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Case Report

Intravitreal bevacizumab in congenital retinal macrovessel with retinal arteriolar macroaneurysm

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Abstract

Congenital retinal macrovessel (CRM) refers to an aberrant vessel, usually a vein, which traverses the macula and supplies both sides of the horizontal raphe. It is a rare condition, mostly asymptomatic and discovered on routine examination. We describe a case of both arterial and venous CRM with a macroaneurysm along the arterial CRM that presented with decreased vision due to prominent lipid exudation at the macula. Treatment with intravitreal bevacizumab resulted in a favourable anatomical as well as functional outcome. To the best of our knowledge, this is the first report of this unusual presentation of CRM, and its successful management with intravitreal bevacizumab.

Keywords: Congenital retinal macrovessel, Retinal artery macroaneurysm, Intravitreal bevacizumab

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Introduction

The term ''congenital retinal macrovessel'' (CRM) was coined by Brown et al. to describe an aberrant retinal vessel, frequently a vein, crossing the central macula and supplying or draining both above and below the horizontal raphe.[1] The condition is usually unilateral, and rarely affects visual acuity.[1,2] We hereby describe an unusual case of arterial as well as venous CRM which was associated with reduction in vision. This was attributed to macular exudation from a macroaneurysm along the arterial CRM. The patient was treated successfully with intravitreal bevacizumab. To the best of our knowledge, this is the first report of intravitreal bevacizumab in retinal arteriolar macroaneurysm associated with CRM.

Case report

A 65 year old female presented with complaints of blurred vision in her left eye since one month before. She had poor vision in her right eye following cataract surgery done 12 years back. She was a hypertensive well controlled by medication.

On examination, her right eye had no light perception, due to aphakic bullous keratopathy and secondary glaucoma. The best corrected visual acuity (BCVA) in her left eye was 20/200. Anterior segment examination including intraocular pressure was unremarkable apart from the presence of a posterior chamber intraocular lens. Dilated fundus examination revealed prominent lipid exudation with aberrant vessels at the centre of the macula (Fig. 1a). Fluorescein angiography showed an arterial CRM arising from the superior branch of the central retinal artery (Fig. 1b) and a venous CRM draining into the inferior branch of the central retinal vein before bifurcation (Fig. 1c). A small macroaneurysm was present along the arterial CRM just temporal to the temporal margin of the optic disc (Fig. 1c and d). There were prominent retinal vascular calibre abnormalities of the superotemporal retinal artery which anastomosed with the numerous branches of the venous CRM. In addition, there was markedly increased

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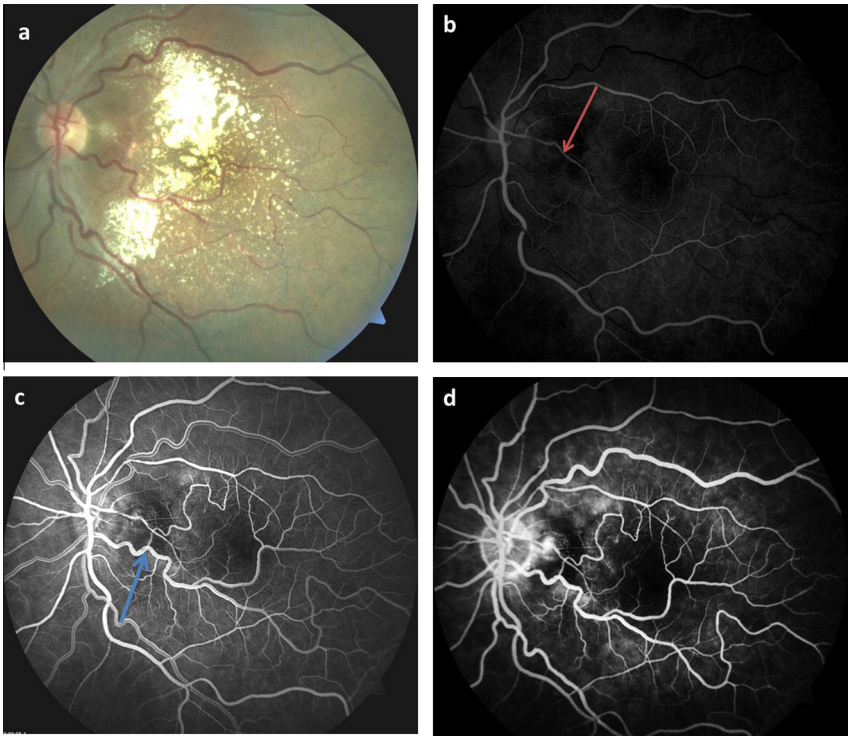

