Optical Coherence Tomographic Features in Idiopathic Retinitis, Vasculitis, Aneurysms and Neuroretinitis (IRVAN)

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Abstract

The purpose of this was to evaluate the Optical Coherence Tomography (OCT) findings in Idiopathic retinitis, vasculitis, aneurysms and neuroretinitis (IRVAN). In this observational case series, three patients with IRVAN were evaluated. The patients underwent complete ophthalmological examination including best-corrected Snellen visual acuity, slit-lamp biomicroscopy, applanation tonometry, fundus photography, fluorescein angiography, and OCT. The main outcome measures were central macular thickness (CMT) and associated morphological changes in different layers of the retina on OCT. In this series there were 2 male patients and 1 female patient. The OCT tomogram of all the three patients demonstrated diffuse retinal thickening with loss of the normal foveal pit contour. The increased thickness in two of the three eyes was associated with focal areas of hyper reflectivity with high backscattering of the inner retinal layers in the macular area suggestive of macular exudation. In the third patient, OCT images demonstrated vitreomacular traction in addition to focal exudative maculopathy. Epiretinal membrane was also observed on OCT scans of two patients. In conclusion, OCT appears to be a valuable tool in evaluating the macular pathological changes in IRVAN. It is helpful in detecting early sub-clinical findings like vitreomacular traction and epiretinal membrane. The identification of these changes can provide further insight to the cause of visual loss in patients with IRVAN.

Keywords: Neuroretinitis; Optical coherence tomography; Aneurysms; Vasculitis

1. Introduction

Idiopathic retinal vasculitis, aneurysms and neuroretinitis (IRVAN) syndrome is a rare clinical disorder first described by Chang et al (1995). It is characterized by three major criteria (retinal vasculitis, aneurysms at arterial bifurcations and neuroretinitis) and three minor criteria
(peripheral capillary non-perfusion, retinal neovascularization and macular exudation). Recently, a clinical staging system has been proposed for IRVAN syndrome by Samuel et al (2007). Although, IRVAN was initially believed to be a benign self-limiting condition, it is now a known fact that it can lead to severe visual loss. Exudative maculopathy and diffuse retinal ischemia account for the loss of vision in patients with IRVAN syndrome (Chang et al., 1995; Samuel et al., 2007). The present case series attempts to correlate the macular changes seen in IRVAN syndrome and the in-vivo ocular coherence tomography (OCT) observations made on sample patients.

2. Methods

This study adheres to the tenets of the declaration of Helsinki. Three patients with IRVAN syndrome from retina clinic of our institution were included in this study. The diagnosis of IRVAN was confirmed only if all the three major criteria were met. In this cross sectional observational case series, the patients underwent a complete eye examination including best-corrected Snellen visual acuity, slit-lamp biomicroscopy, fundus photography, fluorescein angiography, and optical coherence tomography (Stratus OCT, Carl Zeiss Meditec, Dublin, California, USA). The clinical fundus findings were staged as described by Samuel et al. OCT was performed in all cases using line scan and fast macular scan protocol. OCT analysis of the macula was done to assess the extent of macular involvement and its possible clinical correlation.

3. Results

The study population consisted of one female and two male patients. The age of the patients ranged from 11 to 23 years. All patients in our series had bilateral disease and were systemically normal. There were no signs of anterior segment inflammation in all the three patients. The clinical characteristics of these patients are summarized in table 1.

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age (in years)</th>
<th>Snellen Visual Acuity</th>
<th>Clinical Staging</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Male</td>
<td>12</td>
<td>20/120 20/60</td>
<td>III</td>
</tr>
<tr>
<td>2</td>
<td>Female</td>
<td>11</td>
<td>20/400 20/250</td>
<td>II</td>
</tr>
<tr>
<td>3</td>
<td>Male</td>
<td>23</td>
<td>20/60 20/80</td>
<td>II</td>
</tr>
</tbody>
</table>

4. Optical Coherence Tomography Findings

Optical Coherence Tomography parameters of these patients are summarized in table 2.

