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COMMUNICATIONS

CILIARY ARTERY INVOLVEMENT IN GIANT CELL ARTERITIS*†

BY

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THIS condition was first established as a clinical and pathological entity by Horton, Magath, and Brown (1932), who gave it the name temporal arteritis. Although involvement of the temporal arteries is a frequent and often dominant clinical feature, it has long been known that many other arteries may be similarly affected and the term cranial arteritis was suggested from involvement of the branches of the internal and external carotid arteries. Other branches of the aorta may be affected and it has been shown that small arteries are vulnerable as well as larger ones (Crompton, 1959). Frequently there are associated features of a general illness, such as malaise, anorexia, fever, and loss of weight. That the condition is a generalized vascular disease was emphasized by Cooke, Cloake, Govan, and Colbeck (1946) and Heptinstall, Porter, and Barkley (1954) and it is now usually called 'giant cell arteritis', thereby stressing one of its histological features.

The ocular complications of giant cell arteritis have been reviewed by Wagener and Hollenhorst (1958), Parsons-Smith (1959), and Simmons and Cogan (1962). Ocular lesions occur in about 40 per cent. of cases, and complete blindness in one or both eyes follows in about 30 per cent. of cases (Meadows, 1965). In some cases the ophthalmoscopic appearance is that of central retinal artery occlusion. In many cases, however, the loss of vision is much greater than would be expected from the fundus appearance, thus suggesting the possibility of optic nerve ischaemia. Meadows (1965) stated that central retinal artery occlusion occurred clinically in only three out of forty cases in which blindness occurred as a result of giant cell arteritis. Blindness can therefore result from occlusion of the central retinal artery or of the posterior ciliary artery system.

There have been few reports of the *post mortem* changes in the visual pathway in giant cell arteritis. Crompton (1959) refers to three cases in the literature with involvement of the ciliary arteries, in addition to his own case. The purpose of this paper is to present the histological features of a further case and to emphasize the role of ciliary artery occlusion as a cause of blindness in giant cell arteritis, and the importance of considering giant cell arteritis in the differential diagnosis of optic nerve ischaemia in elderly people.

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Case Report*

A 68-year-old man presented with intermittent blurring of vision of the right eye. The next day vision went completely and at this time there was arterial narrowing and some optic disc oedema, but the typical appearance of central retinal artery occlusion was not present; 12 days later there was sudden loss of vision in the left eye and the fundus appearance was similar to that previously seen in the right eye. The patient was hypertensive, B.P. 220/170, and the erythrocyte sedimentation rate was 12 mm./1 hr. The temporal arteries were prominent and tortuous but pulsatile. A provisional diagnosis was made of temporal arteritis and possible obliterative occlusion of the central retinal arteries. Six days later the patient complained of sudden chest pain on the left side and he died 4 hours later. A full *post mortem* examination was carried out and the contents of both orbits were sent to the Institute of Ophthalmology for histological examination.

Pathological Findings.—The cause of death was found to be acute left ventricular failure due to coronary artery disease. The heart weighed 490 g. (normal 300 g.). There was moderate left ventricular hypertrophy but all chambers and valves were normal and there was no visible cardiac infarction. Microscopical examination of the coronary arteries showed diffuse calcification with fibrosis and intimal thickening but no arteritis. The aorta showed marked atheroma of the arch and descending aorta, and microscopical examination showed marked changes of arteritis with intimal proliferation and degeneration of elastic fibres in the areas of more intense cellular reaction. Severe giant cell arteritis was present in the right thyrocervical trunk and in both external carotid arteries with obliteration of the lumen on the right side. Both internal carotids showed patchy involvement whilst severe arteritis with many giant cells and intimal proliferation was present in both temporal arteries. No evidence of arteritis was found in small vessels from the gut, liver, spleen, and kidneys.

