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Benign panretinal uniform radial linear-shaped flecks

Suzanna Airiani, MD*, **Wener Cella, MD***, **Chai Lin Chou, MSc**, **Joaquin Tosi, MD**, and **Stephen H Tsang, MD, PhD**

Bernard & Shirlee Brown Glaucoma Laboratory and Edward S., Harkness Eye Institute, College of Physicians and Surgeons, Columbia University, New York, NY, USA

Our case illustrates a new presentation of benign retina flecks with line-shaped and panretinal, uniform and radial distribution. Compared with previous reports of fleck retina disorders, our case does not show increased autofluorescence deposits, no delay in dark adaptation, neither significant macular nor nasal disc involvement.^{1–5}

A 51-year-old woman with consanguinous parents was referred for evaluation of bilateral retinal lesions. On examination, best-corrected visual acuity was 6/7.5 with moderate hyperopic correction in both eyes. Visual fields were normal. The anterior segments were unremarkable.

Fundus examination revealed multiple yellow-creamy discrete round and mostly linear-shaped flecks scattered throughout the retina in both eyes with relative sparing of the fovea. The flecks distributed in a radial pattern centred around the posterior pole (Fig. 1) and were located well posterior to the retinal vasculature. The discs, maculae and retinal vessels appeared unremarkable. Fundus autofluorescence imaging (cSLO, Heidelberg Retina Angiograph 2; Heidelberg Engineering, Dossenheim, Germany) did not show hyper- or hypo fluorescent flecks (Fig. 2a), but multiple discrete round lesions above the retinal pigment epithelium were apparent in the infrared imaging (Fig. 2b). Corresponding spectral optical coherence tomography (Optivue SD-OCT, Fremont, CA, and Cirrus, Carl Zeiss Meditec Inc., Dublin, CA, USA) revealed that these lesions span across the retinal pigment epithelium complex and photoreceptor inner–outer segment junction layers, without involving the choroid (Figs 3,4). To assess retinal function, ISCEV (International Society for Clinical Electrophysiology of Vision) standardized full-field electroretinography were performed following 20 min and overnight dark adaptation. The electroretinography traces showed that both scotopic and photopic responses were symmetric and within normal limits (Fig. 5).

In contrast to previous reports,^{1–3} the retinal lesions in our patient appeared to be significantly smaller, more of a linear shape(s) and uniform in size(s). These lesions were distributed ubiquitously in the equator and mid and far periphery of the fundus. The centrally located flecks in the posterior pole appeared sparse, small, round and dot-like. Unfortunately, the patient's family members were not available for eye examination. In contrast to the classic findings reported by Audo *et al.*,⁴ where the retinal flecks demonstrated increased autofluorescence, the fundus imaging of our patient did not show any specks of abnormal autofluorescence.

*These authors contributed equally to this report.

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Infrared imaging (Fig. 2b) and optical coherence tomography (Figs 3,4) revealed numerous subretinal dot-like lesions, which correspond to those observed in funduscopy.

Different types of flecked retinal disorders such as dominant radial drusen, fleck retina of Kandori,¹ fundus albipunctatus, retinitis punctata albescens, fundus flavimaculatus and vitamin A deficiency⁵ may be considered as differential diagnosis. However, the lack of drusen on the nasal side of the optic disc, extension of flecks to peripheral retina, absence of high-density autofluorescent deposits on scanning laser ophthalmoscopy imaging and presence of intact *EFEMP1* gene sequence made the diagnosis of dominant radial drusen unlikely. Moreover, normal scotopic electroretinography response excludes fundus albipunctatus. In addition to clinical assessment, electrophysiology testing was essential to exclude fundus albipunctatus and autofluorescent imaging can be a helpful tool to characterize hyper-autofluorescent deposits observed in different variants of benign fleck retina. The normal electrophysiology and visual field testing, in addition to good visual acuity and lack of symptoms justify the condition as benign. Hence, our case represents a different presentation of benign fleck retina,⁴ with linear-shaped flecks and a panretinal, uniform and radial distribution.

References

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Figure 1.

A wide-angle montage of the right (a) and left (b) fundi demonstrates multiple yellow-cream discrete flecks widely distributed in the equator and far periphery of the retina. A few flecks are centred in the posterior pole.

Figure 2.

(a) Fundus autofluorescence of the right eye showing absence of hyper- or hypofluorescent flecks; (b) infrared imaging of the right eye showing multiple discrete round lesions in the para-foveal area.

Figure 3.

Spectral-domain OCT of the right eye (Optivue). (a) Multiple flecks in the posterior pole (yellow and green arrows represent OCT scans on (b) and (c), respectively, and white square represents longitudinal SLO scan on (d)); (b) normal appearing macula; (c) retinal scan near the superior temporal vascular arcade showing that flecks are localized across the layers of the RPE complex and photoreceptors inner–outer segment junction, without involving the choroid (arrowheads pointing flecks and open arrowheads pointing shadows from vessels); (d) longitudinal SLO scan from the macula showing multiple flecks at the level of RPE. IS/OS, inner segment/outer segment; OCT, optical coherence tomography; ONL, outer nuclear layer; RPE, retinal pigment epithelium; SLO, scanning laser ophthalmoscopy.

Figure 4.

Spectral-domain OCT of the right eye (Cirrus Zeiss). (a) Multiple flecks in the posterior pole (yellow and green arrows represent OCT scans on (b) and (c), respectively, and white square represents longitudinal scan on (d); (b) normal appearing macula; (c) retinal scan near the superior temporal vascular arcade showing that flecks are localized across the layers of the RPE complex and photoreceptors inner–outer segment junction, without involving the choroid (arrowheads pointing flecks and group of flecks and open arrowheads pointing shadows from vessels); (d) longitudinal scan from the macula showing a granular aspect on RPE surface. ILM, internal limiting membrane; IS/OS, inner segment/outer segment; OCT, optical coherence tomography; ONL, outer nuclear layer; RPE, retinal pigment epithelium.

Figure 5.

Representative photopic and scotopic electroretinogram recordings compared with normal control after 20 min in the right eye and overnight dark adaptation in the left eye. Full-field electroretinographies were performed with Ganzfeld stimulation on this patient and revealed normal scotopic and photopic responses.