Letter to the Editor

Primary retinoschisis with vascular changes mimicking neovascularization, illustrated with multimodal imaging

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Primary retinoschisis is a common peripheral retinal disease with prevalence ranging from 4 to 7%.(1, 2) Despite the acceptance of its alternative name “senile retinoschisis”, it is not senile or age-related as it has been reported in young patients. It is characterized clinically by a smooth, bullous retinal elevation often associated with inner and outer retinal holes. In addition, primary retinoschisis presents histologically as gradual splitting of the neurosensory retina at the outer plexiform layer, or less commonly at the inner nuclear layer. The diagnosis can be challenging for cases in which the typical features are absent. Here we report two cases of primary retinoschisis presenting with predominantly retinal vascular changes. By illustrating these cases with multimodal imaging, we hope to help clinicians identify cases of atypical retinoschisis earlier, thereby avoiding unnecessary investigations.

In case 1, a 35-year-old female from Cambodia was referred from an optometrist with an incidental finding of presumed retinal neovascularization in the left eye. The patient was asymptomatic. Her medical history was unremarkable, notably there was no history of diabetes or vasculitis. She had no family history of inherited eye disease. The patient was recently cleared of tuberculosis from the immigration department based on a Mantoux test and chest Xray.

On examination, her best corrected visual acuity was 6/9 in the right eye and 6/6 in the left with normal intraocular pressures. As shown in Figure 1, she had fine tortuous, telangiectatic retinal vessels in the superior nasal quadrants. There was no inner or outer retinal hole or crystal present. The retinal vessels were noted to be raised slightly. There was also no sign of intraocular inflammation or vascular sheathing.

The patient underwent extensive investigations with the provisional diagnosis of retinal neovascularization. Branch retinal vein occlusion was excluded because the anomalies were not in the distribution of a venous territory. The vasculitis screen was negative and included the following tests: antinuclear antibody, anti-double stranded DNA, anticardiolipin antibody, angiotensin-converting enzyme, erythrocyte sedimentation rate, c-reactive protein, rheumatoid factor, anti-cyclic citrullinated peptide antibody and syphilis serology. Her fasting glucose and lipids were normal.

The multimodal imaging performed included fundus photos from California ultra-wide field retinal imaging (Optos, UK), optical coherence tomography angiography (OCT-A) with Cirrus 5000 AngioPlex OCT angiography (Carl Zeiss Meditec, Germany) and fluorescein angiogram (FFA) with TRC-50DX (Topcon, Japan). The diagnosis was apparent when OCT-A showed the classic retinoschitic appearance on en-face imaging (Figure 1). The patient had an anaphylactic reaction to fluorescein and consequently limited views of FFA were obtained. Leakage of the telangiectasic vessels was present over the dome of the retinoschisis. No capillary non-perfusion was detected although the area overlying the schisis was partially defocused (Figure 1).

In case 2, a 31 years old Caucasian man was found to have abnormal retinal vasculature by his optometrist on a routine consultation. Ocular and medical histories were unremarkable. Family, social and travel history were also noncontributory.

Examination revealed a best corrected visual acuity of 6/6 in both eyes. Dilated fundal examination revealed an area of retinal aneurysmal dilatation along the inferior arcade, of about 1-1½ disc diameters in size (Figure 2). No sign of intraocular inflammation

was seen. OCT (Sepectralis by Heidelberg Engineering**,** Germany) was performed and revealed a localized retinoschisis(Figure 3).

Here we report two cases of primary retinoschisis which the main feature was telangiectatic retinal vessels. The first case mimicked the appearance of retinal neovascularization, but this diagnosis was excluded on OCT-A in the absence of capillary drop-out.

Although, abnormal vascular features have previously been associated with X-linked retinoschisis(3) and primary retinoschsis(4), this is the first time where the vascular changes associated with primary retinoschisis were documented using multimodal imaging. Our case series disproved that the vascular abnormality was due to neovascularization, as OCT-A for Case 1 did not show capillary non-perfusion around the areas of vascular elements identifiable on the OCT B-scan image. FFA demonstrated leakage only from some abnormal vessels at the height of the retinoschisis, where the vessels were under maximal tension. The vessels which mimicked the appearance of neovascularization did not leak.

Limitations include the retrospective nature of this case series and the limited number of patients. We did not perform FFA and OCT-A for Case 2, as further investigations would not have benefited the patient after the diagnosis became apparent.

Campos in his case series reported the vascular abnormality were neovascularization, based on leaking of the vessels and patchy capillary fillings on fluorescein angiogram.(4) We propose that some of the vascular changes associated with retinoschisis may not be neovascularization secondary to ischaemia. Therefore, those

cases do not need sectoral scatter laser. OCT-A was the key to ascertain capillary non-perfusion. FFA can give equivocal results as stretched vessels also leak and capillary non-perfusion of the raised peripheral retina is difficult to interpret.

