

Optical Coherence Tomography Features in Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis Syndrome

Touka Banaee, MD; Seyedeh Maryam Hosseini, MD

Retina Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

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INTRODUCTION

A 15-year-old girl complaining of reduced visual acuity (VA) in both eyes since 2 months before was referred to our retina service in January 2011. Best corrected visual acuity was 5/10 in both eyes. External eye examination was normal. Biomicroscopy revealed +1 anterior chamber cells and +1 old vitreous cells bilaterally. Other anterior segment structures and intraocular pressure (IOP) were normal in both eyes. Fundus examination revealed the presence of circinate exudative maculopathy, optic disc swelling, multiple tied knot-like aneurysmal dilations of peripapillary and optic disc retinal arterioles (macro-aneurysms), and diffuse retinal vasculitis, bilaterally [Figure 1].

Fluorescein angiography disclosed the presence of saccular aneurysms at the bifurcation of arterioles, optic disc leakage, extensive peripheral non-perfusion areas and vessel wall staining of arteries and veins especially at the border of peripheral non-perfusion areas [Figure 2].

Systemic workup including complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), purified protein derivative (PPD), venereal disease research

laboratory (VDRL) titer, angiotensin converting enzyme (ACE) titer, serum calcium and phosphorus, antinuclear antibody (ANA), C-ANCA, and P-ANCA were normal.

Pan-retinal photocoagulation (PRP) was performed for both eyes to prevent neovascular complications of extensive peripheral non-perfusion and the patient was treated with a short course of Ibuprofen 400 mg three times a day for 2 weeks, with no significant change in VA.

Eye-tracked spectral-domain optical coherence tomography (SD-OCT) (Spectralis HRA + OCT; Heidelberg Engineering, Dossenheim, Germany) was performed from the posterior pole of both eyes. Extensive hard exudate deposits, visible as hyper-reflective lesions, were present in the outer retinal layers [Figure 3]. The papillomacular bundles in both eyes were thrown into folds, with epiretinal membrane (ERM) formation over the fovea in the left eye which was persistent after six months on OCT sections [Figure 3a and b]. There was diffuse retinal thickening at the posterior pole, most severe at the level of the outer nuclear and nerve fiber layers (NFL) [Figures 4 and 5]. There were schisis cavities and vertically oval cystoid spaces underneath the inner limiting membrane (ILM) more prominent around the periphery of the posterior pole [Figures 4 and 5]. Multiple tiny hyper-reflective excrescences were visible on the inner surface of the retina in the left eye on OCT, with no correlation with any specific features on fundus examination except for striations in the NFL [Figure 5].

Correspondence to:

Seyedeh Maryam Hosseini, MD. Retina Research Center, Mashhad University of Medical Sciences, Khatam-al-Anbia Eye Hospital, Ghareni Blvd., 91959, Mashhad, Iran.

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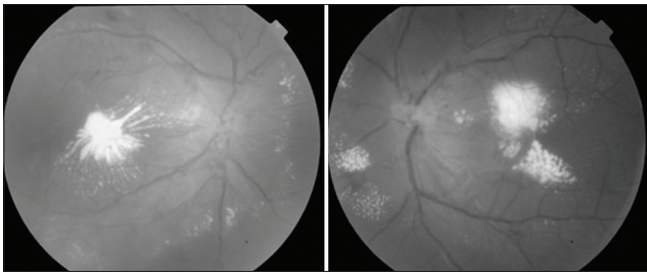


Figure 1. Fundus photography shows circinate exudative maculopathy, optic disc swelling (neuroretinitis) and saccular aneurysms in both eyes.

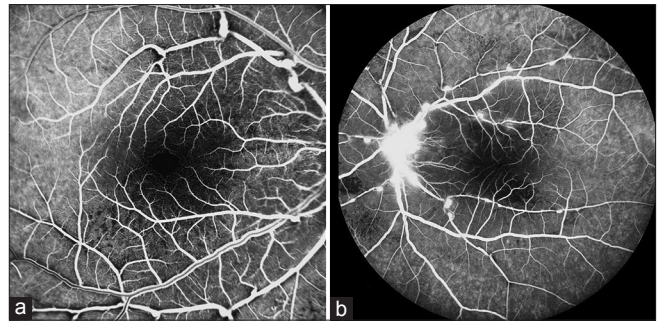


Figure 2. Fluorescein angiography, note multiple tied knot-like aneurysmal dilations of retinal arterioles of the right eye in the early arteriovenous phase. (a) On a later frame from the left eye, leakage from the disc and aneurysms are visible (b).

