Bilateral Retinal Arteritis with Multiple Aneurysmal Dilatations: A Case Report

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Abstract

We report a rare case of bilateral retinal arteritis with multiple aneurysmal dilatations of a thirteen year old female presenting to ophthalmology clinic at King Hussein Medical Center on April 2001 complaining from bilateral painless drop of vision of two weeks duration. Visual acuity was counting fingers up to two meters in the right eye and 6/36 in the left eye. Posterior segment examination revealed bilateral vitreous cells, retinal arteritis and macular edema. Fluorescein angiography showed fusiform aneurysmal dilatation in the main arterial branches and late segmentary staining of the walls; there were neither ischemia, nor venous involvement.

Other systems were normal. All laboratory and radiology investigations were within normal. The patient was treated by oral steroids and immunosuppressive agents. Her final vision stabilized at 6/18 and 6/12 in the right and left eye, respectively; fundus examination showed bilateral macular scars with no active ocular inflammation.

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Introduction

Bilateral retinal arteritis with multiple aneurysmal dilatations was first described in 1983 by Kinacid and Shatz. [1] This disorder occurs in otherwise healthy young patients. Its etiology remains obscure although reported in patients with polyarteritis nodosa, [2- 3] rheumatoid arthritis, [4] and bronchial asthma. [2- 3]

There is controversy as to whether this may be to congenital weakness of focal points in the retinal arteriolar wall, or whether there is some insult to the arteriolar walls, such as that caused by an arteritis-vasculitis, resulting in leakage of lipoproteins with deposition of exudates in the retina leading to non perfusion and neovascularization. [5]

Controversy also exists in the management of these patients. Routes of management include observation, steroids, immunosuppressive agents, laser treatment and surgery.

We report a rare case of bilateral retinal arteritis with multiple aneurysmal dilatations that was treated with oral steroids and immunosuppressive agents.

Case Report

A thirteen year old female not known to have any medical illness presented to ophthalmology clinic at King Hussein Medical Center on April 2001 complaining from bilateral painless drop of vision of a two week duration.

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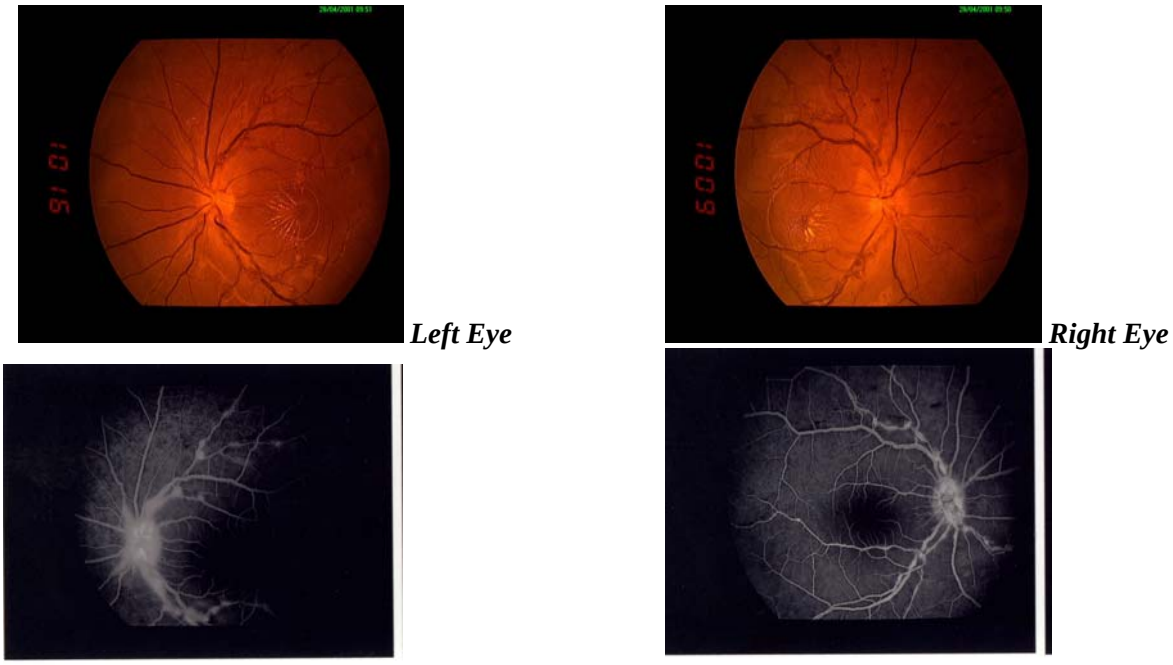
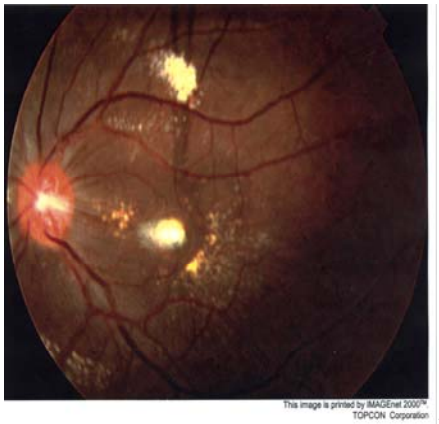
There was no history of previous similar attacks. Visual acuity was counting fingers up to two meters in the right eye and 6/36 in the left eye. Anterior segment examination and intraocular pressure were normal in both eyes. Posterior segment examination revealed bilateral vitreous cells, retinal arteritis and macular exudation and edema. Fluorescein angiography showed fusiform aneurysmal dilatation in the main arterial branches and late segmentary staining of the walls; there were neither ischemia, nor venous involvement (Figure 1). Pediatric consultation confirmed that the patient had only localized ocular disease and that the other systems were normal. Investigations done for the patient included: complete blood count, erythrocyte sedimentation rate, routine chemistry, rheumatic factor, antinuclear antibodies, double stranded DNA, antineutrophil cytoplasmic antibodies, C- reactive protein, complement level (C3 and C4), chest x-ray, chest CT scan, and brucella, toxoplasmosis, and borrelia titers. All results were normal.

