THE **RETINA** PARTNERS

Encino Santa Monica Valencia Thousand Oaks Century City

(877) 3-RETINA

Case of the Month – October 2019

Presented by Christian Sanfilippo, MD

A 63-year-old male was referred to clinic for possible retinal detachment in the right eye noted on routine eye examination. The patient had no visual complaints. Visual acuity measured 20/20 in both eyes. Visual field testing with confrontation revealed a mild nasal visual field defect in the right eye, and was full in the left eye. Intraocular pressures were normal. His anterior segment examination was unremarkable including the anterior vitreous, which was free of pigment in both eyes. Wide field color fundus photographs are shown below.

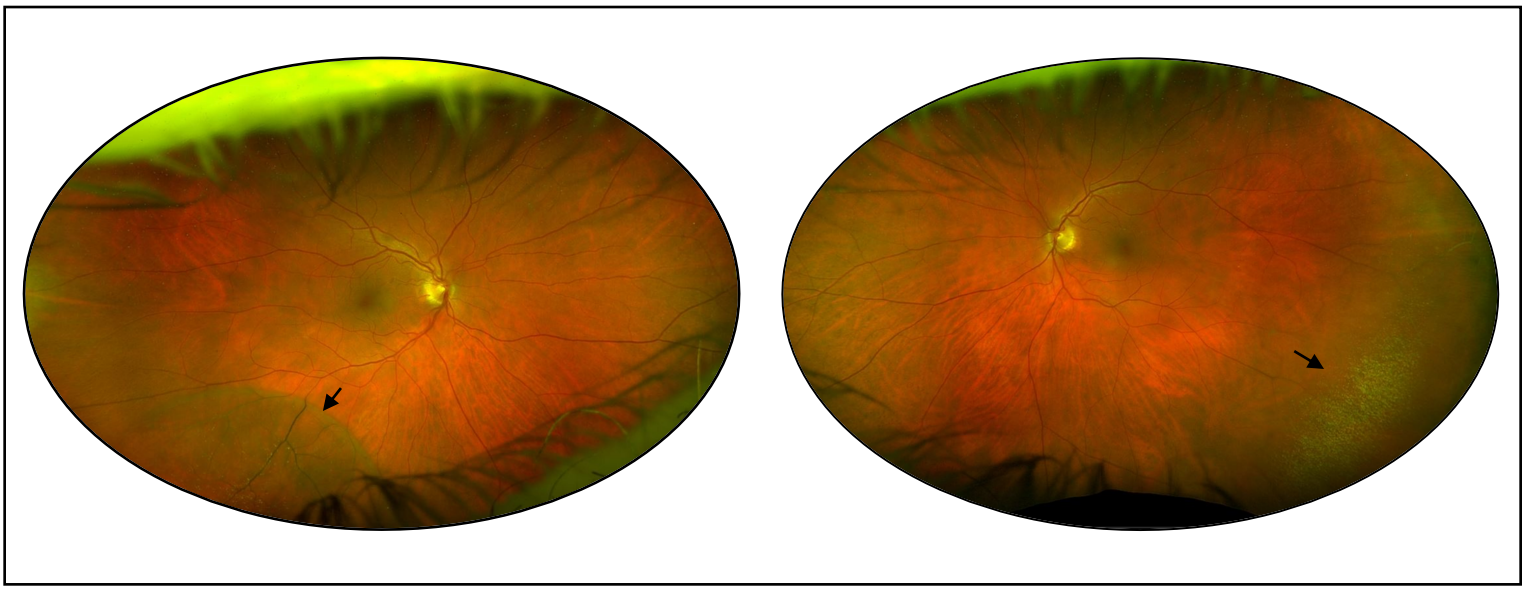

Figure 1: A. Optos color fundus photo of the right eye shows a well demarcated, round area of retinal elevation (arrow). **B.** Optos color fundus photo of the left eye shows a similar, but less well demarcated area inferotemporal with overlying whitish spots consistent with snowflake degeneration.