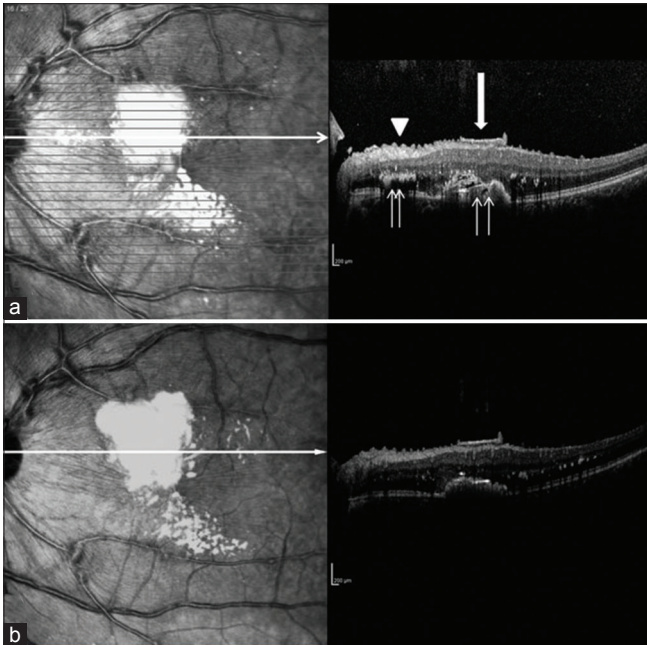


Figure 3. Horizontal cross-sectional spectral-domain optical coherence tomography (SD-OCT) image, the papillomacular bundle of the left eye at first presentation illustrates multiple foldings (arrowhead) on the inner surface of the papillomacular bundle. An epiretinal membrane (ERM) (arrow) and reflectivities in the outer nuclear layer corresponding to hard exudates (double arrows) are also visible (a) Persistence of ERM in the left eye after 6 months (b).

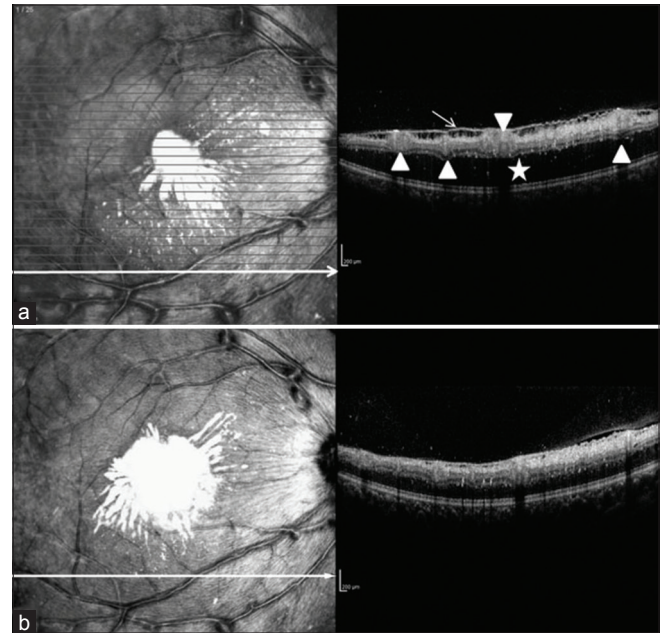


Figure 4. OCT image, the posterior pole periphery of the right eye reveals the presence of schisis cavities and cystoid spaces in the nerve fiber layer (NFL) underneath the internal limiting membrane (ILM, arrow), and severe thickening of the outer nuclear layer (asterisk). The scan goes through multiple sections (arrowheads) of non-aneurysmal parts of vessels (a) The same cross section after 6 months (b).

Sections passing through aneurysms showed oval structures filled with non-homogenous material [Figure 5], which except for size, did not significantly differ from sections of non-aneurysmal vessels [Figure 4].

DISCUSSION

IRVAN syndrome is a rare retinal vascular condition, usually affecting young, healthy women. Major features include multiple tied knot-like aneurysmal dilations of the retinal arteries, exudative retinopathy, diffuse staining of the optic disc, and non-perfusion of peripheral capillaries.^[1,2] No systemic disease has consistently been associated with IRVAN syndrome.^[2]

Vascular dilation and macro-aneurysm formation may be due to an inflammatory process in the walls of retinal arteries, thus inflammatory and infectious diseases namely sarcoidosis, Behcet disease, and collagen vascular diseases must be ruled out.^[1,3] Retinal arterial ectasia similar to macro-aneurysms has been reported in patients with uveitis and sarcoidosis.^[3] Positive P-ANCA test was reported by Soheilian et al. in a man with IRVAN syndrome without any collagen vascular diseases.^[4]

