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Idiopathic Juxtafoveal Retinal Telangiectasis: New Findings by Ultrahigh-Resolution Optical Coherence Tomography

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Abstract

Objective— To investigate the capabilities of ultrahigh-resolution optical coherence tomography (UHR OCT); to compare with the commercially available OCT standard-resolution system, StratusOCT, for imaging of idiopathic juxtafoveal retinal telangiectasis (IJT); and to demonstrate that UHR OCT provides additional information on disease morphology, pathogenesis, and management.

Design— Retrospective, observational, interventional case series.

Participants— Nineteen eyes of 10 patients diagnosed with IJT in at least one eye.

Method— All patients were imaged with UHR OCT and StratusOCT at the same visit. A subset of patients was also imaged before and after treatment of IJT.

Main Outcome Measures— Ultrahigh- and standard-resolution cross-sectional tomograms of IJT pathology.

Results— Using both standard- and ultrahigh-resolution OCT, we identified the following features of IJT: (1) a lack of correlation between retinal thickening on OCT and leakage on fluorescein angiography, (2) loss and disruption of the photoreceptor layer, (3) cystlike structures in the foveola and within internal retinal layers such as the inner nuclear or ganglion cell layers, (4) a unique internal limiting membrane draping across the foveola related to an underlying loss of tissue, (5) intraretinal neovascularization near the fovea, and (6) central intraretinal deposits and plaques. In 63% of cases, the presence of abnormal vessels and a discontinuity of the photoreceptor layer correlated with visual acuity.

Conclusions— Ultrahigh-resolution OCT improves visualization of the retinal pathology associated with IJT and allows identification of new features associated with it. Some of these features, such as discontinuity of the photoreceptor layer, are revealed only by UHR OCT.

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Coats' disease was first described in 1908 as an idiopathic condition of abnormal telangiectatic retinal vessels associated with intraretinal and massive subretinal exudates. In 1956, Reese defined the term *retinal telangiectasis* as a disease primarily of the retinal capillaries.¹ Idiopathic juxtafoveolar retinal telangiectasis (IJT) was first defined in 1982 by Gass and Oyakawa² as a unilateral or bilateral disease associated with incompetent retinal capillaries only in the perifoveal or juxtafoveal area. The initial classification, staging, and pathogenesis of IJT were based largely on clinical examination and fluorescein angiography.^{2,3} Gass^{2,3} defined the following classification: *group 1A*, unilateral congenital parafoveal telangiectasis with telangiectatic capillaries temporal to the fovea; *group 1B*, unilateral, idiopathic, focal juxtafoveal telangiectasis with a small focal area of incompetent capillaries next to the foveal avascular zone; *group 2A*, bilateral idiopathic acquired parafoveal telangiectasis with retinal thickening temporal to the fovea, right-angle venules, retinal pigment epithelial hyperplastic plaques, subretinal neovascularization, and crystalline deposits; *group 2B*, juvenile occult familial IJT; *group 3A*, occlusive IJT with visual loss due to obliteration of perifoveal capillaries; and *group 3B*, occlusive IJT associated with central nervous system vasculopathy.

In clinical practice, the most common forms of juxtafoveal telangiectasis are the unilateral form, which is usually asymptomatic and found typically in men, usually after the age of 40 (type 1B), and the bilateral form, which is found in both men and women, typically between ages 40 and 60 (type 2A). The most common unilateral form of IJT may feature macular edema that affects central visual acuity (VA), whereas the bilateral form presents with parafoveal hemorrhages, retinal pigment epithelium (RPE) hyperplasia, right-angle venules and occasionally choroidal neovascularization, although vision tends to be relatively normal. The most commonly reported subtype¹⁻⁹ is group 2A from the Gass classification, which is now, along with group 1B as the second most common, referred to as *idiopathic juxtafoveal retinal telangiectasis*. The pathogenesis of IJT is not known.^{1,7} Recently, photodynamic therapy has been considered in patients with severe disease and choroidal neovascularization.^{1,6}

Optical coherence tomography (OCT) is employed for the diagnosis and management of various retinal diseases.¹⁰ The commercially available OCT system StratusOCT (Carl Zeiss Meditec, Inc., Dublin, CA) provides noncontact, real-time, cross-sectional in vivo imaging of the retina with an axial resolution of ~10 μm . Optical coherence tomography imaging has proved to be an important clinical tool for understanding the pathogenesis of different retinal diseases.¹¹⁻¹⁷ Ultrahigh-resolution (UHR) OCT with significantly improved axial image resolution has recently been developed and used to image retinal pathologies.¹⁸⁻²¹ Ultrahigh-resolution OCT enhances the visualization of intraretinal architectural morphology such as the ganglion cell layer (GCL), photoreceptor layer, and RPE.¹⁸⁻²¹ Many of these structures undergo physical changes secondary to IJT, and therefore, UHR OCT may be a powerful tool for elucidating the processes occurring in IJT, which has an unknown pathogenesis. We found only 2 previous reports that used OCT to investigate IJT patients.^{8,9} We found one previous study, using an older-generation OCT system,⁸ describing plaques of retinal pigment hyperplasia as intraretinal hyperelective spots associated with shadowing of the reflections from the tissues below. In the most recent case report,⁹ an IJT patient with celiac sprue disease was imaged with StratusOCT, which revealed bilateral retinal thinning of the fovea.

