PostScript

LETTERS

Terson's syndrome: surgical outcome in relation to entry site pathology

Terson's syndrome is a frequent cause of visual loss following subarachnoid haemorrhage (SAH).¹ Vitrectomy is indicated for non-resolving vitreous haemorrhage.^{2,3} We observed a high incidence of entry site (ES) pathology in patients having vitrectomy for Terson's syndrome and recommend routine prophylactic laser or cryotherapy to all entry sites in these patients.

Case series

A case note review identified 11 eyes of eight consecutive patients with Terson's syndrome (age range 27-56 years, M:F = 1:1) referred to the West of England Eye Unit between 1996 and 2004. Surgery was performed for non-resolving vitreous haemorrhage 1-6 months from date of referral. Ten eyes had three port pars plana vitrectomy and one eye had combined phacovitrectomy.

Table 1 summarises the preoperative and postoperative Snellen visual acuity (VA), type of anaesthesia, whether surgery was complicated by intraoperative ES break, and postoperative retinal detachment.

Five of 11 eyes (45.5%) had ES breaks detected intraoperatively. One eye had prophylactic cryotherapy to the entry sites because surgery on the fellow eye was complicated by retinal detachment from an ES break. The remaining five eyes had vitrectomy with no ES prophylaxis. Retinal detachment developed in three eyes between 1-7 months postoperatively, all occurred because of pathology at untreated entry sites.

Comment

Vitrectomy in Terson's syndrome usually results in good improvement in visual acuity,^{2,4,5} as has been demonstrated in our series. Entry site breaks, however, were noted in 45.5% of cases, (95% confidence intervals (CI) 16.8 to 76.7). This compares with a 3.3% incidence of ES breaks (CI 0.7 to 9.4) in 90 eyes of presbyopic patients that underwent

phacovitrectomy for varied pathology by the same surgeons at the West of England Eye Unit.⁶ An internal audit at our unit also showed an incidence of ES breaks of 2.3% of 562 eyes (CI 1.2 to 3.9) having vitrectomy or phacovitrectomy for miscellaneous pathology and 2.9% of 34 eyes (CI 0.1 to 15.3) having vitrectomy to clear vitreous haemorrhage (excluding Terson's syndrome). The same audit revealed 23 patients with a similar age range to our present study (age range 6-50 years, M:F = 17:6) who had vitrectomy or phacovitrectomy for varied pathology excluding retinal detachment and Terson's syndrome. No patient developed either ES tear or postoperative retinal detachment.

Gnanaraj *et al* reported an incidence of 20% of 25 eyes with Terson's syndrome developing intraoperative breaks and 28% of cases had supplementary treatment with laser therapy, cryotherapy, and gas tamponade.⁴ Kuhn *et al* reported no intraoperative complications but two out of 33 eyes (6.1%) had postoperative rhegmatogenous retinal detachments.⁵ A vitrectomy study for non-diabetic vitreous haemorrhage reported ES breaks in one of four eyes with Terson's syndrome.³ These authors treated two of the four eyes prophylactically with cryotherapy and encirclement.

In our case series, patients remained phakic because of their relatively young age and therefore complete clearance of dense basal intragel haemorrhage was difficult. However, an increased incidence of ES breaks was not found in young patients who had vitrectomy for reasons other than Terson's syndrome. Small ES breaks hidden by the residual basal haemorrhage may have been present in the patients developing postoperative retinal detachment and became manifest following further contraction of the residual gel. There may also be an element of intrinsic retinal weakness in patients with Terson's syndrome caused by an episode of retinal ischaemia making them more prone to break formation. Retinal ischaemia has been suggested in explaining the mechanism of intraocular haemorrhage in Terson's syndrome.^{7,8}

We are aware of the limitations of a case note review with relatively small numbers of patients and the use of historical controls, but we recommend routine prophylaxis of all

entry sites in patients undergoing vitrectomy for Terson's syndrome.

