[2]Department of Genitourinary Medicine
Blackpool Victoria Hospital
Whinney Heys Road, Blackpool FY3 8NR, UK

Correspondence: JM Durnian
Tel: +44 1253 303777
E-mail: jon_durnian@hotmail.com

Sir,

Superior macular sparing in central retinal artery occlusion due to sickle cell anaemia

Eye (2004) **18,** 442–443. doi:10.1038/sj.eye.6700684

We describe an unusual manifestation of sickle cell retinopathy, in which part of the macular circulation was spared even though the patient presented with features of a central retinal artery obstruction.

Case report

A 32-year-old African male with homozygous sickle cell disease (SS disease) presented to the eye casualty with a 24-h history of blurred vision in the right eye. He had consumed several pints of alcohol on the two evenings prior to presentation, but had failed to ensure a full rehydration with water. On presentation, his vision was 6/60 in the right eye and 6/6 in the left. There was a right afferent pupillary defect. Conjunctival signs of sickling, first described by Paton,[1] were noted in both the eyes. Fundus examination revealed a milky white retinal appearance consistent with a central retinal artery occlusion (CRAO) but with a zone of normally perfused macula, of approximately 2 clock hours size, superiorly (Figure 1). Grade 1 sickle cell retinopathy was noted in the left eye.

He was admitted for rehydration with normal saline to reduce blood viscosity and improve perfusion. The haematologists commenced prophylactic treatment with hydroxyurea, 2 g daily, to prevent a similar occlusive event in the other eye. Hydroxyurea increases levels of foetal haemoglobin (HbF). HbF interferes with the polymerisation of sickle haemoglobin. The higher its concentration, the lower the sickle haemoglobin. Sickle red blood cells from patients with high HbF are less adherent to vascular endothelium. Hydroxyurea is the only prophylactic drug studied for the prevention of systemic crises in a randomised controlled trial. Its main side effect is myelosuppression requiring two-weekly monitoring of blood counts.[2] Exchange transfusion was also considered to restore vascular perfusion. It has been successfully performed in a 25-year-old patient with bilateral central retinal artery occlusions 8 h postocclusion.[3] In this case, exchange transfusion was not felt to be beneficial because of the late stage of presentation. Reperfusion may have been established, but retinal function in the infracted area would not have recovered.

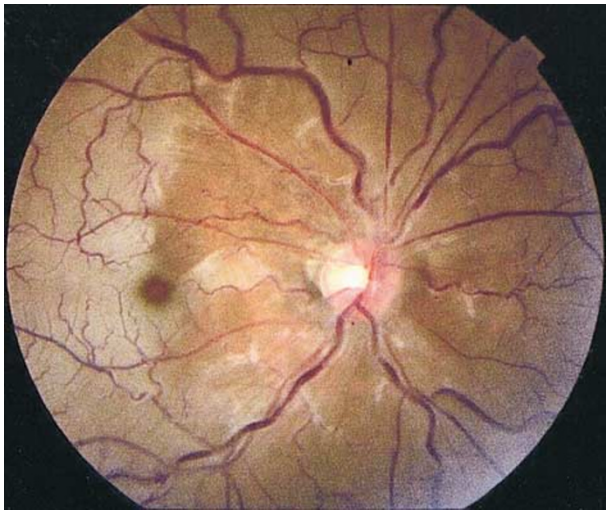

Figure 1 Day 1 fundus appearance showing CRAO with partially perfused macula superiorly.

A fluorescein angiogram (Figures 2a-d) showed features consistent with a central retinal artery occlusion with substantial shutdown of retinal vasculature.

However, there was no visible single thrombus. There was patency of the vessels to fluorescein to a point 1 to 2 disc diameters beyond the disc margin on the temporal side. Part of the macular circulation appeared spared, but this was not due to a patent cilioretinal artery. A late-phase angiogram showed staining of vessel walls, indicating sluggish circulation.

After 2 weeks, vision improved to 6/18 and a month later vision was 6/9 in the right eye. However, he had a very constricted field. Fundus examination showed sequelae of CRAO with optic atrophy and attenuated sheathed blood vessels. (Figures 3a and b). The patient reported using paracentral viewing to maximize visual potential.

