

## Central Retinal Artery Occlusion in a Patient with Pseudoxanthoma Elasticum

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### ABSTRACT

We examined a patient with central retinal artery occlusion (CRAO) who was diagnosed as having pseudoxanthoma elasticum and exhibited angioid streaks. Echocardiography revealed stenosis and plaques of the right carotid artery. Magnetic resonance imaging (MRI) showed multiple cerebral infarctions, which were considered to be the result of vascular-endothelial abnormalities associated with pseudoxanthoma elasticum. Systemic examination of any plaque which may cause CRAO is recommended.

**Key words:** *Pseudoxanthoma elasticum, Angioid streaks, Central retinal artery occlusion, Vascular-endothelial abnormalities*

Pseudoxanthoma elasticum is a hereditary connective tissue disease which involves the elastic fibers of skin, vessels, endo- and/or epi-cardium and ocular fundus. The main ocular involvements are manifested as angioid streaks, which are caused by the degeneration of elastic fibers in the Bruch's membrane. Choroidal neovascularization of macula<sup>1)</sup> and subretinal hemorrhage<sup>4)</sup> are not rare. We report a patient with CRAO and pseudoxanthoma elasticum who demonstrated angioid streaks.

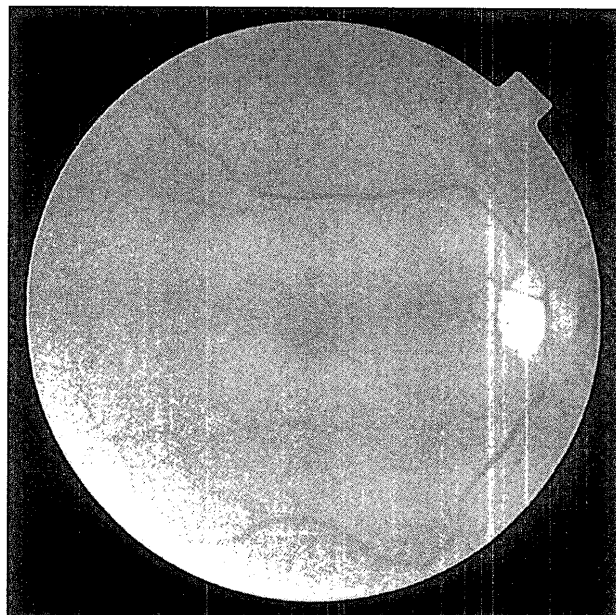
### CASE

A 72-year-old woman with hypertension lost the vision in her right eye when she was having breakfast. On first examination, the corrected visual acuity was hand motion on the right eye and 0.7 on the left eye. The intraocular pressure was normal in both eyes. A relative afferent pupillary defect was distinct in her right eye. The posterior fundus of her right eye was pale and edematous with a cherry red spot (Fig. 1). Angioid streaks were recognized, which extended from the optic nerve disc in each eye (Fig. 2). She had a history of amaurosis fugax, which lasted for an hour, 2 months before this vision loss. Her illness was diagnosed as CRAO, and eyeball massage was performed during and after an intravenous injection of 300 ml of 20% mannitol.

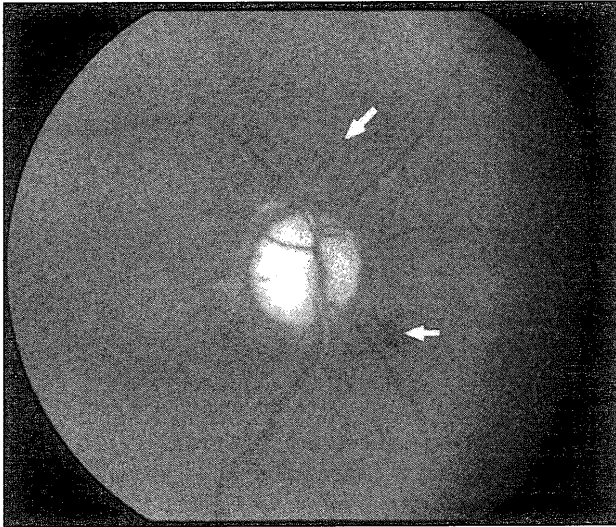
Her neck skin was yellow-white, soft and flabby, with oval- to spindle-shaped papules which were

pea-size and slightly elevated. She was diagnosed as having pseudoxanthoma elasticum histopathologically.

