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Case Report

Idiopathic Frosted Branch Angiitis in Outback Australia

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Abstract

A 40-year old man presented with a two-day history of severe subacute right visual loss and a right superior scotoma. On examination there was right panuveitis, with extensive retinal vasculitis, translucent perivascular exudate, retinal haemorrhage and subretinal exudate. An extensive systemic work-up for an underlying cause was unremarkable. The vitreous biopsy microscopy, culture and PCR for a range of potential causative organisms were all negative. Secondary causes were excluded and a diagnosis of primary idiopathic frosted branch angiitis (FBA) was made. He was commenced on high dose oral prednisolone and gradually improved. His visual acuity remained suboptimal due to residual structural macular damage.

Case Report

A 40-year-old Caucasian tyre-fitter presented with subacute right visual loss over the preceding two days. He described central visual blurring, a superior scotoma, and mild eye redness. His left eye was asymptomatic. Past ocular history was unremarkable. Past medical/social history included intermittent intravenous drug use (IVDU), cigarette smoking and marijuana use. He had multiple tattoos, but no history of immune suppression. He denied any recent illness or exotic travel and systems review was unremarkable. He took no regular medications and had no allergies.

On examination his right best corrected visual acuity (BCVA) was 6/190, and left BCVA was 6/6. Intraocular pressures were within normal range and there was no relative afferent pupillary defect. The right eye revealed mild ciliary injection, fine keratic precipitates, 3-4 + cells in the anterior chamber and 2+ flare. There were 1-2 + anterior vitreous cells present. Right dilated fundus examination showed extensive retinal vasculitis with translucent perivascular exudate (Figure 1). There was haemorrhage involving the posterior pole and inferior arcade, with subfoveal and inferior subretinal exudate (Figure 2). There was no evidence of a retinal vein occlusion in the affected eye. The left eye was unremarkable. A fundus fluorescein angiogram was performed (Figures 3 and 4), as well as serial optical coherence tomographies (OCTs).

Provisional spot diagnosis was that of frosted branch angiitis, either of idiopathic nature or secondary to an infective, inflammatory or infiltrative cause. Infective causes

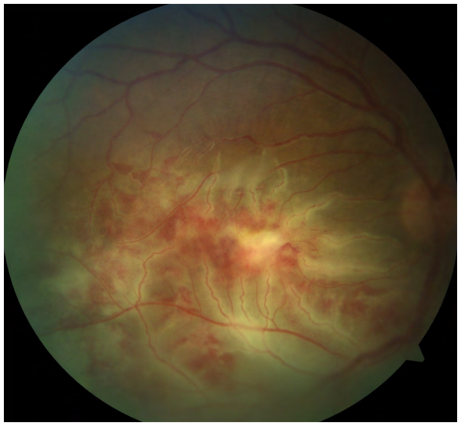

Figure 1: Right posterior pole on initial presentation, showing extensive translucent perivascular exudate, retinal hemorrhage, and central macular exudate.

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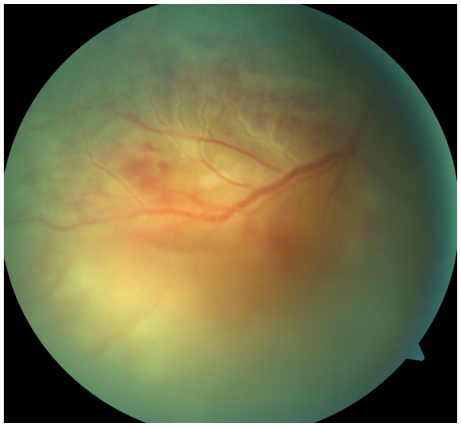

Figure 2: View of the inferior right fundus on initial presentation, outlining the extent of involvement.

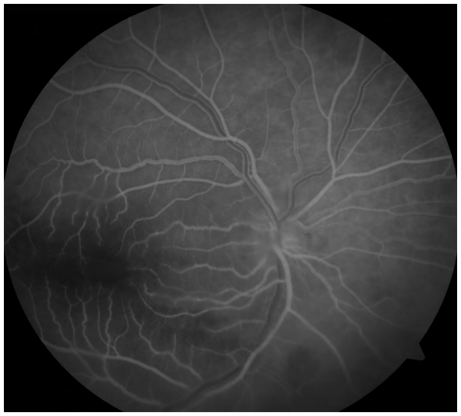

Figure 3: Early arterio-venous phase fundus fluorescein angiogram of the right eye. Only subtle evidence of vascular involvement seen.

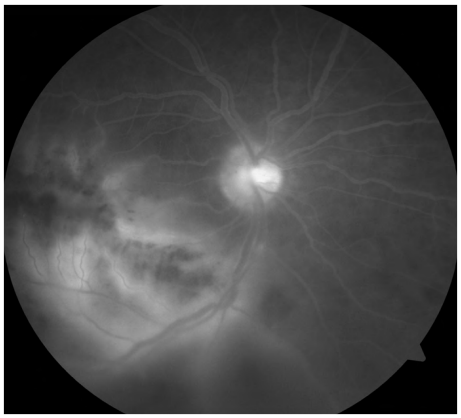

Figure 4: Late phase fluorescein angiogram of the right eye with extensive vascular leakage seen.

of strong consideration included CMV, HSV, VZV, tuberculosis, syphilis and toxoplasmosis, with HIV as a possible underlying condition due to the history of IVDU. Inflammatory/autoimmune causes (such as Behcet's disease, systemic lupus erythematosus, sarcoidosis and inflammatory bowel disease), demyelination (multiple sclerosis) and infiltrative causes were excluded by a negative history, no other suggestive clinical findings and/or negative investigations. Physician review revealed an essentially normal physical examination, including no fever and normal auscultation of heart and lungs. He was commenced on intravenous acyclovir (10mg/kg tds), oral aspirin (150mg daily), topical mydriatic and steroids to the affected eye.

