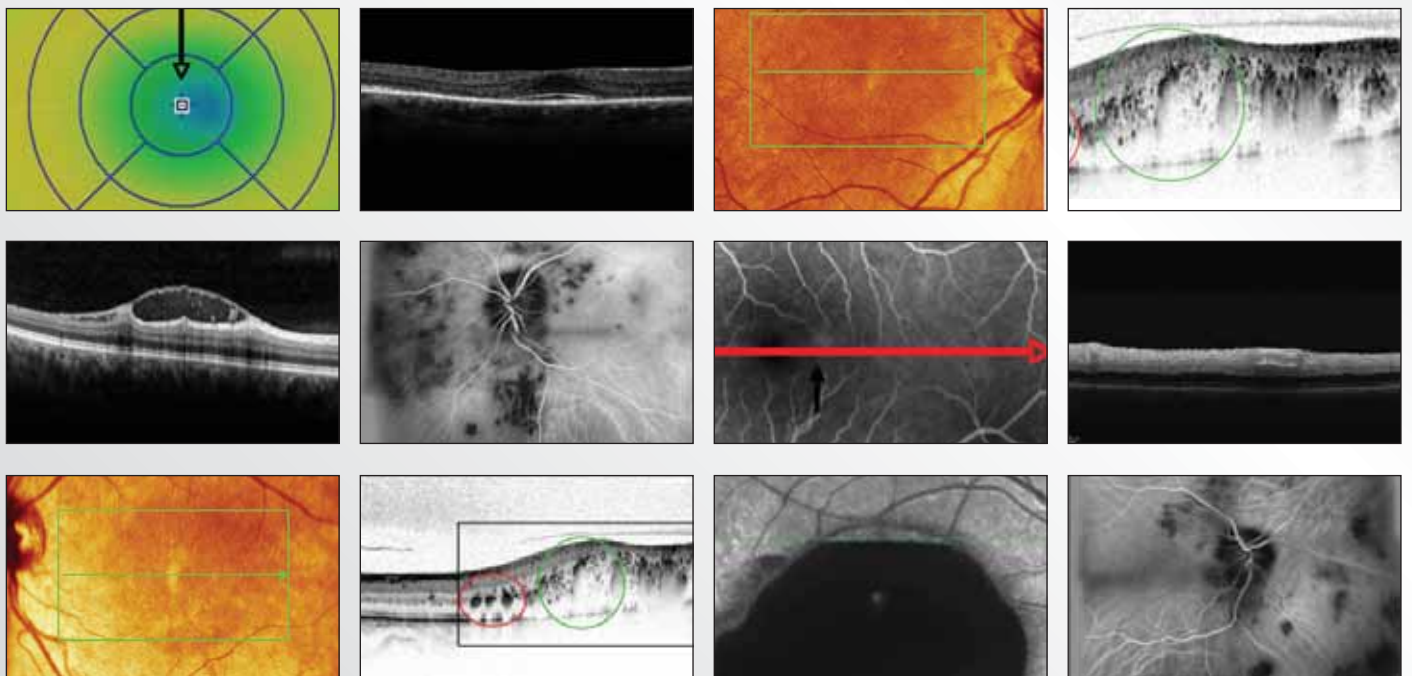


RETINA TODAY

January/February 2010

Multi-modality Imaging of the Retina



Multi-modality Imaging of the Retina



Advanced imaging with spectral-domain optical coherence tomography (SD-OCT) enables clinicians to detect details of chorioretinal pathology like never before. Small, subtle changes in the macula and even in the retinal periphery can now be seen, allowing for more targeted treatment and better outcomes, both medically and surgically. The SPECTRALIS system (Heidelberg Engineering, Heidelberg, Germany) provides OCT images in fine detail and confocal, blue laser autofluorescence (BluePeak) fundus images which are highly sensitive in picking up areas of RPE and retinal abnormalities. In this supplement to *Retina Today*, the assembled faculty present cases where the SPECTRALIS proved essential in detecting different, and at times challenging, pathology. This series of cases includes side-by-side comparisons with standard imaging methods and our panel of experts provide some interesting pearls for interpreting the data that are produced by SD-OCT imaging.

– Lawrence A. Yannuzzi, MD

Dr. Yannuzzi reports no financial relationship with Heidelberg Engineering.

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Hyperreflective Foci in DME

BY URSULA SCHMIDT-ERFURTH, MD

When diabetic macular edema (DME) is classified by findings on optical coherence tomography (OCT), edema in the retina and the macula is most often classified as subretinal or intraretinal; however, upon closer observation, it is apparent that another element is present in this case. In a study that was recently published in *Ophthalmology*,¹ my colleagues and I sought to analyze the hyperreflective foci that seem to be evenly spread throughout the retinal layers in eyes with DME and consistent in eyes with this disease. In this article, I describe these hyperreflective foci, which to our knowledge, is the first description of these black spots associated with DME.

CASE PRESENTATION

We imaged a small sample of patients (n=12) who had treatment-naïve, clinically significant DME with the Stratus OCT (Carl Zeiss Meditec, Jena, Germany), the Cirrus HD-OCT (Carl Zeiss Meditec), and the SPECTRALIS SD-OCT (Heidelberg Engineering, Heidelberg, Germany). The arrows in the OCT slices identify the hyperreflective foci, or black dots, that are seen throughout the retina layers (Figure 1). The foci in the SPECTRALIS image (Figure 1, bottom right) are much more clearly defined.

If you compare the location on the OCT scans with that of the retina, you will see that most of the foci are hard exudates. Based on this observation, we decided that there must be a correlation between exudates and foci.

If one is trying to precisely scan where clinically relevant exudates are present, attention should be paid to areas of confluent aggregates of foci. These aggregates are located in deeper layers (Figure 2) as compared with individual foci that are more isolated and located superficially, suggesting some movement. A shadowing that appears to be a small vessel is present close to the spots, suggesting a close correlation between the two. These vessels can be found, of course, in the superficial layers of the retina. Upon scrutiny, foci or hard exudates can be found in the vessel walls.

We looked to the literature regarding the histology on hyperreflective foci and found some cases with similar findings. Cusick et al² postulated that there is a high concentration of apolipoprotein B around retinal vessels in the context of hard exudates in DME eyes.

Our hypothesis is that these are lipid apolipoprotein

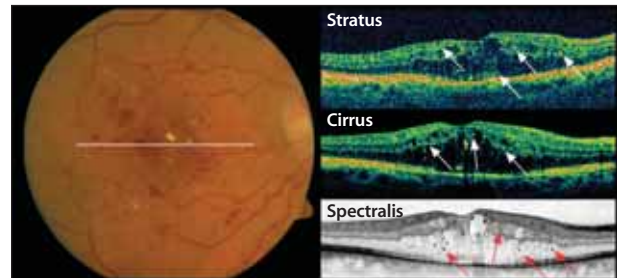


Figure 1. The arrows identify the hyperreflective foci, or black dots, that are seen throughout the retina layers.

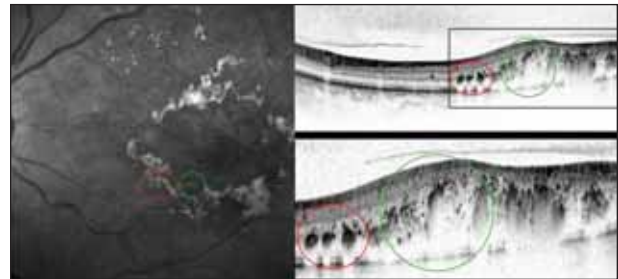


Figure 2. Aggregates of hyperreflective foci are located in deeper layers of the retina.

