Optical Coherence Tomography Findings in Idiopathic Perifoveal Telangiectasia

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Abstract

***Purpose*:** To determine the optical coherence tomography (OCT) findings in eyes with idiopathic perifoveal retinal telangiectasis (IPT)

***Methods*:** This study is a retrospective review of patient charts, OCT, fundus photography, related to 16 eyes (11 patients).

***Results*:** The most consistent finding of idiopathic perifoveal telangiectasia seen in 93.7% of eyes (15 eyes) on OCT was the presence of hyporeflective intraretinal spaces (cysts) in the absence of retinal thickening. Other findings identified in IPT were: loss and disruption of the photoreceptor layer in 87.5% (14 cases), internal limiting membrane draping across the foveola related to an underlying loss of tissue in 25% (4 cases), an abnormal outward disfiguring of outer retinal layers which may be related to outer retinal atrophy in 37.5% (6 cases).

***Conclusion*:** The OCT findings in idiopathic perifoveal telangiectasia were characteristic and are helpful for better understanding its pathogenesis and visual function abnormalities.

***Keywords*:** Idiopathic Juxtafoveolar Retinal Telangiectasis, Idiopathic Macular Telangiectasia, Optical Coherence Tomography, Parafoveal Telangiectasis

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Introduction

The term retinal telangiectasia was first proposed by Reese[1] to describe a retinal vascular disorder characterized by dilation of the capillaries of the retina. If irregular dilations are limited to the capillaries of the foveal avascular zone, it is known as parafoveal telangiectasia.[1]

In 1982 Gass proposed a classification of idiopathic macular telangiectasis.[2] A modified classification of this entity was proposed by Yannuzzi in 2006, in order to produce a better understanding of the disease.[3] Optical coherence tomography (OCT) is a relatively new, noninvasive imaging technology, which has provided to be an important clinical tool for understanding the pathogenesis of different retinal diseases.

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With advances in retinal imaging, our ability to image retinal microarchitecture in vivo has reached an unprecedented, near histologic level. The pathogenesis of idiopathic perifoveal retinal telangiectasis (IPT) is not known. With use of OCT, retinal structural changes secondary to IPT may clarify the processes occurring in this disease. In literature the most consistent finding of IPT in 87.5% eyes on OCT was the presence of intraretinal cysts without retinal thickening. The OCT features in these eyes appears to be predominately retinal atrophy.[4]

There are few published reports of the OCT features of this disease. The current study reports OCT findings in 16 eyes with type II idiopathic macular telangiectasia.

Methods

We reviewed the medical records of consecutive patients in our files referred to us with a diagnosis of perifoveal telangiectasia (according to OCT and fluorescein angiography [FA] findings) from February 2008 to June 2010.

All patients had clear media and good fixation to allow for good quality OCT imaging.

Patients with coexistent diabetic retinopathy, systemic hypertension or a history of radiation therapy to the head were excluded from the study. The study protocol was reviewed in advance of the study by institutional review board. Each participant was informed of its purpose and gave written consent to participate. A chart review of cases who developed type II IPT including data on gender, laterality, age and best corrected visual acuity (BCVA) was provided.

OCTs were done by a single experienced vitreoretinal fellow. For each eye, vertical and horizontal cross-sectional scans (9 mm in length) were obtained, centered on the fovea. Multiple raster scans which covering a 6 mm × 6 mm area also were obtained.

OCT images performed by spectral domain (SD) OCT (Heidelberg spectralis HRA+OCT) were reviewed by two experienced vitreoretinal fellows for retinal thickness in the center of fovea, presence of hyporeflective areas in retinal layers, and arrangement of different layers of sensory retina and retinal pigment epithelium. All patients had FA by Heidelberg HRA FA for detection and confirmation of diagnosis of perifoveal vascular telangiectasia (Heidelberg Engineering, Heidelberg Germany).

Results

Sixteen eyes of 11 patients were included in the study. Mean patient age was 57.87 (range: 48-74). Visual acuity ranged from ($^{20}/_{20}$ - $^{5}/_{100}$). Mean central subfield thickness (CST) was 234 µm (range: 141-329 µm).

