Late onset vitreo-retinal complications of regressed retinopathy of prematurity

A Tufail, A J Singh, R J Haynes, C R Dodd, D McLeod, D G Charteris

Aim: To report the clinical findings, management, and outcomes in eyes undergoing surgery for regressed retinopathy of prematurity (ROP) with vitreoretinal complications.

Method: Retrospective review of 40 eyes of 32 patients with regressed ROP who presented between 1989 and 2001 at two UK referral centres.

Results: Of 29 eyes presenting with rhegmatogenous retinal detachment (RRD), 15 initially underwent a scleral buckling procedure and 14 initially underwent vitrectomy with or without additional buckling. Primary surgery was anatomically successful in 11/15 eyes that underwent a non-vitreoretinal retinal detachment repair and 8/14 that required vitrectomy. The final reattachment rate after reoperation was 28/29 eyes. Median visual acuity improved from 6/60 to 6/36 following retinal detachment repair. A further 11 eyes of eight patients from this series underwent prophylactic surgery, laser, or cryotherapy for predisposing vitreo-retinal pathology and/or retinal breaks, all of which were stabilised.

Conclusions: In eyes with RRD and signs of regressed ROP successful reattachment of the retina can be achieved using either vitrectomy or external surgery with an associated overall improvement in visual acuity. A range of external and closed microsurgical approaches is required to effectively deal with the diverse manifestations of regressed ROP.

The characteristics and natural history of tractional retinal detachment associated with retinopathy of prematurity (ROP) in neonates and infants have been extensively described. The presentation and management of the later sequelae of ROP have however been less well investigated. Retinal changes found in regressed ROP include dislocation of retinal vessels, peripheral vascular changes (incomplete vascularisation of peripheral retina, telangiectatic vessels, vascular arcs with circumferential interconnection, and abnormal branching of retinal vessels), and vitreoretinal interface changes (pigmentary changes, peripheral folds, vitreous membranes, and lattice-like degeneration). A number of early reports described the characteristics and treatment of rhegmatogenous retinal detachment (RRD) in patients with regressed ROP, but the management of such detachments has been less well documented since the advent of pars plana vitrectomy surgery. Two more recent reports have described the surgical management of late onset RRDs associated with regressed ROP in the United States. The purpose of this study was to document the clinical findings, contemporary management, and outcomes for patients with regressed ROP referred for vitreoretinal surgical assessment to two major centres in the United Kingdom.

PATIENTS AND METHODS

A retrospective review was undertaken of the records of 32 patients with regressed ROP who had been referred for vitreoretinal surgical assessment to the Royal Eye Hospital, Manchester or Moorfields Eye Hospital, London during the period 1989–2001. Data were extracted and analysed only in those patients who fulfilled the following criteria: a history of prematurity (defined as being born at 37 or less weeks of gestational age), and/or very low birth weight (less than 1500 g) with or without oxygen administration and stay in a special care baby unit, together with ophthalmoscopic findings consistent with regressed ROP. None of the patients included in the study had previously undergone either posterior or anterior segment surgery or previous treatment such as scatter laser photocoagulation, for the acute manifestations of ROP. Refractive error was also recorded, but axial length data were not available to differentiate between refractive abnormalities due to malposition of the iris/lens diaphragm and abnormal globe size. Eyes could not be staged accurately by the International Classification of Retinopathy of Prematurity, as most of the patients initially presented before the widespread adoption of the classification. Clinical details were recorded and included: Snellen distance visual acuity (initial and final), vitreoretinal findings at presentation, surgery undertaken, and the anatomical and functional outcomes. The mean length of follow up was 40 months (range 2–144 months).

