S Sarangapani, L Chang and K Gregory-Evans

Western Eye Hospital
St Mary's Trust, London

Correspondence: K Gregory-Evans
Western Eye Hospital
St Mary's Trust
Marylebone Road
London NW1 5YE, UK
Tel: 44 (0)207 886 3201
Fax: 44 (0)207 886 3203
E-mail: k.gregory-evans@ic.ac.uk

Sir,

Visual loss due to cerebral infarcts in pseudoxanthoma elasticum

Eye (2002) **16,** 785–786. doi:10.1038/sj.eye.6700173

Pseudoxanthoma elasticum (PXE) is a rare heritable connective tissue disorder with autosomal dominant or autosomal recessive modes of inheritance. It involves the elastic tissues of the eye, skin and cardiovascular system. Ocular features include angioid streaks due to involvement of the Bruch membrane. In patients with PXE and angioid streaks, visual loss is generally due to development of a choroidal neovascular membrane (CNVM). We report a case of PXE with visual loss due to multiple cerebral infarcts.

Case report

A 36-year-old man with a known diagnosis of PXE presented with a 5-day history of sudden onset right sided blurred vision. His best corrected visual acuity was 6/9 in the right eye and 6/6 in the left eye. Anterior segment examination was normal except for a right-sided relative afferent pupillary defect. Intraocular pressure was normal in both eyes. Fundus examination revealed angioid streaks, pigment mottling of the fundus in both eyes and pallor of the right optic disc (Figure 1). Physical examination revealed cutaneous lesions of PXE in the form of yellowish papules on the neck. Visual field analysis showed a right homonymous visual field defect. Fundus fluorescein angiogram confirmed the absence of CNVM. Magnetic resonance imaging (MRI) of the brain and orbits was performed. Axial fluid attenuated inversion recovery (FLAIR) sequence MRI of the brain revealed multiple ischaemic infarcts in the brain, the largest being in the left parieto-occipital lobe (Figure 2). The orbits and optic nerves were however normal.

The patient was young with no risk factors of cerebrovascular disease. He was a non-smoker. Blood tests revealed no evidence of diabetes mellitus, hyperlipidaemia or coagulopathy. A cardiovascular assessment was sought from a cardiologist. This included an electrocardiogram and transthoracic echocardiogram. There was no evidence of any cardiovascular abnormality apart from borderline hypertension, which did not require treatment.

Comment

Apart from CNVM, visual loss in PXE can also be caused by infarction of the visual pathways, most commonly involving the orbital portion of the optic nerve.[1] Our patient showed features of multiple areas of central nervous system ischaemia. The left parieto-occipital lobe infarct was thought to be the cause for the visual field defects. The presence of a relative afferent pupillary defect and optic disc pallor on the right side suggests involvement of the right optic nerve. Though conjectural, these changes are likely to be due to an old infarct of the optic nerve as the optic disc pallor was noted at acute presentation itself (within 5 days of onset of symptoms).

Cerebral ischaemia in PXE is caused by small vessel

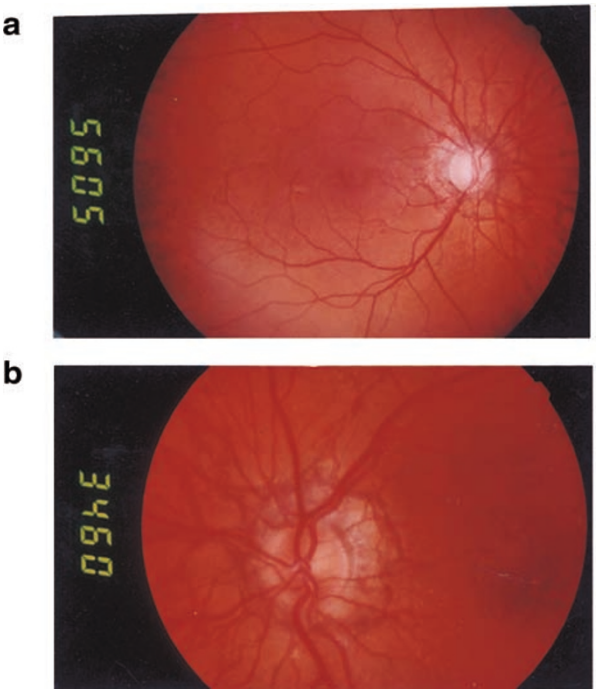

Figure 1 Fundus photographs of right eye (a) and left eye (b) showing prominent angioid streaks and pigment mottling of the posterior pole. Note diffuse optic disc pallor and characteristic peau d'orange appearance temporal to the macula in the right eye.