To evaluate the morphology of IJT and treatment of this disease, comparative imaging with both UHR OCT and StratusOCT was performed on a series of eyes. StratusOCT (~10 μm), with its 4-fold increase in imaging speed or transverse pixel density compared with earlier commercial instruments, provides more detailed cross-sectional information on retinal pathology than previous commercially available ophthalmic diagnostic techniques. The objective of the present study was to identify situations where UHR OCT (~3 μm) provides additional information on morphology, pathogenesis, and management of IJT, and to use UHR OCT as a foundation for better interpretation of standard-resolution OCT images.

Materials and Methods

A prototype clinical UHR OCT system suitable for performing studies in an ophthalmology clinic was developed. A femtosecond titanium:sapphire laser with a ~125-nm bandwidth centered at the wavelength of 815 nm was developed and used as the OCT imaging light source.²² The UHR OCT system achieved axial imaging resolutions of ~3 μm in the eye. The UHR OCT prototype system was integrated on a slit-lamp biomicroscope that was adapted with a charge-coupled device to provide a video image of the fundus. The patient's eye position was established by using either internal or external fixation targets. The UHR OCT image was generated using scans with a 1.5-mm axial depth and 6 mm in the transverse direction, with ~3- μm axial and 15- to 20- μm transverse resolution in tissue (3000 axial and 600 transverse pixels). Pixel spacings were 0.5 μm per pixel in the axial direction and 10 μm per pixel in the transverse direction. The imaging protocol (6 radial macular scans of 6-mm length, at 30° interval angles, centered at the fovea) was the same for both systems to allow a direct comparison of the images.

Optical coherence tomography imaging was performed within the safe retinal exposure limits established by the American National Standards Institute.²³ The American National Standards Institute standard for safe retinal exposure accounts for wavelength, duration, and multiple exposures of the same spot on the retina. For the wavelengths and scanning conditions used in this study, the American National Standards Institute standard for maximum permissible ocular exposure is 1 mW (700-nm center wavelengths) and 1.54 mW (800-nm center wavelengths), assuming 30 consecutive scans in the same spot. In this study, UHR OCT imaging was performed with up to 750 μW of incident optical power in the OCT scanning beam.

The StratusOCT image was generated using standard scans of 2-mm axial depth and 6 mm in the transverse direction, with ~10- μm axial and 20- μm transverse resolution in tissue (1024 axial and 512 transverse pixels). Pixel spacings were 2 μm per pixel in the axial direction and 12 μm per pixel in the transverse direction. The scanning rate of the StratusOCT system is 400 A-scans per second, or about 1.3 seconds per 512-A-scan image.

After scanning of the patient's eye, both the StratusOCT and UHR OCT images were corrected for axial motion using standard reregistration algorithms. These algorithms have been used in all previous prototype and commercial OCT systems.¹²

Imaging was performed using the standard-resolution StratusOCT and the prototype UHR OCT in the ophthalmology clinic of the New England Eye Center at Tufts University School of Medicine. The study was approved by the institutional review board committees of both Tufts–New England Medical Center and Massachusetts Institute of Technology and complies with the Health Insurance Portability and Accountability Act of 1996. After appropriate informed consent, approved by the Tufts–New England Medical Center Institutional Review Board and according to the Declaration of Helsinki, we examined one or both eyes of each of 10 patients. All subjects were voluntarily recruited from the ophthalmology clinic at the New England Eye Center.

The diagnosis of IJT was performed using standard dilated retinal examination and fluorescein angiography. Ultrahigh- and standard-resolution OCT images were obtained from each eye for direct comparison of the images from the 2 instruments. Comparative imaging was performed on 19 eyes of 10 patients (4 male and 6 female) with different classes of IJT (2 patients unilateral and 8 patients bilateral IJT). The mean age of patients in this study was 56 ± 11 years (range, 35–76). Demographics and clinical information of the investigated population are presented in Table 1. Treatment was subsequently carried out in some eyes (2 patients), and further comparative imaging was performed after the treatment. With some eyes (3 patients) a follow-up visit was carried out 2 weeks to 9 months after the first visit.

Results

Standard-resolution and UHR OCT images of a normal macula are presented in Figure 1. The intraretinal layer morphology in the OCT images correlates well with the histological morphology of the retina in the macular region.²⁴ The nerve fiber layer and the plexiform layers are more optically backscattering than the nuclear layers and presented as red, yellow, or bright green false color in the OCT images.^{25–28} In both OCT images, the first highly back-reflecting layer is the nerve fiber layer. The 3 low-backscattering intraretinal layers, presented as blue or black false color in the OCT images, are the GCL, inner nuclear layer (INL), and outer nuclear layer (ONL). The optically backscattering layers, bright green in both images, are the inner plexiform layer and outer plexiform layer. Ultrahigh-resolution OCT can identify fine retinal structures, such as the thin backreflecting external limiting membrane (ELM) just below the ONL, not clearly seen in standard-resolution OCT. The junction between the photoreceptor inner segment and outer segment (IS/OS) is visualized in both images as a first thin highly backreflecting (red) feature in the outer retina, anterior to the RPE and choriocapillaris.^{11,20,21} The RPE is the second highly backscattering layer in the outer retina. A small and highly reflecting region in the inner retina, close to the NFL but not near the foveola, is identified as a normal retinal blood vessel in both standard-resolution and UHR OCT images.

