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Case Report

A Case of Recurrent Vasculitic Vitreous Haemorrhage in One Eye and Idiopathic Choroidal Granulomas in The Fellow Eye of A Young Individual

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Abstract

A 26-year-old male presented with sudden diminution of vision in right eye (OD) for 15 days. Best-corrected visual acuity (BCVA) was finger counting 1-metre in OD and 6/6 in left eye (OS). Fundus examination revealed fresh vitreous haemorrhage (VH) in OD. Investigations including complete haemogram, VDRL, Mantoux test and Contrast-Enhanced Computed Tomography chest were normal. A presumptive diagnosis of Eales disease was made. Subsequently he also developed multiple small choroidal granulomas in OS. He was started on oral corticosteroid and treated with multiple episodes of scatter-laser and intravitreal bevacizumab in OD for recurrent VH. He ultimately required pars-plana vitrectomy in OD for persistent VH despite anti-VEGF and laser. Choroidal granulomas in OS disappeared after treatment with oral steroid. He maintained BCVA of 6/6 in both eyes after 6-months and was kept under close follow-up. Multimodal approach and combination modality of treatment may be required for optimal outcome in such cases.

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Keywords: *Eales Disease, Retinal Vasculitis, Pars Plana Vitrectomy, Vitreous Haemorrhage, Anti-Vegf*

Introduction

Eales' disease (ED), an idiopathic occlusive vasculitis mainly affects the mid-peripheral retina of young males aged 20-40 years. Henry Eales first observed this condition in the 1880s manifesting as recurrent retinal haemorrhages along with headaches, epistaxis, dyspepsia, or chronic constipation in young, otherwise healthy men. Wadsworth (1887) highlighted this condition to be associated with retinal inflammation. The disease is more prevalent in Asian countries particularly India, Pakistan and Afghanistan.[1-4] Elliot (1954) described this condition as 'retinal periphlebitis', after noting the presence of perivascular inflammation with retinal haemorrhages. Hallmark changes are perivascular phlebitis, peripheral non-perfusion and neovascularization. Vision loss is characteristically caused by bilateral (often) recurrent, vitreous haemorrhage (VH).[5]

Current treatment option includes corticosteroids, laser photocoagulation, anti-VEGF injections and surgical management in form of Pars Plana Vitrectomy (PPV). The main indications for vitrectomy include non-resolving VH, tractional retinal detachment involving the posterior pole, multiple vitreous membranes with or without tractional retinal detachment, and combined tractional and rhegmatogenous retinal detachment. Evaluation of vitreous fluid of patients of ED has revealed increased expression of VEGF, highlighting the possible role of intravitreal anti-VEGF in treatment and managing progression of ED.[6,7]

We hereby present the multimodal management of a case of ED in a young individual presenting with recurrent episodes of VH.

Case report

A 26-years-old male presented to the Ophthalmology outpatient department of our Institute with sudden painless diminution of vision in the right eye (OD) for 15 days. This decline in vision was also associated with floaters. He took consultation from a private hospital where he was diagnosed as central retinal venous occlusive disease of suspected inflammatory etiology in OD and was started on oral corticosteroids. He did not have any similar episodes in the past and he had no systemic illness.

Vital signs and systemic physical examination including blood pressure were unremarkable. On initial ophthalmological examination, the best-corrected visual acuity (BCVA) was 1/60 in OD and 6/6 in the left eye (OS). The intraocular pressures were 12 and 16 mmHg in OD and OS respectively. Anterior segment was unremarkable in both eyes. Fundus of OD revealed VH with peripheral vascular sheathing. The ultrasound B-scan of OD revealed few moderately reflective dot like echoes in vitreous cavity suggestive of VH with incomplete posterior vitreous detachment (Figure 1A). OS had clear media with normal optic disc and retinal vessels (Figure 1B). A panel of investigations including complete hemogram, erythrocyte sedimentation rate with C-reactive protein, random blood sugar levels, serum homocysteine levels, coagulation profile, viral markers (HBsAg, anti-HIV and anti-HCV antibodies), antinuclear antibody (ANA), liver and renal function tests, Venereal Disease Research Laboratory test (VDRL), mantoux test and contrast enhanced Computed Tomography (CECT) chest were all found to be within normal limits.

