OPTICAL COHERENCE TOMOGRAPHY FINDINGS IN NONPROLIFERATIVE GROUP 2A IDIOPATHIC JUXTAFOVEAL RETINAL TELangiectASIS

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Purpose: To determine the optical coherence tomography (OCT) findings in eyes with group 2a idiopathic juxtafoveal retinal telangiectasis (IJRT).

Methods: Forty-one eyes of 22 patients with nonproliferative group 2a IJRT were examined. OCT testing including retinal topographic mapping and analysis, and horizontal and vertical line scans, was obtained on each eye.

Results: None of the 41 eyes had a thickened foveal center. The average center foveal thickness was 166 μm (31–264 μm). Stage 1 eyes (n = 2) were normal fellow eyes in patients with contralateral group 2a IJRT. Stage 2 eyes (n = 11) all had parafoveal temporal graying and intraretinal temporal fluorescein leakage, but rarely had photoreceptor disruption (18%) on OCT testing. Stage 3 eyes (n = 14) all had clinical and fluorescein findings similar to or more pronounced than stage 2 eyes. All stage 3 eyes also had one or more foveal cysts at various retinal depths on OCT. Most of these eyes (86%) had photoreceptor disruption and outer retinal atrophy on OCT. Stage 4 eyes (n = 14) all had a black foveal or parafoveal pigment plaque and intraretinal temporal fluorescein leakage. All stage 4 eyes had a hyper-reflective plaque with shadowing on OCT corresponding to the pigment plaque. Most of these eyes had one or more foveal cysts (64%) and all of these eyes had photoreceptor disruption and outer retinal atrophy.

Conclusion: OCT helps in the staging of group 2a IJRT and reveals multiple retinal structural abnormalities.


Patients with group 2a acquired idiopathic juxtafoveal retinal telangiectasis (IJRT) usually present with mild blurring of vision in one or both eyes in the fifth and sixth decades of life.1–6 They commonly have superficial parafoveal retinal crystalline deposits,7 gray discoloration of the temporal parafoveal retina, and right-angle vessels with no exudation. These patients also can develop parafoveal intraretinal pigment migration similar to that seen in patients with retinitis pigmentosa. Fluorescein angiography in these eyes usually reveals temporal parafoveal telangiectatic vessels and intraretinal fluorescein leakage that spares the foveal center.1 These eyes can develop subretinal neovascularization originating from the juxtafoveal telangiectatic vessels.1–6,8 Gass classified group 2a IJRT into five stages based on slit lamp biomicroscopic and fluorescein angiographic findings.

Optical coherence tomography (OCT) is a noninva-
sive commercially available retinal imaging technique that allows evaluation of retinal structures in vivo. OCT allows delineation of retinal structural abnormalities in patients with group 2a IJRT.

We present OCT findings in 41 eyes of 22 patients with nonproliferative group 2a IJRT. We also incorporated the OCT findings into a staging of group 2a IJRT based on the pre-OCT Gass staging of this disease. We then discuss a plausible pathogenesis for this enigmatic disease.

Patients and Methods

A computer search located all consultation reports from two of the authors (S.M.C. and S.E.P.) from January 2002 to December 2005 containing the words “retinal telangiectasis.” Each candidate’s medical records, film fundus photographs, and film fluorescein angiograms were reviewed, and 26 patients with group 2a bilateral IJRT were identified (Table 1). Twenty-three of these patients underwent a comprehensive eye examination and OCT. OCT testing included retinal topographic mapping and analysis, and horizontal and vertical line scans. Five eyes with subretinal neovascularization were excluded from this study. Forty-one eyes with nonproliferative group 2a IJRT of 22 patients are presented in this report.

Staging of each eye was done using a combination of noncontact slit lamp biomicroscopy, film fundus photographs, film red-free photographs, film fluorescein angiograms, and vertical and horizontal OCT line scans through the fovea (Table 2).

Results

The patients’ average age was 71 years (range 52–89). Twelve of 22 patients were female. Median visual acuity was 20/40 (range 20/20–20/400) (Table 3). Twenty-five of the 41 eyes had visual acuity of 20/40 or better. Based on noncontact slit lamp biomicroscopy, fundus photographs, fluorescein angiograms, and OCT line scans, the 41 eyes were classified into four stages (Table 2).

Stage 1 eyes (n = 2), the normal fellow eyes of patients with more advanced IJRT in the other eye, had normal OCT scans. The average foveal thickness was 194 μm (Table 3).

