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Short Communication

Optical Coherence Tomography Angiography for Macular Telangiectasia Type 2

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Abstract

Optical Coherence Tomography Angiography (OCT-A) is an evolving retinal imaging modality for vascular diseases. By avoiding the intravenous fluorescein dye and its associated complications, OCT-A better identifies abnormal vasculature in the retinal and choroidal layers without any interference from fluorescein leakage. This is particularly useful for visualization of pathology in macular telangiectasia. Classified as non-proliferative and proliferative, macular telangiectasia can eventually progress to subretinal neovascularization. In this article, we review its pathology at different stages and corresponding OCT-A findings in different retinal layers: from the superficial and deep retinal capillary plexus, through the retinal pigment epithelium, and down to the choriocapillaris layers.

Keywords

Optical Coherence Tomography Angiography; Macular Telangiectasia; Retina; Choroid; Imaging

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Introduction

Optical Coherence Tomography Angiography

With the advancement of technology in speed and resolution, retinal imaging is not limited to optical coherence tomography or Fluorescein Angiography (FA) alone [1]. New imaging modality that uses motion contrast to provide a non-invasive image of the retinal and choroidal vasculature is called Optical Coherence Tomography Angiography (OCTA) [2]. The principle behind is sequential B-scan images over same retinal location, with subsequent analysis of change in amplitude and phase, inferred as flow of red blood cells in vasculature [3]. This cube scan generates a 3-dimensional image set and its extension provides en-face view of the vasculature at different layers of the retina [4]. Therefore, a flow-based imaging is available on top of the intensity-based B-scan imaging [5].

Spectral Domain (SD) and Swept Source (SS) OCTA are 2 major systems of imaging for commercially available OCTA machines [6,7]. SD OCTA utilizes 840 nm light source with a bandwidth of 50 - 90 nm for imaging [8]. The A-scan rate is of 68,000 - 70,000 scans per seconds, whereas the axial resolution, transverse resolution and A-scan depth are variable among different brands of OCTA machines, but much finer than traditional optical coherence tomography [7,9]. In contrast, SS OCTA uses 1,050 nm light source with A-scan rate of 100,000 scans per second [10]. With the longer wavelength over the invisible infrared spectrum, advantage of SS OCTA is not only limited to more comfortable patient experience during the scan, but also better tissue penetration for better image quality of the choroid [11,12]. Besides, the faster scan speed allows more scans over the same position, thus better suppression of motion artifacts, allowing wider field imaging with greater resolution.

Most commercially available OCTA machines process the function of autosegmentation, which split the 3-dimensional images into “slabs”, to reflect a known anatomic layer of the retinal vasculature [13]. Inner, middle, outer retinal and choriocapillaris slabs are pre-set tissue slabs commonly applied clinically for imaging respectively the Superficial Capillary Plexus (SCP), deep Capillary Vascular Plexus (DCP), outer retina to Retinal Pigment Epithelium (RPE) layer and choriocapillaris layers [14]. Border of computer segmented reference planes are listed in Table 1 and manual adjustment of predefined slabs boundaries are possible to facilitate visualisation on pathology of interest [15].

Other than eliminating the risk of nausea and anaphylaxis from exogenous intravenous dye injection [16,17]. OCTA imaging allows visualization of the true anatomy over the pathological vasculature, without blockage from the leaking fluorescein [18]. This advantage is especially useful in studying vascular pathology involving different layers such as age-related macular degeneration, diabetic macular edema, macular telangiectasia etc. [19]. We will focus on the discussion on macular telangiectasia type 2 in this article.

	Slab	Corresponding retinal layers	Boundary
1	Superficial retinal slab	Superficial retinal capillary plexus	3 μm below the internal limiting membrane to 15 μm below the inner plexiform layer
2	Deep retinal slab	Deep retinal capillary plexus	15 μm below the inner plexiform layer to 70 μm below the inner plexiform layer
3	Outer retinal slab	Outer retina to Retinal Pigment Epithelium (RPE) *(no vasculature normally)	70 μm below the inner plexiform layer to 30 μm below the RPE *(best fit continuous surface under RPE)
4	Choroid capillary slab	Choriocapillaris	30 μm below the RPE reference to 60 μm below the RPE reference

Table 1: Commonly used slabs in Optical Coherence Tomography Angiography.

