Bilateral idiopathic frosted branch angiitis in an older patient

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Background: To report the oldest age to our best knowledge presentation of idiopathic frosted branch angiitis (FBA) in the Asian population and its characteristics.
Methods: Case Report
Results: This is a rare case of bilateral fulminant frosted branch angiitis at older age of presentation and prolonged course of visual recovery with poor visual outcome in one eye. 58-year-old Malaysian female patient, presented with sequential involvement of both eye, initially right eye panuveitis, frosted branch angiitis and subretinal fluid at macula area then the left eye. Bilateral vision was hand movement. The laboratory investigation including autoimmune disease, infectious disease and vitreous sample for viral and tuberculosis PCR were negative. Patient was treated with systemic steroid for a total duration of 6 months. Intravenous Acyclovir was initiated followed by oral Acyclovir. Vitritis reduced, exudative retinal detachment and vasculitis resolved but vision remained poor. Her vision slowly regained after 3 months of treatment and at 6 months her best corrected visual acuity for right eye was 2/60 due to ischemic maculopathy and left eye was 6/9.
Conclusion: Older age groups may present with more severe anterior and posterior inflammation compared to a younger age group, therefore prolonged and timely corticosteroid treatment is crucial for good visual outcome.
Conflicts of interest: The authors report no conflicts of interest.
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Background

Frosted branch angiitis (FBA) is a severe form of vasculitis with characteristic fundus appearance of ‘ frosted branches of a tree ’ due to the infiltration of perivascular space with inflammatory infiltrates. Despite the severe retinal appearance, the prognosis is usually good, with rapid recovering of visual acuity after steroid treatment.

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Case history

58-year-old Malaysian female patient with no known medical illness presented to our eye clinic with right eye sudden blurring of vision for 2 days duration. It was associated with mild eye redness and

discomfort. She gave a history of low grade fever 3 days prior to the blurring of vision. Visual acuity of the right eye and left eye was hand movement and 6/9 respectively. Relative afferent pupillary defect was positive in the right eye. Anterior segment examination revealed anterior chamber cells of 4+ with posterior synechiae and presence of anterior vitreous cells in the right eye. Posterior segment examination showed vitritis, dense perivascular exudates and frosted branches appearance at the periphery (Figure 1).

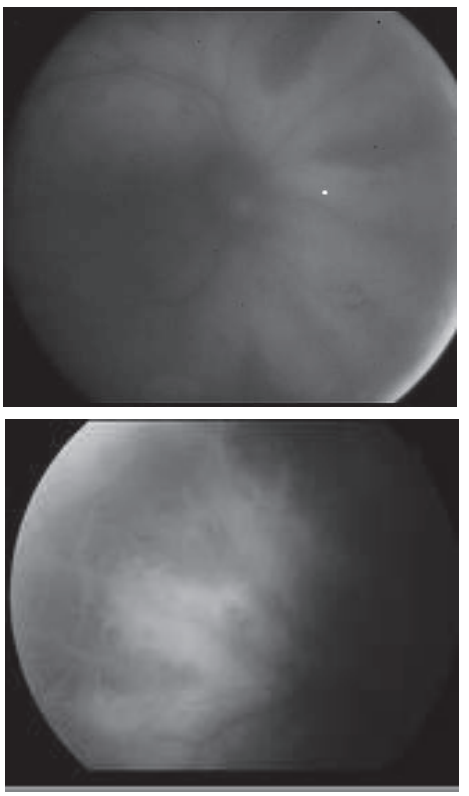

Figure 1: Fundus photos of the right eye

There were scattered small retinal haemorrhages in all 4 quadrants and subretinal fluid at the macula area. Anterior and posterior segment examinations of the left eye were normal (Figure 1). However two days later patient complained of sudden blurring of vision on the left eye with visual acuity dropped to hand movement. Anterior segment examination showed anterior chamber cells of 2+. While posterior segment examination revealed mild vitritis, scattered small retinal haemorrhages, diffuse vascular sheathing with frosted branches appearance at the periphery and subretinal fluid at the macula area. Fundus fluorescein angiography on both eyes showed diffuse leakage from the vessels and discs (Figure 2,3) at late phase with no evidence of vascular occlusion.

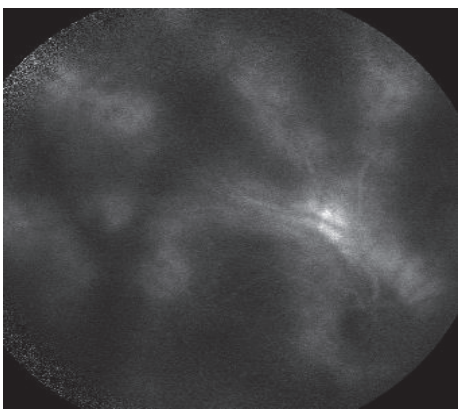

Figure 2: FFA of the right eye showing diffuse vascular leakage at late phase

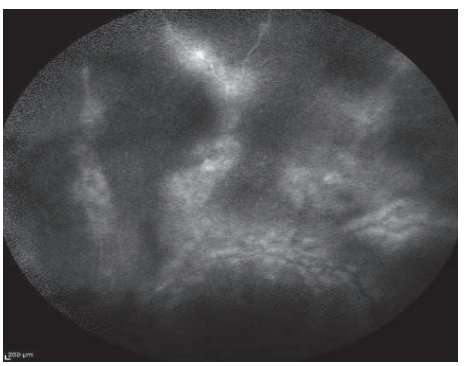

Figure 3: FFA of the left eye showing diffuse vascular leakage at late phase

Vitreous sample was sent for cytomegalovirus, herpes simplex virus, varicella zoster virus and Mycobacterium