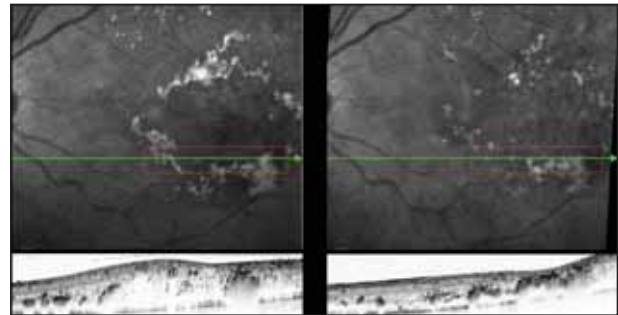


Figure 3. Hard exudates have traveled downward to fill up the deeper layers of the retina.

B aggregates exiting the small vessels and then traveling in the extravascular tissue. Progressive aggregation of these spots causes the development of large plaques that are visible with clinical ophthalmoscopy. The origin of lipid exudation is difficult to determine with their small size and location in the vessel wall, but as soon as the aggregates form, they gravitate to the deeper layers of the retina via the pumping mechanism of the retinal pigment epithelium. Visual function begins to deteriorate once the hard exudates travel to the deeper photoreceptor layers.

FOLLOW-UP AFTER THERAPY

The patient was treated with antivascular endothelial growth factor (anti-VEGF) therapy. At 4 months follow-up the retina appears flatter and the central retinal thickness decreased. The hard exudates, however, remain on pseudohistology; they have traveled downward to fill up the deeper layers of the retina (Figure 3). It is of no surprise that visual function does not improve with anti-VEGF therapy as quickly as it does in age-related macular degeneration (AMD). In AMD, there are no lipids in the retina, just fluid. Anti-VEGF, with its antipermeability effect, may be only half the solution in DME.

CONCLUSION

It is our conclusion that the SPECTRALIS is a useful way to screen for advanced vascular damage in DME that is not

clinically available. This will also allow us to see in the future which patients respond better to therapy for DME. ■

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1. Bolz M, Schmidt-Erfurth U, Deak G, Mylonas G, Kriechbaum K, Scholda C; Diabetic Retinopathy Research Group Vienna. Optical coherence tomographic hyperreflective foci: a morphologic sign of lipid extravasation in diabetic macular edema. *Ophthalmology*. 2009;116(5):914–920.
2. Cusick M, Chew EY, Chan CC, et al. Histopathology and regression of retinal hard exudates in diabetic retinopathy after reduction of elevated serum lipid levels. *Ophthalmology*. 2003;110:2126–2133.

Type 2 Ideopathic Macular Telangiectasia

BY FRANK G. HOLZ, MD

Our goal as doctors is to prevent visual loss via earlier identification of disease, application of prophylactic medicine, and where necessary, earlier treatment. Using fundus autofluorescence (FAF) imaging along with spectral-domain optical coherence tomography (SD-OCT) with the SPECTRALIS system (Heidelberg Engineering, Heidelberg, Germany), I have been able to detect early pathologies that are unable to be seen using other imaging methods. In this article, I present two pairs of siblings with type 2 ideopathic macular telangiectasia (IMT). There is an underlying genetic component to this disease, and with the SPECTRALIS, we are able to isolate several factors that clue us in to the presence of asymptomatic type 2 IMT.

CASE 1

A 42-year-old asymptomatic woman presented to us with 20/20 vision. We classified the fluorescein angiogram (FA) as normal; the blue laser fundus autofluorescence (FAF) and SD-OCT, it also appeared normal when viewed separately. When the FAF was compared to the corresponding SD-OCT scan, however, an anomaly was recognized (Figure 1). Usually the foveal dip appears immediately after the area that corresponds to the hyperreflective band (the little bulge in the fovea seen in Figure 1). For our patient, however, this was not the case. The thinnest part of the fovea, rather, was temporal to where the anatomic fovea sits. After running a volume scan we confirmed that the deepest point of the fovea was not properly

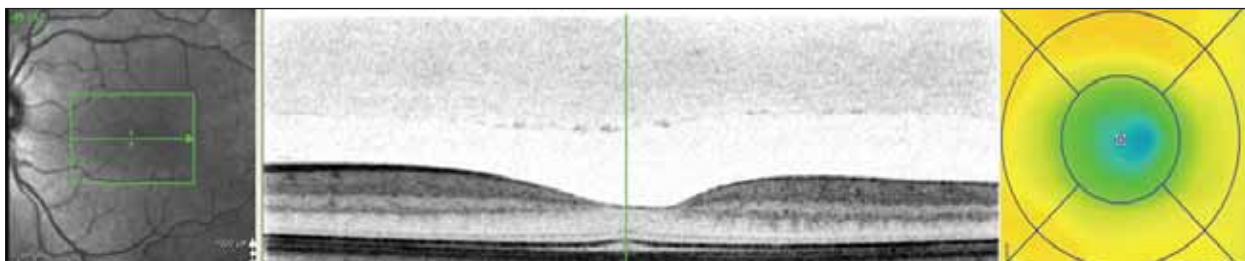


Figure 1. The thinnest part of the fovea was temporal to where the anatomic fovea sits. After running a volume scan we confirmed that the deepest point of the fovea was not properly located.

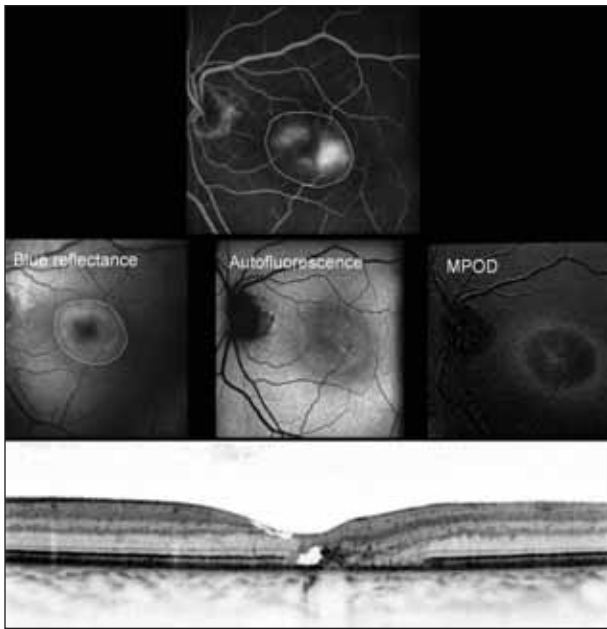


Figure 2. CBR imaging provides an increased signal in a non-invasive manner that picks up any area of blue reflectance.

located (Figure 4).

So why were we taking FA, FAF, and OCTs for a patient with no apparent pathology? The patient's sister had a classic case of type 2 IMT. Although the patient was asymptomatic with 20/20 vision, we suspected that she had type 2 IMT without leakage due to the thinning temporal to the fovea. We found the SPECTRALIS BluePeak blue laser autofluorescence (Heidelberg Engineering) as seen in Figure 2 to be helpful in making this diagnosis in the patient's sister. The confocal red-free imaging provides a markedly increased signal in a noninvasive manner that picks up any area of blue reflectance. FAF shows a maldistribution because of a depletion of luteal pigment, which is pathognomonic to type 2 IMT in the later stages.