Macular Telangiectasia Type 2

First coined by Gass in 1968, classified in system by Gass and Oyakawa in 1982, later modified by Gass and Blodi in 1993, macular telangiectasia is also known as idiopathic juxtafoveal retinal telangiectasia or parafoveal telangiectasia [20-22]. With a prevalence of 0.1%, subject to under diagnosis from its minimal visual symptoms in early stages, this rare disease is thought to be a neurodegenerative disease, originating from Muller cell dysfunction and death causing vascular insult, leading to irreversible loss of inner and outer retinal tissue and photoreceptors, progressing towards macular atrophy [23-27]. At present, widely adopted classification is by Yannuzzi in 2006, based on clinical observation, FA, indocyanine green angiography and spectral domain optical coherence tomography, into idiopathic macular telangiectasia (MacTel) type 1 (aneurysmal telangiectasia) and type 2 (perifoveal telangiectasia) [28,29]. (Table 2) MacTel 2 is further divided into non-proliferative (telangiectasia and foveal atrophy) and proliferative (subretinal neovascular, SRNV) stage.

Starting from the temporal aspect of parafoveal DCP, MacTel 2 later extends circumferentially around fovea with dilated anastomoses between SCP and DCP, invading the Outer Nuclear Layer (ONL) with inner segment/ outer segment/ ellipsoid zone (IS/OS/EZ) loss. The slightly dilated and blunted retinal vessels over temporal parafoveal retina appear to form right angles and travel towards the outer retina [30]. With disease progression, Subretinal Neovascularization (SRNV) originate from both the deep retinal and choroidal circulations [31]. This process which involves all microvascular layers of retina makes it ideal for OCTA imaging.

Patients are asymptomatic or with non-specific complaint of blurring of vision at early stages of disease,[24] but with disease progression, metamorphopsia, paracentral or central scotoma will be symptomatic to affect daily living [32,33]. Clinically, Mac Tel 2 is seen bilaterally, but asymmetrically, as loss of retinal transparency with a grayish coloration over temporal juxtafoveolar area initially with time parafoveal capillary telangiectasia appears mainly on temporal side of macula, progressing to multiple tiny intraretinal golden refractile crystalline deposits near the telangiectasia and stellate foci of intraretinal pigment clumping along the

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right-angled vessels, later cystic cavities or even SRNV membrane, with or without haemorrhages, become obvious on fundus examination.[27,34-38]. With a mean age in 5th to 6th decades of life, MacTel 2 has a slight female gender predilection and a high association with hypertension and diabetes [23,38-43].

Classifications	Gass	Yannuzzi
Type	1A	1
	1B	1
	2A	2
	2B	Deleted
	3A	Deleted
	3B	Deleted

Table 2: Classification of Macular Telangiectasia type 2.

OCTA Imaging of MacTel 2

On the OCTA cross-sectional B-scan, intraretinal hyporeflective spaces are within the inner retina, usually located in the foveal pit, with a predilection for the temporal slope. This inner lamella cyst over the foveolar appears as tissue lost, with the Internal Limiting Membrane (ILM) spanning across or draping over it [24,44]. This ILM drape is characteristically not associated with increased retinal thickening, as if diabetic macula edema or retinal vein occlusion [45]. With disease progression, the inner retinal layers collapse over the cavity and appose onto the RPE. In contrast, hyperreflective intraretinal or inner retinal lesions usually represent the pigment plaques, whereas macula thickening caused by intra- and/or sub-retinal fluid signifies secondary SRNV over the outer neurosensory retinal layers.

En-face OCTA images identify abnormal microvasculature in perifoveal region, especially the temporal DCP of retina. Foveal Avascular Zone (FAZ) appears to be dragged temporally towards the emergency venule. First OCTA images of MacTel 2 was published by Thorell and others in 2014 with the use of SS OCTA using the Optical Coherence Tomography Microangiography (OMAG) algorithm with a scan pattern of 3x3 mm area centered on the fovea [46]. The OMAG algorithm scans a total of 300 B-scan separated 10 µm apart over a 3 mm distance, with roughly 3.8 ms between consecutive scan, to image the microvascular network. Later, few other groups published OCTA images of MacTel 2 with SD OCTA [47,48,49]. With better understanding of the disease with OCTA, a system of grading for MacTel is even available based on OCTA findings [50].