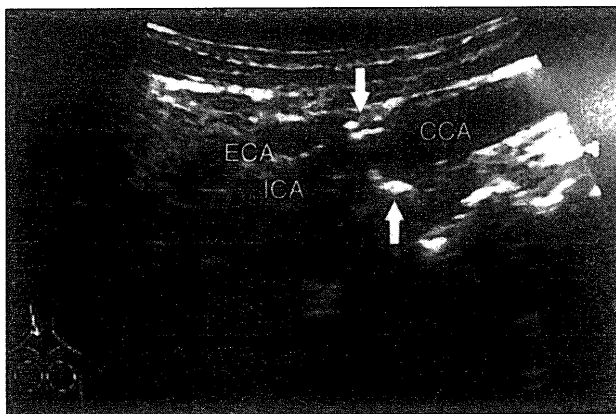
Precordial echocardiography (Color Doppler SSD-2200, Aloka, Japan) showed symmetrical left ventricular hypertrophy. Endocardial plaques were not found, however, and ultrasonography of



**Fig. 1.** Fundus photography of the right eye shows an edematous and pale colored retina, except in the area perfused by the chorioretinal artery.



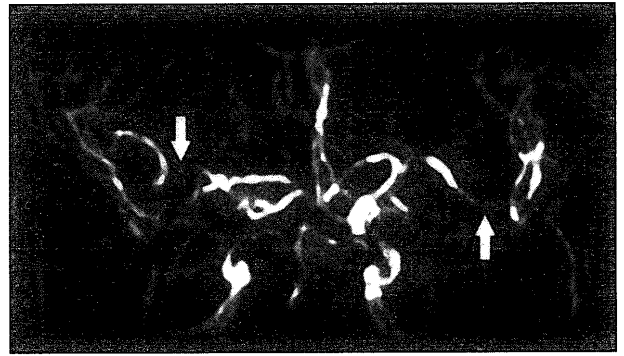
**Fig. 2.** Angioid streaks are spread out radially from the optic nerve head (arrow).



**Fig. 3.** Echocardiography of a plaque (arrow) at the divergence of the right carotid artery. CCA, common carotid artery; ICA, internal carotid artery; ECA, external carotid artery.

the carotid artery revealed stenosis and plaque formation with calcification at the divergence of the right internal carotid artery (Fig. 3). Brain MRI showed multiple cerebral infarctions, and multifocal stenosis of the brain artery, surrounding the bilateral middle cerebral artery, was recognized by magnetic resonance angiography (Fig. 4).

It has been reported that only 2% of patients with amaurosis fugax or CRAO have emboli in the heart<sup>5</sup>. Seventy-seven percent of patients with central/branch retinal infarctions have atherosclerotic stenosis or occlusions of the carotid artery<sup>2</sup>. In our case, echocardiography showed plaques in the internal carotid artery, but not in the heart.



**Fig. 4.** Magnetic resonance angiography shows multifocal stenosis of the brain artery surrounding the bilateral middle cerebral artery (arrow).

Stenosis and plaque formation were confirmed by ultrasonography. Moreover, Schievink et al<sup>3</sup> reported that the characterization of neurovascular disease in pseudoxanthoma elasticum is intracranial aneurysms and cerebral ischemia caused by premature arterial occlusive disease. Our case had multiple stenosis of the brain artery surrounding the bilateral middle cerebral artery.

We considered that these plaque formations and arterial stenosis were the result of changes in the endothelium of vessels associated with pseudoxanthoma elasticum.

Our case indicates that retinal-arterial occlusive disease can occur in patients with pseudoxanthoma elasticum who exhibit angioid streaks.

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