A thorough scleral depressed examination was performed with the indirect ophthalmoscope. No retinal breaks were noted in either eye. Additional imaging with SD-OCT was performed through the lesion in the right eye and is shown below.

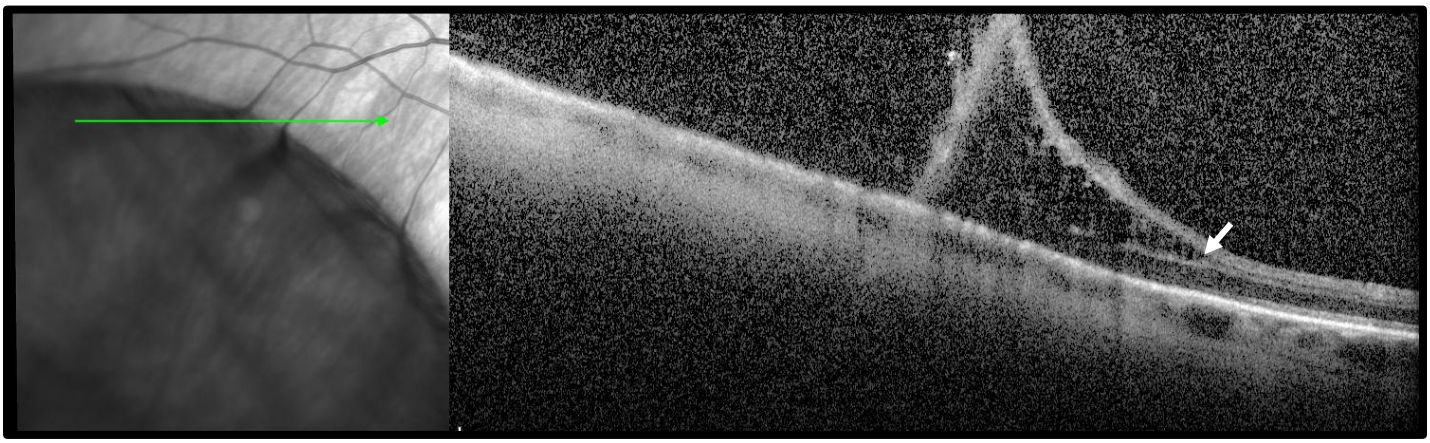

Figure 2: SD-OCT line scan through the inferotemporal lesion shows splitting of the retina at the outer plexiform layer (arrow). There is no subretinal fluid.

Differential Diagnosis: Degenerative retinoschisis, rhegmatogenous retinal detachment, retinoschisis-retinal detachment

Clinical Course:
The patient was diagnosed with degenerative retinoschisis on the basis of examination and SD-OCT findings. Although the posterior edge of the schisis cavity was noted to extend posterior to the equator, he was completely asymptomatic. Observation with serial examination was recommended. Follow up imaging 1 year after initial presentation is shown below.

