

Acta Ophthalmologica

The Official Journal of the Nordic Ophthalmological Societies
The European Association for Vision and Eye Research



Diagnosis/Therapy in Ophthalmology

The misdiagnosis of retinoschisis

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Acta Ophthalmol. 2012; 90: 492–494

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doi: 10.1111/j.1755-3768.2011.02301.x

Senile retinoschisis is a well-recognized clinical entity, which was first described by Bartels (1933). In the majority of cases, it is asymptomatic, infero-temporal, nonprogressive and does not require treatment unless complicated by an outer layer detachment (Byer 1968, 1986). Confusion of the retinoschisis elevation with rhegmatogenous retinal detachment does occur and results in unnecessary laser or surgical intervention. We present four cases in which the diagnosis was in dispute and define characteristics that made for the diagnosis of retinoschisis.

Case #1: A 26-year-old male was referred to the retina service after three retina specialists had diagnosed bilateral retinal detachment and recommended urgent surgery. He was asymptomatic. On examination, his visual acuity was 20/20 with spherical equivalent corrections of -12.00 in both eyes. Funduscopic examination revealed bilateral 360° peripheral retinal elevations anterior to the equator and extending to the ora serrata. In the right eye, there were horseshoe retinal tears at 1:30, 5:30 and 7:00 o'clock near the posterior edge of the retinal elevation (Fig. 1, left). In the left eye, seven horseshoe tears were found near the posterior edge of the elevations at 12:30, 3:30, 4:00, 5:30, 7:00 and 11:00 o'clock (Fig. 1, right). The patient was diagnosed as having retinoschisis with multiple inner layer breaks in both eyes. He was taught how to monitor his peripheral visual field and asked to report any change

without delay. Examination 11 and 22 months later revealed no change in the extent of the retinal elevations, and no pigment lines had developed at the posterior border of the retinal elevations. His visual acuity remained 20/20 in both eyes.

A 360° of retinoschisis in the periphery is not exceptional. The presence of retinal breaks provoked some concern, but considering that the shape of the retinal elevations did not relate to the breaks, did not progress and did not cause a pigmentation line to form over 22 months, reassured us that the breaks were in the inner layers of peripheral schisis.

Case #2: A 35-year-old female was referred to the retina service in October 1989 for treatment of a retinal detachment in her left eye. Her corrected visual acuity with -9.00 in both eyes was 20/20. The fundus examination revealed bilateral peripheral infero-temporal retinal elevations anterior to the equator. There were multiple holes in the elevation in the left eye (Fig. 2, left). Bilateral retinoschisis was diagnosed, and the patient was advised upon how to monitor her visual field and asked to report any changes without delay.

Five years later in June 1994, the patient presented complaining of a shadow developing supero-nasally in the visual field of the left eye. Fundus examination revealed an infero-temporal retinal detachment extending posterior to the schisis elevation and to within a disc diameter of the macula.

The detachment emanated from an outer layer break that had developed at the inferior edge of the temporal retinoschisis (Fig. 2, right). A local buckle procedure with a silicone sponge applied to the outer layer break reattached the retina without drainage of the subretinal fluid. The schisis elevation remained unchanged.

Case #3: A 46-year-old female first came to the retina clinic in February 1999 looking for a second opinion. She had undergone surgery for 'retinal detachment' in the left eye 4 months before and was told that it was not successful. She did not have any visual symptoms and her corrected visual acuity with -7.50 spheres in both eyes was 20/20. Examination of the right eye revealed an inferior bullous retinal elevation extending posteriorly to within two disc diameters of the macula. There was a round hole at 6 o'clock anterior to the equator. A laser test through the hole was positive for retinoschisis (Fig. 3, left). Examination of the left eye revealed an infero-temporal buckle covered with coagulation scars and with retinoschisis on the internal surface that extended beyond the posterior edge of the buckle (Fig. 3, right). No additional surgery was recommended, and the patient was instructed upon how to monitor her visual field.