Figure 1. (a) Colour fundus photograph of the left eye showing macular exudation with aberrant vessels. (b) Fundus fluorescein angiography revealing an arterial CRM arising from the superior branch of the central retinal artery (red arrow). (c) A venous CRM was also seen draining into the inferior trunk of the central retinal vein (blue arrow). Just temporal to the temporal margin of the optic disc, filling of a macroaneurysm along the arterial CRM was seen. (d) Late phase showing leakage from the macroaneurysm, with multiple ill defined leaks in the nasal part of the macula.

branching and anastomosis of vessels in the perifoveal area. This anomalous capillary network contained multiple pin point leaks, especially the nasal to the fovea (Fig. 1d). No capillary nonperfusion areas were noted. Spectral domain optical coherence tomography (SD-OCT) was performed and showed intraretinal hard exudates along with retinal oedema in the nasal part of the macula (Fig. 2a and b).

After being informed about the off-label use of intravitreal bevacizumab as a treatment option, the patient agreed to receive an intravitreal injection of bevacizumab (1.25 mg in 0.05 ml) in her left eye. At one month follow up, BCVA improved to 20/60 with marked decrease in the hard exudates and macular oedema. A second injection of intravitreal bevacizumab was administered. At one month following the second dose, BCVA improved to 20/30 with further reduction in retinal hard exudates (Fig. 2c). SD-OCT confirmed the decrease in macular oedema and hard exudates and revealed normalization of the foveal contour (Fig. 2d). This picture was maintained till two years follow up with no recurrences.

Discussion

A large retinal vessel crossing the horizontal raphe in the macular area is termed as a CRM. Since the first description in 1869 by Mauthner,[3] fewer than 50 cases have been reported. A CRM is usually a retinal vein, rarely it could be an artery or both artery and vein. Our case had both arterial as well as venous CRM; this has been described rarely in literature.[1,2]

CRMs are usually benign and stable and are mostly detected in asymptomatic patients on routine examination. Occasionally, a CRM can present with decreased vision which has been attributed to haemorrhage, foveal cyst, the vessel itself crossing the fovea[1,2] and central serous chorioretinopathy.[4] Our case presented with a macroaneurysm along the arterial CRM and prominent macular exudation, with retinal oedema. Only 2 case reports of arterial CRM with a leaking macroaneurysm affecting visual acuity were found in literature.[5,6] Koizumi et al. also described a retinal arteriolar macroaneurysm and CRM, however, both lesions were distinct and unrelated.[7]

Macroaneurysms are acquired dilatations of the retinal arteries that usually occur in elderly hypertensive women. It has been suggested that the lack of autoregulation in anomalous vessels may contribute to the development of macroaneurysms in CRMs.[6] Symptomatic macroaneurysms may be haemorrhagic or exudative.[8] While spontaneous involution of macroaneurysms with functional recovery is known, it has been suggested that patients with subretinal haemorrhage or exudative manifestations involving the fovea and visual acuity deterioration should be treated to avoid irreversible photoreceptor damage.[9] Conventional laser photocoagulation has been the most commonly employed treatment for symptomatic retinal arterial macroaneurysm, however, in this patient, the location of the microaneurysm in the region of the papillomacular bundle was not amenable to this form of therapy. Since the development of retinal artery macroaneurysm is associated with focal damage to arterial walls, leading to localized ischaemia and VEGF upregulation, the use of anti-VEGF agents has been suggested as a treatment modality. Intravitreal bevacizumab has been shown to hasten resolution of macular oedema and haemorrhage secondary to retinal artery macroaneurysm, with improvement in visual acuity.[10,11]