tuberculosis polymerase chain reactions (PCR). Blood investigations including autoimmune and infectious disease screening were sent. All PCR results and blood investigations were negative. Masquerade condition was also ruled out with negative findings on systemic examination as well as the tumour markers on blood investigations. Based on the clinical presentations and negative investigations for secondary causes, the diagnosis of bilateral idiopathic FBA was made. It was possibly triggered by viral antigen in view of history of low grade fever prior to presentation. Intravenous methylprednisolone was then initiated promptly, 1 g/day for 3 days and then continued with high dose oral prednisolone 1 mg/kg/day with subsequent tapering dose for 6 months. She was also started on intravenous acyclovir 750 mg three times a day for 2 weeks and completed 6 weeks course of oral acyclovir. With treatment, bilateral eye vitritis, subretinal fluid at macula area and vasculitis resolved. However, both eyes vision remained as hand movement. After 3 months, her left eye vision gradually improved with best corrected visual acuity of 6/9 at 6 month. Unfortunately the right eye best visual acuity at 6 month was only 2/60 due to ischaemic maculopathy.

Discussion

Idiopathic FBA predominantly affects the young and healthy patient with female preponderance. It has a bimodal age distribution with one peak in childhood and a second in the third decade. Walker et al [1] reported that, the age of presentation range from 2 to 42 years old. In 2012, the youngest case of FBA at the age of 11 months old had been reported.[2] While the oldest patient reported with FBA was 80 years old from Australia and was associated with infective endocarditis.[3] Another 2 cases reported from Japan with age presentation of 62 and 69 years old. Both were associated with aseptic meningitis and acute chorioretinal insufficiency respectively.[4,5] From the literature review, our patient is the oldest reported case of idiopathic FBA without other ocular or systemic association. In idiopathic FBA, the cause is unknown but suspected to be viral.[6] However the onset of FBA after prodromal illness in 33% of the cases suggest possible hypersensitivity reaction to various infective agents with immune complex deposition.[1] Secondary causes of retinal vasculitis such as multiple sclerosis, acute retinal necrosis, cytomegalovirus, herpes zoster, herpes simplex, HIV and adenovirus infections, pars planitis, Eales disease, syphilis, tuberculosis, and sarcoidosis should be ruled out.[7] In older age group, we need to consider masquerade signs secondary to intraocular lymphoma or leukemia with retinal infiltration.[8] Other than the characteristic fundus of frosted branches of a tree, intraretinal edema, intraretinal hemorrhages, papillitis, vitritis, and iritis can be present. Veins are more affected than arteries. Older patients tend to present with severe anterior and posterior inflammation compare to younger age group.[3,4,5] FFA will demonstrate normal venous flow and delayed filling of arteries in the early phase, then leakage from vessels (veins more than arteries) in the late phase without vascular occlusion or stasis.[11] Visual field test may reveal constriction of visual field or central scotoma secondary to macular edema. Electroretinogram, electrooculogram and visual evoked potential may show reduced amplitudes due to reduced function of the retina and optic nerve.[1] FBA usually responds well to systemic corticosteroid therapy with good and rapid visual recovery.[1,7] Intravitreal and posterior subtenon injection of triamcinolone had been described with success.[2,11] Due to postulated possible viral etiology, acyclovir has been used with unknown effect.[1] In

the more recent report, Adalimumab had been used with good response.[9] Walker et al[1] reported 3 cases without treatment, yet have an excellent visual outcome. In another recent case report, a pregnant woman with bilateral idiopathic FBA had spontaneous clinical improvement without treatment and fully resolved postpartum.[10] However in our case, timely treatment with corticosteroid therapy resulted in good vision in the left eye. Unfortunately for the right eye, there was a delay in treatment for few days which led to poor visual outcome secondary to macular ischemia. Apart from that, our patient had longer recovery of visual acuity despite corticosteroid treatment. This is similar to the two reported cases in Japan.[4,5]

Conclusion

In conclusion, we are reporting the oldest age presentation of idiopathic FBA. In the older age group, they can present with severe panuveitis and require a longer recovery period unlike the younger age patients. Hence, a prolonged course of corticosteroid treatment is needed. With the possibility of complication such as macular ischaemia, immediate administration of corticosteroid therapy is advocated. In such cases may consider anti-tumour necrosis factor such Adalimumab which had been reported to have rapid and long lasting effect on visual improvement however more studies needed to support its use in idiopathic FBA.[9]

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