Dark-without-pressure fundus lesions

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Geographical grey or white areas in the peripheral retina (Schepens, 1952; Rutnin and Schepens, 1967; Karlin and Curtin, 1973; Nagpal, Huamonte, Constantaras, Asdourian, Goldberg, and Busse, in press) have been called white-with-pressure or white-without-pressure depending on the necessity of scleral indentation for them to be visible. Condon and Serjeant (1972a, 1972b) reported mottled brown areas in the retinas of patients with haemoglobinopathies SS and SC. The purpose of this paper is to report homogeneous, geographical, flat, brown areas in the fundi of black patients and to discuss the nature and clinical significance of these lesions.

Case reports

Case 1, a 16-year-old black youth with haemoglobin SS, was first seen at the Sickie Cell Eye Clinic in February 1973. At initial examination he had a visual acuity of 20/20 in both eyes. Slit-lamp examination showed a positive conjunctival sickling sign. Fundus examination showed arteriolar occlusions of the peripheral retina at the 6.30 o'clock position in the right eye and at 2.30 o'clock in the left eye. In June 1973 fluorescein angiography revealed these vessels to be patent. In April 1974 the left fundus showed a small dark brown area at 7.30 o'clock in the midperiphery. It was about 1 disc diameter in size (Fig. 1a) and had sharply demarcated margins. This lesion appeared flat and was a uniform brown in colour. Fluorescein angiography showed neither blockage of retinal or choroidal fluorescence nor any hyperfluorescence. In August 1974 this area was smaller and had faded; in February 1975 it had disappeared (Fig. 1b). A vitreous examination at this time with a Goldmann three-mirror contact lens showed a complete posterior vitreous detachment but without shrinkage—that is, collapse—of the formed vitreous and no other vitreous abnormalities. There were no vitreo-retinal adhesions in the region of the dark brown area.

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Case 2, a 25-year-old black man with haemoglobin SS, was first seen in September 1974, and eye examination revealed a visual acuity of 20/20 in both eyes and a positive conjunctival sickling sign. Fundus examination and fluorescein angiography of the right eye showed an occluded arteriole in the horizontal raphe just temporal to the macular area. Two black sunbursts were seen at 8 and 1 o'clock at the equator. There was a $1\frac{1}{2}$ disc diameter brown area at 2 to 4 o'clock (Fig. 2a) in the midperiphery. The anterior margin was straight; the vertically oriented posterior margin had a dentate border. Fluorescein angiography of this brown area was normal. In November 1974 this area was smaller (Fig. 2b), and the anterior margin had also become irregular. At 12.30 o'clock another brown area $\frac{1}{2}$ disc diameter in size and oval in shape was found. Vitreous examination with Goldmann three-mirror contact lens showed complete, shallow, posterior vitreous detachment without collapse, including the brown areas. The vitreous base remained attached.

Case 3, a 56-year-old black man with haemoglobin SC, was seen in March 1974. Eye examination showed no anterior segment abnormality. The visual acuity was 20/20 in both eyes. Fundus examination of the right eye showed peripheral retinal neovascularization (sea fans) at 8, 9, and 4 o'clock and black sunbursts at 1.30 and 3 o'clock in the equatorial region. A dark brown area 3 disc diameters above and nasal to the disc was about $2\frac{1}{2}$ disc diameters wide (Fig. 3), with irregular margins

FIG. 1a Dark brown area (arrows) of uniform colour in April 1974 in left eye of Case 1. Note crossing of retinal vessels and absence of mottling

FIG. 1b Same area in February 1975. Dark brown area is completely gone. Arrows indicate margins of previous dark-without-pressure

FIG. 2a Dark brown area in right eye of Case 2 in September 1974. Anterior margin is straight (arrows) and posterior margin dentate (arrows). Note crossing of retinal vessels and absence of mottling

FIG. 2b Same area in November 1974. Note irregular anterior margins (arrows) as compared with straight margin (arrows) in A. Posterior margin has also changed

FIG. 3 Dark brown area in right eye of Case 3. This lesion appears almost identical to one pictured by Hamard and others (1974)

FIG. 4 Dark brown area in right eye of Case 6 in December 1974

and a flat appearance. Fluorescein angiography showed normal circulation and fluorescence. Examination of the left eye showed an area of retinal neovascularization at 3 o'clock and a large black sunburst extending from 5 to 6 o'clock. Areas of retinal neovascularization were successfully treated by argon laser photocoagulation. Follow-up examination of the dark brown area 8 months later showed that it was smaller and had faded to light brown. Vitreous examination at this time with the Goldmann three-mirror contact lens showed complete posterior vitreous detachment without collapse. Over and around the dark brown area, the vitreous body was also attached.