Acquired retinoschisis can atypically present with vascular changes. OCT-A is a non-invasive and potentially useful ancillary test in challenging cases. It differentiates anomalous vessels due to traction from neovascularization. We recommend that retinoschisis be included in the differential diagnosis when retinal telangiectasia is found with no apparent cause. We also recommend consideration of OCT-A, where available, as a useful diagnostic modality prior to any invasive investigations.

REFERENCES

1. Buch H, Vinding T, Nielsen NV. Prevalence and long-term natural course of retinoschisis among elderly individuals: the Copenhagen City Eye Study. Ophthalmology. 2007;114(4):751-5.
2. Byer NE. Long-term natural history study of senile retinoschisis with implications for management. Ophthalmology. 1986;93(9):1127-37.
3. Tantri A, Vrabec TR, Cu-Unjieng A, Frost A, Annesley WH, Jr., Donoso LA. X-linked retinoschisis: a clinical and molecular genetic review. Surv Ophthalmol. 2004;49(2):214-30.
4. Campo RV, Reeser FH, Flindall RJ. Vascular leakage, neovascularization, and vitreous hemorrhage in senile bullous retinoschisis. American journal of ophthalmology. 1983;95(6):826-32.

FIGURE LEGENDS

Figure 1: Top. Wide-field retinal photo showing vascular changes associated with retinoschisis. **Middle left**. OCT-A en-face false colour rendering showed no capillary drop out. **Middle right**. OCT-A cross-sectional scan with retinoschisis changes. Vascular elements of the inner retina, as detected by movement from red blood cells, are highlighted in red. **Bottom**. FFA with leak over the dome of retinoschisis.

Figure 2: Aneurysmal changes over the retinoschisis

Figure 3: Cross-section OCT slice showing retinoschisis with the infra-red scout image demonstrating hyporeflectivity of the haemorrhages and telangiectactic elements.