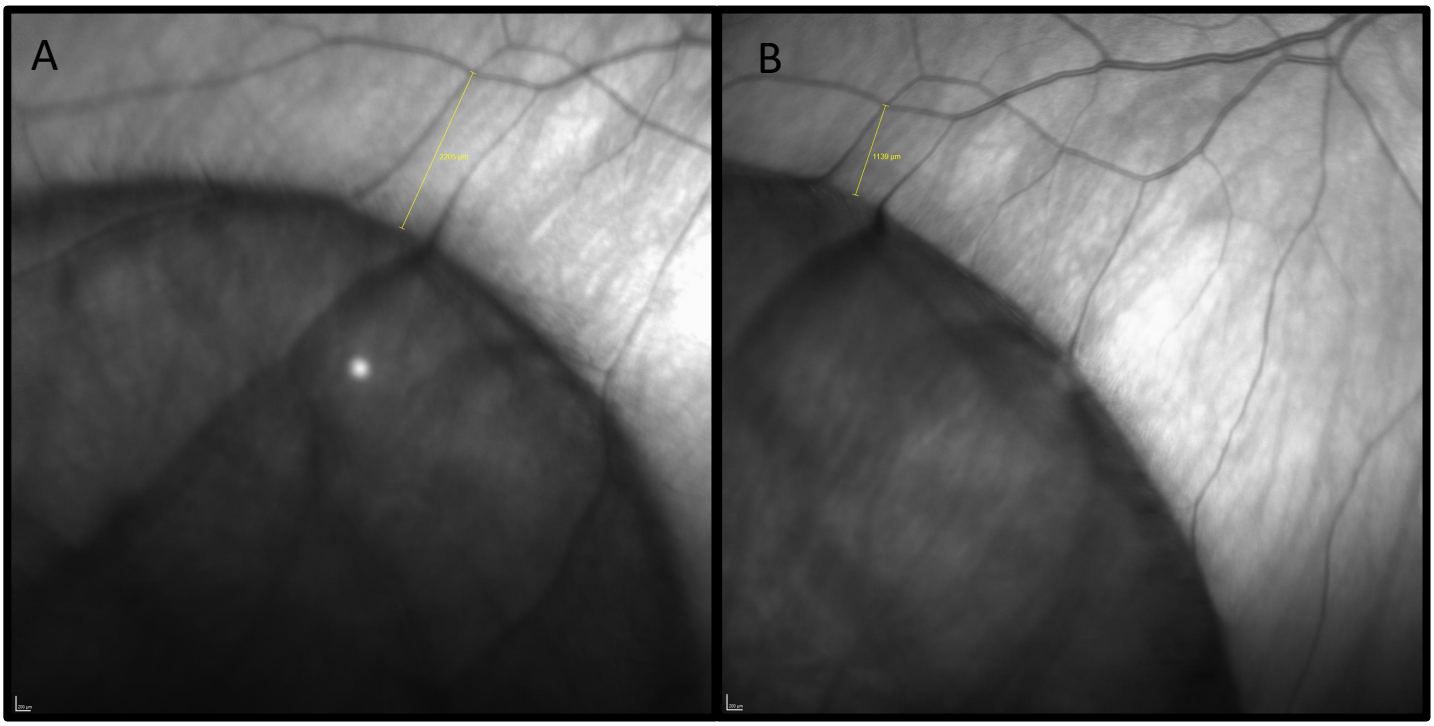

Figure 3: Near infrared imaging of the retinoschisis cavity at initial presentation (A) compared to 1 year follow up (B) shows posterior progression of the retinoschisis cavity as measured from an AV crossing used as a landmark (2205 um vs. 1138 um).

Despite the unusual posterior progression, the patient remained asymptomatic. A discussion was held regarding the best course of management which included laser retinopexy demarcation vs. observation. Because of a lack of data to support any intervention, observation was again elected with shorter interval follow up.

Discussion:

Degenerative retinoschisis is a relatively common, typically asymptomatic and benign finding in the peripheral retina defined as the splitting of the retinal layers. It occurs in up to 4% of individuals over the age of 50 years old, is most often bilateral, has no sex predilection, and is more commonly found in hyperopic eyes. Although the natural history is nearly always benign, retinoschisis may present a challenge to the clinician who must differentiate it from retinal detachment.

Clinically, patients are most always asymptomatic, however formal visual field testing will show an absolute scotoma corresponding to the schisis cavity. Examination will show a well demarcated, dome shaped retinal elevation, most commonly occurring in the inferotemporal quadrant. While, bullous retinoschisis is generally easy to recognize, shallow cavities can be more subtle and more difficult to differentiate from chronic, asymptomatic retinal detachment. Scleral depression can be very helpful in this circumstance. In the former, the entire dome shaped schisis cavity will be displaced equally by the scleral depressor, while in the latter, subretinal fluid is locally displaced leading to flattening of the elevated cavity directly over the depressor. Shallow retinoschisis may also present with whitish dots overlying the cavity commonly referred to as "snowflake degeneration" (Figure 1B, Arrow). These dots are thought to represent degenerating Muller cell footplates. Even with these clues in mind, however, shallow retinoschisis can sometimes be nearly impossible to differentiate clinically from a shallow peripheral retinal detachment. In these cases, OCT imaging can be invaluable. As seen in our patient, OCT line scans obtained through the edge of the schisis cavity can definitively differentiate between retinal detachment and retinoschisis (Figure 2).

