CASE REPORT

Intravitreal ranibizumab in the management of stage 2B Coats disease in a young adult Malay man

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SUMMARY

This paper describes a rare case of Coats disease with late presentation in a young adult. The condition improved with a combination of focal photocoagulation, cryotherapy and intravitreal ranibizumab.

BACKGROUND

We hope to share a rare case of Coats disease presenting in early adulthood. Although complete resolution seems to be unlikely for this condition, the combination treatment given in this case has resulted in partial resorption of intraretinal and subretinal fluid, leading to improvement in visual function.

CASE PRESENTATION

A 20-year-old man, a Malay engineering student, presented with right painless progressive blurring of vision for 4 years with more acute deterioration noticed over the past 3 months. His vision was 1/60, N36 OD. The anterior segment was normal. Fundus examination revealed telangiectatic vessels inferotemporally and extensive perivascular exudation involving the inferotemporal quadrant and macula region (figures 1 and 2). The left eye was entirely normal with vision of 6/6, N6. He was diagnosed to have Coats disease stage 2B in the right eye.

INVESTIGATIONS

Fundus fluorescein angiography of the right eye showed terminal light bulbs with areas of capillary fallout (figure 3).

Figure 2 Fundus examination showing extensive perivascular exudation involving the macula region suggesting Coats stage 2B.

Optical coherence tomography (OCT) on presentation showed macular oedema with 338 μm central foveal thickness (figure 4).

TREATMENT

He was given intravitreal ranibizumab (IVR) injection 0.5 mg/0.05 mL (Lucentis, Genentech, USA) followed by laser photocoagulation to the areas of light bulb and telangiectasiae 1 week later. One month later, he received cryotherapy to anteriorly located tumours and telangiectasiae.

OUTCOME AND FOLLOW-UP

He was last seen 2 months posttreatment. His vision had improved to 6/60, N24 with evidence of reduction in telangiectatic vessels and resorption of

Figure 1 Fundus examination showing telangiectatic vessels and subretinal exudates in the inferotemporal quadrant of the right eye.

Figure 3 Fundus fluorescein angiography showing terminal light bulbs with areas of capillary fallout.

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Figure 4 Optical coherence tomography on presentation showing macular oedema with 338 μm central foveal thickness.

intraretinal and subretinal exudates clinically and on OCT. OCT posttreatment with IVR showed resolution of macular oedema with 138 μm central fovea thickness (figure 5).

His last follow-up was 2 months posttreatment as he subsequently went abroad for further studies.

DISCUSSION

Majority of Coats disease is diagnosed in the first decade of life, although mild forms can present well into adulthood.^{1 2} The commonest treatment modalities are laser photocoagulation and cryotherapy. However, it has been recognised that in advanced stages of Coats, these therapies have limited success with total retinal detachment, neovascular glaucoma or phthisis bulbi, being a feared sequel.³ The levels of vascular endothelial growth factor (VEGF) have been noted to be raised in Coats disease.⁴ Anti-VEGF agents are believed to work by stabilising the blood-retinal barrier hence reducing the amount of subretinal exudation. This allows laser photocoagulation to be more effective.

This case shows that combination treatment incorporating the newly available anti-VEGF agents such as ranibizumab with cryotherapy and laser photocoagulation is safe and improves visual function. It was likewise reported in a patient with recurrent exudation after scleral buckle placement with subretinal fluid drainage, the exudation and retinal detachment nearly completely resolved, and with normalisation of the intraocular VEGF level.⁴

The evidence for usage of anti-VEGF agents in the management of Coats disease is limited to case reports^{5 6} and case series⁷ because of the rarity of the condition and its occurrence in a predominately paediatric population. Our patient is an example of late presentation in the adult population, which has been documented in the literature.^{5 6} Other treatment modalities include the usage of intravitreal triamcinolone combined

with laser or cryotherapy.⁶ Bevacizumab, which is similar to ranibizumab, has also been used either alone^{8 9} or in combination with triamcinolone. Pegaptanib sodium is another alternative anti-VEGF agent which has been shown in a report to have an effect.⁴ Combined therapy has been shown to work in children, teenagers^{7 10} and adults.⁶ Combined therapy has been used for higher stages of the disease¹⁰ although in general when there is macular involvement, the improvements in visual acuity like that in our patient, are modest at best.

This case report shows that in our local population, multimodality and stepwise approach to the treatment of Coats disease, which incorporates adjunctive pretreatment with an anti-VEGF agent, such as ranibizumab, has favourable results.

Learning points

- ▶ Coats disease may present late, that is, in early adulthood where it has the potential to cause loss of an eye in the productive age
- ▶ Intravitreal anti-vascular endothelial growth factor agents are a safe and useful adjunct in the management of Coats disease stage 2B
- ▶ Intravitreal ranibizumab administered together with laser and cryotherapy (multimodality) for Coats disease results in visual acuity and optical coherence tomography improvement

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Figure 5 Optical coherence tomography posttreatment showing resolution of macular oedema with 138 μm central foveal thickness.

Competing interests None.

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