

Fundus signs in temporal arteritis

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SUMMARY A patient with temporal arteritis developed a variety of ischaemic lesions in the eyes. Infarction of the inner retina and optic nerve head was delineated on presentation by white swelling in the retinal nerve fibre layer. The role of interrupted axoplasmic transport in the production of this sign is discussed. Outer retinal infarction was also noted on presentation and subsequently gave rise to striking pigmented scars.

Temporal arteritis often presents with visual loss, and necropsy examination in such cases shows widespread disease of the ophthalmic artery and the extraocular course of its ciliary and retinal branches (Henkind *et al.*, 1970). The medial and lateral posterior ciliary arteries supply the optic nerve head, the outer retina, and, in 20 to 50% of individuals, a variable area of inner retina contiguous with the optic disc (Hayreh, 1969); the central retinal artery supplies the remainder of the inner retina.

We present a patient with a unique combination of fundus signs resulting from arteritic obstruction of the blood supply to each of the above arterial territories. The clinical findings are correlated with recent experimental work on the pathophysiology of ischaemic fundus lesions.

Case report

A 75-year-old woman presented with a 2-week history of 'misty' vision in each eye. She had suffered weight loss, bitemporal headaches, jaw claudication, and aching limbs during the previous 3 months.

On examination vision in each eye was found to be reduced to perception of hand movements only. Pupil reactions were sluggish, and there was a left relative afferent pupillary defect. Examination of the right fundus revealed pallor and swelling of the optic disc and mild translucent swelling of the temporal parapapillary retina (Fig. 1). The ischaemic retina was supplied by two small cilioretinal arterioles and was delineated temporally by a discrete border zone of opaque swelling of the nerve fibre layer (cotton-wool spots). The central arterioles showed irregular narrowing consistent with age, and

the central venous tributaries were of normal calibre and colour. No abnormality of the inner retina was noted in the territory of supply of the central retinal artery.

At first sight the left eye showed a similar ophthalmoscopic picture, with pale swelling of the nasal part of the optic disc and a row of fluffy white cotton-wool spots crossing the papillomacular bundle (Fig. 2). However, there was no cilioretinal arteriolar supply to this eye, and the temporal parapapillary retina was normal in appearance. The remainder of the fundus showed diffuse translucent swelling of the inner retina sparing the fovea, and a band of opacity of the outer retina was noted in the macula (Fig. 2). The central arterioles were attenuated, and the venous blood was markedly deoxygenated.

Fluorescein fundus angiography on the right showed satisfactory filling of the central retinal artery but slow filling of the choroid, optic nerve head, and cilioretinal arterioles. In the left eye markedly impaired perfusion was evident in the territories of supply of both the posterior ciliary arteries and the central retinal artery. Both optic discs showed excessive dye leakage in the late pictures. The right eye had a normal electroretinogram (ERG) and electro-oculogram (EOG), but there were minimal electroretinal responses from the left eye.

The patient appeared to be indifferent to her loss of sight. No other abnormality was found on general examination. The erythrocyte sedimentation rate (ESR) was raised at 116 mm in the first hour. The temporal arteries were palpable but non-pulsatile, and biopsy of the right superficial temporal artery showed inflammatory cell infiltration and thickening of the vessel wall with giant cells, thus confirming the clinical diagnosis of temporal arteritis.

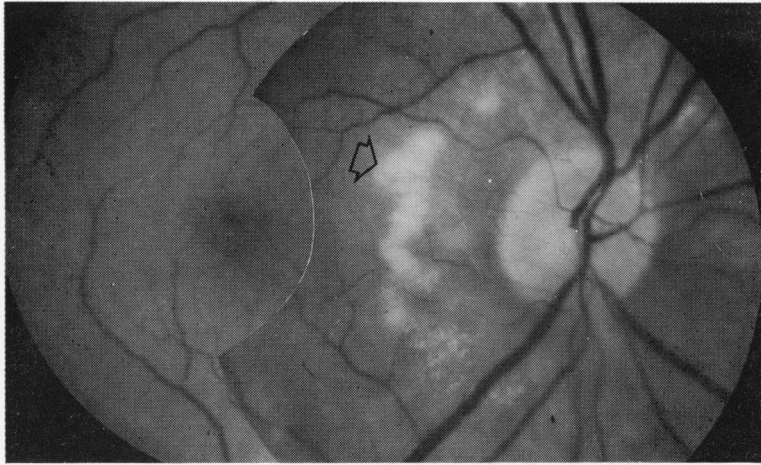


Fig. 1 Composite fundus photograph of right eye on presentation. Cotton-wool spots (open arrow) delineate temporal border of ischaemic cilioretinal arteriolar territory. Nasal part of optic disc is swollen