The patient was treated with oral steroids (40 mg daily) showing improvement in her vision (visual acuity 6/60 in the right eye and 6/24 in the left). The prednisolone was tapered over a three month period to 10 mg daily. An immunosuppressive agent (azathioprine 50 mg daily) was added after three months of receiving steroids as the patient had no further improvement. Her visual acuity improved further to 6/24 and 6/12 in the right and left eye, respectively with no gross leakage on fluorescein angiography. Oral steroids and azathioprine were used for a period of one year. Her final vision stabilized at 6/18 in the right eye and 6/12 in the left; her fundus showed bilateral scattered retinal exudates and macular scars (figure 2). The patient is being followed up till now; figure 3 is colored fundus photography taken on May 2005 showing bilateral macular scars with no active ocular inflammation.

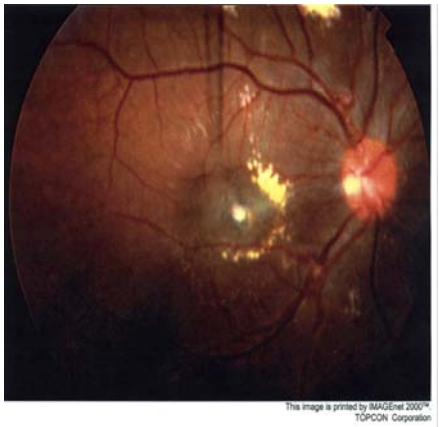


Left Eye ***Right Eye***

Figure 1 Upper: colored fundus photography showing bilateral aneurysmal dilatations in retinal arteries and macular edema, Lower: corresponding fluorescein angiography showing the aneurysmal dilatations and no retinal ischemia.