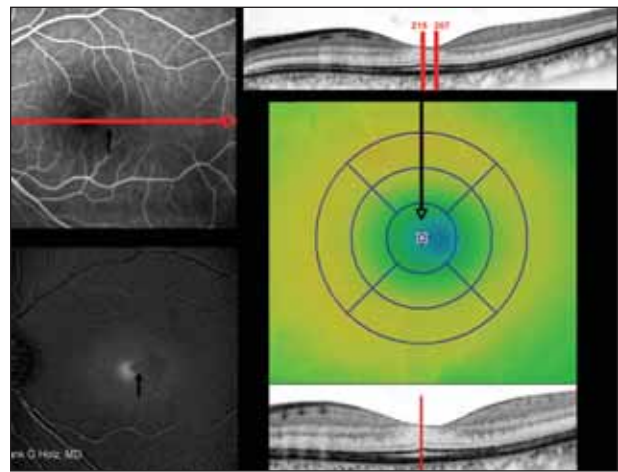


Figure 3. Displacement of the thinnest part of the fovea, suggesting atrophy, loss of cells, or cytoplasmic volume in the neurosensory retina.

CASE 2

In another set of siblings, this time twin brothers, we had similar findings. In Figure 3, there is again displacement of the thinnest part of the fovea, so there must be some atrophy, loss of cells, or cytoplasmic volume in the neurosensory retina. Had we put this automatic volume scan over the deepest pit we would not have picked this up. With the SPECTRALIS, however, the location of the fovea can be moved to where it makes the most sense. The other angiographic features of early type 2 IMT are seen in Figure 4; CBR shows that there is abnormality. In the twin, the angiographic findings show more advanced disease (Figure 5).

DISCUSSION

It is important to diagnose this disease as early as possible. Type 2 IMT is much more common than we previously thought. It is probable that with standard imaging methods we missed a large proportion of patients for whom we could achieve better outcomes



Figure 4. Multi-modality imaging highlights features of early type 2 IMT.

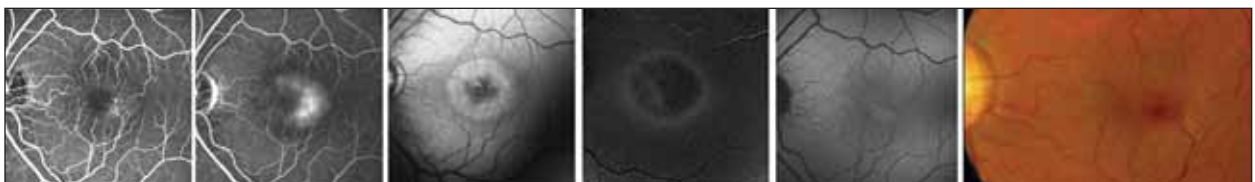


Figure 5. These images show more advanced disease.

had we diagnosed them earlier. In the Spectralis we have three components to improve our diagnostics of this disease. SD-OCT allows us to see thinning temporal to the fovea and FAF aids in the detection of the loss of luteal pigment. Red-free imaging captures increased reflectivity. As result, we can identify the disease earlier before it becomes apparent on FA with the added benefit of avoiding an invasive procedure.

In conclusion, the SPECTRALIS is an important advance in achieving a better understanding of the

genetic components type 2 IMT, both for earlier diagnosis and a better understanding of the disease itself. ■

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Valsalva Retinopathy

BY GIOVANNI STAURENGHI, MD

Valsalva retinopathy is a rare cause of preretinal hemorrhage. Typically, patients present with acute painless loss of vision after performing a Valsalva-type maneuver. Mechanistically, it is thought that an acute rise in intrathoracic/intraabdominal pressure from closure of the glottis leads to an intraocular venous pressure increase and capillary rupture. The patient in this case provided a classic history of straining prior to the onset of symptoms (ie, coughing during asthma attack). Valsalva retinopathy has also been described following weight lifting, vomiting, sexual activity, end stage labor, blowing musical instruments, and compressive injuries.

CASE PRESENTATION

This case is simple, but it is interesting to for the imaging tools used in our clinic. A 40-year-old presented with best-corrected visual acuity of 20/400 in his right eye and 20/20 in his left eye and no other signs or symptoms of significant nature, including normal intraocular pressure in both eyes. The patient had no history of diabetes, hypertension, or any other systemic disease. He was not taking aspirin or any anticoagulant therapy.

His color fundus photo revealed a large preretinal hemorrhage (Figure 1A). His indocyanine green (ICG) angiography (Figure 1C) enabled us to rule out microaneurism, polypoidal choroidal neovascularization, trauma, and choroidal melanoma, leaving us with the diagnosis of valsalva retinopathy.

Valsalva retinopathy can be followed or treated. We chose to use Nd:YAG laser for this patient, which emptied and dislocated the the blood into the vitreous gel 15 minutes after laser. Two days post-laser, the location of the blood is apparent in the spectral-domain optical

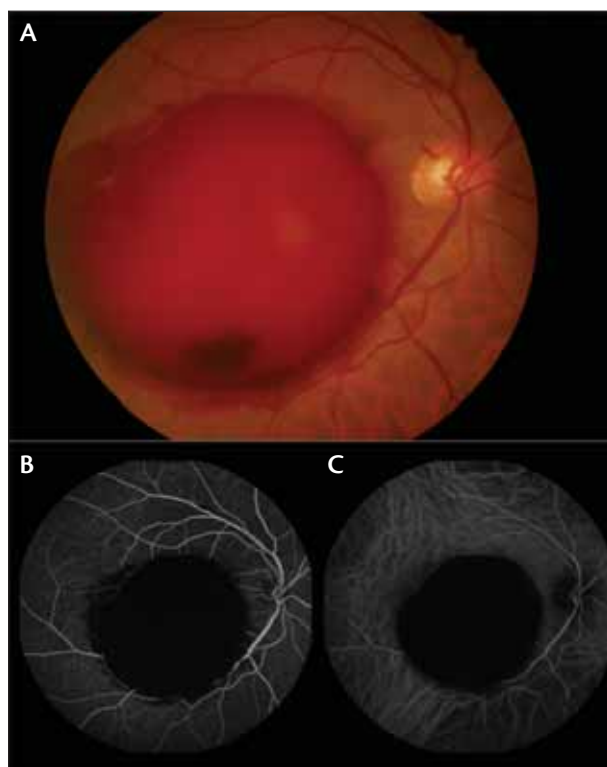


Figure 1. Fundus photography, fluorescein angiography, and ICG of the right eye in a 45-year-old with Valsalva retinopathy show a large premacular hemorrhage involving the posterior pole.

coherence tomography (SD-OCT) on imaging with the SPECTRALIS (Heidelberg Engineering, Heidelberg, Germany). When comparing the fluorescein angiography from the patient from 15 days post-laser progressing to 2 months post-laser. to the SD-OCT scan, the location of the Nd:YAG laser is quite visible in the OCTs

scans. Because the image can be taken at exactly the same point, the SPECTRALIS is useful in following this patient as he improves. ■

Note: The images from this case will appear in a paper that has been recently accepted and that is currently published online: Sabella P, Bottoni F, Staurenghi G. Spectral-domain OCT evaluation of Nd:YAG laser treatment for Valsalva retinopathy. *Graefes Arch Clin Exp Ophthalmol*. 2009 Dec 8. [Epub ahead of print].