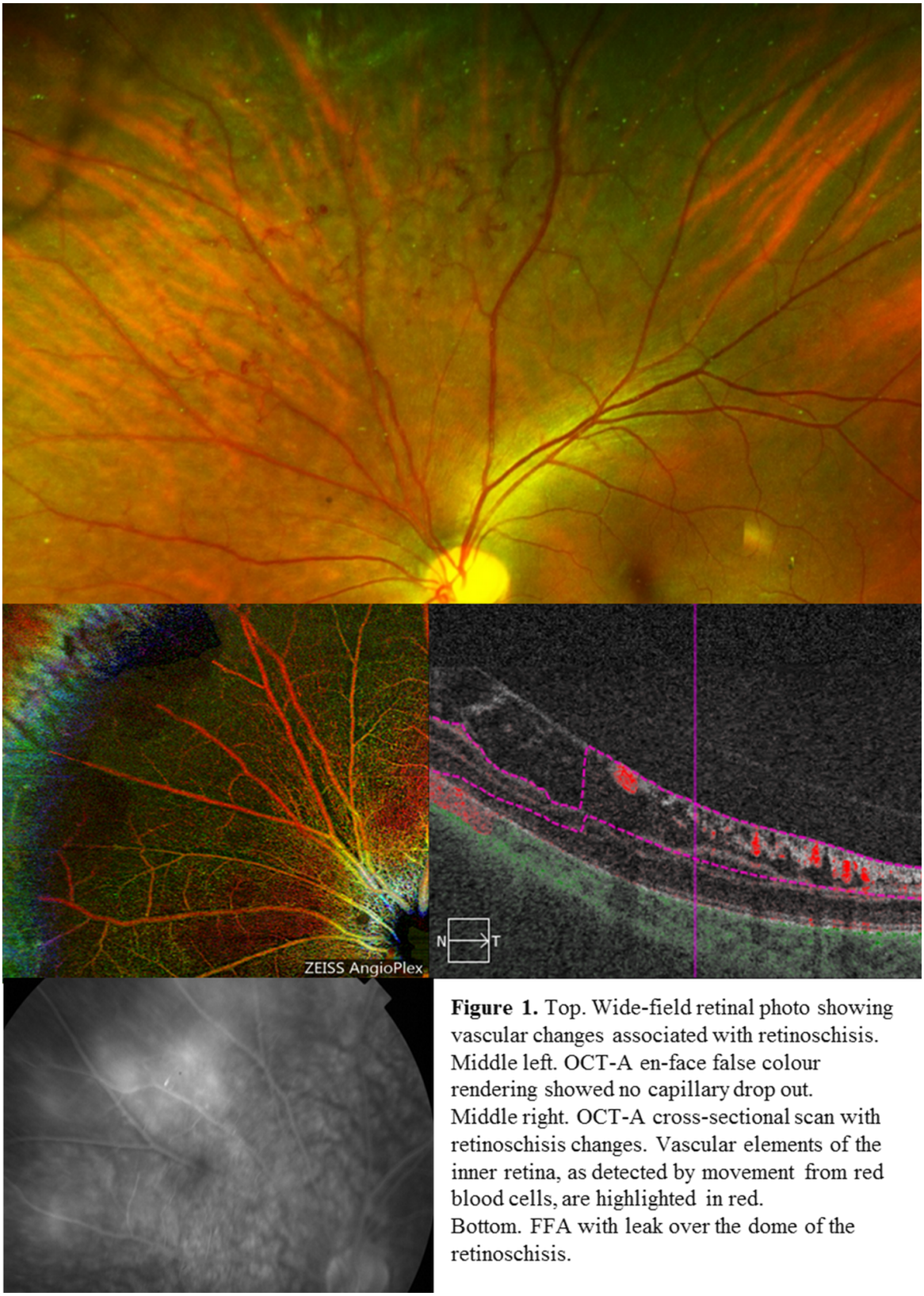

Figure 1. Top. Wide-field retinal photo showing vascular changes associated with retinoschisis. Middle left. OCT-A en-face false colour rendering showed no capillary drop out. Middle right. OCT-A cross-sectional scan with retinoschisis changes. Vascular elements of the inner retina, as detected by movement from red blood cells, are highlighted in red. Bottom. FFA with leak over the dome of the retinoschisis.

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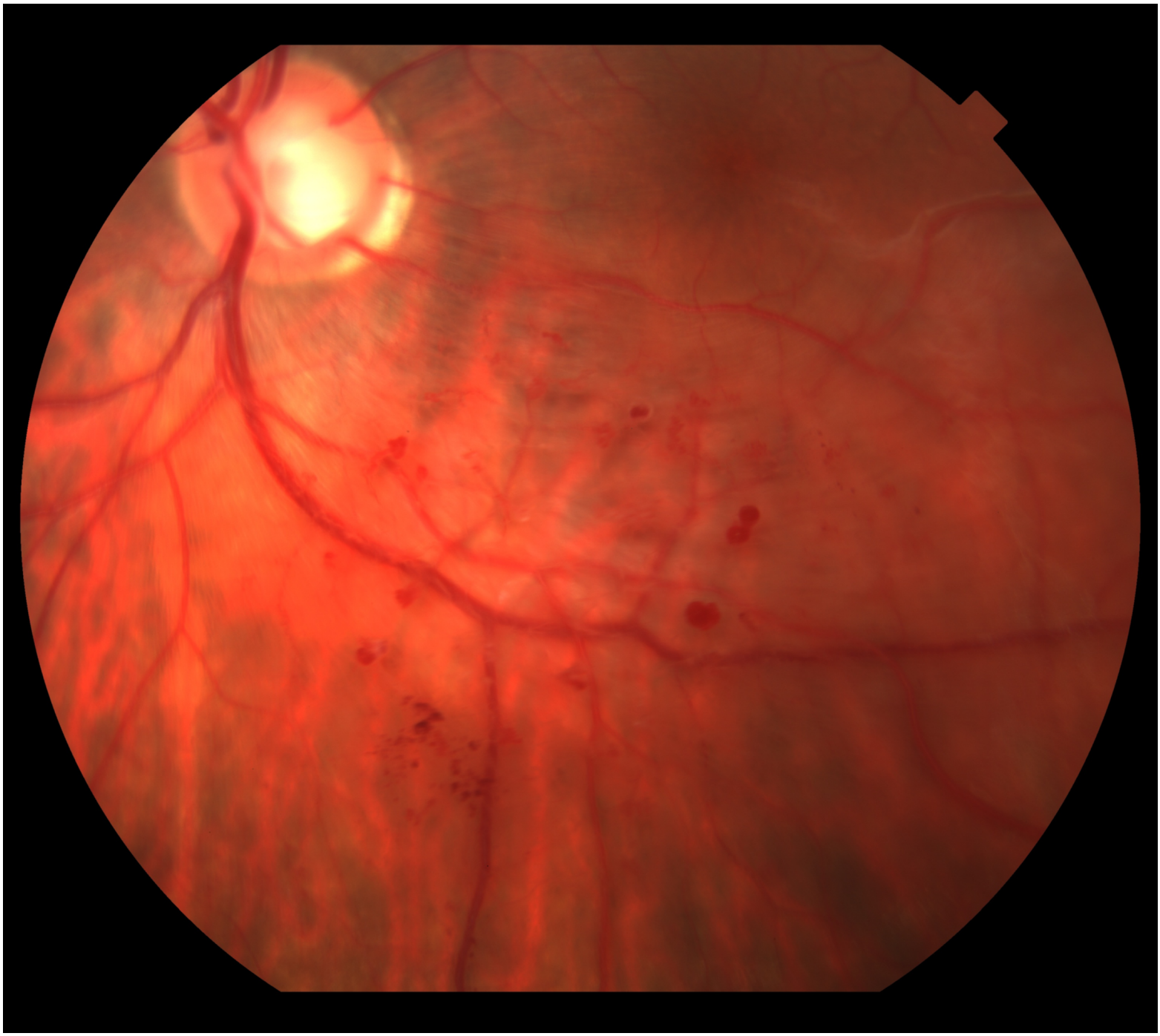

