Case report

A case of frosted branch angiitis in pregnancy: an unusual presentation

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Abstract

Introduction: Frosted branch angiitis usually occurs in children and presents with severe sheathing of all retinal vessels mimicking the appearance of frosted branches of a tree. Objective: To report a case of frosted branch angiitis in a pregnant lady. Case: We, hereby, report a case of a 24-year-old pregnant female who presented to us with the complaints of sudden diminution of vision in the right eye for three days. Fundus examination revealed severe sheathing of all retinal vessels with neurosensory retinal detachment suggestive of frosted branch angiitis. The case was treated with posterior sub-tenon injection of triamcinolone. Conclusion: Similar to the good prognosis reported earlier in most patients with frosted branch angiitis, the outcome in our patient was satisfactory.

Introduction

Frosted branch angiitis is an idiopathic disorder characterized by bilateral (infrequently unilateral) retinal vasculitis usually in young individuals. (Nakai et al, 1992; Harigai, 2002; Sugin, 1991). It can be associated with ocular and systemic disorders such as cytomegalovirus retinitis, toxoplasma retinochoroiditis, systemic lupus erythematosus and acute lymphoblastic leukemia. The vasculitis characteristically extends from the posterior pole to the periphery with uninterrupted severe sheathing of all vessels. Additional fundus findings may include intra-retinal hemorrhage, punctate hard exudates and serous exudative retinal detachment (Geier et al, 1992; Seoet al, 1998; Arnold et al, 1984). The treatment options available for the condition are variable. We describe a case of a 24-year-old pregnant female with frost branch angiitis who was treated with a posterior sub-tenon injection of triamcinolone and termination of pregnancy.

Case report

A 24-year-old pregnant female in the 3rd trimester with the period of gestation of 35 weeks was referred to us for ophthalmic examination in view of sudden diminution of vision in right eye for the past three days. This diminution of vision was not associated with pain, redness or watering. On ocular examination, her vision was hand movements close to face (HMCF) with accurate light projection and 20/20 in OD and OS respectively. Relative afferent pupillay defect (RAPD) was evident in the right eye. On slit-lamp examination, the anterior chamber of right eye showed cells 4+, flare 2+ and retrolental cells. On non-contact tonometry, intraocular pressure was 14 in OD and 12 in OS. Fundus examination of OD revealed optic disc edema with extensive sheathing of both the arteries and veins extending from the posterior pole to the periphery with scattered multiple hard exudates and hemorrhages (Figure 1a). Ocular examination findings of OS were within normal limits. A diagnosis of OD frosted branch angiitis.
was made. Ocular coherence tomogram (OCT) of the right eye revealed neurosensory retinal detachment and was normal in the left eye (Figure 1b). Fluorescein angiography was not performed. General and systemic examination findings were within normal limits except for a blood pressure of 146/100 mm Hg. Systemic investigation performed for HIV, HBS antigen was negative. The ultrasound of the abdomen for obstetric parameters was within normal limits. Laboratory studies including blood cell count, hemoglobin, plasma proteins, urea, and electrolytes revealed no obvious abnormalities except for an iron deficiency anemia. Urine routine, however, was associated with 2+ proteinuria. Examination for TORCH, VDRL and serology for herpetic infections were within normal limits. Labour was induced in view of pre-eclampsia. She gave birth to a healthy baby per vaginally. During the follow-up on the second post-partum day, her visual acuity in OD had not improved although the findings of decreased anterior chamber cells and flare and regressing hemorrhages were present. She was prescribed posterior sub-tenon triamcinolone acetate 0.5 cc (20 μg) in the right eye. During the one-week follow-up, her visual acuity was 20/65 in the right eye, with regressing hemorrhages and inflammation in the fundus and resolution of the neurosensory detachment (Fig 2a and 2b). Her visual acuity even after 12 months of delivery remains the same.

**Legends**

Fig 1a: Fundus photograph at the time of the initial visit revealed optic disc edema with extensive sheathing of both the arteries and veins extending from the posterior pole to the periphery with scattered multiple hard exudates and hemorrhages;

Fig 1b: Ocular coherence tomogram of the macula revealed neurosensory detachment with subretinal fluid accumulation.

Fig 2a: Fundus photograph revealing regressing hemorrhages and inflammation in the fundus.

Fig 2b: Ocular coherence tomogram of right eye revealing resolved neurosensory detachment after treatment.

**Discussion**

Recently, Kleiner classified patients with frosted branch angiitis into three subgroups. The first group consists of patients with lymphoma and leukemia whose disease is due to infiltration of malignant cells. The second group includes patients with associated viral infection or autoimmune disease. The third group is composed of otherwise healthy young patients, with the lack of systemic complications and is referred to as having “acute idiopathic frosted branch angiitis.” (Seo et al, 1998; Arnold et al, 1984).

All possible causes of FBA were evaluated in the patient; however, serology was negative for all infectious pathologies and the counts and peripheral smears were normal. Thus we attributed the clinical presentation to pregnancy. It is however difficult to place this case in the above-described classification as pregnancy is associated with various haemodynamic and hormonal changes. Determining the definite etiology in this case is difficult; however, it may be secondary to pre-eclampsia which is associated with increased oxidative stress, maternal endothelial (lining of blood vessels) dysfunction, and elevated systemic inflammation. (Steegers EA et al, 2010; Al-Jameil, 2013). On reviewing the literature, we
found only a single case of frost branch angiitis in pregnancy; however, it was seen in a 18-year-old female and had bilateral presentation (Sekeroglu HT et al 2012).

Since the patient was pregnant, fluorescein angiography was not performed, which generally shows normal venous flow and delayed filling of arteries in the early phases, while in the late stages, the leakage from vessels (veins more than arteries), perivenular leakage and optic disc hyperfluorescence are characteristic with no signs of vascular occlusion or stasis. Moreover, these possible general findings would not have made any change in our management.

Despite the severe retinal appearance, the prognosis is usually good in cases of acute idiopathic FBA, with rapid recovery of visual acuity after prompt steroid treatment (Geier et al, 1992; Seo et al, 1998; Arnold et al, 1984). Generally, steroids in oral form are recommended - although their role is debated, in the management of FBA. We treated the patient with induction of labour as we thought that pre-eclampsia might be the cause and fetal maturity had already been reached. Additionally, local posterior sub-tenon injection of triamcinolone acetate was preferred over oral therapy since she was lactating and the disease was unilateral.

Conclusion
Frosted branch angiitis in pregnancy can be treated with posterior sub-tenon triamcinolone and termination of pregnancy.

References


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