Prince of Wales Hospital, Kowloon
Hong Kong
People's Republic of China

[2]Department of Ophthalmology
and Visual Sciences
The Chinese University of Hong Kong
Hong Kong Eye Hospital, Kowloon
Hong Kong
People's Republic of China

Correspondence:
W-M Chan

Department of Ophthalmology and
Visual Sciences
The Chinese University of Hong Kong
3/F, Hong Kong Eye Hospital
147K Argyle Street Kowloon, Hong Kong
Tel: +852 2632 2879
Fax: +852 2648 2943
E-mail: cwm6373@netvigator.com

Financial Interest: Nil.
Financial Support: Nil.

Eye (2004) **18,** 1275–1277. doi:10.1038/sj.eye.6701399
Published online 30 April 2004

Sir,

Macular infarction a presentation of sickle cell crisis

A 24-year-old black male with sickle cell disease (SCD) 'SS' presented to the emergency room (ER) in sickle cell crisis with acute painless loss of vision OD, chest, and leg pains since one day. His visual acuity was HM OD and J1 OS with 3+ afferent pupillary defect OD. Anterior segment examination was unremarkable. Ophthalmoscopy OD revealed retinal pallor with dilated tortuous vessels. OS fundus was normal. Systemic evaluation revealed icterus with normal pulmonary, cardiovascular, or neurological examination with no history of comorbid conditions. The haemoglobin was 9.6 gm/dl and haematocrit was 27.9%. Exchange transfusion was performed subsequent to hydration.

At 1 week the visual acuity (VA) was HM OD and 20/25 OS. Ophthalmoscopy OD revealed pale, white, thickened retinal lesion centred on the fovea, arterial attenuation, cherry red spot, and pale optic disc (Figure 1 inset). Fluorescein angiography (FA) OD demonstrated perifoveal arteriole occlusions with enlargement of the FAZ (Figure 1). Vessels had areas of blocked fluorescein in early and late phases (Figure 1a and b), while hyperfluorescein areas (Figure 1b) in the late phases (staining). FA OS was normal.

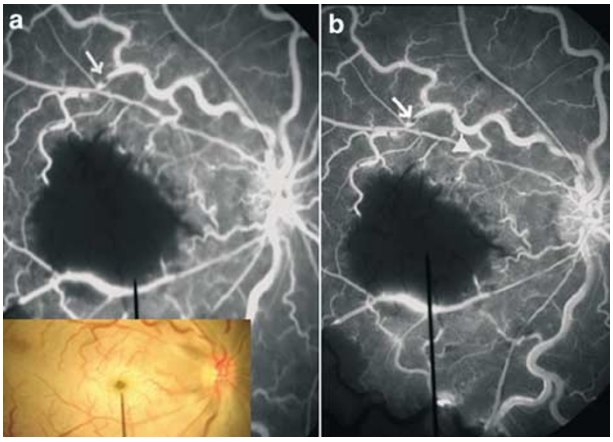

Figure 1 (a) Fluorescein angiogram OD (1 week) early phase showing a marked increase in size of the FAZ due to microvascular occlusion. The white arrow shows that the area of block fluorescence may be due to microemboli. Inset: Photograph of the fundus of the right eye at 1 week. (b) Fluorescein angiogram OD (1 week) late phase showing marked a increase in the size of FAZ due to microvascular occlusion. The white arrow shows the area of block fluorescence, while the white arrowhead shows the area of hyperfluorescence (staining) may be suggestive of microemboli and/or slugging of sickled cells with slowed circulation in the venules.

At 3 months VA was CF 3 feet OD, 20/20 OS. Ophthalmoscopy OD revealed pale disc with marked arterial attenuation and pigmentary changes in the macula (Figure 2a). Macular perimetry (Nidek Technologies, Vigonza, Italy) revealed unstable fixation with absolute macular scotoma OD with normal findings OS (Figure 2b-d). His haemoglobin was 9.5 gm/dl and haematocrit was 26.0%.

Comments

Occlusive disease of the peri-foveal arterioles is known to occur in sickle cell disease.[1,2] We present a dramatic occlusive event of the macula surrounding the foveal avascular zone, resulting in severe and permanent loss of vision. We are unaware of any previous report of FA showing possible microemboli in the retinal vessels with macular infarction in SCD. Direct sickling may cause occlusion in arterioles or in capillary beds creating a 'log-jam' in the arterioles.[3] In this case, the occlusions surrounded the macula from multiple directions, suggesting perifoveal

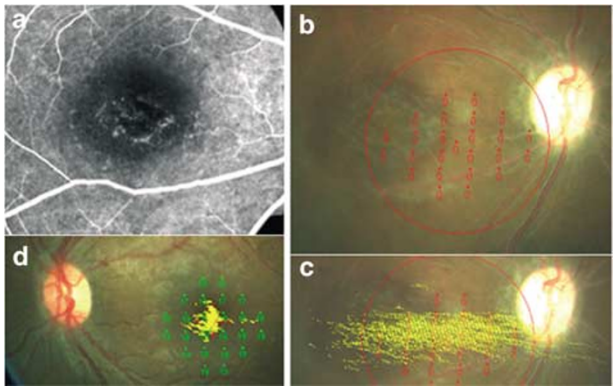

Figure 2 (a) Fluorescein angiogram OD (3 months) late phase showing enlargement of FAZ, with pigmentary changes suggestive of macular infarction. (b) Macular perimetry OD with MP-1 showing absolute scotoma. (c) Fixation stability mapping by the MP-1. The yellow dots are the fixation points during the test showing unstable fixation. (d) Macular perimetry OS by MP-1 showing no scotoma and central stable fixation.

arteriolar occlusion due to microemboli and/or slugging of sickle cells with slowed circulation in the venules.

Our patient presented with an acute visual loss resulting in irreversible macular infarction. His visual recovery was minimal because of delay in presentation.

References

1 Acacio I, Goldberg MF. Peripapillary and macular vessel occlusions in sickle cell anemia. *Am J Ophthalmol* 1973; **75**: 861–866.
2 Welch RB, Goldberg MF. Sickle-cell hemoglobin and its relation to fundus abnormality. *Arch Ophthalmol* 1966; **75**: 353–362.
3 Goldberg MF, Galinos S, Lee CB, Stevens T, Woolf MB. Editorial: Macular ischemia and infarction in sickling. *Invest Ophthalmol*. 1973; **12**: 633–635.

KV Chalam and VA Shah

Department of Ophthalmology
University of Florida College of Medicine
580 W 8th street, Jacksonville, FL 32209 USA

Correspondence: KV Chalam
Tel: +1 904 244 9361
Fax: +1 904 244 9391
E-mail: kvchalam@aol.com

Proprietary Interest: None.
Presented as a poster at the 21st Annual Meeting of the American Society of Retina Specialists, New York, NY, August 2003.

Eye (2004) **18,** 1277–1278. doi:10.1038/sj.eye.6701409
Published online 23 April 2004