RESULTS

Forty eyes of 32 patients underwent either retinal reattachment surgery (n = 29) or prophylactic treatment of vitreoretinal pathology (n = 11). All patients included in this study had either a history of prematurity (mean 28.8 weeks gestation, range 24–34 weeks), and/or low birth weight (mean 1110 g, range 730 to 1600 g) with or without oxygen administration in a special care baby unit (mean length of stay 12 days, range 2–33 days). Three patients had intraventricular haemorrhage related to prematurity, and two of these patients also had necrotising enterocolitis. The mean age at presentation of RRD was 22.3 years (range 8–59 years) though it was notable that most patients fell into two age groups; 8–20 years (62.5%) and 32–45 years (32.5%) (with a single outlier aged 59) (table 1). Myopia was recorded in 90% of eyes (mean 5.9 dioptres, range 0 to −12 dioptres). The retinal vasculature emanating from the optic disc and/or the macula was heterotopic in 33 (82.5%) eyes, temporally displaced in 26 (65%) eyes and nasally displaced in seven (17.5%) eyes. Non-retinal ocular abnormalities noted included four (10%) eyes presenting with cataract, 12 (30%) patients with a history of strabismus, and 11 (27.5%) patients with nystagmus. Fifteen eyes (37.5%) had signs of
lattice peripheral retinal degeneration and nine (22.5%) had peripheral retinal pigmentary changes.

The presenting vitreoretinal signs and management of these patients are summarised in table 2. Retinal breaks were discovered preoperatively or intraoperatively in 28 of the 29 eyes undergoing retinal detachment surgery; the mean number of breaks found was 1.9 (median 1, range 1–6). In patients who required only one vitreoretinal procedure the mean number of breaks found at initial presentation was 1.6, whereas in eyes requiring more than one procedure the mean number of breaks was 2.6. In two eyes, the breaks were posteriorly located while in the remainder they were equatorial. The majority of breaks were located in the temporal retina (62% superotemporal, 22.5% inferotemporal, 10.5% inferonasal, and 7% superonasal (A)). In patients with nasal ectopia (n = 7) of the major disc vessels the majority of the retinal breaks (67% of the total breaks in this subgroup) were also located in the temporal retina.

The first or primary operation was successful in reattaching the retina in 20 of 29 eyes (69%). Nine of the 29 eyes required between one and three further procedures to achieve final overall reattachment in 28 of the 29 eyes. Fifteen eyes had initial conventional scleral buckling surgery, the extent of buckling reflected the extent of identified traction as indicated by the presence of lattice degeneration or other evident areas of non-detachment of the vitreous. Conventional scleral buckling surgery successfully reattached the retina in 11 (73%). The other 14 eyes underwent primary vitrectomy (which was combined with a localised scleral buckle or encirclement in nine). Two of these eyes required vitrectomy to allow treatment of posteriorly located retinal breaks and in one eye vitrectomy facilitated relief of vitreoretinal traction (no break being discovered though the detachment configuration was in keeping with a rhegmatogenous component). Primary vitrectomy surgery was successful in achieving reattachment in 8/14 (57%) eyes. Of the nine eyes in which primary surgery failed, three underwent vitrectomy and oil exchange (patients 8, 10, and 23), one encirclement and oil exchange (patient 20), two vitrectomy and gas insertion (patients 21 and 29), one vitrectomy, retinectomy and oil (patient 19), and two cryotherapy and buckle (patients 4 and 17). Patient 29 redetached 1 month after his initial encirclement and drain procedure and required vitrectomy, gas insertion, and rebuckling to successfully reattach the retina.

At presentation 21 of 29 eyes with RDD had involvement of the macula. The median preoperative visual acuity was 6/60 (6/18 for macula on and 6/60 for macula off detachment) and improving to a median 6/36 (6/18 for macula on, 6/36 for macula off detachment) postoperatively at the last follow up visit. For macula on detachments there was an overall mean one line of vision loss with 3/8 (42%) patients losing one or more lines of vision (range 1–5 lines lost). For macula off detachments there was an overall mean one line of vision gain with 4/21 (19%) patients losing one or more lines of vision (range 1–7 lines).