Case 1. The OCT tomogram of the patient showed diffuse thickening of the retina with absence of normal foveal pit contour. There was confluence and aggregation of the exudates in the nerve fiber layer of the retina. There were corresponding areas of backscattering shadowing the inner retinal layers. Few non reflective spaces were seen with in the neurosensory retina of the left eye corresponding to cysts. A thin reflection over the neurosensory retina was noted in OCT tomograms.
of left eye suggestive of epiretinal membrane along with faint reflection of the posterior hyaloid (figure 1).

Fig 1. OCT and fundus features of case 1
Case 2. The OCT tomogram of the patient also showed diffuse thickening of the retina with the loss of normal foveal pit contour. The focal areas of high backscattering were suggestive of macular exudation. The OCT images demonstrated that the loss of foveal contour was associated with vitreomacular traction and perifoveal detachment of the posterior hyaloid membrane with foveal adhesion. The vitreomacular traction was clearly evident in the right eye as compared to the left eye (figure 2).

Fig 2. OCT and fundus features of case 2

Case 3. The OCT tomogram of this patient also showed diffuse retinal thickening in both eyes and epiretinal membrane in left eye.
Table 2 OCT Parameters of the patients

<table>
<thead>
<tr>
<th></th>
<th>Central Macular Thickness (in microns)</th>
<th>Epiretinal Membrane (ERM)</th>
<th>Vitreo-macular Traction (VMT)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>OD</td>
<td>OS</td>
<td>OD</td>
</tr>
<tr>
<td>Case-1</td>
<td>633</td>
<td>458</td>
<td>Present</td>
</tr>
<tr>
<td>Case-2</td>
<td>633</td>
<td>589</td>
<td>Absent</td>
</tr>
<tr>
<td>Case-3</td>
<td>325</td>
<td>370</td>
<td>Absent</td>
</tr>
</tbody>
</table>

5. Discussion

In this era, non-invasive OCT has evolved as the primary investigative modality for macular disorders, as it gives vivid cross-sectional description of the retinal layers. However, OCT findings in IRVAN to the best of our knowledge have not been reported earlier.

IRVAN is a rare retinal vascular disorder, the natural history of which is yet to be established. There are previous reports in the literature citing the conflicting course of the condition (Chang et al., 1995; Owens and Gregor, 1992). However, there is uniform consensus among the retinal specialists with regard to the loss of vision in patients with IRVAN. The established causes of vision loss in patients with IRVAN are diffuse retinal ischemia and its neovascular sequelae such as vitreous hemorrhage and neovascular glaucoma and exudative maculopathy (Chang et al., 1995; Samuel et al., 2007; Owens and Gregor, 1992). The present case series attempts to correlate the loss of visual acuity and macular changes seen in initial stages of IRVAN syndrome with the in-vivo ocular coherence tomography (OCT) observations made on the sample patients.

In our study, we consistently noted diffuse thickening of the retina with backscattering shadowing of the inner retinal layers corresponding to the areas of macular exudation on color fundus photograph in all the patients. This observation suggests that macular exudation is responsible for initial visual deterioration in patients with IRVAN, which is in accordance with the findings of Samuel et al.

Due to the usually significant macular exudation, subtle macular changes are poorly discernable on clinical examination in patients with IRVAN. OCT may be more useful in detecting such subtle pathological changes. This is exemplified in the cases reported herein where epiretinal membrane (ERM) was seen on OCT scans of both eyes of case-1 (figure-1) and left eye of case-3 and vitreo-macular traction (VMT) was noted in both eyes of case-2. These observations further broaden the spectrum of macular changes in IRVAN and provide us with insight regarding the contributing factors for vision loss in patients with IRVAN. These OCT findings to the best of our knowledge have not been reported earlier.
In conclusion, OCT features show significant correlation with clinical features of IRVAN but is also useful in detecting other changes such as ERM and VMT. The identification of these changes may help explain the additional contributing factors responsible for decreased visual acuity in patients with IRVAN.

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