Non-proliferative MacTel 2

In early stage 1, there is only deep retinal capillary telangiectasia. En-face OCTA images usually demonstrate mildly dilated vessels in retinal DCP (Deep Retinal Slab) from the middle retinal layer predominantly over temporal side of fovea; without alternation in microvasculature in superficial layers, i.e. no abnormal blood flow in the superficial retinal slab (Fig. 1). Capillary density remains normal for both SCP and DCP (Fig. 2). Microcysts other than the large cystic cavities are sometimes observed over the ganglion cell and inner nuclear layers, which correspond those small lacunae in the SCP and DCP [31].

In intermediate stage 2, vascular changes involve both the SCP and DCP, inner retinal capillary telangiectasia sets in. En-face OCTA images show multiple, telangiectatic, microaneurysmal-like dilated vessels residing within the middle retinal layer, extending to inner and outer retina (superficial and deep retinal slabs). One or more sets of dilated draining venules originate at right-angle from temporal part macular telangiectatic vascular net may be observed adjacent to the inner foveal cystic spaces [51]. There is loss of vascular density in both SCP and DCP from capillary dropout, with widening of intervascular spaces, but outer retinal slab is absent from abnormal flow signal [52].

In stage 3, telangiectatic vascular invasion involves the full thickness of the retina. Variable sizes of pigment clumps, if present, will pose an optical shadowing effect. There is greater capillary dropout over SCP and DCP and wider intervascular spaces compared to stage 2. FAZ is dragged temporally towards emergence of the dilated venules, becoming distorted and irregular in shape. However, the FAZ size is variable, depending on the presence of subinternal limiting membrane vascular ingrowth into FAZ or capillary dropout, which give a larger or smaller FAZ respectively. There may also be additional layers of newly formed capillaries invading the avascular ONL, causing IS/OS/EZ disruption [53].

Figure 1: The images of the superficial retinal capillary plexus in optical coherence tomography angiography of a Stage 1 Macular Telangiectasia type 2 eye.

Figure 2: The images of the deep retinal capillary plexus in optical coherence tomography angiography of a Stage 1 Macular Telangiectasia type 2 eye.

Proliferative MacTel 2

In stage 4 disease, SRNV develops with significant pigment clumping. Flow signal blockage by pigment clumping is typical. OCTA en-face images show alternation in juxtafoveal capillary network, with prominent anastomoses extending to outer retina, disrupting the IS/OS/EZ. There is significant capillary dropout, with severe distortion of vascular flow pattern. As SRNV communicates to both retinal and choroidal circulations, blood flow of an SRNV can be detected in the outer retinal slab, or even the choroid capillary slab [54]. Meanwhile, OCTA B-scan demonstrated neurosensory retina elevation, with or without retinal edema, by the SRNV and subretinal fibrovascular pigmented plaque. Hyperreflectivity on OCTA B-scan arises from intraretinal pigment and subretinal haemorrhage may be appreciated.

Although routine anti-Vascular Endothelial Growth Factor (VEGF) is not recommended, efficacy of anti-VEGF in proliferative Mac Tel 2 has been reported and effective in controlling the SRNV to protect patients from severe visual loss [55-61]. With anti-VEGF treatment, OCTA demonstrated decrease in calibers of retinal vessels and their anastomoses [31]. It is also observed that microaneurysms with the telangiectasia vessels disappeared on OCTA, yet whether the lesion is regressed or the blood flow within is diminished to undetectable remains a question.

Late Atrophic Stage

In stage 5, fibrovascular proliferation and subretinal fibrosis develops. OCTA en-face images show significantly distorted and stretched macular vessels, with reduction in vascular volume and capillary dropout on superficial and deep retinal slabs. On the contrary, blood flow from the disciform fibrovascular membrane is well seen on outer retinal and choroid capillary slabs. FAZ is distorted, dragged and enlarged. Eventually, the macula appears abnormally thin from enlargement of intraretinal cysts with collapse of empty spaces. Telangiectasia persists, with a decrease in capillary density.

Conclusion

OCTA has the advantages of non-invasive and repeatable imaging for disease diagnosis and disease progression or treatment response monitoring. It is also a faster and cheaper imaging modality that provides good 3-dimensional image quality on microvasculature of retina and choroid, not obscured by fluorescein leakage.

Conflict of Interest

The authors declare no conflict of interest, financial or otherwise.

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