A presumptive diagnosis of ED was made based upon the normal laboratory studies and an unremarkable systemic evaluation. This initial episode was treated with intravitreal

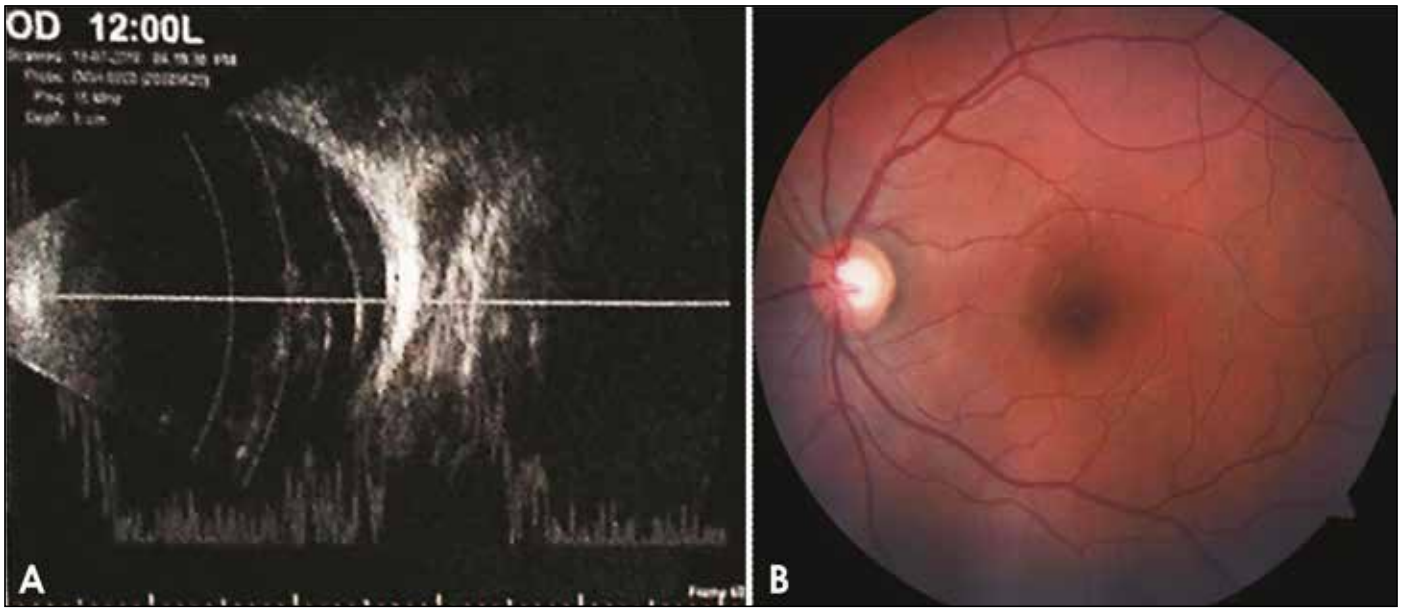

Figure 1: *At presentation (A). Ultrasound B scan (OD) showing moderate vitreous haemorrhage with incomplete posterior vitreous detachment, (B). OS fundus with normal posterior pole.*

bevacizumab (IVB; Avastin®, Roche AG, Basle, Switzerland) 1.25 mg/0.05 ml injected under topical anesthesia in OD after explaining the off-label use, and oral corticosteroid was continued.

After 6-weeks, the BCVA improved to 6/12 in OD and fundus findings revealed a resolving VH. Peripheral limited scatter laser was performed in visible retina and oral corticosteroid was tapered gradually. However, in further follow-ups, he presented with neovascularization of the disc (NVD) and a fibrovascular proliferation extending along supero-temporal arcade in OD (Figure 2A). At this visit, supplemental scatter laser was performed in peripheral retina. Examination of OS was normal (Figure 2B).

