Periocular steroids for macular edema associated with retinal arteriovenous malformation: A case report

Roshni Mohan, Romana Fazal

Retinal racemose hemangiomas (RRH) are vascular malformations comprising of direct arteriovenous communications in the retina. Exudation and neurosensory detachment are some of the complications which may cause decreased visual acuity. Herein, we describe a case of a 38-year-old male presenting with unilateral Group II RRH complicated with macular edema. Initial treatment with intravitreal bevacizumab yielded a poor therapeutic response. Subsequently, he was treated with a posterior sub-tenon injection of triamcinolone acetonide following which there was a prompt decrease in edema with simultaneous improvement in vision. The visual acuity was maintained and no recurrence was seen even after 6 months of successful treatment.

**Key words:** Bevacizumab, periocular steroids, racemose hemangioma, retinal arteriovenous malformation, triamcinolone

Retinal racemose hemangiomas (RRH) or arteriovenous malformations (AVM) are rare vascular anomalies of the retina with only about 167 cases reported in the literature.[1] Approximately 30% of patients have concurrent intracranial...
AVM, the condition is termed as Wyburn Mason syndrome.\textsuperscript{[2]} It may sometimes be associated with macular edema and cause defective vision. Therefore, in the present study, we report the clinical features and management of a case of Group II RRH with macular edema.

Case Report

A 38-year-old male presented with sudden onset drop in his left eye vision for 2 days. His systemic history was insignificant. Best-corrected visual acuity (BCVA) was 20/20 in the right eye and 20/20p in the left eye. Visual axes were parallel while pupillary reaction and intraocular pressures were normal in both eyes. Fundoscopy of the left eye showed an abnormal network of vessels near the supero-temporal arcade with surrounding retinal edema and hard exudates encroaching into the macula [Fig. 1a and 2a]. The first branch of the supero-temporal artery (black arrow) was seen communicating with the supero-temporal vein (star) via an abnormally dilated communicating vessel (green arrow) [Fig. 1b]. The surrounding capillary bed, arteriolar, and venular elements were also dilated. The artery was tortuous, beaded, and the vein showed a kink at its junction with the abnormal communicating vessel. The color of the blood column in the involved segment of the vein was bright red signifying the rapid transit of highly oxygenated blood from the artery into the vein. The segment of the supero-temporal vein distal to the AV communication showed some dilatation and few retinal hemorrhages suggesting a minor degree of branch retinal vein occlusion (BRVO). Fundus fluorescein angiography (FFA) showed the filling of this abnormal vascular communication and the draining vein in the early phases [Fig. 3]. The surrounding vascular elements and the area of kink showed leakage in the late phases. The distal portion of the supero-temporal vein showed delayed filling with AV transit time of 30 s. A diagnosis of Group II RRH with macular edema was made. Optical coherence tomography (OCT) showed intraretinal cysts with subfoveal detachment, few hyperreflective lesions corresponding to hard exudates, and a central retinal thickness (CRT) of 501 µ [Fig. 4a]. Magnetic resonance imaging with angiography of the orbit and brain was unremarkable.

We decided to inject intravitreal bevacizumab (IVB, 1.25 mg/0.05 mL), given macular edema. One-month postinjection, the hard exudates, and edema had increased with CRT of 524 µ and vision had dropped to 20/40 [Fig. 4b and 2b]. Due to the worsening of symptoms and to avoid complications of intravitreal injections, a trial of posterior sub-tenon injection (PST) of triamcinolone acetonide (40 mg/mL) was given. Three weeks later, macular edema showed a significant decrease, with complete resolution and restoration of vision to 20/20 by 6 weeks [Fig. 2c and 4c, d].

