Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN) Syndrome

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Case Report: A 27-year-old female presented to the emergency room in July 2011 complaining of generalized headache and progressive decrease of vision in the right eye for few weeks. She was using oral contraceptives since 6 months. Her medical history was free of any systemic diseases. Examination showed right eye visual acuity (VK): 6/60 unaided, IOP 14mmHg, color vision 7/17 by Ishihara pseudocolourachromatic plates, normal anterior segment, optic disc swelling with secondary macular sub-retinal fluid and sub-foveal sensory detachment. The left eye VA 6/6, IOP 16mmHg, color vision 17/17, normal anterior segment, optic disc swelling (Fig.1 & 2).

Patient was admitted to the medical unit with a presumed diagnosis of benign intracranial hypertension (BICH). Lumber puncture (LP) was done with high closing pressure. Patient treated with Acetazolamide and Furosemide. She was followed in neuro-ophthalmology for BICH. In September 2011, patient presented with decreased vision both eyes. Examination showed bilateral neuroretinitis, bilateral vasculitis and development of subfoveal sensory detachment in left eye, in addition to the progressively increasing right macular edema. She was still on oral acetzolamide. Fundus fluorescein angiography (FFA) showed aneurysmal dilatation of disc arteries and vasculitis (Fig.3 & 4). Complete vasculitis work-up was done and did not suggest any abnormality.

Azathioprine was added lately “on February 2013” due to patient non-compliance to the follow-up.

June 2014 patient developed neovascularization for which pan-retinal photoacoagulation (PRP) was done (Fig.6).

The initial diagnosis of BICH changed to IRVAN syndrome and she was started on oral steroids with no marked improvement (Fig 5).

Last examination in 2016 showed VA 6/9 unaided both eyes, and no macular edema. (Fig 7)

Discussion:

**IRVAN syndrome is a rare disease of unknown etiology. It affects young patients with female predominance. It is not associated with systemic disease.**

The oculus manifestation is classified into: **Major criteria:** 1. Retinal vasculitis 2. Neuroretinitis 3. Aneurysmal dilatation of the optic nerve head and retinal arterioles. **Minor criteria:** 1. Peripheral capillary non perfusion 2. Retinal Neovascularization 3. Macular exudation (1, 2). Our patient had all major and minor criteria.

**Staging of the oculus findings is proposed to evaluate disease progression (2). Stage1. Macroaneurysms, exudation, neuroretinitis and retinal vasculitis. Stage2. Capillary non-perfusion with angiographic evidence. Stage3. Posterior segment neovascularization of the disc or elsewhere and/or vitreous hemorrhage. Stage4. Anterior segment neovascularization (rubecous iritis).** Stage5. Neovascular glaucoma. Applying this system to our patient she was stage 3. Visual impairment can result from exudation, maculopathy or retinal ischemia and its sequelae i.e vitreous hemorrhage and neovascular glaucoma.

Treatment of IRVAN is divided into: 1. Medical. 2. Laser photoacoagulation. 3. Surgical. Laser is indicated to treat the ischemia and neovascularization and the surgery, pars plana vitrectomy, is indicated for vitreous hemorrhage (12, 13). Oral steroids is used but there are many reports confirming its lack of efficacy in IRVAN syndrome (14, 15). Azathioprine has been reported to have good effect on vasculitis (16). In our patient the steroid was not effective, however the Azathioprine showed better effect. Intravitreal injection of anti vascular endothelial growth factor Anti-VEGF are also used to treat macular edema in IRVAN syndrome (16, 17). We did not use Anti-VEGF as the macular edema regressed after the use of Azathioprine.

References