Fig. 2 Composite fundus photograph of left eye on presentation. Cotton-wool spots (open arrow) delineate ischaemic temporal retina. Nasal part of optic disc is swollen. Outer retinal infarction inferotemporal to fovea (fine arrows)

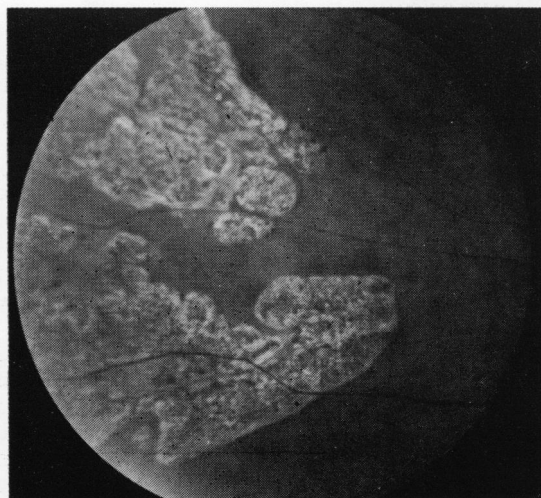
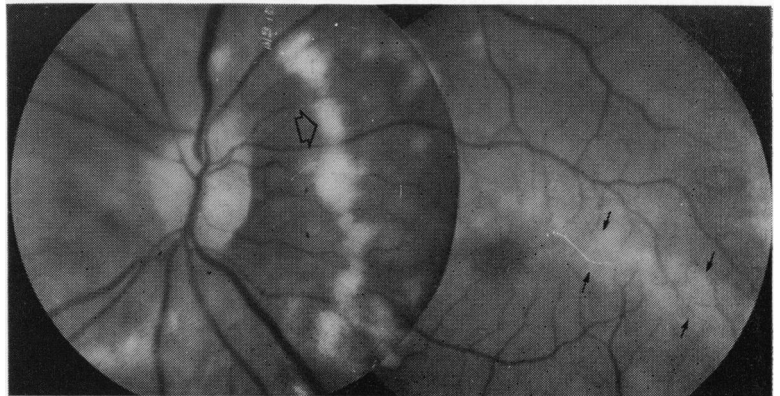


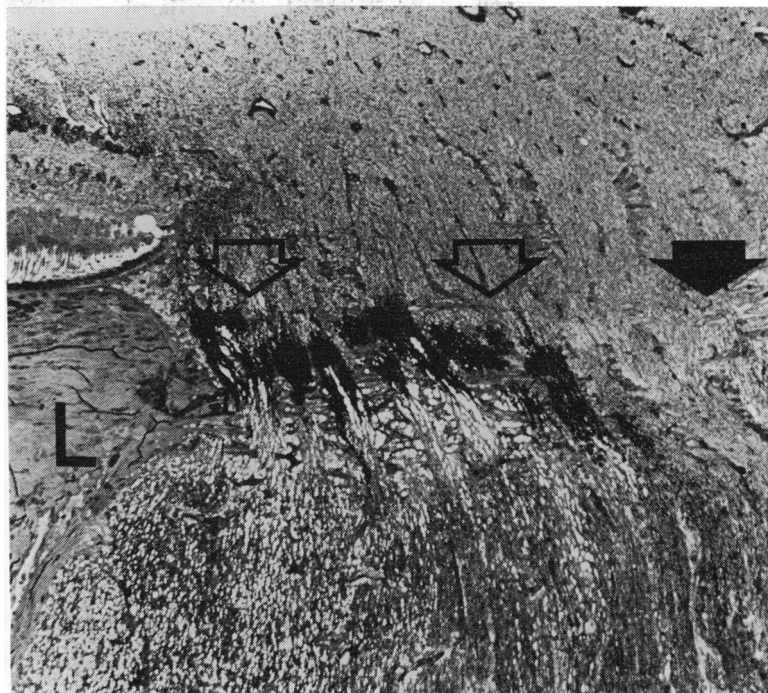
Fig. 3 Fundus photograph of nasal retina of left eye 1 month after presentation. Pigmented lesions of outer retina in territory of medial posterior ciliary artery

Treatment with systemic steroids was instituted immediately. The patient initially received 90 mg of prednisolone daily, and this was gradually reduced as the ESR became normal. A month after she was first seen her vision in the right eye had improved to 6/9 and in the left eye to 6/36. However, the fields of vision were grossly restricted, both eyes showing complete inferior altitudinal defects. The optic discs were pale but not cupped, and the retinal nerve fibre bundle striations were absent, indicating loss of neurones. In the left eye wedge-shaped areas of pigment epithelial disturbance were noted nasal to the optic disc (Fig. 3), and bands of pigmentary change (Siegrist streaks) were also present inferotemporal to the fovea where outer retinal infarction had been seen on presentation. The left relative afferent pupillary defect persisted.