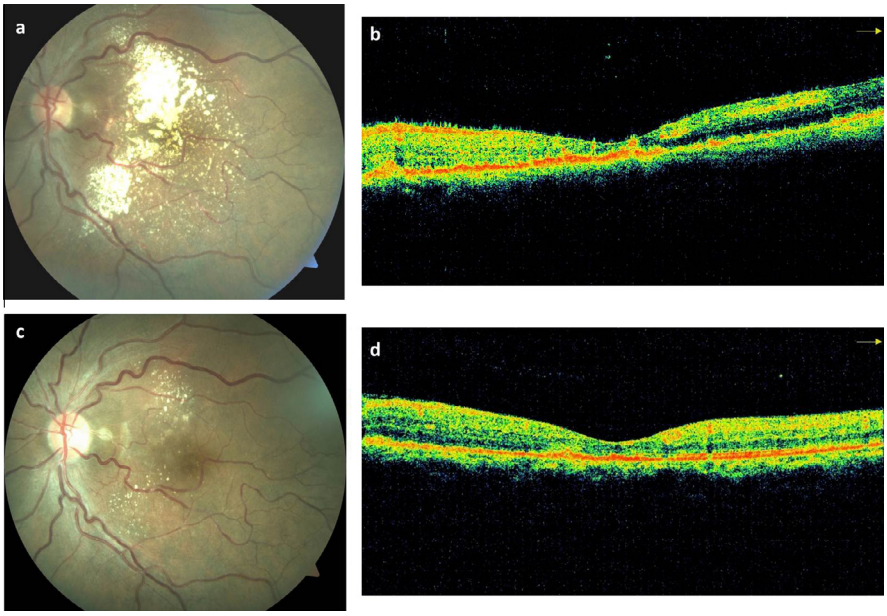

Figure 2. (a) Colour fundus photograph of the left eye at presentation. (b) Horizontal spectral domain optical coherence tomography (SD-OCT) scan showing scattered intraretinal hard exudates (also subfoveal) and macular oedema (nasally). (c) Colour fundus photograph of the left eye 2 months after presentation, following 2 monthly injections of intravitreal bevacizumab. There was considerable reduction in the exudation. (d) Horizontal SD-OCT scan showing decrease in hard exudates and oedema.

VEGF inhibition after administration of intravitreal bevacizumab could have reduced vascular permeability, with consequent resolution of the lipid exudation and retinal oedema in our patient. This resulted in considerable improvement in visual acuity which was maintained till two years of follow up. While the CRM and macroaneurysm persisted, there was no recurrent retinal oedema or lipid exudation. This is the first report of successful treatment of visual impairment due to a macroaneurysm in an arterial CRM with intravitreal bevacizumab.

References

1. Brown GC, Donoso LA, Magargal LE, Goldberg RE, Sarin LK. Congenital retinal macrovessels. *Arch Ophthalmol* 1982;**100**:1430–6.
2. de Crecchio G, Alfieri MC, Cennamo G, Forte R. Congenital macular macrovessel. *Graefes Arch Clin Exp Ophthalmol* 2006;**244**:1183–7.
3. Mauthner L. *Lehrbuch der ophthalmoscopie*. Vienna: Tendler and Co; 1869, 249.
4. Kumar V, Ghosh B, Raina U, Goel N. Central serous chorioretinopathy in a patient with congenital retinal macrovessel. *Can J Ophthalmol* 2009;**44**:e57.
5. Chalam KV, Gupta SK, Vinjamaram S, Shah VA. Clinicopathologic reports, case reports, and small case series: congenital anomalous retinal artery associated with a leaking macroaneurysm. *Arch Ophthalmol* 2003;**121**:409–10.
6. Musadiq M, Gibson JM. Spontaneously resolved macroaneurysm associated with a congenital anomalous retinal artery. *Retin Cases Brief Rep* 2010;**4**:70–2.
7. Koizumi H, Iida T, Mori T, Furuta M, Yannuzzi LA. Retinal arteriolar macroaneurysm and congenital retinal macrovessel. *Ophthalmic Surg Lasers Imaging* 2009;**40**:513–5.
8. Lavin MJ, Marsh RJ, Peart S, Rehman A. Retinal arterial macroaneurysms: a retrospective study of 40 patients. *Br J Ophthalmol* 1987;**71**:817–25.
9. Tsujikawa A, Sakamoto A, Ota M, Oh H, Miyamoto K, Kita M, et al. Retinal structural changes associated with retinal arterial macroaneurysm examined with optical coherence tomography. *Retina* 2009;**29**:782–92.
10. Pichi F, Morara M, Torrazza C, Manzi G, Alkabes M, Balducci N, et al. Intravitreal bevacizumab for macular complications from retinal arterial macroaneurysms. *Am J Ophthalmol* 2013;**155**:287–94.
11. Cho HJ, Rhee TK, Kim HS, Han JI, Lee DW, Cho SW, et al. Intravitreal bevacizumab for symptomatic retinal arterial macroaneurysm. *Am J Ophthalmol* 2013;**155**:898–904.