Despite laser, he developed recurrent VH in OD with BCVA of 6/36 during follow-up at 15-weeks. Further, fundus of OS revealed multiple subretinal yellowish lesions of variable size (¼th disc diameter or smaller) predominantly in the posterior pole without any associated vitritis, suggestive of choroidal granulomas (Figure 3A). Spectral domain Optical Coherence Tomography (SD-OCT) over these lesions in OS showed focal hyporeflectivity within the choroid without any alteration of overlying retinal pigment epithelium-bruch's membrane (RPE-BM) complex suggestive of partial thickness choroidal granuloma (Figure 3B). In view of recurrent episodes of VH, he was treated with multiple sittings of supplemental laser photocoagulation and two injections of IVB in OD, 4-weeks apart. For choroidal

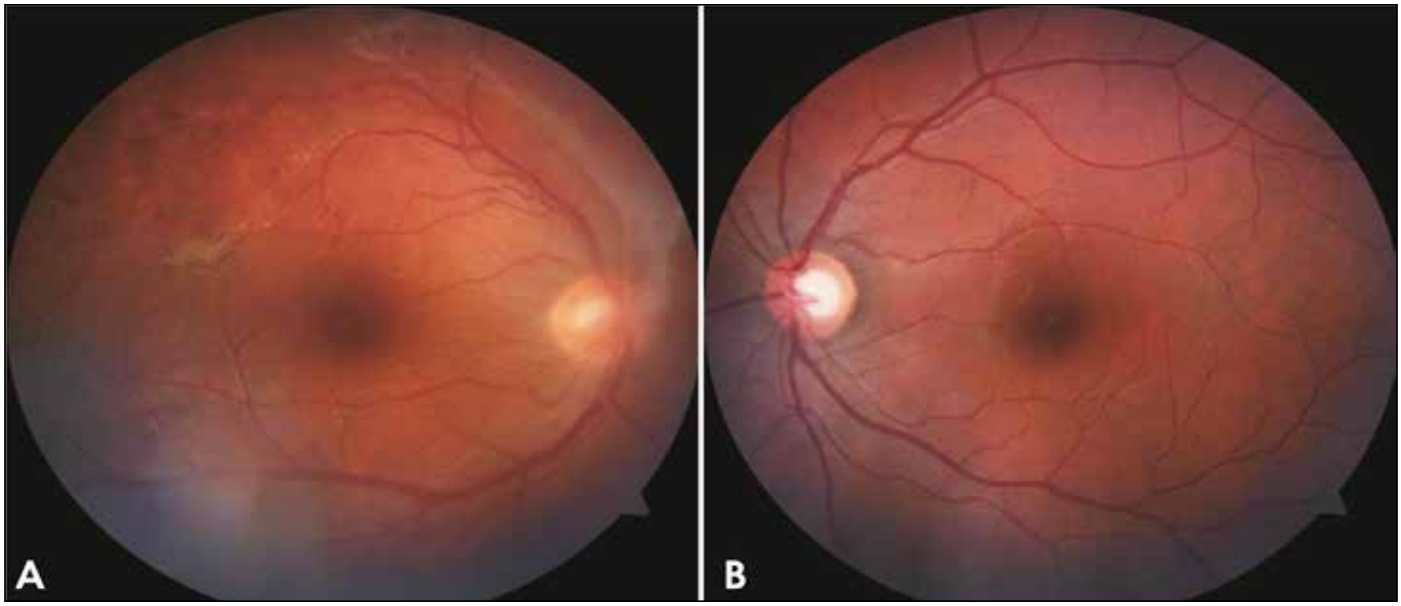

Figure 2: *Follow up at 6-weeks (A). OD showing neovascularization of disc and fibrovascular proliferation extending along supero-temporal arcade and (B). OS within normal limits*