Discussion

Ischaemic optic neuropathy, as seen in the right eye of our patient, is the commonest ocular presentation

Fig. 4 Autoradiograph of horizontal section through the optic nerve head of a monkey. The lateral posterior ciliary artery was occluded and ^3H -leucine was then injected intravitreally and left for 5 hours before enucleation. Ischaemic vacuolation in laminar (L) and retrolaminar regions temporally. Intra-axonal accumulation of label in prelaminar region temporally (open arrows). Label transported normally into nasal part of optic nerve (solid arrow)



of temporal arteritis and results from involvement of the posterior ciliary circulation in the arteritic process (Foulds, 1969; Hayreh, 1969). Experimental occlusion of the posterior ciliary arteries in monkeys resulted in similar disc swelling (Hayreh and Baines, 1972a), but no definitive histological investigation of

the acute disc lesion was carried out. Our recent investigation (McLeod *et al.*, 1978) has substantiated the hypothesis (McLeod, 1975, 1976; Hayreh, 1977) that the disc swelling results from interruption of orthograde axoplasmic transport in retinal ganglion cell axons (Figs. 4 and 5).

In our patient's right eye occlusion of the posterior ciliary arteries also resulted in parapapillary infarction of the inner retina. The cotton-wool spots which delineated the temporal border of the chorioretinal infarct can also be attributed to obstruction of orthograde axoplasmic transport in the retinal nerve fibre layer (McLeod, 1975). Such an accumulation of axoplasm has been demonstrated in autoradiographic studies of retinal infarcts in pigs (McLeod *et al.*, 1977).

The cotton-wool spots in our patient's left eye delineated the nasal border of a large infarct of the inner retina which resulted from partial occlusion of the central retinal artery (Oji and McLeod, 1978). This accumulation of axoplasmic debris corresponds to that seen on the disc side of retinal infarcts produced experimentally (McLeod *et al.*, 1977) and reflects obstruction of retrograde axoplasmic transport in the retina. The band and wedge-shaped infarcts of the *outer* retina resulted from arteritic involvement of the posterior ciliary arterial supply to the left eye. Although such infarcts are readily produced by occlusion of the posterior ciliary

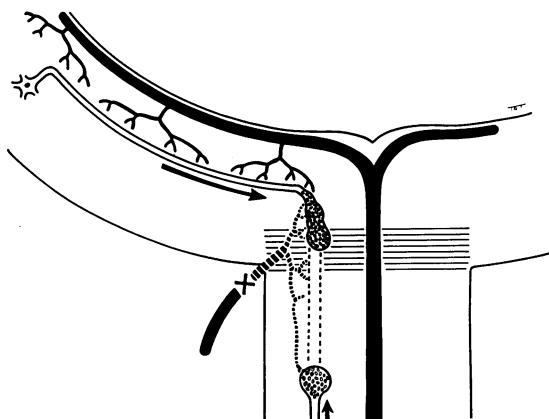


Fig. 5 Diagram of changes in a retinal ganglion cell axon after occlusion (X) of posterior ciliary artery. Ischaemic vacuolation in optic nerve head (axolemma dotted). Axonal swellings from obstructed orthograde (long arrow) and retrograde (short arrow) axoplasmic transport

arteries in monkeys (Hayreh and Baines, 1972b), they are relatively unusual in temporal arteritis, probably owing to incomplete occlusion of the posterior ciliary supply and some capacity of the inner retinal circulation to sustain outer retinal viability during choroidal ischaemia. It is therefore of interest that in our patient, and also in a case previously illustrated (McLeod, 1973), outer retinal infarcts developed in eyes with combined involvement of the central retinal and posterior ciliary circulations.

Finally, the improvement in central vision which followed treatment of our patient with systemic steroids illustrates the remarkable ability of neurones to survive prolonged ischaemia. Recovery of central vision despite infarction of the papillomacular bundle has previously been noted in cases of cilioretinal infarction (McLeod and Ring, 1976) and probably reflects the survival of axons on the vitreal aspect of the retinal nerve fibre layer.

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