Case 4, a 14-year-old black girl with haemoglobin SC, was seen in December 1974. Ocular examination showed a visual acuity of 20/20 in both eyes. Fundus examination of the right eye showed a black sunburst at 10 o'clock in the equatorial region, a few scattered small iridescent spots in the fundus periphery, and a dark brown area at 2 o'clock in the midperiphery. It was flat, horizontally orientated, about 2 disc diameters long and 1 disc diameter wide, and had irregular but sharply outlined margins. Iridescent spots just outside the posterior edge were similar to the haemosiderin-laden macrophages in a resolving salmon patch. The surface of this brown area was smooth with no granularity. Fluorescein angiography of this brown area showed normal perfusion. The area of the old salmon patch at its posterior edge showed possible occlusion of a small precapillary arteriole with faint blockage of choroidal fluorescence. One month later the dark brown area was smaller and had faded. The vitreous showed flat posterior vitreous detachment involving the entire fundus in addition to the region of the brown area.

Case 5, a 26-year-old black woman with SS haemoglobin, was seen in April 1974. The visual acuity was 20/20 in both eyes. Slit-lamp examination of the conjunctiva showed a positive conjunctival sickling sign. Fundus examination of the right eye showed occlusion of retinal arterioles in the equatorial region in several areas. The left eye showed similar changes in the retinal periphery and a small oval, dark brown area (1 disc diameter) at 9.30 o'clock in the midperiphery. Fluorescein angiography of this area was normal. In December 1974 the right fundus had developed a small soft exudate above the foveal area. In both eyes the peripheral retinal arterioles showed the spontaneous remodelling pattern described by Galinos, Asdourian, Woolf, Stevens, Lee, Goldberg, Chow, and Busse (1975). In February 1975 the dark brown area in the left eye had faded. Fluorescein angiography showed that this area appeared normal. The vitreous examination in February 1975 with the Goldmann three-mirror contact lens was normal. There was no vitreous detachment, collapse, or shrinkage.

Case 6, a 12-year-old black girl with haemoglobin SS and a history of cerebrovascular accident, was seen in November 1974. Her visual acuity was 20/20 in both eyes. Fundus examination showed several areas of peripheral arteriolar occlusion in both eyes. The right eye showed a flat dark brown area (Fig. 4), about 2 disc diameters nasal to the disc, extending from 2.30 to 5.30 o'clock. Its width varied from 1 to 3 disc diameters

and it had irregular but sharply demarcated margins. The rest of the retina had a normal appearance. Fluorescein angiography showed normal circulation. The left eye also showed peripheral arteriolar occlusions and a dark brown area extending from 7 to 10 o'clock 2 disc diameters nasal to the disc. It was flat and 3 to 5 disc diameters wide. The distinct margins were surrounded by normal retina. Fluorescein angiography showed normal retinal circulation and normal choroidal fluorescence. The vitreous examination was normal in both eyes, showing no vitreous detachment or vitreous collapse.

Case 7, a 46-year-old hypertensive black woman, was examined in January 1975. Her systemic blood pressure reading was 220/120 mm Hg. Haemoglobin electrophoresis showed haemoglobin AA. Fundus examination revealed mild generalized narrowing of retinal arterioles and a few linear flame-shaped haemorrhages. In the right eye 2 disc diameters below the disc there were two dark brown areas which were flat, irregular, and clearly demarcated. The surrounding retina was normal. Fluorescein angiography showed normal retinal circulation. Except for attachment to the disc and vitreous base, the vitreous was shallowly detached without collapse. No vitreo-retinal adhesions were seen over this brown area or surrounding the retina.

