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AM Abu El-Asrar<sup>1</sup>, S Jestaneiah<sup>1</sup> and AM Al-Serhani<sup>2</sup>

<sup>1</sup>Department of Ophthalmology  
College of Medicine  
King Saud University  
Riyadh, Saudi Arabia

<sup>2</sup>Department of Otorhinolaryngology  
College of Medicine  
King Saud University  
Riyadh, Saudi Arabia

Correspondence: AM Abu El-Asrar  
Department of Ophthalmology, King Abdulaziz  
University Hospital, Airport Road, PO Box 245  
Riyadh 11411, Saudi Arabia  
Tel: +966 1 4775724/4775741  
E-mail: abuasrar@KSU.edu.sa

Sir,

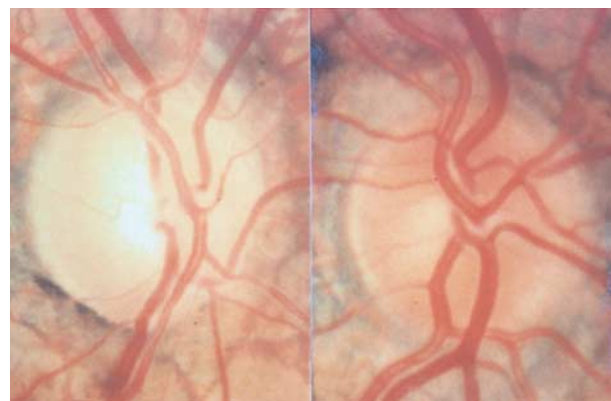
### Pseudoxanthoma elasticum and nonarteritic anterior ischaemic optic neuropathy

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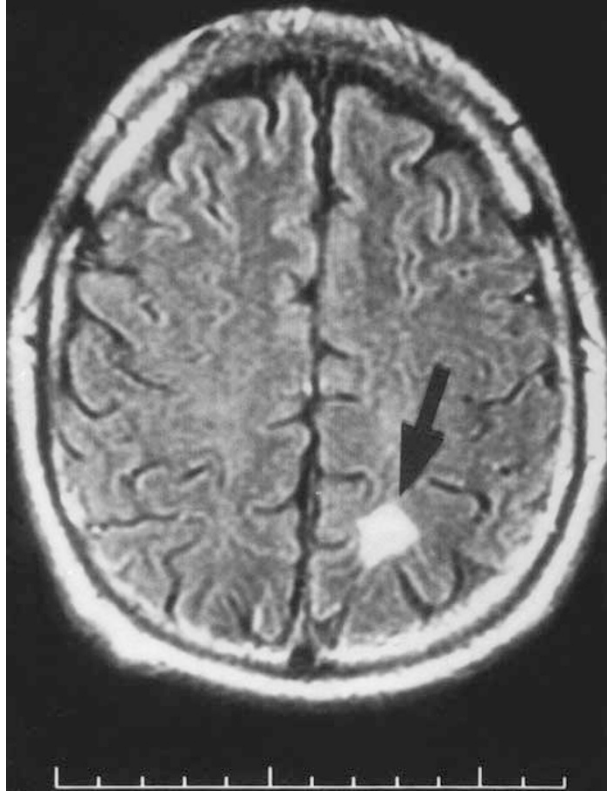
Patients with pseudoxanthoma elasticum can have extensive arteriosclerotic changes at an early age. This vascular pathology can result in premature coronary artery disease and cerebrovascular disease.<sup>1–3</sup> We present a young patient with pseudoxanthoma elasticum, who we believe developed nonarteritic anterior ischaemic optic neuropathy secondary to arteriosclerotic changes in the optic disc vasculature.

#### Case report

A 36-year-old gentleman presented with blurred vision in his left eye. His best visual acuities were 6/9 OD and 6/18 OS. There was a generalized pallor of the optic disc in the right eye with the cup disc ratio being 0.6 in the right eye and 0.3 in the left eye (Figure 1). There was no relative afferent pupillary defect. In the left eye, a superior temporal branch retinal vein occlusion at an arteriovenous crossing was noted along with macular oedema. Angioid streaks were noted in both the fundi. The intraocular pressures were unremarkable at 16 mmHg OD and 18 mmHg OS. Visual field examination revealed superior and inferior arcuate field defects in the right eye and a paracentral scotoma in the left eye. He was also found to have yellow papules arranged in a linear pattern on his neck. A diagnosis of pseudoxanthoma elasticum (PXE) was made. MRI revealed the presence of high signal lesions, suggestive of ischaemic areas, in the left high parietal lobe and adjacent to the anterior horn of the right and left lateral ventricle (Figure 2). There was no evidence of any compressive



**Figure 1** Optic disc photographs showing the pale, cupped disc in the right eye (right) and the normal optic disc in the left eye (left).



**Figure 2** MRI scan showing the presence of a high signal lesion (black arrow), suggestive of an ischaemic area, in the left high parietal lobe.

pathology in the anterior visual pathway. There was no history of ocular trauma.

Blood pressure, full blood count, serum lipid profile, and fasting blood sugar were normal. Carotid doppler was unremarkable. Thrombophilia screen did not reveal any abnormalities and antiphospholipid antibodies were absent.

To summarize, a patient with PXE presented with a pale, cupped disc in the right eye and a branch retinal vein occlusion in the left eye.

### Comment

The disc pallor and cupping in the right eye of this patient might have been caused by anterior ischaemic optic neuropathy, which in turn could have resulted from arteriosclerotic changes in the optic disc vasculature secondary to PXE. Similar arteriosclerotic changes in the cerebral vessels might have also resulted in cerebral infarcts and the corresponding high signal MRI lesions.<sup>4</sup>

Arteriosclerosis is frequently seen in elderly population, but extensive arteriosclerotic changes can be seen at an early age in patients with PXE.<sup>1</sup> The pathogenesis appears to involve abnormalities of elastic

fibres with aberrant calcification.<sup>1</sup> It is believed that the elastic media is the primary site of vessel calcification. This is then followed by intimal calcification. Later intimal fibrous tissue proliferation and atheromatous plaques occlude the lumen.<sup>1</sup> Although the vascular manifestations of PXE affect primarily the middle-sized arteries, it can also result in diffuse 'small vessel' arteriopathy.<sup>2</sup> The presence of arteriosclerotic changes in our patient is supported by the branch retinal vein occlusion and the arteriovenous crossing changes in the fundi.<sup>5</sup>

In addition to glaucoma, optic disc cupping has been reported to occur following anterior ischaemic optic neuropathy and compressive lesions of anterior visual pathway.<sup>6</sup> Normal tension glaucoma is unlikely in our patient as it is usually a disease of the elderly; the mean reported age is in the 60s.<sup>6</sup> Also the greater neuroretinal rim pallor, as seen in our patient, goes in favour of nonglaucomatous cause for the disc cupping.<sup>6</sup> There was no compressive lesion noted on the MRI in our patient.

To summarize, we believe that PXE can cause arteriosclerotic changes in the optic disc vasculature, which can result in a nonarteritic anterior ischaemic optic neuropathy.

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S Murthy and S Prasad

Arrowe Park Hospital, Wirral, CH49 5PE, UK

Correspondence: S Murthy  
Birmingham and Midland Eye Center  
Birmingham B18 7QH, UK  
Tel: +44 121 6 27 8 349  
Fax: +44 121 6 27 8 789  
E-mail: ynsms@hotmail.com