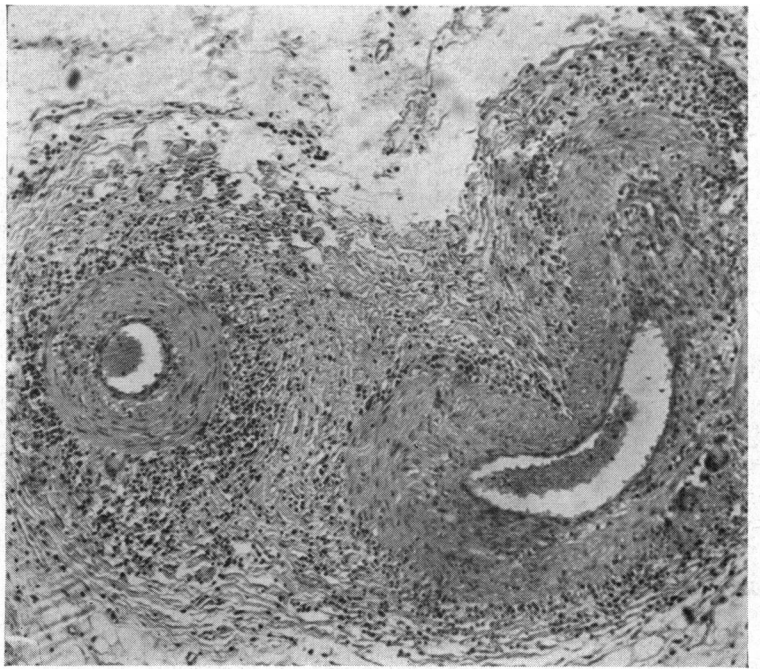

FIG. 1.—Section of ophthalmic artery and a branch. The artery on the left is relatively normal apart from some inflammatory cell infiltration in the adventitia. The artery on the right shows marked intimal proliferation and reduction of lumen and three giant cells one of which is in the adventitia. Haematoxylin and eosin. × 64.

*The clinical details of this case have been reported elsewhere (Cullen, 1963).

RIGHT ORBIT

Macroscopical examination showed extreme atheroma of the internal carotid artery. The ophthalmic artery and all its branches were enlarged and thickened, particularly at the origin of the central retinal artery, where there was a nodular swelling and abnormal angulation of the vessels.

Microscopical examination of serial cross-sections through the ophthalmic and central retinal artery showed that both vessels were greatly thickened and fibrosed with an intense chronic inflammatory infiltration in the adventitia. In some sections giant cells and medial calcification were prominent and others showed marked fibroblastic proliferation of the intima (Fig. 1). The central retinal artery was markedly involved in the inflammatory process and in some sections the lumen was almost completely occluded.

RIGHT EYE

Macroscopical examination showed some oedema around the disc with collapse of all retinal arteries.

Microscopical examination showed no gross abnormality of the cornea, iris, or angle of the anterior chamber. Apart from marked anterior cystic degeneration of the retina, significant abnormalities were present only in the posterior segment of the globe where there was marked oedema of the retina, particularly at the macula and disc. The retinal vessels were generally collapsed and occasionally showed perivasculitis, but no signs typical of giant cell arteritis, and the classical picture of central retinal arterial occlusion was not present. There was generalized loss of ganglion cells but *post mortem* changes only were seen in the rod and cone layer. The choroidal vessels showed arteriosclerotic changes only. The short ciliary arteries, however, both outside the globe and in the sclera, showed advanced arteritis with intimal proliferation, medial necrosis, giant cell reaction, and intense inflammatory cell infiltration in the adventitia (Fig. 2, and Figs 3 and 4, overleaf).