In the OCT images, we found a unique feature of the retina that we call the internal limiting membrane (ILM) drape. This feature represents loss of the outer plexiform layer due to a central cystoid at the base of the fovea, with retina having a normal foveal contour and foveal thickness, no perifoveolar edema, and no cystoids outside the focal area. The ILM drape is seen in the OCT images as a thin ILM layer that crosses above the cystoid space in the focal area of the retina.

All the subjects diagnosed with IJT and included in this study were imaged with both standard-resolution OCT and UHR OCT. The selected IJT cases presented in this study are classified as groups 1B (patient 1) and 2A (patients 2–5) according to Gass.^{2,3}

Selected Case Reports

Patient 1—A 47-year-old man with 20/25 vision in his left eye was diagnosed with unilateral IJT, classified as group 1B upon clinical examination (Fig 2A). Two UHR OCT images obtained at different scan orientations are presented (Fig 2B, C). In both UHR OCT images, the presence of a central cystoid and fluid structure is evident. This structure increases the retinal thickness centrally. Abnormally large intraretinal blood vessels located near the fovea and deep in the ONL, which are characteristics of IJT, can be visualized in the UHR OCT image (Fig 2C). Both UHR OCT images also show additional smaller cystoidic structures located in the INL; however, the ONL seems to be intact. In one UHR OCT image (Fig 2B), a low reflective signal that spans the entire retina is present inside of the central cystoid/fluid structure. This fine retinal spanning structure may be Müller cell bodies that span the separation between the ONL and the outer plexiform layer. It was visualized only with the imaging resolution provided by UHR OCT. In addition, the UHR OCT images showed no involvement of the outer retina (ONL, ELM, IS/OS), which remained intact and juxtaposed to the RPE, which was not clearly discernible in the StratusOCT image (images not shown here) due to its lower resolution. Furthermore, the photoreceptor segments appear intact, which helps explain this patient's good VA (20/25).

Patient 2—A 76-year-old woman with 20/25 vision in her right eye (Fig 3A) and 20/50 vision in her left eye (Fig 3C) was diagnosed with bilateral idiopathic juxtafoveal telangiectasis, classified as group 2A. The UHR OCT macular scans from both the right eye (Fig 3B) and the left eye (Fig 3D) clearly showed foveal thickening due to cystoidic intraretinal structures

bilaterally. In the left eye, very small cystoidic structures present in the INL can be visualized in the UHR OCT image (Fig 3D). Ultrahigh-resolution OCT enhances visualization of the smaller structures in the IS/OS and photoreceptor layer in both eyes (Fig 3B, D). In addition, the ILM is detected as a structure spanning the foveola region of the left eye. This feature, the ILM drape, which may be unique to IJT type 2A, is clearly seen in the UHR OCT image (Fig 3D) but is not clearly seen with StratusOCT. The UHR OCT image enables visualization and identification of the intact ELM throughout the entire retina in both eyes. Ultrahigh-resolution OCT has the ability to identify small retinal features and abnormal changes such as the disruption in the IS/OS near the foveal region of the left eye (Fig 3D). This disruption can be an important indicator of photoreceptor integrity and allows for an explanation of the mildly abnormal VA of the patient's left eye in contrast with the right eye, in which this retinal layer is shown by the UHR OCT image to be intact (Fig 3B). The findings in the UHR OCT images correspond to the VA deterioration in both eyes.

Patient 3—A 54-year-old woman with 20/30 vision in her left eye and 20/25 vision in her right eye (Fig 4A) was diagnosed with bilateral IJT, classified as group 2A. The eyes had very similar presentations; therefore, only the right eye is shown. For comparison, a standard-resolution OCT macular image (Fig 4B) and a UHR OCT one (Fig 4C) with the same scan orientation are shown. In both eyes, the StratusOCT macular scans showed a normal retinal contour and normal neurosensory retinal layers. Ultrahigh-resolution OCT can enhance the visualization of smaller retinal structures such as the photoreceptor layer and the IS/OS integrity. Ultrahigh-resolution OCT was able to detect a slight foveal thinning in both eyes. The improved resolution of the UHR OCT image (Fig 4C) also enables visualization and identification of the ELM, which seems to be intact throughout the entire retina in both eyes. The ELM is not seen clearly in the StratusOCT image (Fig 4B). In the foveola region of the UHR OCT macular scans (Fig 4C), the photoreceptor segments appear to be disrupted with a small cystlike structure, which is not visualized in the standard-resolution OCT image (Fig 4B) obtained in the same location. The UHR OCT image shows a low-backreflecting signal spanning the ONL and connected to the rest of the sensory retina right above the cystoidic structure, which might indicate Müller cells, justifying the normal VA in both eyes of this patient.

