Case report

Rapid “epiretinal membrane” development following intravitreal bevacizumab for Coats’ disease

Andrew W. Kama,b, Michelle Hui, Svetlana Cherepanoff, Michelle Huic, Svetlana Cherepanoff Le, Adrian T. Fungf,g,h,*

1. Introduction

Coats’ disease is a sporadic, typically monocular disorder characterised by retinal telangiectasias (Stage 1) which can progress to lipid exudation (Stage 2) and retinal detachment (Stages 3–5) in late disease.1–3 It has a strong male predominance (3:1) and although it may be recognised at any age, most cases present within the first decade of life.2,3

Management depends on disease severity. Laser photocoagulation and/or cryotherapy to the telangiectatic vessels is recommended when lipid exudation threatens the macula. In recent times, anti-vascular endothelial growth factor (VEGF) agents have also been used as an adjuvant to treat Coats’ disease. This is based on the observation that VEGF is known to be markedly elevated in eyes affected by Coats’ disease.1,2 We describe a potential adverse effect - rapid development of “epiretinal membrane” (“ERM”) following a single injection of intravitreal bevacizumab in a patient with Stage 2 juvenile Coats’ disease. Informed consent for this report was provided by the patient and his parents.

2. Case report

A 7-year-old boy with good general health was referred with telangiectatic vessels, aneurysms and intraretinal lipid temporal to his left fovea confirmed on optical coherence tomography scans (OCT, Fig. 1A). The rest of the ocular examination including the right eye was normal and given the male gender, age and clinical findings a diagnosis of Stage 2 Coats’ disease was made. Since he was asymptomatic and the visual acuity was 20/20, the patient and his parents opted for close observation.

The patient was followed twice a year for three and a half years, during which time his disease remained stable. However, at four years he started to notice blurring of his vision which had dropped to 20/30 due to development of cystoid macular edema (Fig. 1B). Fundus fluorescein angiography was attempted but aborted due to difficulty with cannulation. Argon laser photocoagulation was applied to the
temporal aneurysms and affected retina but four months later following this there was still no resolution of the edema and his vision had worsened to 20/60 (Fig. 1C). An OCT raster through the fovea showed trace thickening of the internal limiting membrane/"ERM". A single injection of intravitreal bevacizumab (Avastin; Genentech, San Francisco, CA, 1.25mg/0.05mL) was given under topical and sub-conjunctival anaesthesia.

At the 4-week post-injection follow-up visit the vision had dropped to 20/150 and a dense "ERM" was noted at the macula (Fig. 1D). At 8 weeks post-injection the vision had deteriorated to 20/400 with further progression of the "ERM". The central foveal thickness had increased from 333μm at the time of the bevacizumab injection to 579μm. The patient underwent 25-gauge pars plana vitrectomy with "ERM" and internal limiting membrane (ILM) peeling using trypan blue dye (0.06%). The posterior hyaloid was found to be extremely thickened. Supplemental endolaser was applied to the temporal and inferior aneurysms, telangiectasias and retina. Interestingly, histopathological analysis demonstrated highly folded, paucicellular membrane favouring ILM. Four months post-operatively the macular edema and vision has partially recovered to 20/30. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Fig. 1. Colour fundus photographs (left) and optical coherence tomography scans with corresponding horizontal rasters through the fovea (right) at different time points. A) At baseline there is a patch of circinate hard exudate and mild edema temporal to the fovea with surrounding telangiectasias. Optical coherence tomography demonstrates hyper-reflective hard exudate temporal to, but not involving the fovea. Vision is 20/20. B) Four years after baseline the patient is noticing blurring of his vision and the acuity has declined to 20/30. There is lipid deposition migrating towards the fovea and optical coherence tomography demonstrates cystoid macular edema. Focal argon laser photo-coagulation is applied to telangiectasic vessels temporal to the fovea. C) Four months after focal argon laser photo-coagulation, there has been no improvement in the cystoid macular edema and the vision has declined further to 20/60. A trace thickening of the internal limiting membrane/"epiretinal membrane" is just visible on optical coherence tomography. D) Four weeks following intravitreal bevacizumab injection, there has been marked growth of a dense "epiretinal membrane" over the macula. There is thickening of the retina and loss of the foveal dip on optical coherence tomography scans. Vision is 20/150 and worsened to 20/400 by 8 weeks following intravitreal bevacizumab injection. E) Four months following pars plana vitrectomy and “epiretinal membrane” peeling, there has been marked reduction in the macular edema and vision has partially recovered to 20/30.
vitrectomy and membrane peeling in the setting of Coats’ disease has been reported rarely.\(^8,9\) It was felt that expedient management would likely prevent the permanent macular changes that may be seen if ERMs are left untreated for longer periods.\(^6,7\) This timely intervention is likely to have prevented more severe visual acuity deterioration in our patient.

In summary, we report a case of rapid macular “ERM” development following intravitreal bevacizumab for juvenile Coats’ disease. Caution is advised when considering anti-VEGF agents for the management of this disease.

**Patient consent**

The patient and his parents provided written informed consent for the report.

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**Conflicts of interest**

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**Authorship**

All authors attest that they meet the current ICMJE criteria for Authorship.

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**Appendix A. Supplementary data**

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.ajoc.2018.06.002.

**References**