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1. Ezra E, Dowler JG, Burgess F, Sehmi K, Hamilton PA. Identifying maculopathy after neodymium: YAG membranotomy for dense diabetic premacular hemorrhage. *Ophthalmology*. 1996;103(10):1568–1574.
2. Raymond LA. Neodymium:YAG laser treatment for hemorrhages under the internal limiting membrane and posterior hyaloid face in the macula. *Ophthalmology*. 1995;102(3):406–411.
3. Shukla D, Naresh KB, Kim R. Optical coherence tomography findings in valsalva retinopathy. *Am J Ophthalmol*. 2005;140(1):134–136.

White Dot Syndrome

BY SRINIVAS SADDA, MD

A 45-year-old woman with scotoma in both eyes that had been enlarging over the course of 1 week was referred to me by her treating ophthalmologist. The patient denied the presence of other symptoms including headaches, tinnitus, transient visual obscurations, and nausea or vomiting. She had a history of hypertension. She was screened by her referring physician for syphilis, Lyme disease, and Bartonellosis with negative results for all. Her ophthalmologist had diagnosed the patient with papilledema.

The patient's visual acuity was 20/20 in the right eye and 20/25 in the left eye and her color vision was intact. The only significant finding was that she had a bit of anterior vitreous cell. The patient's fundus images showed significant disc edema in each eye—more on the right than the left, as well as faint whitish spots deep in the retina in a peripapillary distribution.

The patient's fluorescein angiogram (FA) showed early blocked fluorescence corresponding to the areas of these white dots with some staining in the later phases. FA also showed some disk leakage in both eyes.

One week later, the patient complained of further enlargement of her scotoma and blurring of her vision. Her visual acuity was relatively unchanged. Her visual acuity was relatively unchanged. We did notice a few fine keratic precipitates in her cornea and she had some cell in her anterior chamber. Static perimetry revealed enlargement of blind spots in both eyes.

Fundus exam of the right eye at this visit (now 1 week after first presentation) now demonstrated peripheral white dots (Figure 1) as well as small areas of apparent retinitis—there was whitish change in the retina as well as some hemorrhage. The left eye demonstrated similar peripheral findings. The FA now demonstrated granular areas of hyperfluorescence in the peripapillary regions



Figure 1. White spots are apparent on the patient's right-eye fundus photo.



Figure 2. Hyperfluorescence is seen in the peripapillary regions on FA.

(especially nasally) in each eye (OD, Figure 2). Indocyanine green (ICG) angiography showed hypofluorescence corresponding to these peripapillary lesions (Figure 3).

Spectral-domain optical coherence tomography (SD-

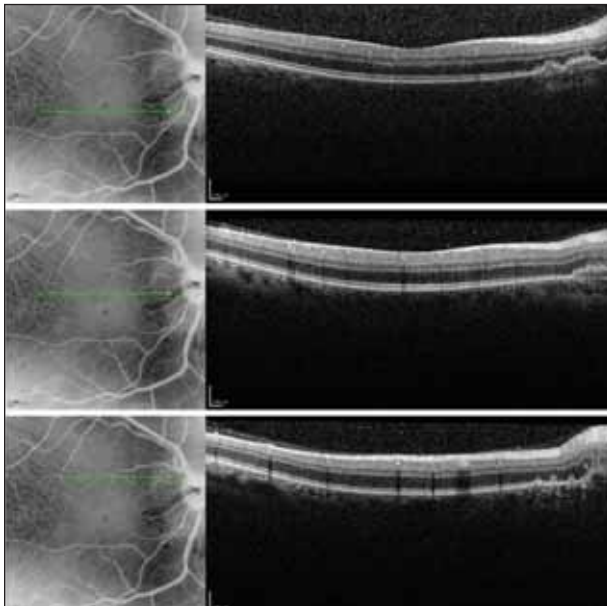


Figure 4. The OCT reveals areas of elevation underneath the RPE layer that corresponded to the peripapillary white-dot lesions on the fundus photos.

OCT) scans from the SPECTRALIS system (Heidelberg Engineering, Heidelberg, Germany) for this patient are shown in Figure 4. The OCT revealed areas of elevation underneath the retinal pigment epithelium (RPE) layer that corresponded to the peripapillary white-dot lesions.

The patient returned 2 weeks later and reported that her vision was improving with the blurriness subsiding and her blind spots becoming smaller. Her fundus images (Figure 5) and OCTs (Figures 6) showed that sub-RPE deposits had nearly disappeared and the disc edema had resolved.

CONCLUSION

We are still unsure as to the precise diagnosis for this patient, although we know that it falls within the spectrum of white dot syndrome. Two specific diagnoses that one might consider are multiple evanescent white dot syndrome (MEWDS) or acute posterior multifocal pigment placoid epitheliopathy (AMPPE). MEWDS, however, is usually unilateral, and there usually is no accumulation of inflammatory material below the RPE, just disruption of the photoreceptor layer. In addition, retinal whitening has not been previously described in patients with MEWDS. The case is also atypical for AMPPE as there was never a typical placoid appearing lesion. However, we did uncover a previous atypical case of AMPPE reported in 1972 by Kirkham et al¹ that described the connection with papillitis, vasculitis, and retinal whitening. Overall, when considering the spectrum of white dot syndromes, this patient seems to fit best somewhere between AMPPE and multifocal choroiditis. Perhaps, as we evaluate this patient over

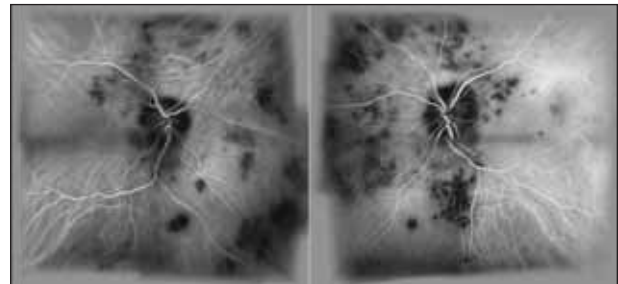


Figure 3. ICG shows corresponding hypofluorescence.



Figure 5. The white dots have largely resolved.

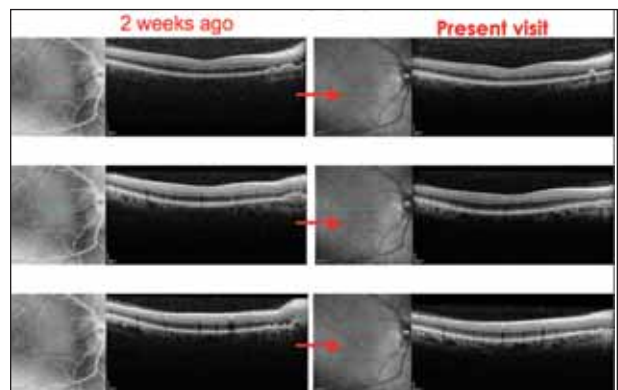


Figure 6. The OCTs show that sub-RPE deposits had nearly disappeared and the disc edema had resolved.

time, a more specific diagnosis will be possible. Regardless, multimodal imaging with the SPECTRALIS—in particular our ability to correlate the angiographic findings with the sub-RPE lesions on OCT—was very helpful in studying the disease course in this patient and distinguishing between diagnoses. ■

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1. Kirkham TH, Flytche TH, Sanders MD. Placoid pigment epitheliopathy with retinal vasculitis and papillitis. *Br J Ophthalmol.* 1972;56:875–880.