Stage 2 eyes (n = 11) had a slight graying and loss of transparency of the parafoveal retina (100%), parafoveal telangiectasis, and late temporal fluorescein leakage (100%) (Table 3). Eyes with foveal hyporeflective zones on OCT were excluded from this group. Four (36%) of these eyes also had superficial retinal crystals. The mean foveal thickness in these eyes was 195 μm (156–232 μm). OCT line scans showed photoreceptor disruption and outer retinal atrophy in two eyes (Figure 1, Table 3).

Stage 3 eyes (n = 14) had dilated and blunted right-angle vessels (69%) in addition to graying of the temporal retina (100%), retinal crystals (57%), and telangiectasis seen with slit lamp biomicroscopy and fluorescein angiography (Table 3). Rarely, a circular abnormality was seen in the fovea (29%) that looked like a lamellar macular hole and correlated with the OCT finding of a foveal cyst. Eyes with foveal hyporeflective zones were included in this group. Eyes with a black, superficial, stellate plaque, usually temporal to the fovea, were excluded from this group (Table 2). The mean foveal thickness in these eyes was 210 μm (156–232 μm). Optical coherence tomography showed a foveal cyst in all of these eyes with an average width of 250 μm (50–700 μm) and an average height of 75 μm (50–150 μm). Most of these

<table>
<thead>
<tr>
<th>Stage</th>
<th>Minimum findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Fundus examination, fluorescein angiography, and optical coherence tomography are normal and fellow eye has stage 2–5 group 2a IJRT</td>
</tr>
<tr>
<td>2</td>
<td>Fluorescein angiography shows juxtafoveal leakage sparing the fovea, optical coherence tomography shows no foveal thickening and no foveal cyst</td>
</tr>
<tr>
<td>3</td>
<td>Optical coherence tomography shows foveal cyst</td>
</tr>
<tr>
<td>4</td>
<td>Fundus examination shows black, stellate foveal or juxtafoveal pigment plaque</td>
</tr>
</tbody>
</table>

