Spontaneous peripheral chorioretinal neovascularisation in association with sickle cell anaemia

JAMES C. LIANG AND LEE M. JAMPOL

From the University of Illinois Eye and Ear Infirmary, 1855 W. Taylor Street, Chicago, Illinois 60612, USA

SUMMARY Chorioretinal neovascularisation was noted in the midperipheral fundus of a young black girl with sickle cell anaemia. Rapid-sequence fluorescein angiography showed choroidally fed neovascularisation in the centre of a large black sunburst (chorioretinal scar). We suspect that a massive midperipheral retinal haemorrhage from the sickle cell disease caused the sunburst and altered Bruch's membrane, thus allowing the ingrowth of choroidally fed vessels.

Peripheral retinal neovascularisation is a well described finding in patients with haemoglobin SC disease, sickle cell β-thalassaemia, and homozygous sickle cell anaemia. Peripheral chorioretinal and choriotireal neovascularisation has been reported to occur following argon laser or xenon arc photocoagulation for proliferative sickle cell and diabetic retinopathies. To our knowledge such neovascularisation has not been noted to occur spontaneously in either of these conditions. We report a case of spontaneous chorioretinal neovascularisation in association with sickle cell anaemia in the peripheral fundus of a 14-year-old girl.

Case report

A 14-year-old black girl with sickle cell anaemia was referred to the Sickle Cell Eye Clinic at the University of Illinois Eye and Ear Infirmary for a routine eye examination. She had no ocular complaints and denied previous visual difficulties. There was no history of ocular trauma, surgery, or photocoagulation.

The best corrected visual acuity with a minor hyperopic correction was 20/25 (6/7.5) OU. Results of external, ocular motility, and slit-lamp examinations were normal except for 3+ conjunctival sickling signs bilaterally.

The maculas and discs appeared normal bilaterally.

The right fundus had multiple black sunbursts (peripheral chorioretinal scars) from one-half to 2 disc diameters in size located at the 3, 4, 5, and 6 o'clock equators. In the left eye black sunbursts from one-half to 2 disc diameters in size were at the 7, 8, 9, and 11 o'clock equators. In addition, a very large 6 by 7 disc diameter black sunburst was located just posterior to the equator from the 3 to 5 o'clock meridians (Fig. 1) with a one-half by one disc diameter patch of fibrous tissue in its centre.

No obvious areas of retinal neovascularisation were apparent with indirect ophthalmoscopy. However, rapid-sequence fluorescein angiography showed choroidally fed neovascularisation in the fibrous tissue at the 4 o'clock position in the large black sunburst of the left eye (Figs. 2, 3, 4).

Blood tests gave the following results: haemoglobin, 10.1 g/dl; haematocrit, 27.9%; white blood cell count, 8.9 x 10^9/l. A quantitative haemoglobin electrophoresis showed S haemoglobin 88.8%; A2 haemoglobin 3.9%, and fetal haemoglobin 7.3%. The irreversible sickle cell count was 5.9%.

The patient has been followed up for more than a year; there has been no change in the area of neovascularisation. No photocoagulation or other therapy has been attempted because of the stability of the lesion and the disappointing results previously described following treatment of this type of neovascularisation.

Discussion

Spontaneous peripheral chorioretinal neovascularisation...
Figure 1  Fundus photograph showing the very large black sunburst in the temporal periphery of the retina of the left eye. The arrow indicates a tuft of fibrous neovascularisation.

Asdourian and colleagues have convincingly shown that black sunbursts evolve after occlusion of midperipheral retinal arterioles by plugs of sickled erythrocytes. Ischaemic necrosis of the vessel wall ensues, which in turn can result in intraretinal or preretinal haemorrhage. If the blood dissects to the level of the retinal pigment epithelium (RPE), it may stimulate proliferation and migration of the RPE, which forms a black sunburst. Black sunbursts may be seen in patients with sickle cell anaemia, haemoglobin SC disease, or sickle cell \( \beta \)-thalassaemia.

In our patient we suspect that a massive intraretinal and subretinal haemorrhage occurred with resultant
Spontaneous peripheral chorioretinal neovascularisation in association with sickle cell anaemia

Fig. 2 Early-phase fluorescein angiogram showing filling of the fibrous neovascularisation (arrow) from the choroidal circulation prior to filling of nearby retinal vessels.

Fig. 3 Later-phase fluorescein angiogram showing filling of nearby retinal vessels. There is no vascular connection between the retinal circulation and the area of neovascularisation (arrow).

Fig. 4 Late-phase fluorescein angiogram showing leakage from the area of neovascularisation (arrow).

formation of a huge black sunburst. We suggest that mechanical factors or the cellular response to the large intraretinal or subretinal haemorrhage altered the extracellular matrix or fibrous component of Bruch's membrane in some way, and that this altered Bruch's membrane, allowing the development of spontaneous chorioretinal neovascularisation.

Choroidal ischaemia following focal photo-coagulation was noted in 13 of 19 patients with sickling haemoglobinopathies who subsequently developed choroidally fed neovascularisation. Choroidal occlusive disease in sickling haemoglobinopathies has been described following posterior ciliary artery occlusions. Characteristic wedge-shaped or triangular sectors of yellow-brown pigment mottling are seen, but focal occlusions of choroidal precapillary arterioles (Elschnig's spots) or capillaries have not been described. In our patient fundus findings, as well as any fluorescein angiographic indications of choroidal occlusive disease, were absent.

Two recent reports of choroidally fed neovascularisation following feeder vessel photo-coagulation of proliferative sickle cell disease suggest that these new vessels are best left untreated unless absolutely necessary. At the University of Illinois 6 of 7 treated eyes were unchanged or worse following xenon, argon, or cryopexy treatment of choroidally fed neovascularisation. At the University of the West Indies 7 of 11 eyes were unchanged or worse following xenon or cryopexy treatment of choroidally fed new vessels. In our patient the lesion is apparently stable, and no treatment is contemplated at present.

The study was supported in part by Core Grant EY 01792 from the National Eye Institute and Research Grant PHS HL 15168 from the National Heart, Lung, and Blood Institute, Bethesda, Maryland.
References