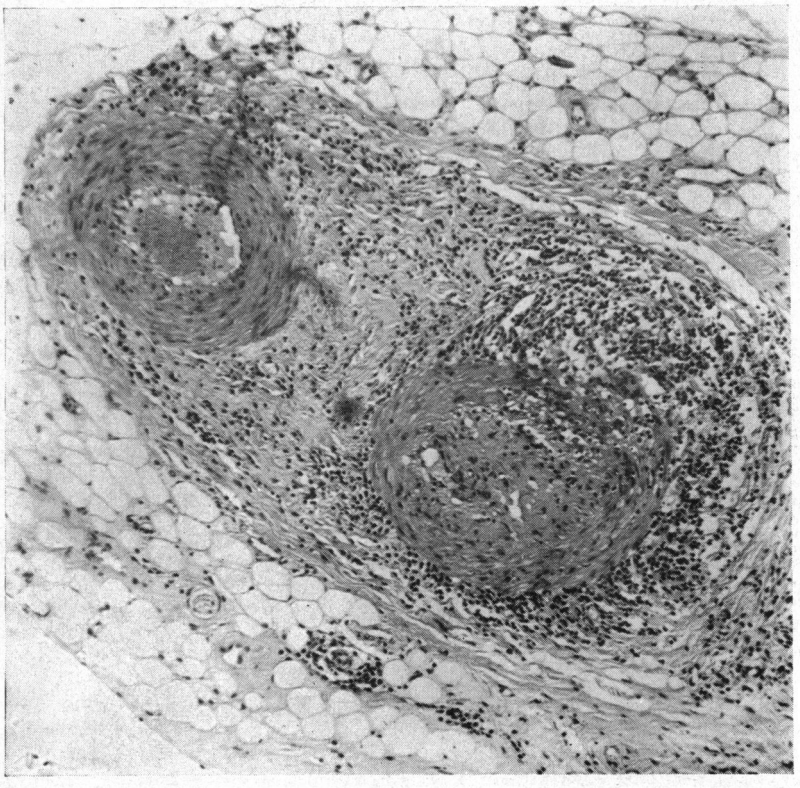

FIG. 2.—Section of posterior ciliary arteries. The artery on the left appears to be normal. The abnormal artery on the right shows occlusion and beginning recanalization of the lumen and is surrounded and infiltrated by chronic inflammatory cells. Haematoxylin and eosin. × 64.

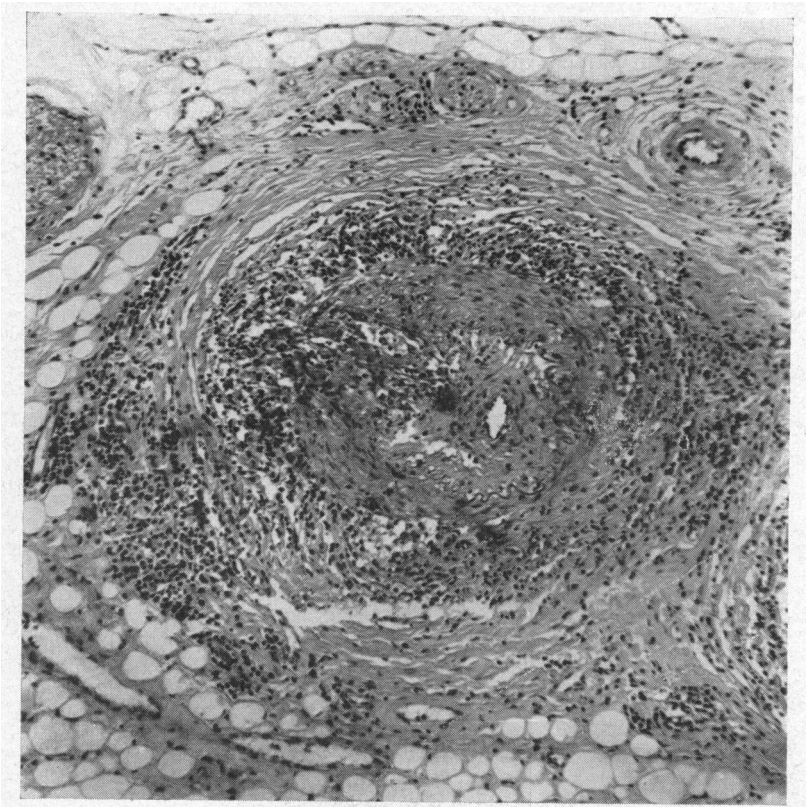

FIG. 3.—Section of posterior ciliary artery. There is occlusion of the lumen, infiltration with chronic inflammatory cells, and fragmentation of the internal elastic lamina. Haematoxylin and eosin. × 64.