OCT features for each patient are depicted in table 1.

In our study, we reported OCT findings in four types:

1: Retinal cyst

We use the term cyst for all hyporeflective areas evident on OCT (Figures 1, 2 and 3). The most consistent finding of idiopathic perifoveal telangiectasia seen in 93.7% (15 of 16 eyes) of eyes on OCT was the presence of hyporeflective intraretinal cyst.

2: Internal limited membrane drape

It is seen in the OCT images as a thin layer above the cystoid space in inner layers of the retina (Figure 1) (in 4 of 16 eyes: 25%).

3: Loss and disruption of the photoreceptor layer

We found the photoreceptor layer involvement in this disease locally (Figures 1 and 3) (in 14 of 16 eyes: 87.5%).

4: Outward disfiguring

We found an abnormal deviation of retinal layers in some OCT patterns. It may be related to thinning and atrophy of outer layers resulting in this outward deviation of retinal structures (in 6 of 16 eyes: 37.5%) (Figure 2).

None of the eyes showed reduced backscatter such as presence of intraretinal fluid in cystoid macular edema.

Table 1. Findings in eyes with idiopathic perifoveal retinal telangiectasis (best corrected visual acuity, age, fluorescein angiography, optical coherence tomography)

Number	BCVA	Age	FA finding	OCT finding
1	OD: 2/10	63	Leakage (T,N)	Cyst, photo dis, ILM drape
2	OS: 1/10	63	Leakage (T,N)	Cyst, photo dis, ILM drape, outward figure
3	OD: 7/10	58	Leakage (T)	Cyst, photo dis
4	OS: 10/10	58	Leakage (T)	Cyst
5	OD: 5/100	48	Leakage (T)	Cyst, photo dis
6	OD: 1/10	58	Leakage (T)	Cyst, photo dis, outward figure
7	OS: FC 2m	74	Leakage (T)	Cyst, photo dis
8	OD: 2/10	50	Leakage (T, N)	Outward figure, photo dis
9	OS: 5/10	50	Leakage (T)	Cyst, photo dis
10	OD: 1/10	58	Leakage (T)	Cyst, photo dis, outward figure
11	OD: 7/10	55	Leakage (T)	Cyst, photo dis
12	OS: 8/10	55	Leakage (T)	Cyst, photo dis
13	OD: 1/10	63	Leakage (T)	Cyst, photo dis, ILM drape
14	OD: 7/10	57	Leakage (T)	Cyst, photo dis, Outward figure
15	OS: 3 /10	57	Leakage (T,N)	Cyst, photo dis, ILM drape, outward figure
16	OS: 7/10	59	Leakage (T)	Cyst

BCVA: Best corrected visual acuity
FA: Fluorescein angiography
OCT: Optical coherence tomography
T: Temporal
N: Nasal
Photoreceptor dis: Photoreceptor disruption
ILM: Internal limited membrane