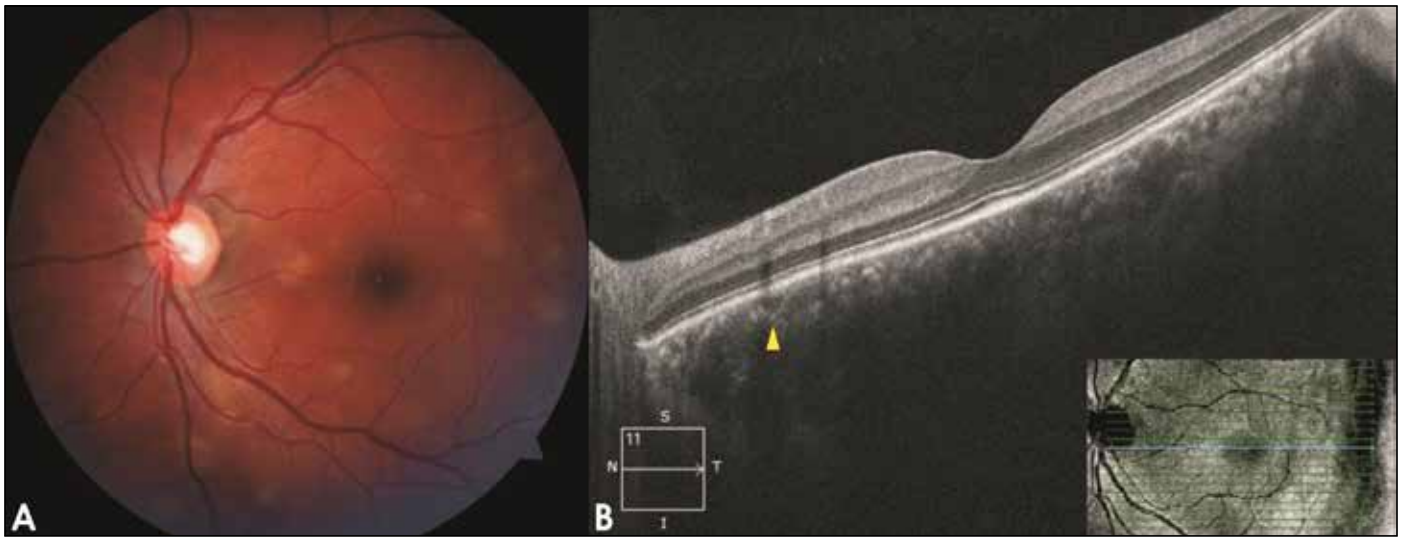

Figure 3: *Follow up at 15 weeks (A). OS showing multiple small yellowish subretinal lesions predominantly in the posterior pole suggestive of choroidal granulomas and (B). SD-OCT line scan passing through one such granuloma showing hyporeflective lesion within the choroid without alteration of RPE-BM complex (yellow arrow).*

granulomas in OS, oral corticosteroid was restarted at the dose of 1mg/kg of body weight. At 28-weeks, however, the visual acuity declined to 2/60 due to another episode of VH in OD (Figure 4a). Choroidal granulomas in OS regressed without any pigmentation or scarring and oral steroids was tapered (Figure 4B). Although advised PPV, he refused surgery at this moment and agreed upon a trial of one more injection of IVB. However, He showed no improvement at 30 weeks' follow-up.

With a diagnosis of retinal periphlebitis with persistent VH despite multiple sittings of laser photocoagulation and IVB, he ultimately underwent 25-gauge PPV with endolaser in OD. Intraoperatively, a fleshy fibrovascular frond was noted from the peripapillary area to superotemporal arcade, which was dissected with the help of cutter. On post-operative follow-up of 3 months, he maintained BCVAy of 6/9 in OD; fundus showed regressed NVD and trimmed fibrovascular fronds. At final follow-up of 6 months, he had a BCVA of 6/6 in both eyes without any reactivation of vasculitis or choroidal granulomas in either eye (Figure 5A and B). He was kept under two-monthly follow up.

