Sympathetic ophthalmia presenting with signs of frosted branch angiitis: Report of a case

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Abstract:
Sympathetic ophthalmia is an ocular inflammatory disease commonly associated with penetrating trauma. We report a case of sympathetic ophthalmia who had Dalen-Fuchs nodules on presentation and developed a frosted branch angiitis (FBA)-like picture which is not a known feature of this disease. A 25-year-old male patient was seen 8 months following penetrating trauma to the left eye. He was treated with oral steroids and immunosuppressives. Fundus fluorescein angiography confirmed the presence of perivascular retinitis; optical coherence tomography showed retinal thickening and ultrasonography showed peripapillary choroidal thickening. Atypical features such as FBA in cases of sympathetic ophthalmia can be present and can indicate an ongoing active inflammation. Early aggressive management is required to prevent visual morbidity.

Keywords:
Dalen-Fuchs nodules, frosted branch angiitis, immunosuppressives, perivascular retinitis, sympathetic ophthalmia

Introduction
We report a case of sympathetic ophthalmia in whom perivascular retinitis and a frosted branch angiitis (FBA)-like picture was found on examination along with Dalen-Fuchs nodules. The typical features of exudative retinal detachment were not present.

Case Report
A 25-year-old Asian Indian male patient was seen by us 8 months following penetrating trauma to his left eye. He had developed defective vision of 4-month duration in the left eye and 3 months in the right eye. He had been treated with topical steroid drops but continued to have diminishing vision. He developed pain and defective vision in the right eye. He had undergone a vitrectomy with endolaser and intraocular foreign body removal in his left eye elsewhere before he was referred to us. He had a history of eruptive Lichen planus several years ago for which he was treated with a course of oral prednisolone. His best-corrected visual acuity was 6/9 and N6 in the right eye and no perception of light and less than N36 in his left eye. Applanation tonometry was 10 mm in the right eye and not recordable in the left eye as it was very soft. Slit-lamp examination of the right eye revealed mutton-fat keratic precipitates and aqueous cells 1+ and aqueous flare 1+. The cornea and pupil were normal. Left eye had pigment on the corneal endothelium, early cataract, and posterior synechiae but no anterior chamber reaction. Fundus examination of the right eye showed 1+ vitreous haze and disc hyperemia with multiple Dalen-Fuchs nodules, perivascular retinitis, and extensive vascular sheathing, and the left eye had no view of the fundus. [Figure 1]. Fundus fluorescein angiography showed a perivasculitis with an FBA-like picture. Optical coherence tomography in the right eye revealed an altered foveal.

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contour, an epiretinal membrane, retinal thickening nasal to the fovea with posterior vitreous detachment, and a bumpy retinal pigment epithelial cell layer. Ultrasonography in the right eye showed peripapillary choroidal thickening of 1.6 mm. Investigations for sarcoidosis and tuberculosis was negative. As the patient required immunomodulators and immunosuppressives such as cyclosporine and azathioprine, respectively, his blood urea, creatinine, blood pressure, and full blood count were done and found to be normal. There was no evidence of systemic infections. We started the patient initially on tablet azathioprine 50 mg 3 times daily and tablet prednisolone 60 mg along with antacid and calcium supplements.

The inflammation continued and tablet cyclosporine, 150 mg twice daily was added to his treatment. He improved significantly, and examination showed no anterior chamber and vitreous cells. The perivasculitis and hyperemia of disc resolved after 4 months of treatment. The vision at the end of 3 months of treatment is now 6/6, N6 in the right eye, and inaccurate projection of light in the left eye. The immunosuppressives are being tapered now, and the patient is being followed up at regular intervals. Follow-up of 1 year showed no reactivation.

Discussion

Sympathetic ophthalmia, a bilateral granulomatous panuveitis which develops as a complication of penetrating ocular injury or surgery and presents with granulomatous diffuse panuveitis. The pathophysiology is believed to be due to an autoimmune hypersensitivity reaction against ocular antigens in the injured eye. The diagnosis is evident from history and clinical examination but can be further substantiated by fundus fluorescein angiography which reveals optic disc hyperemia and dye leakage, hyperfluorescent areas of dye pooling and multiple hyperfluorescent foci of choroidal leakage which correspond to areas of Dalen-Fuchs nodules. Ultrasonography aids in diagnosis as it shows choroidal thickening in posterior pole.

FBA indicates the white perivascular retinal sheathing around blood vessels with lymphocytes and plasma cells and is most frequently seen with cytomegalovirus retinitis. In lymphoproliferative diseases such as lymphoma and leukemia, it is due to malignant cells. Accumulation of inflammatory cells causes FBA in systemic lupus erythematosus, inflammatory bowel disease, toxoplasmosis, and acquired immunodeficiency syndrome. A report of vasculitis resembling FBA in association with HIV has been reported in a child. Acute idiopathic FBA due to a hypersensitivity reaction toward a variety of infectious agents has been reported in young healthy people who respond well to oral corticosteroids. Toxoplasma retinochoroiditis has also been reported to be associated with FBA and is seen as a common diffuse sheathing of arteries and veins with submacular exudation. Exudative retinal detachment or pigmentation an evidence of resolved detachment which is a frequent feature of sympathetic ophthalmia was not seen in this patient. Apart from trauma, retinal surgery, cataract surgery, pars plana vitrectomy, enucleation, evisceration, cryotherapy, and irradiation have been implicated as other causes. Other reports of atypical presentations of sympathetic ophthalmitis following 23-gauge vitrectomy have been reported. Steroids are the mainstay of treatment and are combined with immunosuppressives such as azathioprine. Lesser success has been noted with methotrexate and immunomodulatory drugs such as chlorambucil, infliximab, and etanercept. In long-standing refractory cases, mycophenolate mofetil, the dose of 1 g twice daily has been found to work well in combination with steroids. As our case had persistent inflammation with FBA, we added cyclosporine to azathioprine. This case is reported for the development of FBA-like features which has not been seen in association with sympathetic ophthalmia in the past. The fact that perivasculitis can be a feature of sympathetic ophthalmia even in the absence of more typical signs such severe granulomatous anterior uveitis and exudative detachment gives an insight of varying clinical presentations even in of posterior uveitis that follows trauma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.
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Conflicts of interest
There are no conflicts of interest.

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