Discussion

The lesions described by Condon and Serjeant (1972a, 1972b) occurred in patients with homozygous sickle cell disease and haemoglobin SC. These areas were characterized as having a mottled brown surface surrounded by a halo of pale retina. They were often associated with copper-coloured, iridescent, glistening spots. Fluorescein angiography showed normal circulation in these areas, but adjacent areas showed arteriolar occlusive disease. They suggested that the mottled brown areas represented the sequelae of deep subretinal haemorrhages. Okun (1969) showed a chorio-retinal scar surrounded by a brown area developing from a salmon patch in a patient with sickle cell disease. The lesions reported by Condon and Serjeant (1972a, 1972b) and Okun (1969) probably represent sequelae of salmon patch haemorrhages that dissected deeply into the retina and into the subretinal space—that is, they probably represented early stages of black sunbursts.

We have seen seven patients with brown lesions (four with haemoglobin SS, two with haemoglobin SC, and one with hypertension and haemoglobin AA) of the fundus that had uniform colour without any mottling. These areas appeared to be unrelated to the residue of salmon patches such as schisis cavities, iridescent spots, or sunburst lesions. These flat lesions varied in size from 1 to several disc diameters. Some had regular, round, or straight margins; whereas others had irregular,

serrated, or dentate margins. Location and orientation of these lesions also varied. The lesions we saw were located near the posterior pole or in the midperiphery. They were orientated either radially or circumferentially. Most of the lesions were transient, and when followed-up for weeks to months, changed shape and sometimes disappeared completely. Fluorescein angiography in our patients did not reveal any vascular abnormalities in these areas, although Case 4 showed occlusion of a small precapillary arteriole posterior to the brown area. Our cases resembled one pictured by Hamard, Coquelet, Jaeger, Malmejac, and Mondon (1974) which they identified as a salmon patch. We do not feel that the lesion shown by these authors represented a salmon patch, because salmon patches are bright red (later yellow), much smaller, and usually cover retinal vessels. This lesion appeared several disc diameters in size and had a brown homogeneous colour with normal overlying retinal vessels. Unfortunately, no fluorescein angiogram of this lesion was presented. These dark-without-pressure areas cannot be related to preretinal, intraretinal, subretinal, or choroidal haemorrhages because normal choroid is visible in these dark areas by ophthalmoscopy and fluorescein angiography.

The dark brown areas may represent an island of normal fundus surrounded by extensive white-without-pressure changes. However, some of these lesions are very small, representing only about 2 to 5 per cent of the total retinal surface; white-without-pressure lesions involving 95 to 98 per cent of the retina appear to be only a remote possibility. These dark brown areas, however, are comparable in some respects to areas of white-without-pressure. Like white-without-pressure, they are flat, vary in shape, and can occur in numerous locations throughout the eye. The white-without-pressure lesion has been noted to be migratory in nature and can disappear with passage of time (Nagpal and others, in press). Similarly, dark-without-pressure lesions are migratory and can also fade in time. Both can be seen in

the fundus without any scleral indentation. We labelled these brown lesions as dark-without-pressure, because, like white-without-pressure, they do not need scleral indentation to be seen and are analogous to white-without-pressure lesions in the aspects noted above.

There are several differentiating features. The white-without-pressure lesions are white or grey; they occur mostly in the equatorial region or peripheral to the equator and, according to our previous observations (Nagpal and others, in press), represent changing vitreo-retinal adhesions. In contrast, dark-without-pressure lesions are brown and occur most commonly near the posterior pole or in the midperiphery. The status of the vitreous appears to be unrelated to the presence of the dark areas. In three of our patients the vitreous was normal, and in four patients there was shallow detachment of the vitreous without liquefaction, shrinkage, or collapse.

These dark brown lesions may represent subtle changes or an altered reflex in the pigment epithelium in the internal limiting membrane of the retina, or in other anatomical regions of the fundus. The factors leading to these changes are not known. Future histopathological examination may help identify the nature of these lesions.

Summary

Seven black patients had dark brown homogeneous geographical areas of the fundus. Six cases were associated with sickle cell haemoglobinopathies and one was associated with systemic hypertension. These flat lesions were uniform in colour and occurred in the posterior pole or in the midperiphery. They appeared to be transient and often disappeared leaving no residue. The cause is unknown. By analogy with white-without-pressure fundus lesions, we have called these areas dark-without-pressure.

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