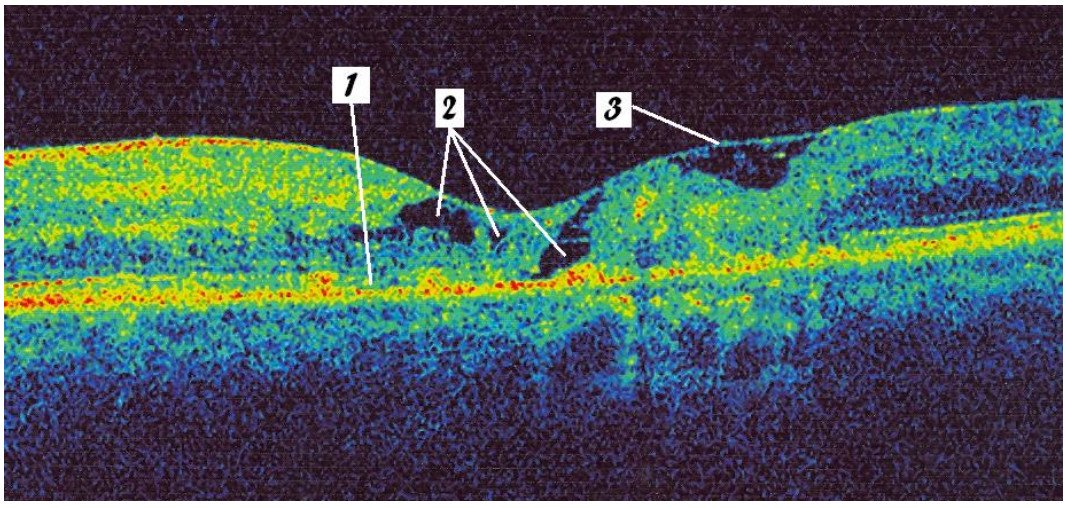

Figure 1. Three patterns of OCT in IPT. 1: shows an area of photoreceptor disruption. 2: shows multiple cysts at various retinal depths (in superficial, middle and deep layer). 3: depicts internal limited membrane draping, which is a thin layer over a superficial retinal cyst. It may represent a preserved foot plates of the Muller cells.

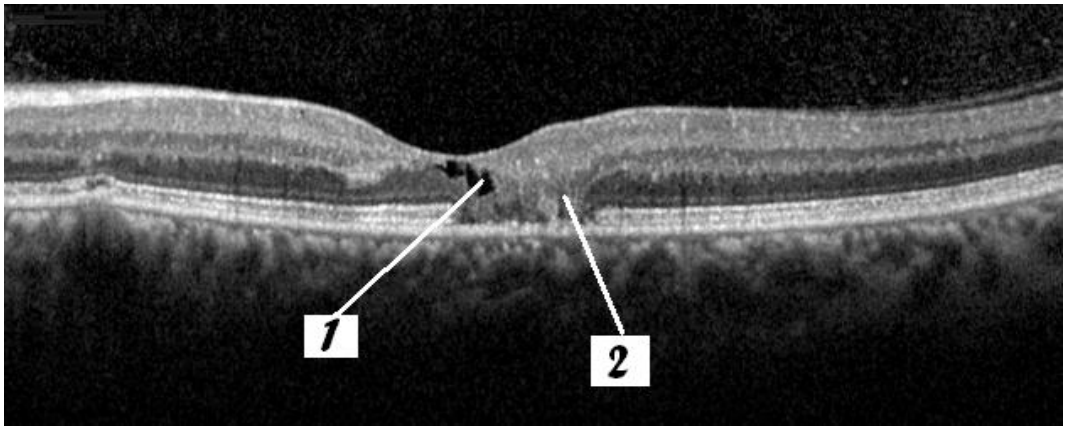

Figure 2. 1: shows retinal cyst. 2: Outward deviation of retinal layers, may be due to outer retinal atrophy.