Age-related Macular Degeneration Treated With Anti-VEGF

BY KARL CSAKY, MD, PhD

The SPECTRALIS SD-OCT system (Heidelberg Engineering, Heidelberg, Germany) is highly advanced and is particularly useful for identifying challenging diagnoses. It is also a technology that is valuable in day-to-day management practice, even in our more straightforward cases. This case, a 72-year-old man with neovascular age-related macular degeneration (AMD), illustrates this.

The patient's visual acuity in the right eye was counting fingers at four feet for several years and vision in the left eye was 20/50. The patient had been referred to me for suspected retinal pigment epithelial tear in the left eye. The referring doctor had given 15 bevacizumab injections (Avastin, Genentech, Inc.) since January of 2008 and in August of 2009, the patient underwent triple therapy with verteporfin photodynamic therapy, bevacizumab, and triamcinolone acetonide (Kenalog, Bristol-Myers Squibb). The referring doctor saw a change in the subsequent fundus photo and was concerned.

The photos of the patient's eye (Figures 1A and B) show areas of both hypo- and hyperpigmentation. The fluorescein angiograms (FA; Figures 2A and B) show corresponding areas of hyper- and hypofluorescence. The SPECTRALIS SD-OCT image shows a tremendous disruption of the retina with areas of subretinal fluid. Figure 3 shows a comparison of images taken of the same slice of retina with the Stratus OCT (Carl Zeiss Meditec, Jena, Germany) vs the SPECTRALIS showing areas of subretinal fluid. The SPECTRALIS image shows the areas of photoreceptors and the preservation of the retinal layers and inner segment/outer segment junction, explaining why this patient has 20/50 vision in that eye. The Stratus is not able to provide such definition.

DISCUSSION

One of the advantages of the SPECTRALIS is the definition that it provides of the retinal structures. We know that preservation of these layers is a critical predictor of visual acuity. If these structures are intact over a fairly disrupted subretinal space, we want to treat aggressively with anti-VEGF therapy.

Additionally, the images taken with the SPECTRALIS can

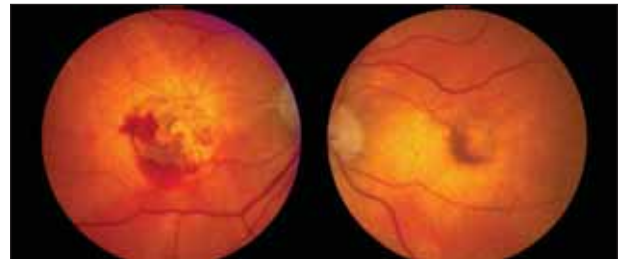


Figure 1. Fundus photos show hyperpigmentation.

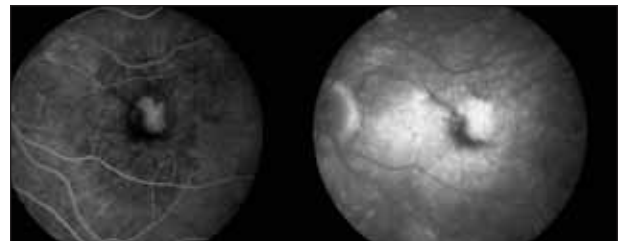


Figure 2. FA shows corresponding areas of hyper- and hypofluorescence.

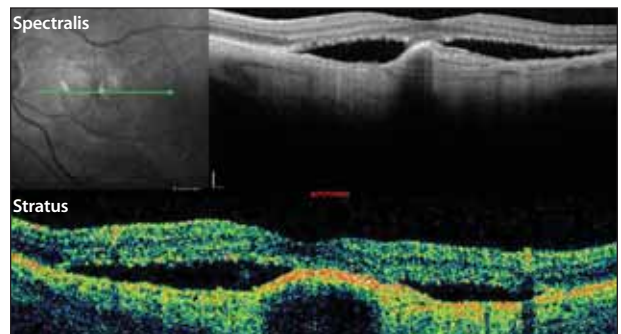


Figure 3. The Spectralis vs Stratus.

be helpful in management of these cases because they can be used in patient education by providing evidence to support continuing therapy after multiple injections. ■

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Type 2 Idiopathic Macular Telangiectasia

BY DIANA V. DO, MD

This case illustrates the value of spectral-domain optical coherence tomography (SD-OCT) for both understanding the structural changes underlying visual loss in type 2 idiopathic macular telangiectasia (IMT).

A 63-year-old woman presented with decreased vision and upon examination, she had perifoveal changes in the retinal pigment epithelial (RPE). Fluorescein angiography (FA) showed bilateral leakage from the retinal vessels. These diagnostic findings resulted in a diagnosis of type 2 IMT.

TD-OCT VS SD-OCT

Time-domain OCT (TD-OCT; Figure 1) of her right eye shows hyporeflective spaces underneath the fovea. SPECTRALIS SD-OCT (Heidelberg Engineering, Heidelberg, Germany) at a similar plane shows these hyporeflective spaces (Figure 2) but also reveals other abnormalities in the area of telangiectasia that are not evident with TD-OCT. Figures 2 and 3 are additional SD-OCT scans from different planes that demonstrate clear differences between the TD- and SD-OCT technologies. In the SPECTRALIS image (Figure 3), the bottom hyper-

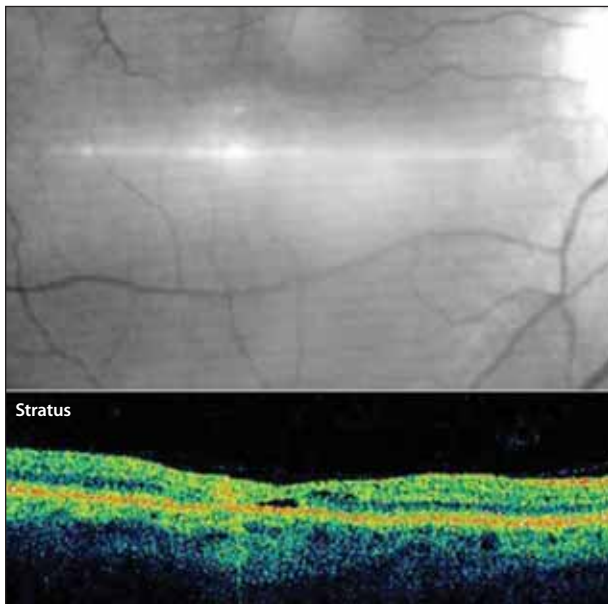


Figure 1. TD-OCT showed hyporeflective spaces underneath the fovea.

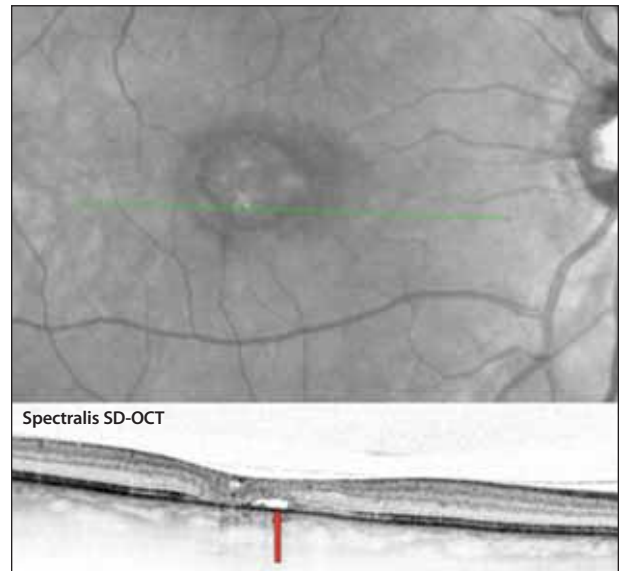


Figure 2. SD-OCT image also showed hyporeflective spaces but it also revealed other abnormalities in the area of telangiectasia that are not evident with TD-OCT.

