Detecting Lesions with Widefield Color and AF Images and Diagnosing the Detected Lesions with OCT

Jerome Sherman

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Note:
The term retinoschisis should not be used to indicate a retinal detachment. Instead it should be used to indicate a splitting of the sensory retina. This is a more precise and accurate term for the phenomenon. Retinal detachment should be used to indicate a detachment of the neurosensory retina from the retinal pigment epithelium.

Retinoschisis or Retinal Hole?

BOTH! See RR #22

Two slices from one eye. Is it a Retinoschisis or Retinal Detachment?

BOTH!

Case 2: High Definition OCT and Topcon 3D Image of Right Eye

Case 2: The horizontal OCT section and 3D image document the detachment of the neurosensory retina temporal to the macula. Note the cystic type gaps on either side of the outer plexiform layer.
Notice multiple splits within neurosensory retina.
PIL still contiguous with the RPE.

A retinoschisis appears as multiple splits of the neurosensory retina with a portion of the outer retina still visibly attached to the underlying RPE in OCT. In contrast, a retinal detachment appears as a complete separation of the neurosensory retina from the RPE. The distinction between the two disorders is apparent when comparing the OCT images.

Consideration of the junction between the inner and outer segments a.k.a. the Photoreceptor Integrity Line (PIL) may be useful.

Although the photoreceptor integrity line, or the PIL (defined as the junction between the inner and outer segments) is barely visible in most histological sections, it is highly prominent in normal SD OCTs. The PIL, as shown above, should be continuous throughout the entire scan in normal eyes. The PIL is considered by some as a mere artifact that is due to the difference in the index of refraction of the inner and outer segments but this artifact is remarkably useful in SD OCT interpretation.
Normal High-Speed Ultra High Resolution OCT

Ultra-high-resolution OCT systems produce normal images such as shown above. The RPE appears in many scans to be a multi-layered structure. The anterior border is Verhoff’s membrane and the posterior border is Bruch’s membrane. As will be demonstrated in some of the cases to follow, Bruch’s membrane becomes visible in SD-OCT scans as the RPE is attenuated in select disease states. In this ultra-high resolution, “Connecting cilia” corresponds to our PIL/junction and “Lamina basalis choroideae” is Bruch’s membrane.

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BACKGROUND

Purpose: To assess the specificity and sensitivity of the iWellnessExam™ protocol in a cohort of normal subjects and subjects with confirmed disease.

The iWellnessExam report provides:
- eight high-resolution cross-sectional retinal images
- data analysis results
- full retinal thickness map
- ganglion cell complex map
- report on superior/inferior symmetry within the eye and symmetry between eyes

Sensitivity: the proportion of actual positives which are correctly identified as such.

e.g. the percentage of sick people who are correctly identified as having the condition

Specificity: the proportion of negatives which are correctly identified as such.

e.g. the percentage of healthy people who are correctly identified as not having the condition

Normal Subjects:
- Retinal and/or optic nerve disease:
- Retinal Pathology:
- Optic Nerve Pathology:

Specificity 99%
Sensitivity 96%
Sensitivity 95.5%
Sensitivity 90%

SPECIFICITY & SENSITIVITY

The Osterberg Graph

Only 5% of photoreceptors in the retina are cones, and these are concentrated at the fovea. As reported by Osterberg in 1935, the density of cones drops off rapidly just several degrees from the fovea, and rods begin to predominate outside of the central 5 degrees or so. This known distribution of rods and cones is important in the interpretation of the PIL in SD-OCT scans. The depth of the OCT section (6mm) is denoted on the Osterberg graph by the vertical dashed green lines.


Spectral-Domain (High-Speed) OCT System

* OCT Angiography features are not available for sales in US
Enhanced HD Line

12mm EHD Line. 52 YrO Male, 20/20
Tracking and Y-axis zoomed

13mm EHD Line. 45 YrO Female, Pacific Islands, -6.5D,
Scanned through contact lenses. No Y-axis zoom

BEYOND OPHTALMOSCOPY

Peripapillary Retinoschisis: A Novel Clinical Entity Revealed by OCT

Case 1

OPTOS-OPTOMAP

P 200 Dx

DAYTONA

Optos Ultra Wide-field Auto Fluorescence
See what you can’t see with any other technology

What novel information does AF give us?
What novel information does it give us?