Screening blood tests were unremarkable, including a negative inflammatory and infective screen. Quantiferon TB Gold, serology for syphilis, hepatitis B, hepatitis C and HIV, were all negative. FBC with differentiation, ACE, ANA and anti-DNA were unremarkable. The urine was clear. Chest X-ray, CT brain/chest/abdomen revealed no abnormalities. MRI brain was normal. Transthoracic echocardiogram was normal.

Prompt vitreous biopsy and intravitreal injection of ganciclovir 2mg/0.1ml, voriconazole 0.05mg/0.1ml, vancomycin 1mg/0.1ml and ceftazidime 2.25mg/0.1ml was performed. The vitreous biopsy was sent for PCR of HSV-1, HSV-2, VZV, CMV, Enterovirus and Toxoplasma gondii, which were all negative. Microscopy and culture was negative. Unfortunately he developed an iatrogenic posterior cataract, requiring a lensectomy, and later a secondary IOL.

Once infective, inflammatory and infiltrative causes were excluded, a diagnosis of primary idiopathic frosted branch angiitis (FBA) was made. He was commenced on high dose oral prednisolone (1.5mg/kg), and tapered over 2 months according to a rapid improvement of his retinal appearance. At one month his right BCVA had only improved to 6/48, despite dramatic resolution of inflammation, due to significant macular structural damage (Figures 5 and 6). Right BCVA after secondary IOL (sulcus) was 6/30.

Discussion

FBA is a rare entity describing a condition with distinctive panuveitis and widespread retinal vasculitis with florid translucent perivascular exudate [1]. FBA can be either idiopathic or secondary, with angiographic findings according to aetiology. FBA was first reported in 1976 in a Japanese boy [2], with subsequent reports predominating in the Japanese population. There is a bimodal age distribution-one peak in childhood and a second peak in the third decade. Prevalence favours females over males, and is more commonly bilateral [1]. With an unknown

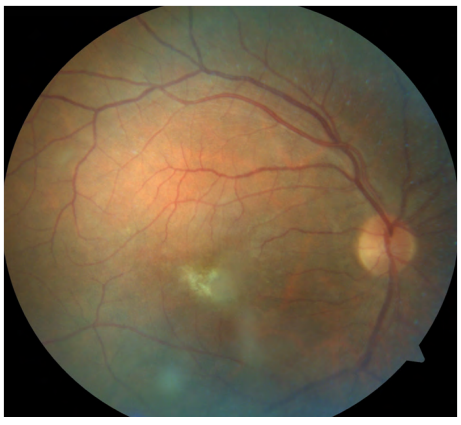

Figure 5: Right eye posterior pole fundus photo three weeks after initial presentation. Residual submacular material seen, but the florid angiitis has settled.

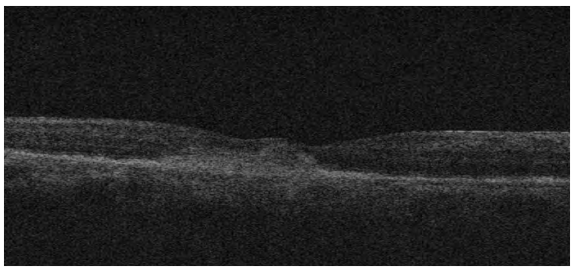

Figure 6: Right macular optical coherence tomography (OCT) three weeks after initial presentation. Significant central submacular hyperreflective material is seen, as well as loss of the outer retinal structure.

aetiology, a hypersensitivity to various infective agents, leading to common pathway of immune-complex deposition has been suggested [1-4]. No prodromal illness was identified in our case.

The majority of reported cases have been treated with systemic steroids, with good visual recovery, unless significant macular involvement is present, such as the case was with our patient. Recently, there has also been a report of recurrent idiopathic FBA treated with an anti-tumour necrosis factor agent, with good effect in a child with significant adverse effects to systemic steroids [5]. Suspected cases of FBA require thorough investigation to exclude secondary causes that may give an FBA-like appearance-including various infective (e.g. CMV, HSV, VZV, rubella, toxoplasmosis), inflammatory/autoimmune (e.g. Behcet's disease, systemic lupus erythematosus, Crohn's disease) and infiltrative causes.

To the best of our knowledge this is the first case of primary idiopathic FBA reported in Australia.

Conclusion

FBA is a rare entity describing a condition with distinctive panuveitis and widespread retinal vasculitis with florid translucent perivascular exudate. Suspected cases of FBA require thorough investigation to exclude secondary causes, including various infective, autoimmune and infiltrative causes. Visual prognosis is usually good, unless there is significant macular involvement, as seen in this case. To the best of our knowledge this is the first case of primary idiopathic FBA reported in Australia.

Acknowledgements

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