Patient 4—A 35-year-old woman with 20/30 vision in her right eye and 20/60 in her left eye was diagnosed with bilateral IJT, classified clinically as group 2A. Upon clinical examination of the left eye (Fig 5A), a yellow deposit is noticed in the central macula. The UHR OCT image (Fig 5B) shows the thickening of the retina, especially in the foveal region, and a highly reflective yellow spot in the ONL underneath the inner retina in the foveola region. This yellow spot is characteristic of type 2A IJT.^{1–3} A small portion of the sensory retina detached from the RPE in the foveola region. The region of photoreceptor detachment is marked in the UHR OCT image by the missing or reduced reflective portion of the ELM signal and the usually highly reflective signal that represents the IS/OS. In addition, the UHR OCT image shows small cystoidic changes visible in the GCL and INL. Due to its lower resolution, the standard-resolution image may be interpreted incorrectly as a complete loss of photoreceptor retinal tissue in the region of the foveola, without the presence of the exudate.

Patient 5—A 61-year-old man with 20/70 vision in his both eyes was diagnosed with bilateral IJT, classified as group 2A (Fig 6A, C). In both eyes, the UHR OCT images showed highly reflective areas indicative of blood vessels with the characteristic shadowing of the OCT signals from the underlying structures (Fig 6B, E). In the right eye, the UHR OCT image showed retinal thinning, with a disruption of the photoreceptor layer in the fovea that is demarcated by the loss of the photoreceptor segment signals in the OCT image (Fig 6B). A small portion of the ILM drape spanning the foveola can also be seen in this OCT image. The left eye's UHR

OCT images (Fig 6D, E) show similar findings—deep intraretinal neovascularization and photoreceptor layer disruption in the foveal region—but the disruption of the photoreceptor layer is to the extent of almost creating a lamellar hole. Both UHR OCT images (Fig 6D, E) also show an ILM drape extending over the fovea, above the lamellar hole. In addition, the UHR OCT image shows cystoidic structures close to the foveal region in the GCL, INL, and ONL of the right eye and in the GCL and INL of the left eye. The high-resolution images allow differentiation of the ELM and IS/OS, which are interrupted in the foveal and perifoveal region in both eyes. In the left eye, a foveal pit detachment is noted in the OCT image (Fig 6D). Disruption of the photoreceptor layer that is present and clearly evident in both eyes probably accounts for the moderate VA loss (20/70).

Patient 6—A 64-year-old woman with vision that is counting fingers from 4 feet in the right eye and 20/60 in the left was diagnosed with bilateral IJT (group 2A). Corresponding to the patient's VA, the fluorescein angiography of the right eye (Fig 7A) suggested more leakage than in the left eye (Fig 7C). In the right eye, the UHR OCT macular scans (Fig 7B) demonstrate a complete loss of the foveal pit with cystoidic spaces in the GCL and INL and disruption of the photoreceptor layer and RPE. In the foveal region, the sensory retina has hyperelective spots in the outer retina that seem to be due to plaques of retinal pigment hyperplasia. Gass initially noted this as a feature of IJT type 2A.^{2,3} The UHR OCT image also indicates that the architectural morphology of the ELM and IS/OS is preserved in the perimacular region but not in the foveal region. In the left eye, the UHR OCT images illustrate disruption of the photoreceptor layer with probable fluid accumulation in the outer retina at the foveola. The fluid accumulation is located between the IS/OS and RPE, with some preservation of the IS/OS and photoreceptor layer. This finding helps to explain the moderately reduced vision in the left eye (20/60) relative to the right eye. The UHR OCT image shows a small ILM drape extending over the fovea. In addition, a thin backscattering ELM layer is present in the UHR OCT image. The photoreceptor inner and outer segments are lifted away from their anatomical position against the RPE, but remain intact. Due to their lower resolution, it is difficult for standard-resolution images to discriminate the photoreceptor segment morphology in this detail.

In our study, 4 common major characteristics seen in UHR OCT were found in a large number of the IJT patients (19 eyes of 10 patients): foveal thinning was found in 58% of the patients, intraretinal cystoids in 63%, the ILM drape in 42%, and photoreceptor disruptions in 84% (Table 1).

Discussion

A comparative imaging study was performed using the UHR OCT and StratusOCT systems on patients with clinically diagnosed IJT. Both OCT systems can noninvasively acquire retinal images that are capable of differentiating most major intraretinal layers, such as the retinal nerve fiber, inner plexiform, inner nuclear, outer plexiform, and outer nuclear, as previously demonstrated on other pathologies.^{9,10,20,21,25,26} In our study, the StratusOCT, at its best resolution, is capable of visualizing IJT characteristic features such as large to medium-size intraretinal cystoids, blood vessels, and foveal deposits, as well as the newly defined feature ILM drape. In addition, UHR OCT is capable of visualizing smaller structures such as the ELM and the IS/OS.^{20,21,26} The ability to visualize the ELM and the IS/OS can be an important indicator of photoreceptor integrity or impairment.^{18,21} The UHR OCT images also reveal morphological changes in IJT such as small cystoidic structures, exudates, neovascularization, and pigment plaques that are not well differentiated with standard-resolution StratusOCT images. The ability to visualize photoreceptor morphology as well as track small structural changes associated with IJT may play a role in further understanding IJT's pathogenesis, which has yet to be elucidated.