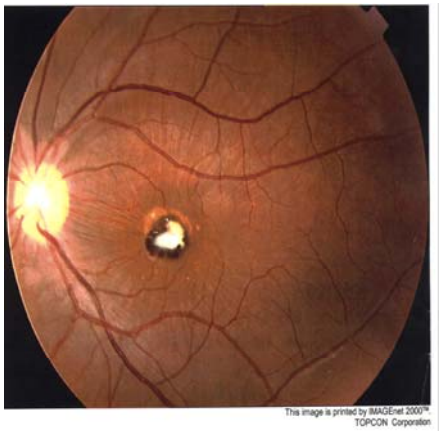


Left Eye

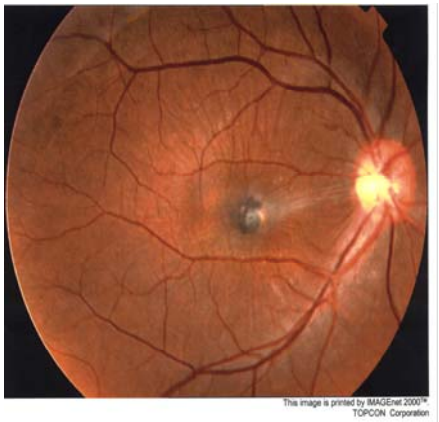


Right Eye

Figure 2: Bilateral macular and retinal exudation.



Left Eye



Right Eye

Figure 3: Bilateral macular scars and no active ocular inflammation.

Discussion

Bilateral retinal arteritis with multiple aneurysmal dilatations can lead to retinal ischemia causing neovascularization, hemorrhage, and subretinal fibrosis if not properly managed. Retinal ischemia and neovascularization leading to vitreous hemorrhage had been previously reported [1, 3, 5-6] Macular involvement had also been reported.[1]

Subretinal fibrovascular proliferation and epiretinal membrane is a sequel of chronic macular edema and serous retinal detachment.[2] Our patient had neither retinal ischemia nor neovascularization but showed bilateral macular involvement. Bronchial asthma was reported to be associated with this condition suggesting Churg-Strauss syndrome (allergic granulomatous angiitis) as a possible etiology.[2-3]

This syndrome is charachterized by segmentary vasculitis involving small blood vessels. Ocular manifestations include scleritis, uveitis, optic neuritis and amaurosis fugax.[7] Bronchial asthma was excluded in our patient on the basis of clinical and radiological investigations. Neuroretinitis is another condition to be considered. Work up includes borrelia, toxoplasma and brucella titers, complements level, antinuclear antibodies and double stranded DNA.[8] All were within normal in our patient.

The few cases reported in the literature showed different management guidelines. Different strategies had been allocated including observation, medical treatment, laser and surgery. Controversies exist on where and when to apply laser treatment. Tomita and his colleagues advised for early retinal photocoagulation to the areas of non-perfusion even without neovascularization.[9] On the other hand, Shatz et al. (1998) suggested laser treatment only in cases of neovascularization or when exudation spreads to the macula. He also showed the opinion of three experts on how to manage such patient: Dr. Gass suggested a walling-off laser treatment temporal to and avoiding exudates, Dr. Brown advised for observation of regression or light laser treatment over aneurysms and Dr. Mieler advised for scatter laser to areas of non-perfusion.[5]

Disappearance of retinal aneurysms has been reported after scatter retinal photocoagulation on the non-perfusion retinal area.[6, 10] Even without treatment, aneurysms may also disappear.[11-12]

Surgery may be needed in patients with non resolving vitreous hemorrhage and subretinal fibrosis. Salvador et al. (1996) described a patient with subretinal fibrosis who did not improve on systemic steroid and immunosuppressive treatment; he needed pars plana vitrectomy, epiretinal membrane dissection, and posterior hyaloid dissection.[2]

Oral steroids are not indicated for management of retinal lesion if not associated with vasculitis.[10, 11, 13]

We did not do laser photocoagulation for our patient as she did not show areas of retinal non-perfusion or neovascularization. Ought to the presence of retinal vasculitis, she improved initially on oral steroids and showed further improvement on immunosuppressive treatment. After a four year period of follow up, her ocular condition remained stable without showing any complication or recurrence.

Our patient was successfully treated and was not complicated by retinal ischemia or neovascularization. We think this is attributed to the early administration of steroids and immunosuppressive agents. Therefore, we recommend early steroid and even immunosuppressive agents administration for the treatment of patients with retinal arteritis and multiple aneurysmal dilatations.

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