Deciphering dark retinal patches

BY BLANCA FLORES AND ANASTASSIOS KOSTAKIS

We present a case of bilateral extensive dark without pressure (DWP) located outside the retinal vascular arcades in a young female. This retinal finding is benign but, in some instances, darkened retinal patches could be associated with potential sight-threatening conditions. Multimodal imaging can help in the identification of key clinical features to guide the necessary diagnostic approach and management plan.

Case report

A 15-year-old female of African descent was referred for evaluation to the paediatric ophthalmology department due to the presence of bilateral darkened retinal areas found incidentally during a routine optometrist visit. She denied loss of vision, flashing lights or floaters. Her past ocular history included bilateral mild myopia but was negative for trauma or prior ocular pathology. There were no systemic comorbidities, and she was not taking long-term medication.

Visual acuity was 6/6 bilaterally with myopic correction (spherical equivalent of -3.00 in each eye). Pupillary reactions were symmetrical in both eyes. Colour vision tested with Ishihara plates was full. Biomicroscopy revealed unremarkable anterior segment with no signs of inflammation or media opacity.

Dilated fundus examination showed extensive geographical patches of flat dark-brown retina with irregular, but well-defined margins nasal to the disc, that extended outside the trajectory of temporal arcades (Figures 1, 2 (A)). The left eye had a greater area of darkening compared to the left. The macula was spared. There were no associated optic nerve or retinal vessel abnormalities.

Optical coherence tomography (OCT) of the dark areas showed a clear and abrupt reduction of the reflectivity of the ellipsoid zone band of the inner photoreceptor layer (Figures 1,2 (B)). No subretinal fluid or choroidal abnormalities were noted. The fovea was anatomically intact. Fundus autofluorescence images showed a corresponding relative hypo-autofluorescence that was homogeneous throughout the areas of retinal darkening (Figures 1,2 (C)). Static perimetry testing was full for both eyes (Figures 1,2 (D)). These features were compatible with DWP.

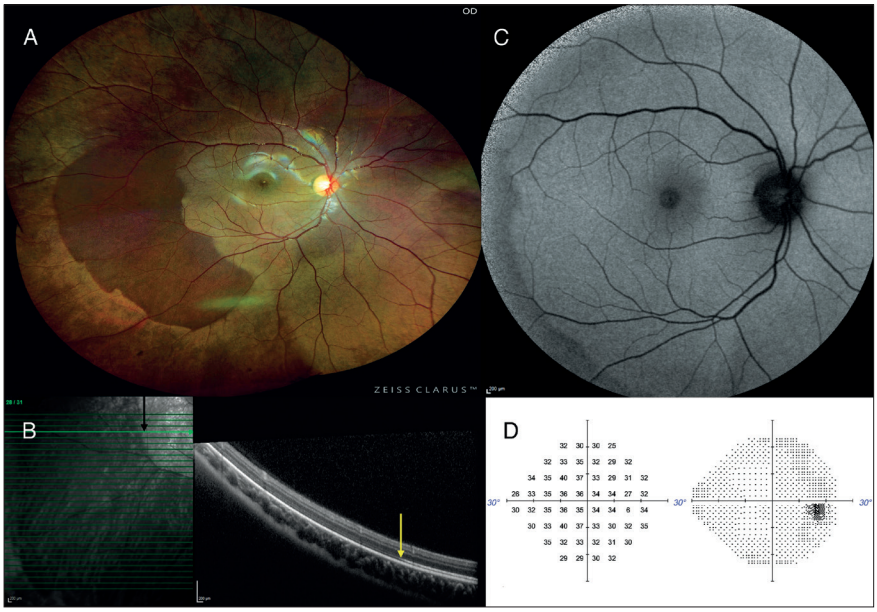

Figure 1: Right eye: (A) Montage photograph of posterior segment shows extensive areas of densely pigmented retina (dark without pressure) with well-demarcated borders, extending over the equator; (B) Corresponding fundus autofluorescence image reveals hypo-autofluorescence of the darkened retinal patches outside the temporal arcades. The macular area and optic disc exhibit normal autofluorescence patterns; (C) OCT demonstrates the sudden reduction in reflectivity signal of the ellipsoid zone band at the edge of the retinal dark without pressure (arrows); (D) normal Humphrey 30-2 visual field testing.