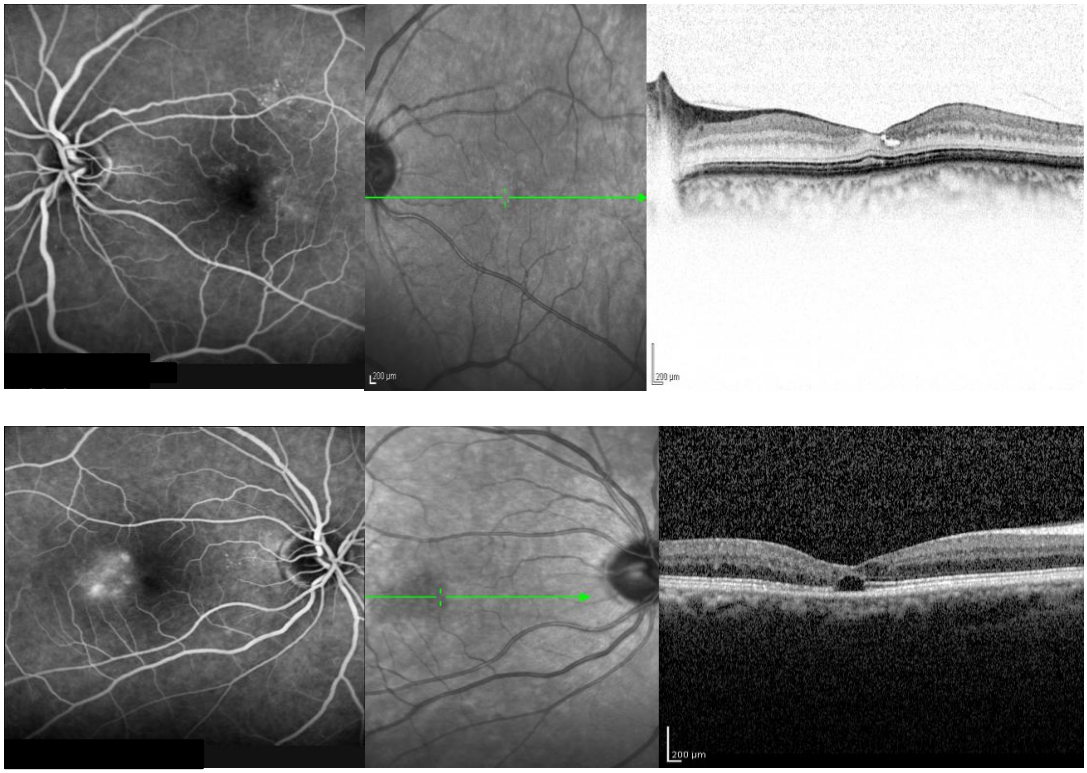

Figure 3. Bilateral perifoveal telangiectasia in a 58-year-old man. Best corrected visual acuity in right was $^{20}/_{30}$ and in left $^{20}/_{20}$. Staining or leakage in temporal side in both eyes in fluorescein angiography (right more than left). Upper right figure shows retinal cyst in inner layer in left eye and lower right figure is a deep retinal cyst with photoreceptor disruption in horizontal optical coherence tomography scan.

Discussion

Other studies such as Gaudric,[5] Surguch[6] and Koizumi et al[7] show multiple retinal changes in IPT by OCT, such as intraretinal cystoids space in lack of foveal edma, photoreceptor layer damage in juxtafoveal area, intraretinal hyperreflective lesion and defect in outer retina.

The foveal cysts seen in idiopathic perifoveal telangiectasia are not as typical cysts seen in cystoid macular edema because they lack associated clinical and angiographic findings, and exhibit without associated retina thickening.[8] There are multiple structural differences between such cysts and the ones which are due to interrupted retinal circulations in diabetes or other retinal vascular diseases (Figure 4).

Internal limited membrane (ILM) drapping is special finding that could be find mainly in IPT and we believe that it is different from usual intraretinal cystic spaces in other situation such as diabetic macular edema and retinal vein occlusion. We believe that such hollow spaces in retina are the result of retinal cell loss that may be caused by longstanding macular hemodynamic and nutritional changes in IPT (Figure 5).

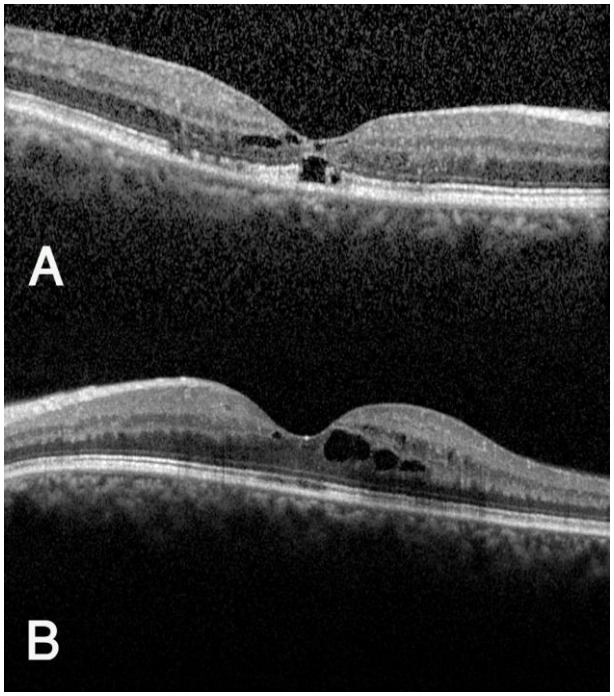

Figure 4. This compound compares between cystic changes between idiopathic perifoveal retinal telangiectasis and early diabetic macular edema. “A” part shows spindle shape intraretinal cysts without considerable foveal thickening and accompanied by Internal limited membrane drapping in central fovea and photoreceptor layer attenuation in left side of central fovea and segmental loss of outer retina in subfoveal area. In part “B” which is Optical coherence tomography cross sectioning from macula in early diabetic macular edema, multiple cystic changes as multiple spheroid spaces in outer nuclear layer are noted and accompanied by localized retinal thickening. Retinal layers arangement are preserved.