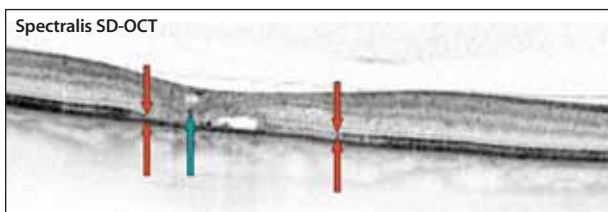


Figure 3. The IS/OS of the photoreceptors, which are represented by the space between the two red arrows, are disrupted in the area of telangiectasia and that most likely accounts for the decrease in visual acuity in the right eye.

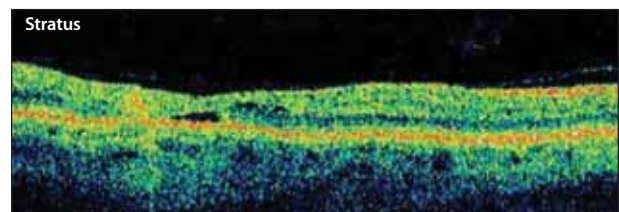


Figure 4. The areas of moderate reflectivity, which could either represent areas of pigment migration or areas of old, inactive, subretinal neovascularization that are seen in Figure 3 are not well distinguished in the TD-OCT.

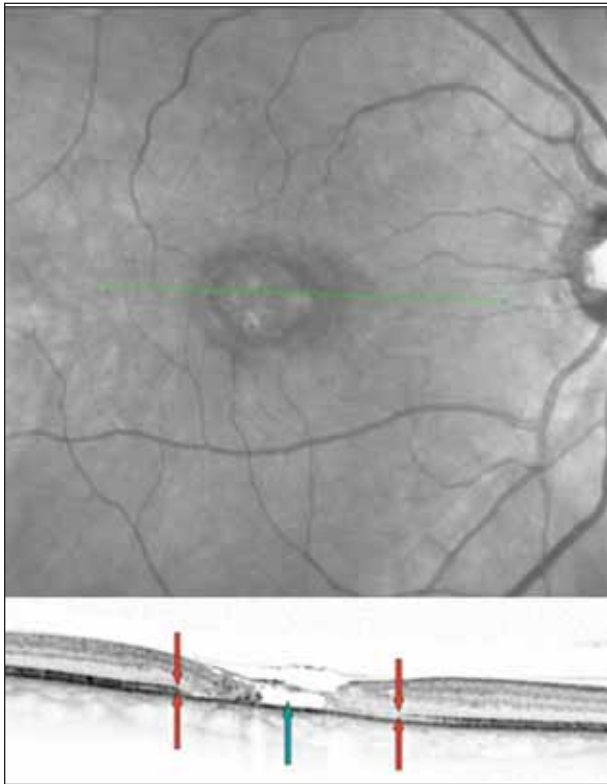


Figure 5. In a more superior horizontal line scan, we continue to see increased areas of cavitations as well as disruption of the photoreceptor layer.

reflective line is the RPE and the line just above it is the intersection between the outer and inner segments of the photoreceptor layer. The inner segments and outer segments of the photoreceptors, the space between the two red arrows (Figure 3), are disrupted in the area of telangiectasia and that most likely accounts for the decrease in visual acuity in the right eye. We also see areas of moderate reflectivity, which could either represent areas of pigment migration or areas of old, inactive, subretinal neovascularization. These layers are not well distinguished in the TD-OCT (Figure 4).

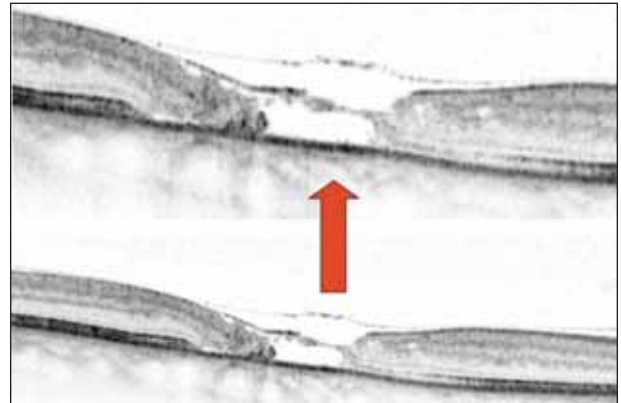


Figure 6. Magnified view of the central area, providing a closer look at the separated vitreous, the hyporeflective structure under the fovea, and the disrupted photoreceptor layer.

In another SPECTRALIS image in a more superior location (Figure 5), we continue to see hyporeflective spaces within the retina as well as disruption of the photoreceptor layer. Figure 6 is a magnified view of the central area, providing a closer look at the separated vitreous, the hyporeflective structure under the fovea, and the disrupted photoreceptor layer.

DISCUSSION

This case clearly illustrates the utility of SD-OCT, particularly in macular diseases where there is disruption of the photoreceptors. Although SD-OCT is not critical in the diagnosis of type 2 IMT, it is necessary in order to understand why the vision loss has occurred and can be helpful in educating patients about the vision loss. ■

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Purtcher's Retinopathy Secondary to Sickle Cell Disease

BY JENNIFER I. LIM, MD

One of the complications of sickle cell disease is retinopathy caused by the deprivation of blood and oxygen due to the inability of sickling red blood cells to access the retinal vasculature.

Our patient, a 19-year-old black woman who had been diagnosed with sickle cell disease at 3 years of age presented with 20/20 vision in both eyes, peripheral neovascularization, and a history of pneumonias in her right eye. The color fundus photographs (Figure 1) show noticeable venous tortuosity, retinal neovascularization, and arterial whitening.

OCT IMAGING

On a SPECTRALIS (Heidelberg Engineering, Heidelberg, Germany) spectral-domain optical coherence tomography (SD-OCT) scan taken superior to the fovea, we saw a slight depression sign (Figure 2). Figure 3 shows the central macula. SD-OCT scans of the inferior portion of the retina show the inner segment/outer segment junction and the retinal pigment epithelium are intact. The inner retina, however, looks as if it has been "crimped" down (Figure 4). The thinning goes throughout the inferior retina and then superiorly through the macular area again with more significant "crimping."