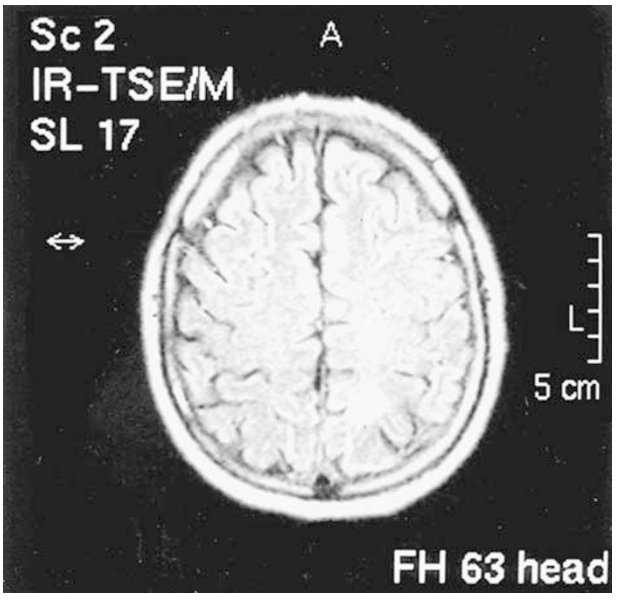

Figure 2 Axial FLAIR sequence MRI of the brain shows high signal area at the left parieto-occipital boundary. Appearance suggests ischaemia.

occlusive disease with hypertension, also common in PXE acting as an accelerating factor.[2–4] In a study of 100 cases of PXE,[4] patients who developed ischaemic stroke were found to have only small vessel involvement.

Ischaemic stroke in patients with PXE is usually not seen until the fifth decade of life unlike our patient.[2,3] Relative risk of developing ischaemic strokes is estimated to be 3.6 in PXE patients less than 65 years compared with the general population.[4] Other neurological complications quoted in the literature include intracranial aneurysms, subarachnoid and intracerebral hemorrhages, progressive intellectual deterioration, mental disturbances and seizures.[5] This case reminds the ophthalmologists about the systemic nature of this condition and draws attention to the need for neuroimaging for unexplained visual loss in patients with PXE.

References

1 Johnson MW, Kincaid MC, Trobe JD. Bilateral retrobulbar optic nerve infarctions after blood loss and hypotension: a clinicopathological case study. *Ophthalmology* 1987; **94**: 1577–1584.
2 Fasshauer K, Reimers CD, Gnau HJ *et al*. Neurological complications of Gronblad–Strandberg syndrome. *J Neurol* 1984; **231**: 250–252.
3 Mayer SA, Tatemichi TK, Spitz JL *et al*. Recurrent ischemic events and diffuse white matter disease in patients with pseudoxanthoma elasticum. *Cerebrovasc Dis* 1994; **4**: 294–297.
4 van den Berg JS, Hennekam RC, Cruysberg JR *et al*. Prevalence of symptomatic intracranial aneurysm and ischaemic stroke in pseudoxanthoma elasticum. *Cerebrovasc Dis* 2000; **10**: 315–319.
5 Iqbal A, Alter M, Lee SH. Pseudoxanthoma elasticum: a review of neurological complications. *Ann Neurol* 1978; **4**: 18–20.

AKV Aralikatti[1], MW Lee[1], ME Lipton[2] and GG Kamath[1]

[1]Department of Ophthalmology
Arrowe Park Hospital
Wirral, UK

[2]Department of Radiology
Arrowe Park Hospital
Wirral, UK

Correspondence: AKV Aralikatti
Department of Ophthalmology
Arrowe Park Hospital
Arrowe Park Road
Upton, Wirral
Merseyside CH49 5PE, UK
Tel: +44 (0) 151 678 5111
Fax: +44 (0) 151 604 7152
E-mail: anildipali@hotmail.com

Sir,

Reattachment of extensive Descemet's membrane detachment following uneventful phaco-emulsification surgery

Eye (2002) **16,** 786–788. doi:10.1038/sj.eye.6700079

Descemet's membrane detachment (DMD) is an uncommon condition with a wide range of possible aetiologies. Probably the commonest cause is a localised detachment occurring at cataract extraction surgery. More extensive detachments give rise to a 'double anterior chamber' after perforation of Descemet's membrane in deep lamellar keratoplasty.[1] Large DMD after cataract extraction is rare. Non-surgical factors that could predispose to DMD are traumatic, congenital glaucoma and corneal ectasias, among others.

DMD detachment can be classified as:

Planar: <1 mm separation from the stroma

- Peripheral detachment only
- Combined peripheral and central detachment

Non-planar: >1 mm separation from the stroma

- Peripheral detachment only
- Combined peripheral and central detachment.[2]