Discussion

Eales' Disease (ED) manifests with overlapping stages of venous inflammation (vasculitis), occlusion, and retinal neovascularization. It has a propensity to cause neovascularisation at the junction of perfused and non-perfused retina leading to recurrent episodes of VH.[8]

Abraham et al. in their study evaluated the fundus of 144 eyes with ED, which showed periphlebitis and sheathing of vessels (84%) as the commonest presentation, followed by

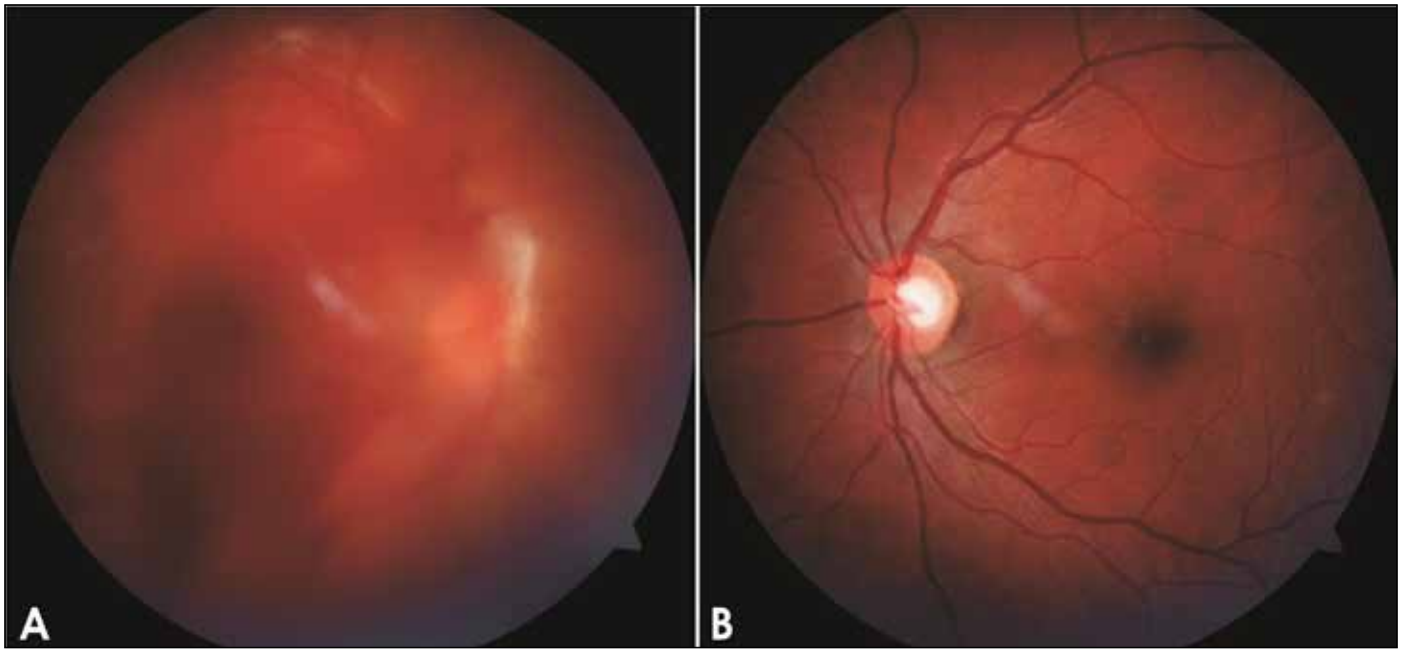

Figure 4: *Follow up at 28 weeks A. OD showing recurrence of vitreous haemorrhage, and B. OS showing regressed choroidal granulomas.*