An additional 11 eyes without RRD underwent prophylactic treatment. In four eyes of two patients, an encircling procedure was performed either for a peripheral traction detachment or for lattice degeneration with vitreoretinal traction. Eight eyes underwent cataract and/or indirect laser photocoagulation for flat retinal breaks or lattice degeneration (table 2). There was no progression to RRD or tractional retinal detachment in any of these eyes during a mean follow up of 14 months (range 6–40 months).

**DISCUSSION**

This series of patients with regressed ROP serves to illustrate the variable clinical features of the condition, which can lead to problems in recognition and management. A history of
premature birth with oxygen treatment may be not be volunteered many years later and clinicians should always consider regressed ROP in unusual cases of retinal detachment (particularly in children and young adults with other ocular abnormalities such as myopia, strabismus, nystagmus and cataract). Signs of chronicity, such as subretinal strands, tide marks, retinal atrophy and fibrosis are typical of regressed ROP retinal detachments. This may not be related to regressed ROP itself since chronicity is a recognised feature of RRD in younger patients. An absence of reported symptoms is also characteristic of RRD in this age group and was seen in a number of cases in this series.

The pathological changes found in chronic ROP have been described by Foos and the clinical features by Tasman. The cases we report have clinical features consistent with this pathology. For example, the elevated white ridge in the retina is thought to represent an earlier onset of ROP, and the presumed associated nasal ectopia of the disc vessels has been recorded in other cases of late ROP. It is notable that in our cases of nasal ectopia there did not seem to be any increased frequency of nasal rather than temporal retinal breaks, suggesting that vitreoretinal traction has its primary effect in the temporal retina. The majority of retinal breaks were found temporally, consistent with previously reported findings. The level of myopia (mean −5.9 dioptres) has previously been reported in patients with both treated and untreated ROP.

The failure rate of primary surgery in our series was relatively high (31%) and is similar to the failure rates for RRD (23% primary failure) reported by Kaiser and co-workers, and an initial failure rate of 50% with one procedure (12% final failure rate) by Sneed and co-workers. The complex vitreoretinal association with widespread variable vitreoretinal adhesions together with multiple retinal breaks may account for the high failure rate.

All the extensive retinal detachments presenting for assessment were considered to have a rhegmatogenous fundus abnormality. The final outcome was considered to have a rhegmatogenous fundus abnormality. Therefore, the fundal features might not always correspond to textbook descriptions of ROP. For example, ectopia of the macula and disc vessels is generally towards the temporal periphery but seven of the patients in this series had nasal dragging. Disease preferentially affecting the nasal retina is thought to represent an earlier onset of ROP, and the nasal ectopia of the disc vessels has been previously reported in patients with both treated and untreated ROP.
component, none being apparently analogous to the series of tractional retinal detachments reported by Machemer. Nevertheless, it appears that abnormal vitreoretinal traction is an important associated feature of the RRDs in ROP due to either the variable residue of retrolental fibroplasia (which may extend posterior to the vitreous base in some ROP eyes), or to variable tractional manifestations at the equator with or without lattice-like features. Evidence of vitreoretinal traction does not preclude a successful surgical outcome from conventional scleral buckling surgery but where traction is marked or in the presence of posteriorly located retinal breaks, vitrectomy (possibly in conjunction with scleral buckling) should be considered. Thus, a range of external and closed microsurgical approaches is required to effectively deal with the diverse manifestations of regressed ROP.

As the screening procedures for acute ROP improve in the future, affording the opportunity for timely cryosurgery or laser photocoagulation to the ischaemic retinal periphery, presentations of RRD from spontaneously regressed ROP might be expected to decline in frequency. However, RRD can also occur in eyes with previously treated ROP and the late vitreoretinal sequelae of treated acute ROP may come to replace spontaneously regressed ROP as a vitreoretinal surgical challenge in the future.

References