The UHR OCT images of IJT pathologies revealed important findings in the pathogenesis of this disease, some of which were not described by previous studies. A summary of demographic information and clinical and UHR OCT findings for all the evaluated patients diagnosed with IJT is presented in Table 1, and the statistics of the main findings (with percentages) are presented in Table 2.

From our evaluated set of 19 eyes of 10 patients clinically diagnosed with IJT, we found the following:

Retinal Thickness Is Variable in This Disease and Does Not Necessarily Correlate with the Degree of Fluorescein Leakage Seen in Fluorescein Angiography (e.g., Patient 6)

Retinal thickness was found to increase (patients 1 and 2) when there was a moderate to large cystoid, when fluid accumulated under the retinal layers (patient 6), and/or when yellow deposits were found in the central retina (patient 4). In previous reports, yellow deposits were noticed in the central fovea in IJT.^{1–4} Table 2 shows that in some cases (16% of eyes) there was no correlation between the retinal thickening and the leakage seen in fluorescein angiography. In other cases (patients 3 and 5), retinal thickness decreased centrally due to a decrease in the photoreceptor layer thickness in the foveal region.

StratusOCT measures the foveal thickness as the average thickness at the intersection point of the 6 linear OCT scans centered on the foveola. Previous studies^{29,30} found foveal retinal thickness values on normal subjects, by StratusOCT, to be between $164 \pm 21 \mu\text{m}$ and $180 \pm 23 \mu\text{m}$. We found that the qualitative findings of retinal thinning or thickening seen in UHR OCT correlate well with the quantitative measurements of foveal thickness from the StratusOCT, with a few exceptions where the StratusOCT scans were not well centered in the fovea and gave falsely higher values. The foveal thicknesses by StratusOCT are presented in Table 1.

The Photoreceptor Layer Is Locally Disrupted in Some Idiopathic Juxtafoveal Retinal Telangiectasis Cases

The photoreceptor layer involvement of this disease is either the thinning (patient 1; patient 2, right eye; patient 5) or disruption (patient 2, left eye; patients 3–6) of this layer (Table 1). In cases of a thin photoreceptor layer a moderate VA loss could be present; however, these patients may (patient 5) or may not (patient 1; patient 2, right eye) experience VA loss. The standard-resolution OCT imaging of IJT with photoreceptor layer involvement may classify IJT findings as normal retinal tissue (patient 3). In some cases of a disrupted photoreceptor layer (patient 2, left eye; patients 4–6), a correlation with VA loss is found in 84% of eyes (Table 2). In addition, the disruption of the photoreceptor layer may also correlate with the leakage found on fluorescein angiography, as in patient 6. The photoreceptor layer disruptions can be of various causes, such as cystoids in the central fovea (patient 3), deposits (patient 4), fluid accumulation, or RPE plaque (patient 6). The origin of the photoreceptor layer disruption seen in patient 6 is still controversial. The disruption is generally associated with choroidal neovascularization, plaques of retinal pigment hyperplasia, or chronic venous stasis. From our study, the origin of this RPE disruption is unclear. Previous studies also described findings such as RPE plaques.^{1–3,8}

In our study, we suspect that the disruption to the photoreceptor layer seen in UHR OCT may be a finding of great importance because it can better guide treatment decisions in the IJT cases. Treatment of fluid accumulation and VA loss in patients affected by IJT with photoreceptor layer disruption would not be expected to result in improvement of the visual outcome. Some disruptions in ONL are also present in some cases, sometimes even with minimal thickness of this layer (patients 1 and 5).

Gass^{2,3} concluded that, in most patients with IJT, VA loss is caused by the intraretinal edema and exudate. In our study, we found that in some cases (patient 1 and 2) there is edema, but the VA is normal in eyes where the photoreceptor layer is intact and moderately decreased in eyes where there is a small disruption of the photoreceptor segments.

A recent study (Spaide RF, Virgili G, Leys A. Morphologic correlations of bilateral idiopathic juxtafoveal retinal telangiectasis and visual acuity. Poster presented at: American Academy of Ophthalmology meeting, November, 2003; Anaheim, California) reported finding an overall morphologic correlation between OCT findings for patients with IJT and their VA changes. Our study shows that the VA loss correlates with the integrity of the photoreceptor segments in 63% of eyes (Table 2).

Cystoids Are Present in Idiopathic Juxtafoveal Retinal Telangiectasis

In our study, 12 of 19 eyes presented cystoids (Table 2). Cystoids of various sizes are present in IJT (mentioned previously).¹⁻³ Some of the cystoids, usually large to moderate in size, are located in the central retina and in the foveola (patients 1 and 2). Some of the cystoids, usually small, are present in the INL (patients 2, 4, and 6) or in other retinal layers such as the GCL (patients 4 and 5). The large to moderate cystoids are likely to be found with standard-resolution OCT imaging as well; however, small cystoids are revealed only by the UHR OCT images, due to their increased resolution. In some cases, pseudocystoids in the foveola (patient 2; patient 5, right eye; patient 6) are present due to the presence of the ILM drape.