Eight years after the initial presentation, the retinoschisis posterior to the buckle in the left eye was unchanged. The right eye developed

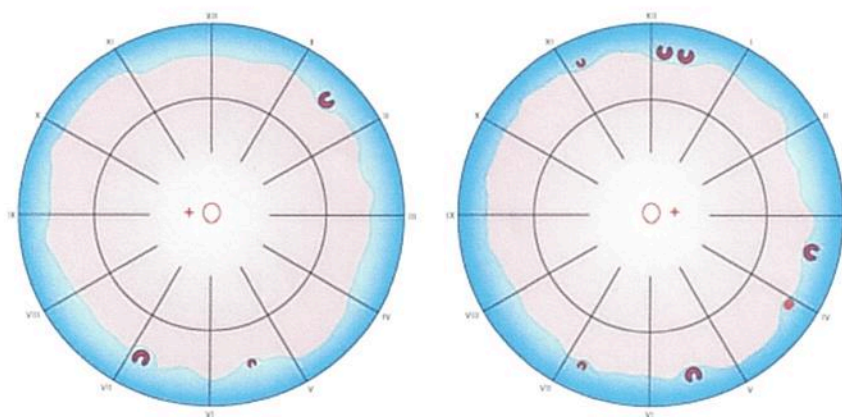


Fig. 1. Case 1: Bilateral peripheral retinoschisis with three tears in the right eye (left) and seven tears in the left eye (right).

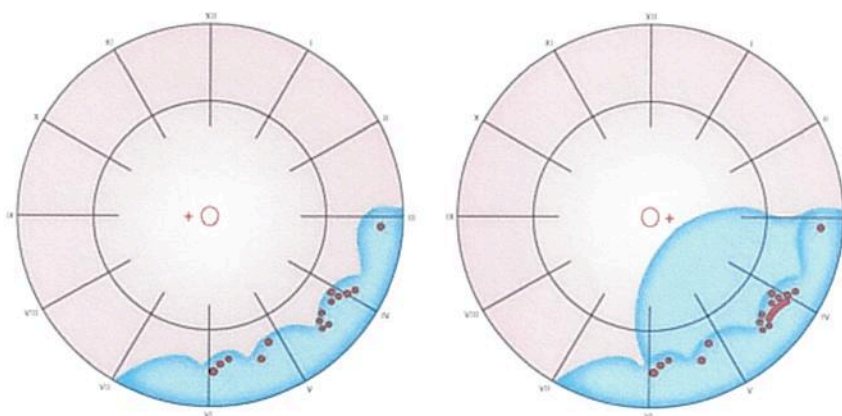


Fig. 2. Case 2: Infero-temporal retinoschisis with multiple holes in the inner layers (left) and a retinal detachment secondary to a large tear in outer layers that occurred 5 years later (right).

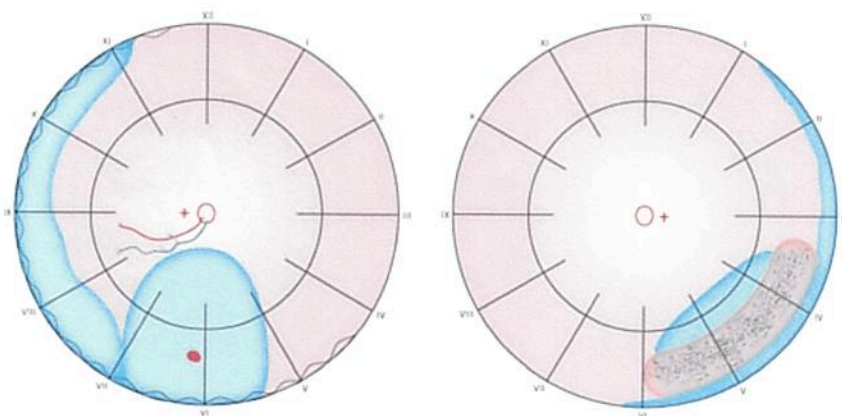


Fig. 3. Case 3: Inferior schisis with a hole at 6 o'clock (left) and retinoschisis persisting anteriorly and posteriorly to a scleral buckle (right).

some pigment dysplasia at the posterior edge of the schisis. An OCT of the posterior edge revealed some pigment epithelial dystrophy. The visual acuity continued to be 20/20 in both eyes.

Case #4: A 26-year-old asymptomatic female came to the retina service

on a day prior to a scheduled retinal surgery elsewhere looking for a second opinion. A retina specialist 2 days before had diagnosed a retinal detachment in her right eye. Her mother and an aunt had undergone surgery for repair of retinal detachment. The cor-

rected visual acuity with -9.50 and -8.75 spherical equivalent corrections was 20/20 in both eyes. Fundus examination of the right eye revealed an infero-temporal retinal elevation that extended posteriorly beyond the equator with a round hole anterior to the equator near 6 o'clock (Fig. 4, left). The temporal extent of the elevation appeared excessive in relation to the position of the retinal hole for a rhegmatogenous detachment. A laser application through the hole gave a positive response in the outer layers (Fig. 4, right). An examination of the retina in the left eye revealed retinal schisis in the inferior periphery. Bilateral retinoschisis was diagnosed, and the patient was advised upon how to monitor her visual field and asked to report new visual symptoms without delay. The patient has been followed for 18 years with no change except for some pigment epithelial dysplasia at the posterior edge of the schisis cavity in the right eye. The vision remains 20/20.