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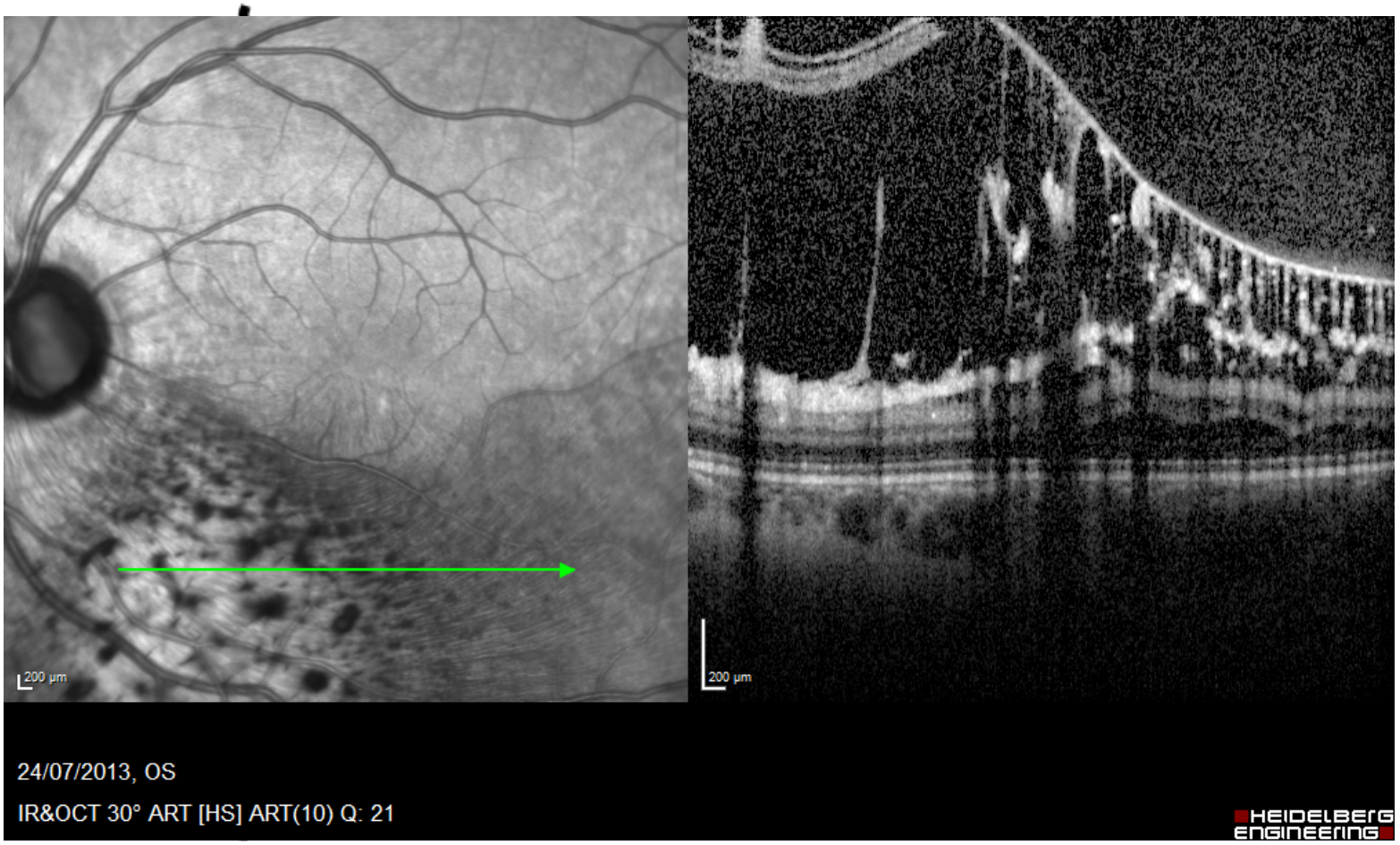

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