SELECTIVE THINNING OF THE INNER RETINA

My colleagues and I are working on a study for which we have imaged approximately 60 patients with sickle cell retinopathy using the SPECTRALIS SD-OCT. Some of the patients have no clinically visible arterial occlusions or other macular abnormalities, yet they have OCT findings consistent with arterial infarction. Almost all have significant temporal (or nasal) macular thinning. The impetus for this study was that the presumed notion that infarcts, which occur when the red blood cells sickle in the eye, often go unnoticed because the inner retina is most affected, sparing the photoreceptors. The SPECTRALIS is particularly useful in detecting these infarcts. The ischemia that occurs, however, causes irreparable damage to the inner retina, resulting in selective thinning of the inner retina. We have found this to be true compared with our controls. Similar findings have been reported in the literature.¹ ■

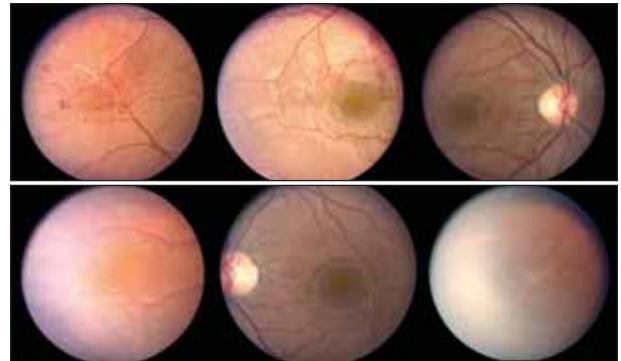


Figure 1. Color fundus photographs show noticeable venous tortuosity, retinal neovascularization, and arterial whitening.



Figure 2. SD-OCT superior to the fovea, a slight depression sign is visible.



Figure 3. SD-OCT of the central macula.

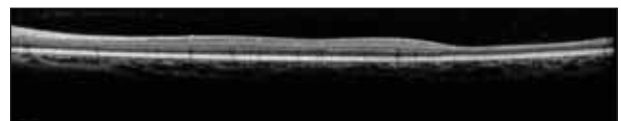


Figure 4. The inner retina appears to have been "crimped" down.

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1. Witkin AJ, Rogers AH, Ko TH, Fujimoto JG, Schuman JS, Duker JS. Optical coherence tomography demonstration of macular infarction in sickle cell retinopathy. *Arch Ophthalmol*. 2006;124:746-747.

Hydroxychloroquine Retinopathy

BY DAVID M. BROWN, MD

A white 49-year-old woman was referred to our office complaining of visual distortion in her right eye present for 2 weeks. Her past medical history included severe Sjogren's syndrome and she was on multiple medications, none of which she brought with her. Her symptoms were loss of central vision and overall blurred vision with visual acuity of 20/25 in the right eye and 20/30 in the left eye; her pupils were normal. Her optometrist and referring ophthalmologist found no abnormalities.

Our initial exam included fundus photography, fluorescein angiography (FA), blue laser fundus autofluorescence (FAF), and spectral-domain optical coherence tomography (SD-OCT) using both the Cirrus HD-OCT system (Carl Zeiss Meditec, Jena, Germany) and SPECTRALIS SD-OCT (Heidelberg Engineering, Heidelberg, Germany).

The fundus photo showed no abnormality and her FA and FAF also appeared normal. The Cirrus shows blunt-

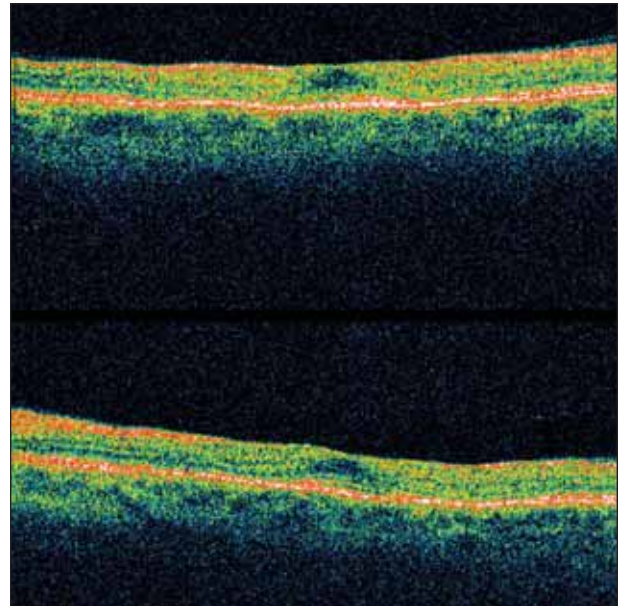


Figure 1. Cirrus OCT shows blunting in the foveal reflex.

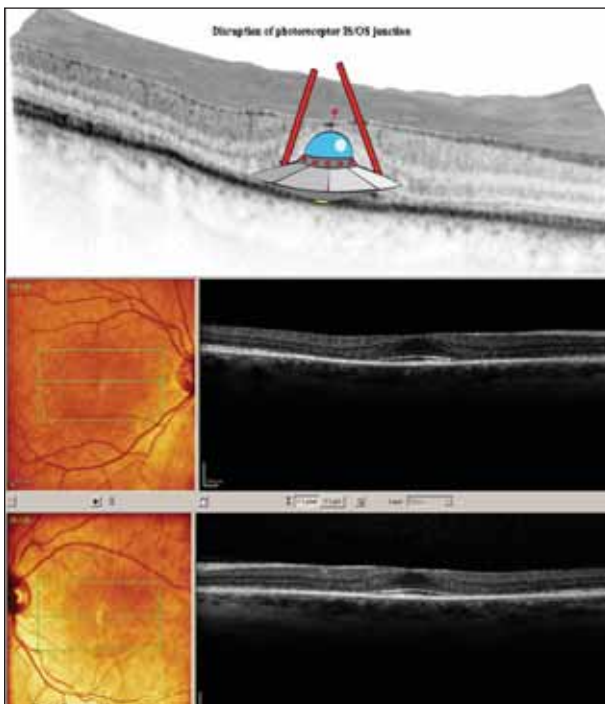


Figure 2. SD-OCT with the SPECTRALIS further delineates a "flying saucer" elliptical shape that causes disruption of the photoreceptors at the IS/OS junction.

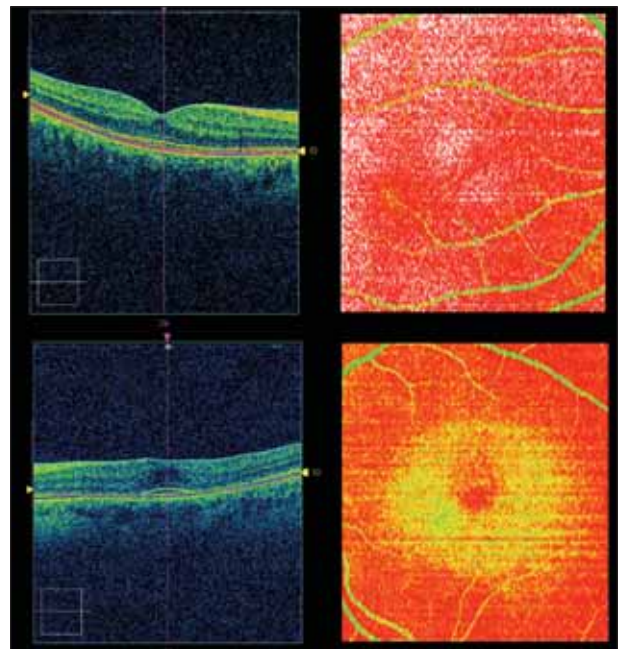


Figure 3. On en-face SD-OCT, an early bulls eye is apparent, suggesting a toxicity to hydroxychloroquine toxicity.

ing in the foveal reflex but does not confirm a diagnosis (Figure 1). The SPECTRALIS SD-OCT image shows a “flying saucer” elliptical shape with disruption of the photoreceptors at the inner segment/outer segment junction (Figure 2). On en face SD-OCT an early bull’s eye is apparent, suggesting hydroxychloroquine toxicity (Figure 3).