The differentiation of senile retinoschisis from rhegmatogenous retinal detachment is usually not difficult. In an effort to avoid misdiagnosis and unnecessary surgery of retinoschisis, the retina specialist should take into account the semi transparency, the shallowness of the elevation and the absence of a gravity component. Retinoschisis is asymptomatic as it was on presentation in all of our cases. There is no history of flashes or spots or the development of a shadow. It is useful to demonstrate the extent of the deficit in the visual field caused by the schisis to the patient. Patient #2 had no symptoms on the initial visit, but 5 years later when she complained of a shadow developing beyond her schisis defect, it signalled the development of a retinal detachment emanating from an outer layer break in the area of schisis.

Shape: retinoschisis is most frequently found in the infero-temporal periphery. Retinal breaks are infrequent and usually in a position unrelated to the shape of the retinal elevation. The occurrence of horseshoe tears in the peripheral retinoschisis of the first patient is unique and prompted three retinal surgeons to recommend surgical repair. The absence of symptoms, the shape of the retinal elevation, which conformed to

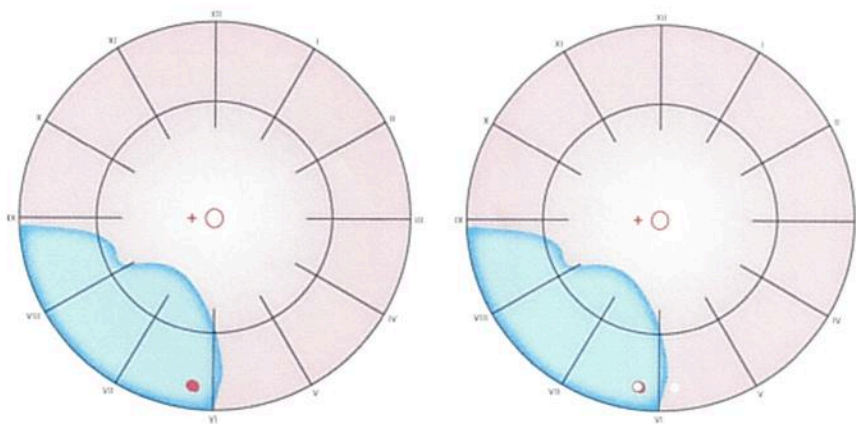


Fig. 4. Case 4: Temporal retinoschisis with a hole at 6 o'clock unchanged over 18 years (left). A laser test spot applied with minimum intensity adjacent to the schisis and with the same intensity through the retinal hole yielded a similar response, confirming the presence of attached outer layers (right).

peripheral schisis and was unchanging, and the failure of pigment lines to develop over time confirmed that the breaks were only in the inner layers of peripheral retinoschisis.

Laser test and OCT: the shape of the retinal elevations in cases 3 and 4 was inconsistent with the position of the round holes for rhegmatogenous retinal detachment. A laser application through the breaks gave a positive response in the outer layers similar to a test spot in adjacent attached retina and was an additional diagnostic test that confirmed schisis (Lincoff et al. 2003).

In the infrequent case of the retinoschisis that approaches the posterior

pole, an OCT can define retinoschisis, i.e. the presence of attached outer layers of the retina and elevated inner layers as in case #3.

Peripheral retinoschisis with inner layer break(s) is asymptomatic and does not develop pigment demarcation lines over time. A laser test through a break gives a positive response in the outer retinal layer and is diagnostic. However, a retinoschisis patient should be advised to test the visual field regularly and report the development of a shadow beyond the schisis defect.

Acknowledgements

This study was supported by The Edward L. Grayson Retinal Research

Fund, West Orange, New Jersey; The Samuel I. Newhouse Foundation, New York, New York; The Kuri Family, Mexico City, Mexico.

The authors have no proprietary interest in this study.

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Received on September 13th, 2011.

Accepted on September 20th, 2011.

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