IJRT = idiopathic juxtafoveal retinal telangiectasis.
Table 3. Findings

<table>
<thead>
<tr>
<th>Findings</th>
<th>All</th>
<th>Stage 1</th>
<th>Stage 2</th>
<th>Stage 3</th>
<th>Stage 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total patients</td>
<td>22</td>
<td>2</td>
<td>8</td>
<td>11</td>
<td>9</td>
</tr>
<tr>
<td>Age, y, mean (range)</td>
<td>69 (49–82)</td>
<td>64 (50–77)</td>
<td>70 (57–77)</td>
<td>71 (50–82)</td>
<td>71 (49–81)</td>
</tr>
<tr>
<td>Diabetes mellitus, n (%)</td>
<td>8 (36)</td>
<td>0 (0)</td>
<td>2 (25)</td>
<td>3 (27)</td>
<td>4 (44)</td>
</tr>
<tr>
<td>Examination</td>
<td>41</td>
<td>2</td>
<td>11</td>
<td>14</td>
<td>14</td>
</tr>
<tr>
<td>Snellen BCVA, median (range)</td>
<td>20/70 (20/20–20/400)</td>
<td>20/25 (20/20–20/30)</td>
<td>20/30 (20/25–20/50)</td>
<td>20/40 (20/25–20/200)</td>
<td>20/60 (20/30–20/400)</td>
</tr>
<tr>
<td>Crystals, n (%)</td>
<td>17 (41)</td>
<td>0 (0)</td>
<td>4 (36)</td>
<td>8 (57)</td>
<td>5 (38)</td>
</tr>
<tr>
<td>Temporal gray, n (%)</td>
<td>33 (80)</td>
<td>0 (0)</td>
<td>11 (100)</td>
<td>14 (100)</td>
<td>8 (57)</td>
</tr>
<tr>
<td>Visible foveal cyst, n (%)</td>
<td>7 (17)</td>
<td>0 (0)</td>
<td>2 (18)</td>
<td>4 (29)</td>
<td>1 (7)</td>
</tr>
<tr>
<td>Pigment plaque, n (%)</td>
<td>14 (34)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>14 (100)</td>
</tr>
<tr>
<td>FA findings</td>
<td>35</td>
<td>2</td>
<td>11</td>
<td>13</td>
<td>9</td>
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<tr>
<td>Right-angle vessels, n (%)</td>
<td>26 (74)</td>
<td>0 (0)</td>
<td>8 (73)</td>
<td>9 (69)</td>
<td>9 (100)</td>
</tr>
<tr>
<td>Telangiectasis, n (%)</td>
<td>28 (80)</td>
<td>0 (0)</td>
<td>8 (73)</td>
<td>11 (85)</td>
<td>9 (100)</td>
</tr>
<tr>
<td>Temporal leakage, n (%)</td>
<td>33 (94)</td>
<td>0 (0)</td>
<td>11 (100)</td>
<td>13 (100)</td>
<td>9 (100)</td>
</tr>
<tr>
<td>OCT findings</td>
<td>41</td>
<td>2</td>
<td>11</td>
<td>14</td>
<td>14</td>
</tr>
<tr>
<td>Foveal thickness, μm, mean (range)</td>
<td>199 (93–286)</td>
<td>194 (191–196)</td>
<td>195 (156–232)</td>
<td>210 (161–286)</td>
<td>191 (93–253)</td>
</tr>
<tr>
<td>Foveola thickness, μm, mean (range)</td>
<td>166 (31–264)</td>
<td>152 (149–155)</td>
<td>166 (119–263)</td>
<td>172 (104–264)</td>
<td>163 (31–236)</td>
</tr>
<tr>
<td>Focal reflective spots, n (%)</td>
<td>9 (22)</td>
<td>0 (0)</td>
<td>5 (45)</td>
<td>4 (29)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>ILM drape, n (%)</td>
<td>17 (41)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>10 (71)</td>
<td>7 (50)</td>
</tr>
<tr>
<td>Foveal cyst, n (%)</td>
<td>23 (56)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>14 (100)</td>
<td>9 (64)</td>
</tr>
<tr>
<td>Cyst diameter, μm, mean (range)</td>
<td>220 (50–700)</td>
<td>250 (50–700)</td>
<td>200 (75–700)</td>
<td>Unreliable</td>
<td></td>
</tr>
<tr>
<td>Cyst thickness, μm, mean (range)</td>
<td>80 (50–150)</td>
<td>75 (50–150)</td>
<td>75 (50–100)</td>
<td>Unreliable</td>
<td></td>
</tr>
<tr>
<td>Photoreceptor disruption, n (%)</td>
<td>28 (68)</td>
<td>0 (0)</td>
<td>2 (18)</td>
<td>12 (86)</td>
<td>14 (100)</td>
</tr>
<tr>
<td>Outer retinal atrophy, n (%)</td>
<td>28 (68)</td>
<td>0 (0)</td>
<td>2 (18)</td>
<td>12 (86)</td>
<td>14 (100)</td>
</tr>
</tbody>
</table>

BCVA = best-corrected visual acuity; FA = fluorescein angiography; OCT = optical coherence tomography; ILM = internal limiting membrane.
eyes had thinning and loss of the normal architecture of the outer retina (86%) (Figure 2, Table 3).

Stage 4 eyes (n = 14) had dilated and blunted right-angle vessels (100%) in addition to graying of the temporal retina (100%), retinal crystals (38%), and telangiectasis seen with slit lamp biomicroscopy and fluorescein angiography. All of these eyes had a black, superficial, stellate plaque, usually temporal to the fovea. The mean foveal thickness in these eyes was 191 μm (93–253 μm). The OCT scans also showed hyper-reflective intraretinal plaques with shadowing that corresponded to the pigment plaque. In addition, all of these eyes had photoreceptor disruption and outer retinal atrophy. Most of these eyes had foveal cysts (64%) with an average width of 200 μm (75–700 μm) and an average height of 75 μm (50–100 μm) (Figure 3, Table 3).

Discussion

The Gass staging of group 2a IJRT predates OCT. We included OCT in our staging of this disease because it reveals the microstructure of the retina in greater detail than can be perceived by even the most astute observer (Table 2). With its 10 μm resolution, OCT can detect microscopic pockets of intraretinal and subretinal fluid. In retinal diseases, like group 2a IJRT, where leakage of fluorescein from retinal vessels is central to the disorder, OCT can be instrumental at delineating subtle pockets of intraretinal and subretinal fluid as well as structural alterations of the retinal layers.

We found intraretinal or subretinal pockets of fluid in 23 of 41 of the eyes in this study using OCT. Only six of these cysts were visible ophthalmoscopically. In one case, a central cyst evident on fundus examination was not present on OCT scanning. This eye had a pseudocyst that appeared at the edge of the circular parafoveal gray zone.