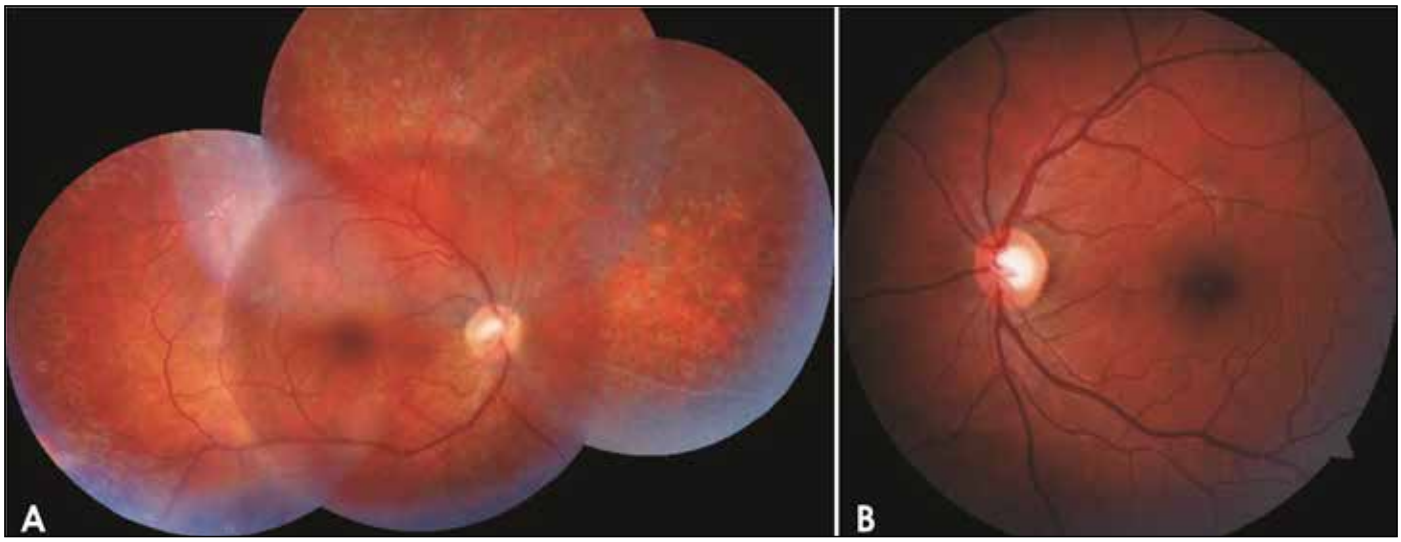

Figure 5: *Post-operatively after 6 months A. OD showing peripheral laser scars and trimmed fibrovascular fronds. The neovascularisation at the disc has regressed. B. OS showing complete regression of choroidal granulomas*

kinky tortuous venules and irregular calibre of vessels (37%), healed chorioretinitis (35%), surface NVE or neovascular vitreous fronds (50%), and VH (34%). Our patient presented with perivascular sheathing accompanied by multiple episodes of VH from neovascular fronds.

Magesan et al. in a case report described a 39-year-old male with a 6-year history of ED and a normal systemic work-up at the initial presentation but on a follow-up visit after 6 years, fundus examination of the right eye revealed a choroidal tuberculoma, which responded well to anti-tubercular treatment (ATT) and oral steroid. Ocular tuberculosis can be associated with Eales disease and often manifest later in the course of the disease.[9] Our patient presented with small choroidal granulomas of unknown aetiology in OS in the course of treatment which responded well to oral corticosteroids. ATT was not added in our patient due to absence of any definitive evidence of intraocular or extraocular tuberculosis.

Several investigators have demonstrated favourable results with laser photocoagulation applied to the non-perfused retina, and to the junction of perfusion and non-perfusion.[10,11] However, Chanana et al. in his retrospective, interventional case series concluded that IVB injections might be effective as an adjunctive or alternative treatment of retinal neovascularization in ED, where inadequate visualization precludes laser photocoagulation.[12] In the present case, laser photocoagulation could not be effectively accomplished in the initial visits because our patient had poor media clarity due to VH. He subsequently received several intravitreal injections of anti-VEGF. However, Patwardhan et al.[13] have shown that repeated IVB in patients with ED with dense VH may not hasten the resolution of VH or reduce the need for vitrectomy. Our patient finally underwent PPV with endolaser in OD, which ultimately resulted in definitive treatment of his condition.

A final visual acuity of 6/6 in the affected eye was achieved in our patient following vitrectomy. As regards to the prognosis of ED, the visual acuity in patients ranges from normal to no-light perception, but most eyes are known to retain good acuity.[14,15] Hence, patients should be counselled to consider vitrectomy early in the course of the disease especially in the setting of persistent VH despite laser and anti-VEGF.

To conclude, multimodal approach and combination modality of treatment in the form of laser/anti-VEGF/vitrectomy may be required for optimal outcome in an individual with recurrent vasculitic VH. Long-term follow-ups are required in such patients to look for any recurrence.

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