A Unique Feature, Internal Limiting Membrane Draping, Was Found in Idiopathic Juxtafoveal Retinal Telangiectasis

The ILM drape feature can create cystlike structures or macular holes (patient 5, left eye). The ILM drape feature can be significantly extended over the foveola and is easily visible with OCT (patient 2; patient 5, left eye), or it can be small and seen only in UHR OCT (patient 5, right eye; patient 6). The large ILM drape features are detected by standard-resolution OCT; however, the small ILM features are only visible in the UHR OCT images, due to their increased resolution. The ILM drape feature was found in 42% of eyes (Table 2).

Large Intraretinal Blood Vessels Are Found in Idiopathic Juxtafoveal Retinal Telangiectasis

Intraretinal blood vessels were found in 21% of eyes in our study (Table 2). Large blood vessels characteristic of IJT, as mentioned by Gass,^{2,3} are visualized near the foveola in the OCT images. Our UHR OCT system is capable of locating large blood vessels to the INL and ONL. The origin of these vessels is not known, and theories are considering enlarged deep vessels or vessels due to neovascularization.¹⁻⁷

This small retrospective study (19 eyes) presents characteristic features in IJT, whose pathology is still unknown, shown by both StratusOCT and UHR OCT. In addition, the novelty of this study consists in new features that are present only in UHR OCT images. Further studies are warranted to increase the number of IJT patients imaged for better statistical results of the findings presented here. A follow-up study of the IJT patients intended to stage this disease better would also be of great value for the understanding of this disease.

The current diagnosis method for IJT is fluorescein angiography. However, considering OCT's new IJT findings, we hypothesize that it may be possible to diagnose IJT and follow-up IJT patients by OCT images alone. Our study is still limited and cannot answer this question clearly because of the small sample size (10 patients).

In summary, the first extended imaging study of idiopathic juxtafoveal telangiectasis with UHR OCT provides useful information on the unknown pathogenesis of IJT. Ultrahigh-resolution

OCT demonstrated the ability to detect smaller changes and describe the localization of these changes in the intraretinal layers better. Ultrahigh-resolution OCT allows detailed imaging of the photoreceptor morphology associated with different types of IJT. The following major findings were found in retinal images of the IJT patients: retinal thickness did not necessarily correlate with leakage found on fluorescein angiography, large blood vessels near the foveola, the presence of cystlike structures and deposits in the outer retina, and the presence of ILM drape and RPE plaque. Features such as small cystoids and a small ILM drape as well as thinning or disruption of the photoreceptor layer are only visible with UHR OCT. Therefore, UHR OCT imaging has the potential to improve the understanding of IJT pathogenesis and the causes of visual loss secondary to IJT as well as to enhance the diagnosis and treatment of this disease.

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Figure 1.

A, A commercially available optical coherence tomography (OCT) standard-resolution system (StratusOCT) image of a normal human macula. **B**, Ultrahigh-resolution (UHR) OCT of the normal macula at the same location. The images demonstrate the ability to visualize intraretinal layers that can be correlated with retinal anatomy. Most of the major intraretinal layers can be visualized in the StratusOCT image, but the ganglion cell layer (GCL) and external limiting membrane (ELM) are much better visualized in the UHR OCT image. INL = inner nuclear layer; IPL = inner plexiform layer; IS = inner segment; NFL = nerve fiber layer; ONL = outer nuclear layer; OPL = outer plexiform layer; OS = outer segment; RPE = retinal pigment epithelium. Red, highly backscattering layers; blue, low-backscattering layers.

Figure 2.

Patient 1. **A**, Color fundus photography of the left eye of a unilateral patient with idiopathic juxtafoveal retinal telangiectasis (IJT) classified as group 1B (Gass JD, Oyakawa RT. Idiopathic juxtafoveal retinal telangiectasis. *Arch Ophthalmol* 1982;100:769–80) (Gass JD, Blodi BA. Idiopathic juxtafoveal retinal telangiectasis. Update of classification and follow-up study. *Ophthalmology* 1993;100:1536–46). The color photography depicts some macular edema. **B**, **C**, Ultrahigh-resolution optical coherence tomography image of a patient with unilateral 1B (IJT). Scans taken at 180° (**B**) and 240° (**C**). Both images demonstrate a foveal cystoid, small intraretinal cystoids, and fluid accumulation under the sensory retina. A Müller cell is shown in **B**, and intraretinal blood vessels are shown in **C**. The photoreceptor layer is shown intact in both images. ELM = external limiting membrane; IS/OS = junction between the photoreceptor inner segment and outer segment.

Figure 3.