Comment

Ocular complications in SS disease are unusual. They are much more common in sickle cell thalassaemia (Sthal) and sickle cell C disease (SC).[4] The postulated reason is that in SS disease, the severe anaemia and thus low haematocrit lead to a lower blood viscosity than in Sthal or SC where anaemia is less marked and the haemoglobins exist in a higher concentration. Sickling has

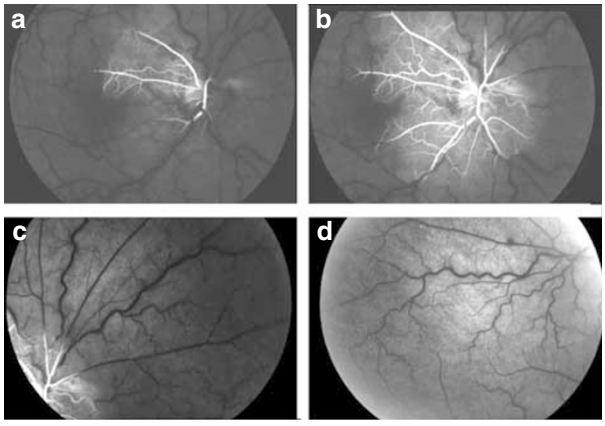

Figure 2 (a,b) Angiograms at 0:21.9 and 1:07.1, respectively, showing shutdown of peripheral vasculature with partial patency of macular circulation. (c) Angiogram at 1:33.6 showing no perfusion of nasal circulation. (d) Angiogram at 2:09.7 showing no perfusion of peripheral temporal circulation.

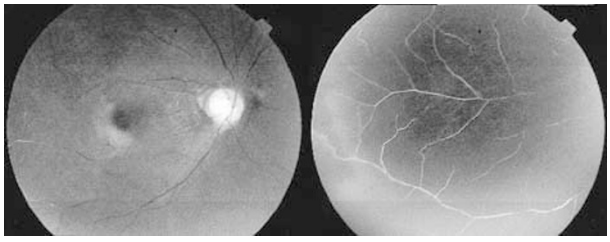

Figure 3 One-month appearance showing disc pallor and sheathed peripheral blood vessels.

a greater effect on blood viscosity and predisposes to vascular occlusion.[5]

There have been case reports of central retinal artery occlusions in association with SS disease.[6,7] This case is unusual because of the superior zone of macular sparing, which is not due to cilioretinal artery sparing. It appears to be due to patency of part of the macular circulation originating from the superotemporal retinal vessels. Vascular obstruction in sickle cell disease occurs due to RBC sickling leading to sluggish blood flow, erythrocyte aggregation, activation of coagulation, and eventual vaso-occlusion.[8] In our patient, we hypothesise that the original insult was dehydration, leading to an increase in plasma viscosity which caused sickling in the central retinal artery. The ensuing panretinal hypoxia led to further sickling downstream of the initial obstruction with sludging and total occlusion of peripheral retinal vasculature. This may have been compounded by the smaller vascular calibre and lower oxygen tensions of the peripheral vessels. The macular sparing was due to a patent macular vessel before the superotemporal vessel became occluded.

Thus, this case illustrates a previously unreported ocular manifestation of SS disease, due to which the patient lost a substantial amount of peripheral vision but retained a narrow field of central vision.

It highlights the prophylactic use of hydroxyurea and the possible therapeutic role of exchange transfusion in managing this problem. These options merit further investigation.

References

1 Paton D. Conjunctival sign of sickle cell disease. *Arch Ophthal* 1961; **66**: 90.
2 Ballas S. Sickle cell anaemia, progress in pathogenesis and treatment. *Drugs* 2002; **62**: 1143–1172.
3 Weissman H, Nadel AJ, Dunn M. Simultaneous bilateral retinal arterial occlusions treated by exchange transfusions. *Arch Ophthal* 1979; **97**: 2151–2153.
4 Kanski J. *Clinical Ophthalmology,* 3rd edn. Butterworth-Heinemann: Oxford, 1994, p 370.
5 Harry J, Misson G. *Clinical Ophthalmic Pathology,* Ist edn. Butterworth-Heinemann: Oxford, 2001, p 293.
6 Mansi IA, Alkhunaizi AM, Al-Khatti AA. Bilateral central retinal artery occlusion secondary to sickle cell disease. *Am J Hematol* 2000; **64**: 79–80.
7 Fine LC, Petrovic V, Irvine AR, Bhisitkul RB. Spontaneous central retinal artery occlusion in haemoglobin sickle cell disease. *Am J Ophthalmol* 2000; **129**: 680–681.
8 Jakobiec FA, Albert DM. *Principles and Practice of Ophthalmology.* WB Saunders Co: London, 1994, pp 1006.

C Sanghvi, A Laude, A Smith and CL Dodd

Department of Ophthalmology
Manchester Royal Eye Hospital
Oxford Road
Manchester M13 9WH, UK

Correspondence: CA Sanghvi
20, Stablefold
Worsley
Manchester M28 2ED, UK
Tel.: +44 161 288 8659
Fax: +44 161 273 6354
E-mail: casanghvi@yahoo.co.uk

Sir,

Deposition of gold in ocular structures, although known, is rare. A case of ocular chrysiasis in a patient of rheumatoid arthritis on gold treatment is presented

Eye (2004) **18,** 443–444. doi:10.1038/sj.eye.6700694

An asymptomatic 50-year-old white man was referred for corneal evaluation by his optician. His past medical