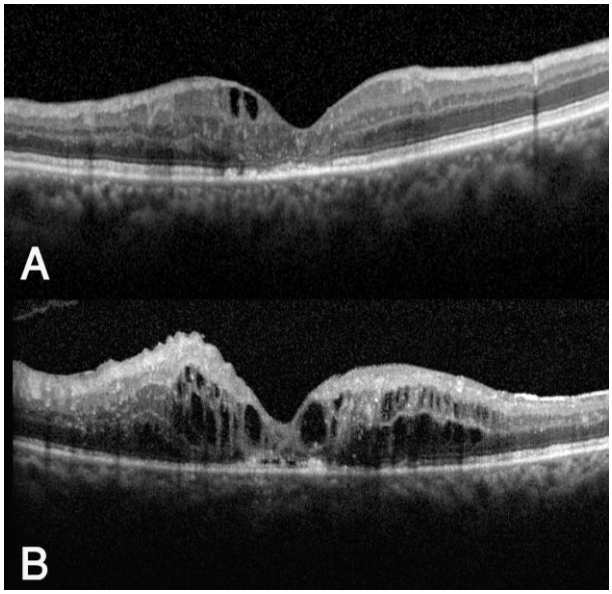

Figure 5. This figure has two parts. Part “A” belongs to a patient with idiopathic perifoveal retinal telangiectasis and part B to one with diabetic macular edema. In part “A”, Internal limited membrane drapping represented az superficial retinal cystic spaces. Outer layers are homogeneous, but in “B” part that belongs to patient with diabetic macular edema, hollow spaces that represent retinal cystic changes are deeper and in outer layers. Outer retinal layer edema with cystic changes are notable which never seen in idiopathic perifoveal retinal telangiectasis patients.

In all of eyes, retinal layer thickening which represent retinal edema didn’t exist. The coexistant foveal atrophy indicated the possibility of atrophic changes in the retina with fluid collection or empty cysts.

Consistent with Gass and Blodi’s proposed pathogenesis of foveolar atrophy in idiopathic perifoveal telangiectasia, the cysts may result from cellular atrophy in the middle retinal layers, perhaps with subsequent fluid accumulation.[9] They could represent a schisis-like cavity with loss of the cellular components of the Muller cells and preservation of their footplates (internal limiting membtane).[10,11]

In our study we detected that the photoreceptor layer is locally disrupted in some IPT cases. In previous studies by Gaudric et al[5] photoreceptor layer damage and disruption was mentioned in IPT.

In our patients with IPT two important features could be demonstrated: ILM draping because of underlying tissue loss and also an abnormal outward disfiguring of outer retinal

layers which may be related to outer retinal atrophy.

The outer retinal atrophy in these eyes could not be caused by retinal vascular abnormalities alone, because the outer retina derives oxygen and nutrients from the choriocapillaris and not the retinal circulation.

Muller cell dysfunction, on the other hand, could lead to outer retinal atrophy and degeneration. Muller cells normally maintain the health of the surrounding neurons including the outer retinal neurons (photoreceptors).[12]

The photoreceptors are lost due to nutritional atrophy of overlying retinal cells, which permits retinal pigment epithelial cells to migrate along the retinal venules resulting in the formation of black satellite plaques.[4]

Conclusion

Although OCT measures and represent retinal anatomy, but such changes in IPT maybe match up with physiological theory for IPT. In summary, OCT is a powerful new tool for evaluating macular disorders and OCT findings specially in idiopathic macular telangiectasia type II In such patients OCT findings maybe very characteristic and are helpful for better understanding its diagnosis, pathogenesis and visual function abnormalities.

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