Patient 2. **A**, Red-free photography of the right eye of a bilateral patient with idiopathic juxtafoveal retinal telangiectasis (IJT) classified as group 2A (Gass JD, Oyakawa RT. Idiopathic juxtafoveal retinal telangiectasis. *Arch Ophthalmol* 1982;100:769–80) (Gass JD, Blodi BA. Idiopathic juxtafoveal retinal telangiectasis. Update of classification and follow-up study. *Ophthalmology* 1993;100:1536–46). Photography is enhanced in the foveal region to show cystoid structure. **B**, Ultrahigh-resolution optical coherence tomography (OCT) image of the right eye of a patient with bilateral 2A IJT. The image demonstrates a foveal cystoid. All the other intraretinal tissue layers, including the photoreceptor layer, are shown intact. **C**, Red-free photography of the left eye of a bilateral patient with IJT classified by Gass (see above) as group 2A. Photography is enhanced in the foveal region to show cystoid structure. **D**, Ultrahigh-resolution OCT image of the left eye of a patient with bilateral 2A IJT. The image demonstrates a foveal cystoid and small intraretinal cystoids in the temporal side of the fovea. The photoreceptor layer is shown with a disruption in the foveola region; however, the peripheral photoreceptor layer is intact. The external limiting membrane (ELM) is shown intact throughout the macula. In addition, the internal limiting membrane (ILM) is shown across the macula, forming what we call the ILM drape. The photoreceptor segment disruption correlates with the larger visual acuity loss in the left eye.

Figure 4.

Patient 3. **A**, Color photography of the right eye of a bilateral patient with idiopathic juxtafoveal retinal telangiectasis (IJT) classified as group 2A (Gass JD, Oyakawa RT. Idiopathic juxtafoveolar retinal telangiectasis. *Arch Ophthalmol* 1982;100:769–80) (Gass JD, Blodi BA. Idiopathic juxtafoveolar retinal telangiectasis. Update of classification and follow-up study. *Ophthalmology* 1993;100:1536–46). A normal retina is depicted. **B**, Commercially available optical coherence tomography (OCT) standard-resolution system (StratusOCT) image of a bilateral patient with group 2A IJT. The image shows a normal-looking retinal scan and perhaps a thinner fovea. Quantitative measures indicate a thinner macula, including the foveola. **C**, Ultrahigh-resolution (UHR) OCT image of a bilateral patient with group 2A IJT. The image shows a normal-looking retina in the peripheral macula with some thinning of the photoreceptor layer and a thinner fovea. In the foveola, a small disruption of the photoreceptor layer and a cystlike structure can be visualized. A Müller cell connecting the inner and outer retinal layers is also visible. Both StratusOCT and UHR images are taken at a 90° scan orientation.

Figure 5.

Patient 4. **A**, Fluorescein angiography of the left eye of a patient with bilateral group 2A idiopathic juxtafoveal retinal telangiectasis (IJT). The red-free photograph shows a yellow central spot in the foveola. Angiography shows some late leakage. **B**, Ultrahigh-resolution optical coherence tomography image of the left eye of a patient with bilateral group 2A IJT, taken at a 150° scan orientation. The image demonstrates a yellow deposit and small cystoids in the ganglion cell layer and inner nuclear layer with elevation of the inner retina. Loss of the photoreceptor segment junction (IS/OS) signal is shown centrally where the photoreceptor junction is intact peripherally. ELM = external limiting membrane.

Figure 6.

Patient 5. **A**, Late-phase fluorescein angiography of the right eye of a patient with bilateral group 2A idiopathic juxtafoveal retinal telangiectasis (IJT). The angiography shows leakage near the foveola. **B**, Ultrahigh-resolution optical coherence tomography (OCT) image of the right eye of a patient with bilateral group 2A IJT, taken at a 30° scan orientation. The image demonstrates a large blood vessel near the fovea, a small cystoid in the ganglion cell layer (GCL), an internal limiting membrane (ILM) drape across the fovea, and complete loss of the photoreceptor layer centrally, with a thinning of the retina but intact photoreceptor layer peripherally. **C**, Late-phase fluorescein angiography of the left eye of a patient with bilateral group 2A IJT. The angiography shows a pattern similar to that of a macular hole, with some leakage near the foveola. **D, E**, Ultrahigh-resolution OCT image of the left eye of a patient with bilateral group 2A IJT, taken at 150° (D) and 180° (E) scan orientations. Both images demonstrate large blood vessels near the fovea, small cystoids in the GCL and inner nuclear layer, an ILM drape across the fovea creating a lamellar hole, and complete loss of the photoreceptor layer centrally but an intact photoreceptor layer peripherally.

Figure 7.

Patient 6. **A**, Late-phase fluorescein angiography (FA) of the right eye of a patient with bilateral group 2A idiopathic juxtafoveal retinal telangiectasis (IJT). The FA shows changes similar to that of choroidal neovascularization (CNV). **B**, Ultrahigh-resolution optical coherence tomography (OCT) image of the right eye of a patient with bilateral group 2A IJT, taken at a 90° scan orientation. The image demonstrates changes near the fovea that could be correlated with retinal pigment epithelium (RPE) plaque and CNV. Small cystoids are present in the inner nuclear layer, and complete loss of the photoreceptor layer is detected centrally. The retina is thickened, which is associated with CNV. **C**, Late-phase FA of the left eye of a patient with bilateral group 2A IJT. The FA shows hyperfluorescence associated with leakage. **D**, Ultrahigh-resolution OCT image of the left eye of a patient with bilateral group 2A IJT, taken at a 90° scan orientation. The image demonstrates fluid accumulation in the fovea underneath the junction between the photoreceptor inner segment and outer segment, but with an intact external limiting membrane and RPE. A small internal limiting membrane (ILM) drape is shown in the foveal region.