- Fundus Autofluorescence (FAF or AF) is a novel, non-invasive imaging procedure that often yields abnormalities that are invisible to ophthalmoscopy and standard color fundus photography.
- FAF is likely due to lipofuscin, the “wear and tear” pigment found in retinal cells, especially RPE cells.
- The normal retinal pigment epithelium (RPE) yields a slightly granular AF glow in contrast to the optic disc and retinal blood vessels which appear black.
- Hyper-AF: The accumulation of lipofuscin, often due to lysosomal dysfunction, results in increased AF and suggests RPE dysfunction or stress.
- Hypo-AF: Decreased FAF suggests loss of RPE cells as well as ganglion photoreceptors and correlates to reduced levels of lipofuscin.
- As revealed in a series of cases to follow, Panoramic or ultra-wide field FAF is now possible and appears to reveal abnormalities throughout the retina, often invisible to other imaging modalities.

BEYOND OPHTHALMOSCOPY

Indirect Ophthalmoscopy

No form of ophthalmoscopy for fundus AF exists!

AF abnormalities have myriad etiologies

V - vascular, vitamin deficiency
I - Infectious, inflammatory
T - trauma, toxic
A - autoimmune, allergic
M - metabolic, mass lesions
I - inherited, idiopathic
N - neurodegenerative
E - endocrine, environmental
S - senile, stress

Figure: VITAMINESx2
This mnemonic is helpful to remember possible etiologies of AF abnormalities.

Next Case

Exemplary cases of Hyper and hypo AF
Exemplary cases of hyper and hypo AF

Hyper AF
Hypo AF

Next Case
Choroidal nevus
(In addition to hyper and hypo AF, there is also iso AF)

OS Composite
Composite View
Green Separation View
Red Separation View

OS AF
OS Composite vs. AF
This lesion is an amelanotic choroidal mass that could represent inflammatory granuloma (sarcoid, syphilis, TB), metastasis, nevus, or melanoma. It is sometimes impossible to differentiate a stable granuloma from amelanotic nevus. Some would include this lesion under the umbrella of solitary idiopathic choroiditis.

Example of hyper-AF
Next Case

Congenital Hypertrophy of the RPE (CHRPE)

OS Composite vs. AF

CHRPE: Congenital Hypertrophy of the Retinal Pigment Epithelium

Example of hypo-AF
A study describing the choroid-retinal vascular anatomy overlying CHRPE using OCT revealed overlying retinal thinning and photoreceptor loss directly over CHRPE in 10 consecutive patients. A study describing the choroid-retinal vascular anatomy overlying CHRPE using OCT revealed overlying retinal thinning and photoreceptor loss directly over CHRPE in 10 consecutive patients.
Case 1 (8 yo son): Fundus Photography

A comparison of standard fundus photos to AF images clearly reveals unmistakable abnormalities in the AF images only. Note that the small hyper-AF lesions are often not round and many are tail shaped or fish tail shaped, suggesting perhaps the fish tail lesions in Stargardt's disease with fundus flavimaculatus. (see RR #14) The symmetry of the AF findings are quite typical of inherited retinal degenerations. (see RR 54 part 1 and part 2)

Case 2 (5 yo daughter): Topcon Fundus Photos OD and OS

The images above reveal normal white glistening reflections off the internal limiting membrane in each eye. This is an expected finding in young children. The fundus images were interpreted as essentially normal in both eyes.

Case 2 (5 yo daughter): Optomap® FAF Image OD and OS

The AF images reveal a subtle macula bull's eye in each eye. Note the hyper-AF ring around the macula in each eye. This pattern is abnormal.