Histologically, degenerative retinoschisis shares many similarities with the more commonly seen peripheral cystoid degeneration. Peripheral cystoid degeneration describes the histological findings of cystoid spaces found in the far anterior retina which are present in nearly all adults. These spaces have little to no clinical significance, but can sometimes be observed with scleral depressed examination, or in histology slides. In its most common form termed "typical" cystoid degeneration, cysts occur within the outer plexiform layer. Less commonly, cystoid spaces may be seen within the nerve fiber layer, in which case it is termed "reticular" cystoid degeneration. Although the exact pathogenesis of degenerative retinoschisis remains uncertain, it may be the result of coalescence of the cystoid spaces of cystoid degeneration into a single cavity. Like its more common relative, degenerative retinoschisis is also subdivided between the more common "senile" form, again occurring within the outer plexiform layer, and the less common "reticular" form, occurring within the nerve fiber layer. OCT imaging of our patient nicely demonstrates senile retinoschisis with clear splitting through the outer plexiform layer (Figure 2).

Despite the generally excellent prognosis, there are a few complications that may occur in retinoschisis patients. Breaks within the inner wall or outer wall of the schisis cavity are not infrequent. Inner wall breaks, are usually small and difficult to see clinically. Luckily, these breaks are most often clinically insignificant as long as they occur in isolation. This is because on their own, they do not provide a path for fluid to access the subretinal space. In contrast, outer wall breaks are typically larger, more posterior and easier to see clinically. They occur in 11-24% of schisis cases. Outer wall breaks do provide a potential pathway for schisis fluid to accumulate under the retina, a complication termed "schisis-detachment". While over 50% of schisis patients with outer retinal breaks may develop localized schisis-detachment, they nearly always remain asymptomatic and do not require intervention. Progression of schisis-detachment is very slow and usually not clinically significant, probably because schisis fluid is very viscous and therefore does not easily track under the retina. Very rarely, both an inner wall and an outer wall break can occur together. This combination provides a potential pathway for liquified vitreous to travel through the schisis cavity and into the subretinal space. This rare complication called "retinal detachment associated with retinoschisis" is estimated to occur in only 0.05% of all schisis cases. However, like retinal detachment, it can be vision threatening and most often does require surgical intervention with primary scleral buckle, vitrectomy or both.

In the absence of the above infrequent complications, degenerative retinoschisis is rarely progressive or vision threatening. In the longest and largest natural history study of degenerative retinoschisis, Dr. Norman

Byers observed more than 200 cases over a 9 year period. Importantly, only 3% of cases were reported to progress posteriorly over this time period, and none required treatment of any kind. This is what makes our case rather unusual. Serial imaging taken at a 1 year interval clearly demonstrates posterior progression (Figure 3). Fortunately, the cavity remains completely outside of the macula, and the patient is still asymptomatic with 20/20 acuity. In the rare case when posterior progression threatens the central macula, several treatment strategies have been proposed. These include laser demarcation, cryotherapy demarcation, scleral buckling with external surgical drainage and vitrectomy with internal drainage of the schisis cavity. However, no treatment has been shown to definitively halt the progression of the schisis cavity. Therefore, with rare exception, most patients are best served with observation. After a detailed discussion of the different treatments available to our patient, we opted to continue with observation with the option to intervene in the future if the macula becomes involved.

Take Home Points

- Degenerative retinoschisis is a relatively common peripheral retinal finding which is typically asymptomatic, non-progressive and most often does not require treatment.
- Differentiating retinoschisis from shallow retinal detachment is important and can be difficult. OCT imaging is a valuable tool which reliably can differentiate these two entities.
- Rare complications associated with outer wall breaks can lead to two types of combinations of retinoschisis and retinal detachment. Typically, treatment is only necessary when both inner and outer breaks are present.

Thomas Hanscom Robert Engstrom Hajir Dadgostar Amir Guerami Christian Sanfilippo Stavros Moysidis