Table 1
Summary of Individual Data, Including Demographics, Clinical Data, and Ultrahigh-Resolution Optical Coherence Tomography (UHR OCT) Findings in 10 Patients with Idiopathic Juxtafoveal Retinal Telangiectasis (IJT)

Patient No.	Age (yrs)	Gender	VA (Right Eye)	VA (Left Eye)	StratusOCT Foveal Retinal Thickness (μm)		IJT Class	UHR OCT Findings
					Right Eye	Left Eye		
1	47	M	20/15	20/25	197±18	461±14	1B	Left eye: foveal thickening; foveal cyst; small cysts in INL; Müller cell spanning fovea; blood vessels near fovea in ONL.
2	76	F	20/15 20/25	20/15 20/50	195±13 185±3	320±17 187±4	2A	Right eye: foveal thickening; foveal cysts; disruption of the photoreceptor layer centrally. Left eye: foveal thickening; foveal cysts; cysts in INL; ILM drupe; disruption of the photoreceptor layer centrally.
3	54	F	20/30 20/30	20/40 20/25	No StratusOCT 125±4	No StratusOCT 134±11	2A	OU: foveal thinning; disruption of the IS/Left eye; small cysts in the photoreceptor layer; Müller cell.
4	35	F	20/30	20/60	143±4	244±10	2A	Right eye: foveal thinning. Left eye: foveal thickening; foveal yellow spot; small cysts in GCL and INL; detachment of the IS/Left eye; loss of photoreceptor layer.
5	61	M	20/25 20/70	20/60 20/70	211±31* 180±48*	155±21 [†] 252±14 [‡]	2A	Right eye: foveal thinning; large subretinal blood vessel near fovea; ILM drupe; small cysts in GCL, INL, and ONL; disruptions of photoreceptor layer. Left eye: same as right eye; lamellar hole.
6	64	F	CF 4 feet	20/60	250±14	223±16	2A	Right eye: foveal thickening; loss of foveal pit; cysts in GCL and INL; disruption of the photoreceptor layer; RPE plaque. Left eye: foveal thickening; intraretinal fluid due to choroidal neovascularization; ILM drupe; disruption of the photoreceptor layer.
7	58	F	20/40	20/40	213±1	140±1	2A	Right eye: foveal thickening; loss of foveal pit; small disruption of the RPE; ERM. Left eye: foveal thinning; photoreceptor layer thinning.
8	60	M	CF 7 feet	20/20	103±4	122±13	2A	Right eye: foveal thinning; cysts in INL and ONL; ILM drupe; disruption of the photoreceptor layer. Left eye: foveal thinning; inner retinal detachment; ILM drupe; disruption of the photoreceptor layer.
9	52	M	20/60	20/100	160±21	232±21	2A	Right eye: foveal thinning; ILM drupe; disruption of the IS/left eye and photoreceptor layer. Left eye: foveal thickening; foveal cyst; ILM drupe; disruption of the IS/left eye and photoreceptor layer.

Patient No.	Age (yrs)	Gender	VA (Right Eye)	VA (Left Eye)	StratusOCT Foveal Retinal Thickness (μ m)		LIT Class	UHR OCT Findings
					Right Eye	Left Eye		
10	58	F	CF 4 feet	20/20	4103 $\pm$ 2	164 $\pm$ 4	2A	Right eye: foveal thickening; small and large cysts; intraretinal layer structure missing centrally; loss of photoreceptor layer and perhaps RPE. Left eye: foveal thinning.

CF = counting fingers; ERM = epiretinal membrane; F = female; GCL = ganglion cell layer; ILM = internal limiting membrane; INL = inner nuclear layer; IS/OS = junction between the photoreceptor inner segment and outer segment; M = male; ONL = outer nuclear layer; OU = both eyes; RPE = retinal pigment epithelium; StratusOCT = commercially available OCT standard-resolution system; VA = visual acuity.

The StratusOCT values for thicknesses correlate with the findings in UHR OCT, except for a few eyes:

- * No correlation was found due to scan alignment errors in the StratusOCT scans.
- [†] One more patient had his left eye injected locally with kenalog and a decrease in thickness was observed.
- [‡] No correlation was found due to algorithm detection errors of the retinal borders, falsely detecting the ILM drape as the retinal border.

Table 2

Idiopathic Juxtafoveal Retinal Telangiectasis (IJT) Main Findings by Ultrahigh-Resolution Optical Coherence Tomography in 10 Patients

IJT Characteristics	No. of Eyes	% of Eyes
1. No correlation between retinal thickening and FA leakage	3	16
2. Photoreceptor disruption	16	84
3. Intraretinal cysts	12	63
4. ILM drape	8	42
5. Intraretinal foveal neovascularization	4	21
6. Foveal deposits	2	11

FA = fluorescein angiography; ILM = internal limiting membrane.