There are several reasons we included these cysts in the staging of nonproliferative group 2a IJRT (Table 2). First, the cysts likely represent an important stage in the disease evolution when there is a buildup of intraretinal or subretinal fluid and surrounding retinal atrophy. Second, these foveal cysts are readily detectable on OCT. Finally, eyes with detectable foveal cysts may respond to new treatments geared toward diseases like cystoid macular edema and central serous retinopathy where abnormal accumulations of fluid in or under the retina cause retinal damage and vision loss.

In their report, which included nine eyes with group 2a IJRT, Paunescu et al propose different names for OCT hyporeflective voids present at different retinal depths. A superficial hyporeflective void is called cystlike and ILM drape, and a deeper hyporeflective void is called a cyst, cystoid, and pseudo-cystoid. In our discussion we use the term cyst for all hyporeflective voids evident on OCT. In Table 3, we do separately report those voids Paunescu et al call ILM drapes. Nevertheless, we also include these ILM drape voids in our overall accounting of foveal cysts. Any eye with a foveal cyst evident on OCT was excluded from stage 1 and 2 disease in this study.

Group 2a IJRT has clinical, fluorescein angio-
graphic, and OCT findings unlike other macular diseases. Although fluorescein angiography demonstrates leaky telangiectatic parafoveal vessels, OCT scanning does not demonstrate parafoveolar retinal cysts or parafoveolar retinal thickening. Although 23 eyes had foveal cysts, none had a thickened fovea. The average central foveal thickness in our 41 eyes was 166 μm (31–264 μm) (Table 3).

The pathogenesis of group 2a IJRT is controversial. Based on fluorescein angiographic findings, Gass initially suggested a primary role of the leaky retinal capillaries with subsequent chronic nutritional damage to Müller cells in eyes with group 2a IJRT. Later, Gass commented that "this disorder (group 2a IJRT) is not primarily a leaky retinal blood vessel disease," but rather "the primary abnormality may reside in one or both of the parafoveolar retinal neural or Müller cells."14

In an attempt to increase the understanding of this disease, Green et al described the electron microscopic findings of one eye with probable group 2a IJRT.15 This clinical pathologic report showed intracellular and intercellular edema primarily in the inner retinal layers. Foveal cysts comparable to those seen on OCT were not described, and the integrity of the Müller cells was not noted. The primary electron

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**Fig. 2.** A, Color fundus photograph of an eye with stage 3 group 2a idiopathic juxtafoveal retinal telangiectasis showing retinal crystals. B, C, Early and late fluorescein angiogram. Early frames show temporal parafoveal telangiectasis with right-angle vessels. Late frames show leakage. D, Optical coherence tomography horizontal line scan of the same eye shows focal hyperreflective spot, deep retinal atrophy, and foveal cysts.

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**Fig. 3.** A, Early fluorescein angiogram of an eye with stage 4 group 2a idiopathic juxtafoveal retinal telangiectasis. Hypofluorescent spot temporal to the fovea is the plaque of retinal pigment epithelial migrations and hyperplasia. Retinal capillaries are telangiectatic and incompetent. B, Optical coherence tomography horizontal line scan shows thinning of the outer retina, occult foveal cyst, and a broad superficial hyper-reflective area (pigment) with shadow.
microscopy finding in this eye was capillary endothelial cell degeneration and regeneration.\textsuperscript{15} Interestingly, the unusual capillary endothelial abnormalities seen in this eye could be secondary to Müller cell dysfunction, since Müller cells in the retina, like astrocytes in the brain, are critical to proper function of the retinal capillary endothelium.\textsuperscript{16–19} A clinical pathologic report by Fine and Brucker of electron microscopy findings in eyes with cystoid macular edema nicely demonstrates the role of Müller cell swelling and necrosis in the pathogenesis of cystoid macular edema.\textsuperscript{20}

The primary locus of group 2a IJRT, Müller cells, retinal neurons, or retinal capillaries, has yet to be determined. OCT offers a new opportunity to try to understand this puzzling retinal disease by correlating structural information gleaned from OCT with functional information gleaned from clinical examination and fluorescein angiography.

We propose that Müller cell dysfunction may explain the clinical, fluorescein angiogram, and OCT findings in patients with IJRT type 2a: retinal crystalline deposits, gray discoloration of the temporal parafoveal retina, parafoveal telangiectasis, retinal atrophy, black stellate superficial retinal plaques, foveal cysts, and subretinal neovascularization. Each of these findings and their possible relationship to Müller cell dysfunction is described below.

