CASE REPORT

Two-staged en bloc excision of a retinal haemangioma

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SUMMARY

A 31-year-old man presented to us with diminution of vision in the right eye which he noticed since 10 days, with a visual acuity of hand motions only. On examination, festooned pupil and a complicated cataract were noted with no view of the retina. Ultrasonography of the right eye showed retinal detachment in all quadrants with suspected areas of traction and membranes in the vitreous. Left eye examination revealed a normal anterior and posterior segment. Intraoperatively, he underwent a pars plana lensectomy following which an inferior large retinal haemangioma was noted with dilated and tortuous feeder artery and draining vein. The haemangioma was managed by a two-staged procedure including ligation of the feeder artery in the first surgery and the en bloc excision of the angioma at the time of the second procedure after significant shrinkage in size of the tumour.

BACKGROUND

Retinal angiomas (aka haemangiomas/haemangioblastomas) are characteristic benign vascular hamartomas most commonly associated with von Hippel-Lindau (VHL) syndrome. They are one of the earliest manifestations of VHL making the role of an ophthalmologist very important in screening and management of such cases. They often present during the second or third decade of life with exudative complications and can be a challenge to manage.¹ Traditionally, retinal haemangiomas have been treated by laser photocoagulation, cryotherapy or plaque brachytherapy. Rarely, photodynamic therapy (specifically for juxtapapillary tumours), radiotherapy, transpupillary thermotherapy and recently intravitreal antivascular endothelial growth factor agents have been reported for treatment with mixed results.^{2–5} Surgical management of haemangioma by transretinal feeder vessel ligation or in rare instances by en bloc excision has been described by few mainly in the setting of an associated retinal detachment.^{6–10} We hereby describe a case of haemangioma presenting with retinal detachment managed surgically in a two-step approach

festooned pupil and a normal intraocular pressure without any view of the retina. Left eye was asymptomatic and the patient had an unremarkable systemic and family history.

INVESTIGATIONS

Ultrasonography of the right eye showed a retinal detachment in all quadrants with suspected areas of traction (figure 1). The left eye was within normal limits. A diagnosis of chronic retinal detachment secondary to uveitis or unnoticed trauma was considered and surgical management was planned.

TREATMENT

The patient underwent a pars plana lensectomy with a 20-gauge vitrectomy. A large inferior retinal capillary haemangioma with dilated and tortuous feeder artery and draining vein was noted subsequently with an associated retinal detachment (figure 2). Both subretinal and preretinal membranes surrounding the haemangioblastoma were dissected meticulously. A plane was created beneath the feeder artery using intraocular forceps while dissecting the membranes. A 10–0 prolene suture was introduced through one of the sclerotomies and passed beneath the feeder artery, and ligation was performed using two intraocular forceps (figure 3). Immediately after placement and tightening of the knot, shrinkage of the angioma and reducing calibre of the vessels were noted. A retinotomy was made inferotemporally after adequate dissection and retina was settled under air. After adequate endolaser and cryotherapy, silicon oil (1000 centistokes equivalent to 10 stokes or m^2/s) was injected for tamponade and patient was advised strict prone positioning. Postoperative fundus fluorescein

Figure 1 Preoperative ultrasonography of right eye showing medium reflective membranes with suspected areas of traction.

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CASE PRESENTATION

A 31-year-old man presented to us with diminution of vision in his right eye noticed since 10 days. At presentation, he had a best-corrected visual acuity of hand motions only with accurate projection of rays. Right eye had a complicated cataract with a

Figure 2 Intraoperative view of the haemangioma with dilated and tortuous feeder artery and draining vein.

Figure 3 Intraoperative view of ligation of the feeder artery.

angiography confirmed the ligation and reduced flow in the feeder vessels (figure 4).

Retina remained attached postoperatively with visual acuity improvement to 20/80, however, 1 month postoperatively, an inferior recurrent retinal detachment with attached macula was noted and patient was taken up for resurgery.

Figure 4 Postoperative fundus fluorescein angiography confirming the non-filling of the draining artery postligature.

Figure 5 Photomicrograph of the excised specimen in H&E stain showing thin capillary-like channels lined with endothelial cells separated by foamy stromal cells.

During the second surgery in toto excision of angioma was carried out with a retinectomy done around the lesion, ensuring it was anterior to the suture taken earlier, thus achieving a bloodless field. Histopathological examination confirmed the diagnosis of retinal angioma (figure 5).

Systemic evaluation of the patient including MRI (brain and spine), urine vanillylmandelic acid, abdominal sonography were done and were within normal limits.

OUTCOME AND FOLLOW-UP

Patient was doing well during the postoperative period with visual acuity maintained at 20/200 with attached macula, however, inferior recurrent retinal detachment was again noted at the 6-monthly follow-up. No reproliferation of the angioma was noted.

DISCUSSION

Exudative complications of retinal angiomas are the the most common cause of visual loss, but retinal detachment associated with these angiomas has been rarely reported as a cause of visual loss. The surgical technique for management of these complex detachments, which are often tractional or combined detachments is complex indeed. Kwan *et al*¹⁰ described a similar case of combined tractional and rhegmatogenous retinal detachment in their report and advised on the careful excision of the base of these angiomas as they often have associated chorioretinal vascular anastomoses which may bleed.

Khurshid⁸ and Schlesinger *et al*⁹ reported en bloc resection of these tumours without prior ligation. They noted that these tumours may have a higher incidence of sporadic occurrence than anticipated explaining the sporadic occurrence of angioma in our patient.

We performed a two-staged procedure to ensure a bloodless field. By the time of the second surgery, the size of the

Learning points

- ▶ Retinal haemangiomas are the the most common and earliest manifestations of von Hippel-Lindau syndrome but sporadic solitary haemangiomas can also occur.
- ▶ Treatment of these haemangiomas is complex and is governed by their size and the related ocular morbidity.
- ▶ Surgical management can be considered as a two-staged procedure to ensure easy removal of the tumour and better management of intraoperative bleeding.

angioma had significantly reduced with shrinkage in size of feeder vessels, thus making our intraoperative manipulations easier.

One limitation of this case report was that such a large-sized haemangioma was overlooked preoperatively on the ultrasound, probably due to low index of suspicion and presence of extensive membranes and a large tractional retinal detachment. Thus, this should be borne in mind as a differential diagnosis for preoperative assessment of young adults with retinal detachment with no view of retina and ultrasound features of chronic retinal detachments like intraretinal cysts and membranes.

Contributors MJ and KA contributed to literature search and drafting of the manuscript. JC was involved in active management of the patient as the operating surgeon as well as helped in drafting and revision of the manuscript. PKR contributed in drafting and revision of manuscript.

Competing interests None declared.

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