Case 3 (4 yo daughter): Fundus Photography

No marked abnormality is obvious in the color fundus photos or in the AF images in the 4 yo daughter.
Next Case
The PIL/junction is relatively intact in the central 10 degrees or so as depicted here in vertical sections. Outside the central 10 degrees OU, the PIL is virtually non-existent. See RR #25.

These horizontal sections reveal essentially no viable PIL/junction. Bruch’s membrane becomes more prominent as the RPE becomes attenuated.

The PIL/junction is not present in these two horizontal scans superior and inferior to the fovea OD.

Retinitis Pigmentosa

- Topcon 3D OCT revealed the PIL to be present but disorganized in the fovea of each eye.
- Outside of the central 5 degrees, the PIL began to fade gradually and disappear.
- In a disorganized PIL, the well-defined border between the hyperreflective PIL and the hyporeflective outer segments is blurred and the PIL appears to merge into the RPE.

Topcon 3D OCT Horizontal Scan Images of Right Eye

The horizontal OCT scans superior (top image) and inferior (bottom image) to the macula reveal an absent PIL. The only region where the PIL was present was a small segment under the fovea (middle image).

Retinal vessels are usually not apparent in OCT scans, but when pigment cells migrate and surround the vessels, they are more easily visualized.
Three slightly different horizontal OCT sections reveal a lesion that appears to contain a lumen. This arterial vessel is one of several macroaneurysms detected on FA which are sometimes found in hypertensive retinopathy.

- Topcon 3D OCT 6mm scans through a macroaneurysm revealed it as oval and surrounded by a hyperreflective border. On both sides of the macroaneurysm, the PL was present and intact.
- The large size of the macroaneurysm and its subtle emanation into the outer retinal layers such as the RPE and choriocapillaris obstructed and prevented the PL and RPE from being continuous.

The horizontal 3D OCT section through the macroaneurysms (red arrow) corresponds to the same macroaneurysm indicated on the color fundus and FA images.
X-Linked Retinoschisis

Notice radial pattern on fundus

Next Case
60 y/o HM
VA OD 20: sc 70+
VA OS 20: 25+
Next Case

Retinal toxicity invisible to ophthalmoscopy
Plaquenil or Accutane
Case 1: A normal appearing retina in a 25 year old complaining about a new visual problem that has slowed his reading. VA is 20/20 or 6/6 in each eye.

With AF a bull’s eye maculopathy present with AF but not with standard imaging. The left eye reveals a per-foveal ring of hypo AF as well.

A horizontal section through the fovea of the left eye reveals similar findings as displayed previously in the right eye. A small, but intact, PIL is present under the fovea and a perifoveal absence of the PIL is documented. With loss of the PIL, the intact external limiting membrane (ELM) appears to drape over the missing tissue.

As in the right eye, only the AF image reveals an abnormality in the left eye.
Peripapillary Retinoschisis: A Novel Clinical Entity Revealed by OCT

Case 1

Case 2

Case 3

Case 3 continued

Case 4
Peripapillary Retinoschisis: A Novel Clinical Entity Revealed by OCT

Case 5

Note the web of new vessels (red arrows) extending from the disc into the vitreous.
Exudates in and around the outer plexiform layer are common in diabetic retinopathy but also found in hypertensive retinopathy, vein occlusion, macular edema, etc.

In a patient with early non-proliferative retinopathy, SD-OCT revealed about 22 exudates within a 6x6mm box of the horizontal scan. SD-OCT confirmed the location of the exudates in the outer plexiform layer OS. They did not interfere with the clearly defined PIL.

In the horizontal section below the macula only a few very small drusen are visualized. Note the elevation of the retina due to the large underlying drusen in the macula.
Drusen:
Dome-shaped structures called drusen cause the elevation of the RPE. The vast majority of cases with drusen are localized under the RPE. The PIL/junction is mildly compromised above the drusen. As a general rule, it appears that the larger the drusen, the more compromised is the PIL. Note the large drusen with an attenuated PIL, but the smaller drusen has a nearly normal PIL.

Next Case
Dry to Wet AMD
Next Case

Congenital Toxoplasmosis

?reactivation

H/O congenital toxo

Explanation for hyper AF ring?