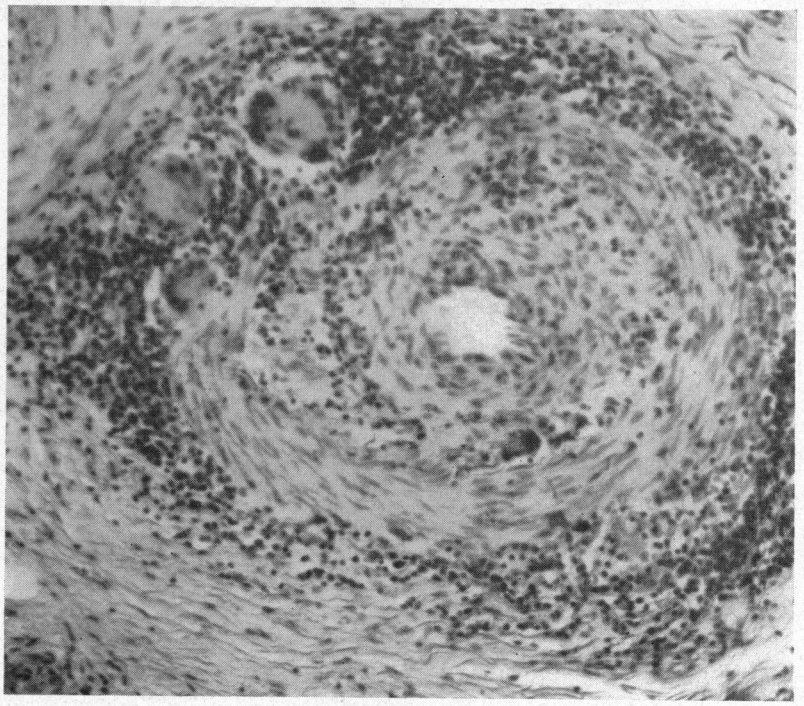

FIG. 4.—High-power view of short posterior ciliary artery. There is marked narrowing of the lumen with proliferation of the intima. Three giant cells are present in the media between 9 and 11 o'clock and another is related to the fragmented elastic lamina at 5 o'clock. Haematoxylin and eosin. × 113.

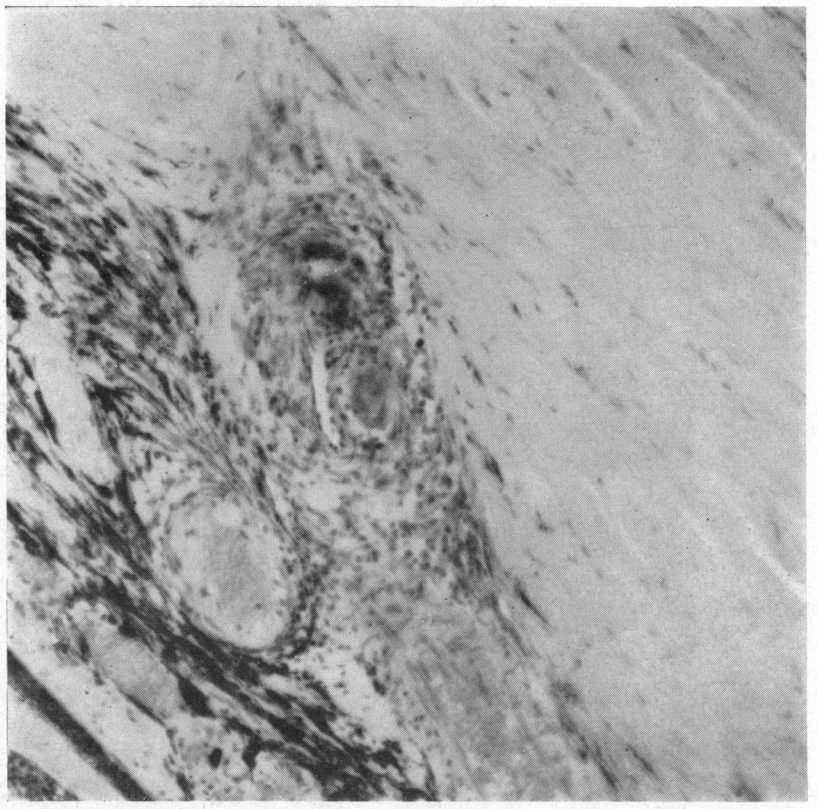

FIG. 5.—High-power view of short ciliary artery entering the choroid. There is narrowing of the artery and several giant cells are present in the vessel wall. The inflammatory changes cease abruptly at the choroid. Haematoxylin and eosin. ×113.