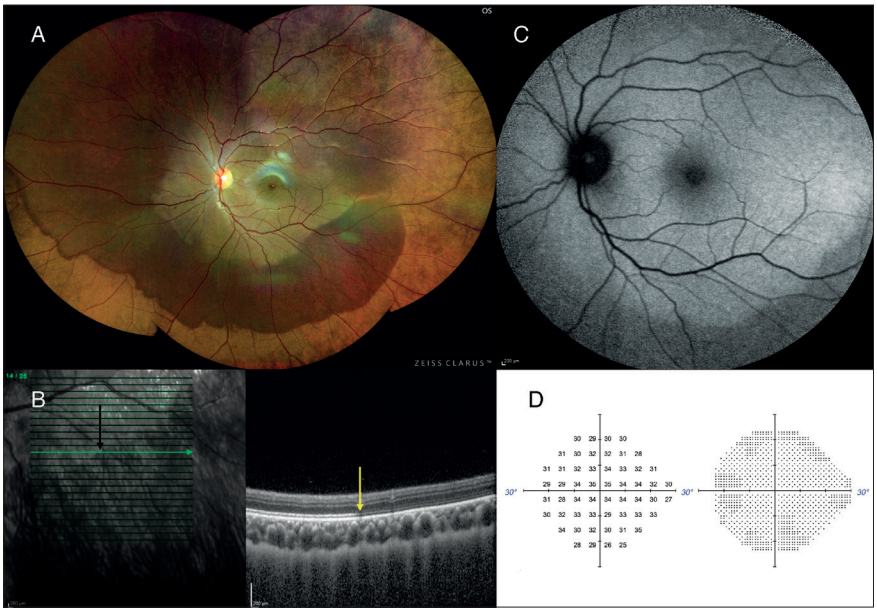

Figure 2: Left eye: see Figure 1 caption, A–D.

The patient underwent haemoglobinopathy screening, which was negative for sickle cell disease or trait. After one year of follow-up, the patient remained asymptomatic with unchanged patches of retinal darkening.

Discussion

Historically, mottled brown patches of the retinal fundus were first described in Afro-Caribbean patients with sickle cell disease or systemic hypertension. It was suggested that these areas could correspond to healed retinal tissue adjacent to underlying patches of capillary occlusion, frequently evident in fluorescein angiography [1]. In more recent publications, areas of retinal darkening have also been described in the context of isolated ocular disease, including congenital hypertrophy of the retinal pigment

epithelium, choroidal osteoma and infectious diseases [2,3]. It is believed that they arise as a result of the local retinal insult.

Isolated patches of darkened retina can also be found in healthy patients without comorbidities; they occur more commonly in those with a higher level of skin pigmentation, including African, Asian or Hispanic origin. The term DWP refers to circumscribed areas of brown retina that are readily visible on biomicroscopic examination without the need to indent the sclera, as it occurs with its homologous 'white without pressure'. They commonly appear near the posterior pole in the mid periphery, and do not seem to be related to any vitreoretinal interface abnormalities [4].

The darkening of the retina appears to arise from a change in photoreceptor reflectivity at the level of the ellipsoid zone band. It has been hypothesised that this change in reflectivity corresponds to a very dense level of photopigment that absorbs more light in the short wavelength spectrum [4]. This in turn translates into reduced hyporeflectivity of the ellipsoid zone confined to the area of retinal darkening, visible in OCT scans. Another hypothesis considers the role of localised cellular metabolic dysfunction in the darkened area, given that the ellipsoid band is highly packed with mitochondria [3]. Since there is no histologic correlation available yet, the true aetiology of this intrinsic photoreceptor variant remains unknown. Nevertheless, isolated areas of DWP appear to be benign without any visual consequence [4]. Our patient was asymptomatic, and we corroborated the absence of vision loss with normal results of field testing.

The clinical behaviour of DWP is variable, with reports of lesions changing in extent or shape over time, by either regressing, migrating or even enlarging at different timeframes [4,5]. It is also not clear if these are congenital anomalies. In our patient, there was no change observed after one year of follow-up.

Most cases of DWP are incidentally found, and it is important to determine if these correspond to an isolated feature, or if there could be an underlying ocular condition. In the latter case, additional signs of ocular inflammation, retinal ischaemia or tumour would be evident on initial examination. A thorough ophthalmic assessment complemented with multimodal imaging can determine the need of systemic investigations (e.g. sickle cell disease); although the majority of cases are benign and do not require any active management. A correct identification of this clinical entity can avoid unnecessary patient concern.

References

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AUTHORS

Blanca Flores, MD, FRCOphth,

Specialty Registrar in Ophthalmology, Sheffield Teaching Hospitals NHS Foundation Trust, UK.

Anastassios Kostakis, FEBO, FRCOphth,

Consultant Ophthalmic Surgeon & Paediatric Ophthalmologist, Doncaster and Bassetlaw Teaching Hospitals NHS Foundation Trust, UK.

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