of the silicone oil. The others did not have any macular fluid during follow-up.

The visual acuity at presentation ranged between 6/12 and 6/60 (mean 6/18). This improved to 6/6 and 6/36, respectively (mean 6/9), with the most significant recovery in one patient being 6/12 from 6/60. There was visual improvement of two or more lines on the Snellen chart in four patients.

The mean follow-up period was 9.1 months (range 5 to 14 months). Data of each patient are shown in detail in Table I.

### DISCUSSION

Optic disc pits have been considered to be a part of the embryologic abnormality relating to incomplete closure of the fetal fissure (1, 2). Abnormal vitreous structure terminating in the pit has been shown by scanning laser ophthalmoscopy (14). This is thought to be a part of an

<table>
<thead>
<tr>
<th>Patient Data and Results After Treatment</th>
<th>Case no.</th>
<th>Age, yr/sex</th>
<th>Presenting visual acuity</th>
<th>Final visual acuity</th>
<th>Internal tamponade</th>
<th>Repeat surgery</th>
<th>Follow-up, mo</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11/M</td>
<td>R 6/18+3</td>
<td>6/12 SF₆ 25% Vitrectomy, laser, gas injection</td>
<td>9</td>
<td>Re-accumulation of serous fluid postoperatively. Fluid resolved after second surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>51/F</td>
<td>L 6/60</td>
<td>6/12 SF₆ 20% None</td>
<td>10</td>
<td>Minimal persistent fluid inferior to disc</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>25/M</td>
<td>R 6/24</td>
<td>6/36 Silicone oil ROSO + cataract surgery with IOL implant</td>
<td>6</td>
<td>4 previous surgeries, postoperatively high IOP and cataract formation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>31/M</td>
<td>R 6/60</td>
<td>6/18 SF₆ 25% Vitrectomy, laser, gas tamponade</td>
<td>11</td>
<td>Re-accumulation of serous fluid postoperatively. Fluid resolved after second surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>8/F</td>
<td>R 6/60</td>
<td>6/36 SF₆ 25% None</td>
<td>5</td>
<td>Amblyopic eye</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>38/M</td>
<td>R 6/12</td>
<td>6/6 C₃F₈ 16% None</td>
<td>9</td>
<td>Macular pigmentary change</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>63/F</td>
<td>R 6/24</td>
<td>6/12 C₃F₈ 14% Gas injection</td>
<td>14</td>
<td>Persistence of parafoveal fluid requiring additional gas injection</td>
<td></td>
<td></td>
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</tbody>
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All patients underwent vitrectomy, posterior hyaloid peel, and laser. ROSO = Removal of silicone oil; IOL = Intraocular lens; IOP = Intraocular pressure
anomalous Cloquet canal, which exerts traction on the pit and may be a significant factor in the development of the serous detachment. In addition to this, optical coherence tomography studies detected vitreous abnormalities such as vitreomacular traction, vitreous strands over the optic disc, and complete or partial posterior vitreous detachment associated with optic disc pit maculopathy (25, 26). This observation supports the role of abnormal vitreous over the macula and optic disc in the pathogenesis of macular elevation associated with optic disc pit. Several theories have been suggested about the origin of the serous fluid. These include communication of the subretinal space with the cerebrospinal fluid (5, 7, 9, 15, 16), communication with the vitreous (7, 9, 18, 19), leakage of the peripapillary circulation through the border tissue of Elschnig (7, 15), and communication through a defect in the optic nerve sheath via a disruption of the intermediary tissue of Kuhnt (15). It also been suggested that there might be a combination of all of the above factors in producing the sensory retinal detachment at the macula (17).

Lincoff et al (20) first suggested that the retinal elevation that communicates with optic nerve pits is frequently a separation of the inner layers of the retina. They also suggested that the outer layer separation appeared to be a secondary phenomenon, which starts at the macula but may not communicate with the pit. Based on slit-lamp examination and stereoscopic transparencies viewed with magnification and projection, they have suggested that an alternative explanation for the maculopathy associated with optic disc pits is that fluid from the pit elevates the nerve fiber layer as it enters the disc, causing a schisis-like separation of the inner layers of retina.

Since then, optical coherence tomography findings (21) have substantiated the fact that fluid from the pit elevates the nerve fiber layer, causing a schisis-like separation. The natural history of the sensory retinal detachment associated with optic disc pit is not clear. However, withholding treatment seems to lead to a poor visual outcome (1, 8, 11). Longstanding sensory retinal detachment can cause irreversible loss of visual acuity due to macular pigment epithelial changes (3, 8, 11, 13). Therefore any form of treatment to reattach the retina early may be preferable to the natural progression of the disease. Spontaneous reattachment of the sensory retina has been reported (2, 12, 13), albeit rarely.

Numerous treatments have been tried in the past with variable amount of success. No one treatment has been uniformly successful. Schatz and McDonald (12) first suggest-
ead a systematic treatment plan for this condition which since then has been shown to give good results.

Since then the macular buckling technique (22) has had good success rates in terms of both visual and anatomic outcomes.

A recent publication has also reported successful outcome with peeling of the internal limiting membrane in these cases (23).

In our experience, pars plana vitrectomy combined with posterior hyaloid peel, intraocular gas injection, and laser treatment to the temporal margin of the disc across the papillomacular bundle is successful in flattening the sensory retinal detachment in some cases and in all cases following supplementary procedures.

Interestingly, the laser uptake on treating over the papillo-macular bundle is a slight blanching as is seen in normal retinal laser, rather than pigment dispersion. This further confirms the accuracy of Lincoff et al’s suggestion (20) of retinoschisis like cavity with the outer retinal layers still in contact with the retinal pigment epithelium. In our series the low intensity laser applications did not seriously affect the papillomacular bundle; hence the good visual outcome. Posturing of the head postoperatively is also important to enhance resolution of the serous macular detachment. In recurrent cases, intraocular tamponade with silicone oil may be helpful in preventing a re-accumulation of submacular serous fluid. As demonstrated in all of our cases, posterior hyaloid peel seems to play an important part in the success of the surgery.

Four out of seven of our patients required a second surgery for the retinal fluid to resolve completely. Following that they had a significant improvement in their final visual acuity. We could not find a cause for this failure of initial surgery and re-accumulation of retinal fluid, but it could be that the first laser treatment did not obliterate the communicating channel between the disc pit and the submacular space and that inner layer separation persists centrally after a gas tamponade and continues to provide access for the flow of fluid from the pit (24).

In conclusion, serous retinopathy associated with optic disc pit is a rare condition which benefits from surgical intervention. Although our series is one of the largest reported in the literature, cumulative data are required to define the management of this condition.

The authors have no proprietary interest in any of the products mentioned.
REFERENCES

12. Schatz H, McDonald HR. Treatment of sensory retinal detachment associated with optic nerve pit or coloboma.