DIAGNOSIS

We learned that this patient had been taking hydroxychloroquine sulfate 200 mg (Plaquenil, Sanofi-Aventis) for 8 years. A 10-2 visual field test with the Humphrey Visual

Field Analyzer (Carl Zeiss Meditec) confirmed a toxicity to hydroxychloroquine. In this case, SPECTRALIS SD-OCT provided early detection of hydroxychloroquine toxicity, before any visual acuity loss had occurred. ■

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Artery Occlusion

BY JEFFREY S. HEIER, MD

A 51-year-old man with a 2-year history of disabling vitreous floaters in his left eye was referred to me by one of my practice partners. The patient had been carefully followed with documentation and had persistent hyperplastic primary vitreous (PHPV) and amblyopia in his right eye with 20/80 vision. He was 20/20 in his left eye, but the floaters were impairing his

ability to work, so he underwent 23-gauge pars plana vitrectomy with retrobulbar block in his left eye. There were no complications during surgery, but 6 hours post-operative the patient reported the onset of photopsias. He described his symptoms as fireworks going off in his eye. The patient reported no pain, no coughing, no headache, and no sneezing. His vision in the left eye was reduced to hand motion and his intraocular pressure measured 10 mm Hg. The color fundus photo is shown in Figure 1. Figure 2 is the postoperative fluorescein

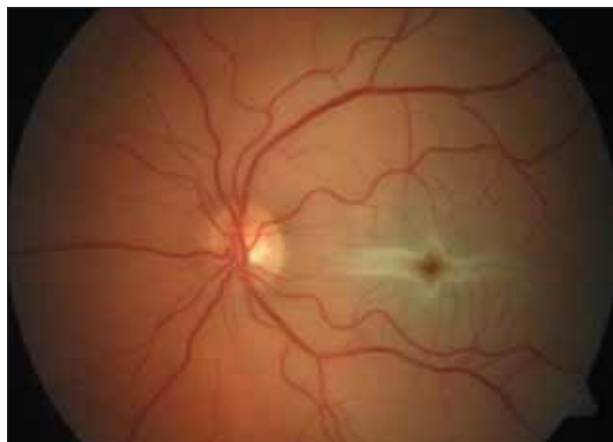


Figure 1. Color fundus photo.

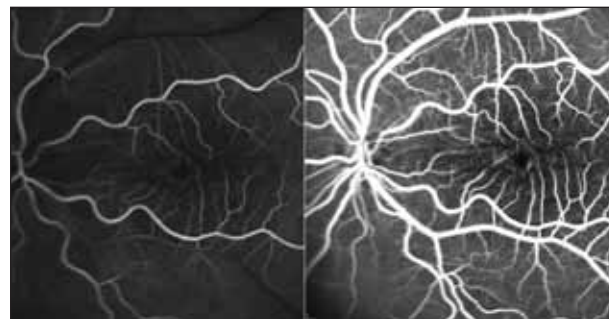


Figure 2. FA shows normal filling time, suggesting that the vessels may be slightly dilated throughout the eye.

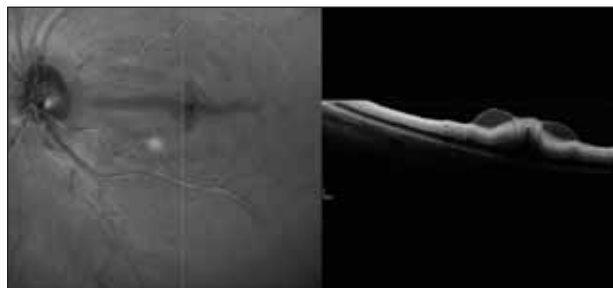


Figure 3. Folding in the macula is clearly seen on SD-OCT with Spectralis.

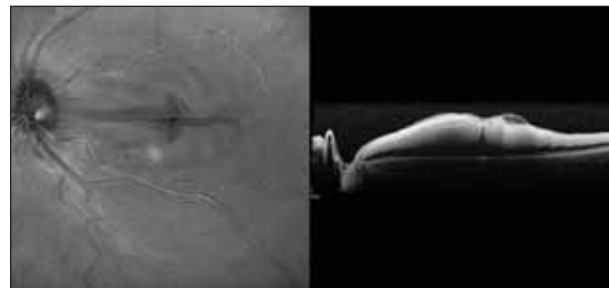


Figure 4. Folding in the macula as seen with Spectralis SD-OCT.

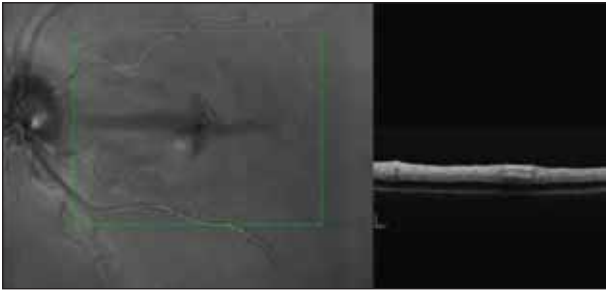


Figure 5. Cube scan from the volume scan did not observe any evidence of VMT.



Figure 7. ICG images showed also showed no change.

angiography (FA), which shows normal filling time, suggesting that the vessels may be slightly dilated throughout the eye. Additionally, there is evidence of folding in the macula. Upon reviewing the patient's spectral-domain optical coherence tomography (SD-OCT; Figures 3 and 4) taken with the SPECTRALIS SD-OCT (Heidelberg Engineering, Heidelberg, Germany), we clearly saw folding of the macula.

Initially, we thought that this could be an unusual case of vitreomacular traction (VMT) where it appeared that there was tenting of the retina. We evaluated the vitreomacular interface of the volume scan using the vitreous option in the software (Figure 5) and did not observe any evidence of VMT. It was then suggested that this patient

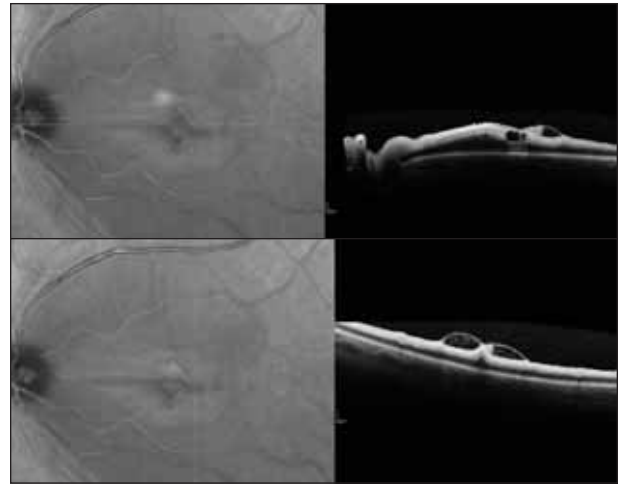


Figure 6. At day 5 postoperative, the OCT images showed no change and the patient's vision continued to be hand motion.

might have hypotony maculopathy, but what I thought was key to making a diagnosis was the whitening of the retina throughout.

At day 5 postoperative, the OCT (Figure 6) and indocyanine green (Figure 7) images showed no change and the patient's vision continued to be hand motion. We determined that the patient may have suffered an artery occlusion the night after surgery. We recently saw this patient back for follow-up and the eye appears normal with an ischemic nerve and sclerotic vessels. ■

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