The retinal crystalline deposits in some eyes with IJRT, and not in any other retinal vascular disease, are thought to be the footplates of degenerated Müller cells.\textsuperscript{1} The composition of these crystalline deposits is unknown. Crystalline intracytoplasmic inclusions measuring 10 \(\mu\)m to 40 \(\mu\)m in diameter sometimes form in central nervous system astrocytes.\textsuperscript{21–24} These crystalline deposits are called Rosenthal fibers and form under chronic stress conditions.\textsuperscript{21–24}

The gray discoloration of the temporal parafoveal retina may be from ischemia and nutritional damage to the middle layers of the retina. The foveal and parafoveal retina has the densest concentration of photoreceptors in the retina. The high metabolic activity of these photoreceptors requires high levels of oxygen and nutrients. The Müller cells, which help provide nutrition to retinal neurons, are relatively sparse in the parafoveal retina.\textsuperscript{25} A similar gray discoloration of the retina has been reported in an eye recovering from a severe vascular occlusion.\textsuperscript{26} OCT did not reveal any abnormality in the area of gray discoloration. This suggests that the alterations in retinal structure in this gray zone are too subtle to be imaged using the current technology.

The parafoveal telangiectasis and concomitant fluorescein leakage would be expected with breakdown of the capillary endothelial blood–retinal barrier. In addition to their structural role, Müller cells confer barrier properties to the retinal capillary endothelium and help regulate retinal blood flow.\textsuperscript{18,19} Since Müller cells maintain the integrity of the blood–retinal barrier, Müller cell degeneration or dysfunction would be accompanied by a breakdown of the blood–retinal barrier and the observable alterations in the parafoveal retinal capillaries of patients with type 2a IJRT.\textsuperscript{18}

The outer retinal atrophy seen with OCT in these eyes (Figures 2 and 3) could not be caused by retinal vascular abnormalities alone, because the outer retina derives oxygen and nutrients from the choriocapillaris, not the retinal circulation. Müller cell dysfunction, on the other hand, could lead to outer retinal atrophy and degeneration, because Müller cells maintain the health of the surrounding neurons including the outer retinal neurons (photoreceptors).\textsuperscript{19,25,27–29}

The foveal cysts seen on OCT are not specifically mentioned in Gass' description of this disease. With slit lamp biomicroscopy, these areas, when visible, look like lamellar macular holes. They have a distinct, often circular, margin and central retinal thinning. These voids with adjacent retinal atrophy form within the boundaries of the foveal avascular zone (Figures 2–4). The voids were localized in the foveola in all 22 eyes and never extended beyond the edge of the capillary free zone. These intraretinal voids are unlike those seen in patients with macular edema caused by retinal vein occlusion, diabetes, and inflammation because they are not associated with increased retinal thickness.\textsuperscript{12}

These voids probably represent pockets of intraretinal and subretinal fluid. OCT scans distinguish tissue layers based on their optical properties. Light used by the OCT scanner can be transmitted, absorbed, or scattered by retinal tissue.\textsuperscript{12} The intraretinal and subretinal dark zones seen in our patients represent areas where light is transmitted through the tissue with no backscattering. Given the reflective margins around these spaces, they probably represent intraretinal and rarely subretinal accumulations of fluid.

The accumulation of fluid in the fovea of these patients is unexpected and difficult to explain. The breakdown of the blood–retinal barrier occurs in the parafoveal retinal capillaries. Therefore, one would expect a fluid accumulation in the parafoveal inner retina. Yet the OCT scans show pockets of fluid inside the foveal avascular zone that are sometimes deep and sometimes superficial (Figures 2–4). These localized patches of fluid may be caused by pertinacious fluid that leaks from the parafoveal retinal capillaries and migrates to the fovea. Such proteinaceous fluid could accumulate within the foveal avascular zone because
of the lack of a capillary system there to remove them. The accumulation of fluid in the fovea of these patients suggests that the proteins responsible for the foveal intraretinal fluid pocket are not readily eliminated by the retinal pigment epithelium. Selective accumulation of subfoveal fluid is sometimes seen in patients after repair of macula-off retinal detachments and from distant retinal vascular disorders suggesting that the retinal circulation may play a role in the elimination of subretinal fluid in some situations.\(^{30,31}\) We propose that patients with group 2a IJRT may have defective Müller cells that selectively degenerate in the parafoveolar area over time and eventually give rise to all of the abnormalities seen in this disease.

**Key words:** idiopathic juxtafoveal retinal telangiectasis, optical coherence tomography, macula, Müller cells, parafoveolar telangiectasis.

**References**