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Table 1 Surgical outcome post-vitrectomy for Terson's syndrome

Eye	Visual acuity		Anaesthesia	ES break treated at time of surgery	Postoperative retinal detachment
	Preoperative	Postoperative			
1	PL	6/12, amblyopia	LA	Yes, one ES treated only	Yes
2	PL	6/24	LA	No	Yes
3	PL	6/7.5	GA	No	No
4	HM	6/9	GA	Yes	No
5	HM	6/6	GA	No	No
6	HM	6/4	LA	Yes	No
7	HM	6/9	LA	Yes	No
8	HM	6/9	LA	Yes	No
9	HM	6/9	LA	No	No
10	HM	6/9	GA	No	No
11	CF	6/6	LA	No	Yes

HM, hand movement; CF, counting fingers; PL, perception of light; LA, local anaesthesia; GA, general anaesthesia; ES, entry site.

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Bilateral aplasia of the optic nerve, chiasm, and tracts in an otherwise healthy infant

Optic nerve aplasia is a rare developmental anomaly characterised by the congenital absence of the optic nerve, central retinal vessels and retinal ganglion cells that is seen most often in a unilaterally malformed eye.^{1,2} Unilateral optic nerve aplasia is generally associated with otherwise normal brain development while bilateral optic nerve aplasia is usually accompanied by severe and widespread congenital central nervous system (CNS) malformations.^{1,2} We report here a unique presentation of a unilateral microphthalmos associated with bilateral optic nerve, chiasm, and tract aplasia without any CNS or systemic abnormalities.

Case report

An 11 month old girl was referred to the neuro-ophthalmology clinic because of unequal size of the eyes. She was born to non-consanguineous parents at 39 weeks gestation with a birth weight of 2750 g. The neonatal period was unremarkable. Developmental condition other than ocular findings was normal. She was the first child of her parents and there was no history of significant ocular disease in her family. Her parents' ocular examinations were normal.

Physical examination was normal, the patient's weight, length, and head circumference were normal for her age. Neurological examination was normal except for vision.

On examination she had no behavioural response to bright light and the pupils were not reactive to it. Right corneal diameter was 10 mm and left 7.5 mm. Both corneas were clear and anterior chambers were formed. Iris hypoplasia was found in both eyes. There was a congenital cataract in the left eye but the lens was clear in the right eye. On dilated fundus examination, the right eye showed absence of optic nerve and central retinal vessels (fig 1). Numerous large, round to oval, circumscribed, whitish to yellow areas of retinochoroidal depigmentation were visible in the periphery (fig 1).

B-scan echography showed a small left globe (axial length 14.80 mm), normal size right globe (axial length 22.30 mm), and absence of optic nerve on both eyes.

Figure 1 Fundus photograph of the right eye shows absence of optic disc, optic vasculature, and areas of retinochoroidal depigmentation.

Magnetic resonance imaging (MRI) of the orbits and brain disclosed marked asymmetry of globe size and bilateral absence of the optic nerve, chiasm and tract (fig 2A-D). There was only a rudimentary optic nerve behind both globes.

TORCH titres were negative and genetic evaluation including DNA analysis was unremarkable. Serum level of growth hormone, TSH, thyroxin, and glucose were all within normal limits.

Comment

Optic nerve aplasia denotes complete absence of the optic nerve and disc, retinal ganglion and nerve fibre layer, and optic nerve vessels. Histopathological examination usually demonstrates a vestigial dural sheath entering the sclera in its normal position, as well as retinal dysplasia with rosette formation.^{1,2} The pathogenesis of optic nerve aplasia remains speculative. It may be due to defective formation of the embryonal fissure, failure of the mesenchymal anlage of the hyaloid system to enter the embryonal fissure, or primary agenesis of the retinal ganglion cells.³

Bilateral optic nerve aplasia is rare, with most reported cases having major CNS malformations.^{1,2,4,5} To our knowledge there is only one previous report of bilateral optic nerve aplasia in an otherwise normal infant.⁶

Microphthalmos of the affected eye was a consistent finding in previous reports of unilateral or bilateral optic nerve aplasia,^{1,2,3,5-7} which was found only on our patient's left eye. To the best of our knowledge, this is the first case with bilateral optic nerve, chiasm, and tracts aplasia in an otherwise healthy infant with one microphthalmic and one normal size eye.

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Figure 2 (A) T1 weighted axial MRI demonstrates right normal size globe with a rudimentary optic nerve and left microphthalmic eye. (B) Axial MRI of chiasmatic section shows absence of chiasm. (C) T1 weighted axial MRI of optic tract section shows absence of tract signals. (D) T1 weighted sagittal MRI shows absence of optic chiasm.