Discussion

RRH is a rare sporadic phacomatoses, classified by Archer et al. into three groups. Group I lesions show an intervening capillary bed between the AV communication, Group II lesions lack a capillary bed, and in Group III, the arteries and veins cannot be differentiated.\textsuperscript{[3]} The arterial pressure is directly transmitted to the veins via the AV communicating vessel. Reports of macular edema associated with AVM suggest that the hyperdynamic flow characteristics of such vascular anomalies predispose the surrounding capillaries or sometimes even the AVM itself, to start leaking due to excessive back pressure from the elevated venous transmural pressure.\textsuperscript{[4,5]} IVB has been successfully used in treating such cases.\textsuperscript{[6,7]} The use of periocular triamcinolone and focal laser photocoagulation to the leaking AVM segment has also been rewarding.\textsuperscript{[4,5,8]} Chuang et al. even documented regression of a Group II AVM after three injections of IVB.\textsuperscript{[9]} Our case was a Group II lesion with macular edema probably due to leakage from the surrounding capillary bed. The junction of the AV communicating vessel with the vein showed structural changes and leakage probably due to an abrupt increase in the venous transmural pressure. The distal segment of the vein showed a minor degree of functional branch vein occlusion due to the same reason. Anti-vascular endothelial growth factor (anti-VEGF) has well-documented actions on decreasing edema by decreasing vascular permeability. Steroids, perhaps by acting on a broader group of mediators causing vascular hyperpermeability help to decrease macular edema. However, the inability to treat the underlying cause may lead to recurrent episodes of decompensation and therefore, such entities have to be regularly monitored for life or until spontaneous regression.

Figure 1: (a) Fundus photo showing Group II RRH with macular edema. (b) Magnified image showing the tortuous beaded supero-temporal artery (black arrow), AV malformation (green arrow), involved segment of the supero-temporal vein (star)

Figure 2: Comparative fundus images of the left eye. (a) At presentation. (b) 1-month post IVB. (c) 6-weeks post PST
has been unequivocally documented. The events which can precipitate decompensation are still unclear. The influence of age, systemic conditions, or the chronic hyperdynamic condition itself may lead to microvasculature abnormalities and subsequent loss of vascular integrity and leakage. The component of ischemia due to highly unsaturated blood bypassing the capillaries and causing tissue hypoxia may be another explanation for the surrounding leaking vasculature. Vascular changes in an old BRVO may be a differential of our case. However, in BRVO, collaterals appear thinner and FFA did not reveal peripheral vascular and retinal capillary changes characteristic of BRVO.

We followed the patient up to 6 months with no recurrences being documented. We do not know why the macular edema increased after the anti-VEGF injection or whether it would have responded to multiple anti-VEGF injections. This report highlights the efficacy of periocular triamcinolone in treating macular edema associated with AVM showing suboptimal response to anti-VEGF.

Conclusion

Macular edema associated with RRH can be treated with periocular steroids. Its natural course has not been well-elucidated, therefore, long-term follow-up is necessary.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Qin XJ, Huang C, Lai K. Retinal vein occlusion in retinal racemose hemangioma: A case report and literature review of ocular complication in this rare retinal vascular disorder. BMC Ophthalmol 2014;14:101.
2. Wyburn-Mason R. Arteriovenous aneurysm of midbrain and retina, facial nevi and mental changes. Brain Dev 1943;66:40.
3. Archer DB, Deutman A, Ernest JT, Krill AE. Arteriovenous communications of the retina. Am J Ophthalmol 1973;75:224-41.
4. Onder HI, Alisan S, Tunc M. Serous retinal detachment and cystoid macular edema in a patient with Wyburn-Mason syndrome. Semin Ophthalmol 2015;30:154-6.
5. Soliman W, Haamann P, Larsen M. Exudation, response to photocoagulation and spontaneous remission in a case of bilateral racemose haemangioma. Acta Ophthalmol Scand 2006;84:429-31
6. Winter E, Elsås T, Austeng D. Anti-VEGF treating macular oedema caused by retinal arteriovenous malformation – A case report. Acta Ophthalmol 2014;92:192-3.
7. Pangtey BP, Kohli P, Ramasamy K. Wyburn–Mason syndrome presenting with bilateral retinal racemose hemangioma with unilateral serous retinal detachment. Indian J Ophthalmol 2018;66:1869-71.
8. Federici T, Battle I. Periocular triamcinolone acetonide as treatment for macular edema secondary to branch vein occlusion associated with retinal arteriovenous malformation. Retina 2006;26:1079-80.
9. Chuang LH, Wang NK, Chen YP, Wu WC, Lai CC. Mature vessel occlusion after anti-VEGF treatment in a retinal arteriovenous malformation. BMC Ophthalmol 2013;13:60.