The precise nature of IRVAN syndrome is still ambiguous. If left untreated, it may cause bilateral severe visual loss.^[5]

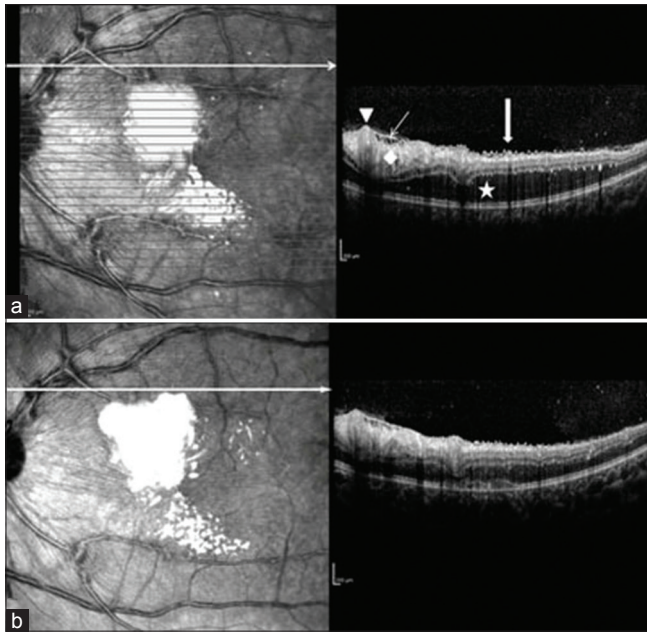


Figure 5. OCT image, the periphery of the posterior pole of the left eye shows increased thickness of the nerve fiber layer (NFL, diamond) and outer nuclear layer (asterisk). There are tiny excrescences on the temporal inner retinal surface (large arrow), not corresponding to any gross ophthalmoscopic feature except for NFL striations. The scan also passes through one of the aneurysms showing oval cross section of the vessel filled with non-homogenous material (arrowhead). Oval cystoid spaces are also visible under the NFL temporal to the aneurysm section (small arrow) (a) OCT after six months at the same retinal area (b).

In this photo essay, we present the OCT features of a case of IRVAN syndrome. To the best of our knowledge, there have been no reports to comprehensively describe SD-OCT features in this disease.

OCT images in the current case, revealed increased thickness of the neurosensory retina especially in the outer nuclear layer. This finding was not out of expectation in the presence of severe exudative retinopathy, which is the result of leakage from macro-aneurysms and the presence of inflammation and neuroretinitis. Increased retinal thickness was also present in all three patients in a series by Naithani et al.^[6]

The most prominent OCT feature of this case was the presence of oval cystoid spaces in NFL, which can be the consequence of severe NFL edema, ischemia or other unknown pathologies in this syndrome. We call them schisis cavities. We have no explanation for the pathologic processes causing these schisis cavities. They may be due to severe intra-retinal edema, an inflammatory process, a degenerative process, or some unknown etiology. They may merely be a cross sectional view of clefts along the arcuate course of the NFL at the periphery of the posterior pole.

Multiple tiny excrescences over the retinal surface in the left eye of our patient were also notable. The excrescences may be remnants of the schisis cavities which have become unroofed due to degeneration or disruption of their inner wall.

Another explanation for tiny excrescences on the retinal surface can be hyper-reflective signals due to precipitation of inflammatory cells, which was ameliorated upon treatment with NSAIDs.

There were some undulations and foldings in the NFL in the region of papillo-macular bundle, which presumably resulted from optic disc swelling, pushing the bundle temporally.

Epiretinal membrane (ERM) was another finding in the left eye of this patient which was also present in the OCTs taken 6 months later [Figures 3a and b].

The presence of ERM and vitreomacular traction in IRVAN syndrome has previously been reported by Naithani et al.^[6] Retinal undulations and folds can be caused by an ERM or a thickened ILM. But it seems unlikely to be the cause of retinal undulations in our case as after treatment with NSAIDs, the undulations and cystic spaces faded in spite of persistence of the ERM and/or the thickened ILM.

This observation also attests to the inflammatory nature of the process.

In the OCT scans, cross section of the aneurysms presented as oval structures filled with non-homogenous material. This view was similar to those of regular vessels except for their larger size.

In summary OCT findings in the present case include severe retinal edema, cystoid spaces in the NFL and excrescences over the retinal surface in the periphery of the posterior pole suggesting some degree of axonal loss and schisis clefts in the NFL.

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Conflicts of Interest

There are no conflicts of interest.

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