The changes seen in the intrascleral arteries of the right eye ceased abruptly at the choroid (Fig. 5).

LEFT EYE

Macroscopical examination showed marked swelling of the disc.

Microscopical examination showed no gross abnormality of the anterior segment. There was marked anterior cystic degeneration of the retina. In general the changes were similar to those seen in the posterior segment of the right eye but with rather more optic nerve oedema. Cross-sections of the optic nerve showed typical giant cell arteritis in the ciliary vessels around the nerve (Fig. 6, overleaf), while the nerve itself showed oedema, collapse of the capillaries, and ischaemic degeneration behind the lamina cribrosa. Although the intrascleral arteries were involved, no changes typical of giant cell arteritis were seen in the choroidal vessels which showed arteriosclerotic changes only.

Discussion

The basic lesion responsible for the ocular manifestations in giant cell arteritis is a progressive occlusive process affecting the ophthalmic artery and its branches to the optic nerve, retina, and extra-ocular muscles. Blindness in one or both eyes is the most serious and most common end-result of ocular involvement. Although central retinal artery occlusion has been demonstrated both ophthalmoscopically and histologically, ischaemic optic neuritis or infarction of the optic nerve is probably the commonest cause of visual loss in giant cell arteritis. Absence of the typical fundus appearance of retinal artery occlusion in the presence of profound visual loss could be explained by occlusion of the artery far back in its course at or near its origin from the ophthalmic artery.

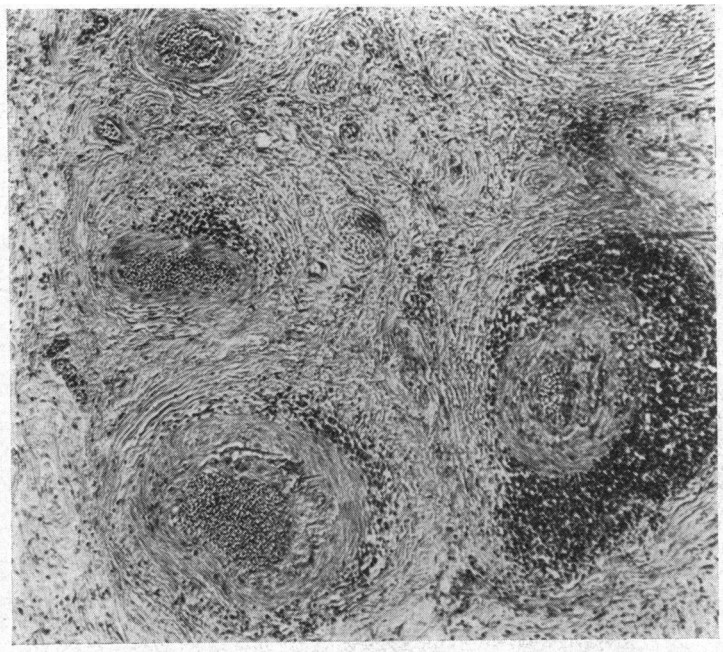

FIG. 6.—Group of short posterior ciliary arteries behind the nerve head. One vessel shows marked fragmentation of the internal elastic lamina and there is an intense inflammatory cell reaction around the artery on the right. Haematoxylin and eosin. × 64.

The severity of functional loss resulting from ischaemia depends on the degree of obstruction to the blood supply of the affected part and on the existence of a collateral blood supply. The intra-orbital part of the optic nerve receives its arterial supply from the ophthalmic artery directly and *via* the pial branches of the long and short posterior ciliary arteries, and also from the central retinal artery. The macula is supplied only by posterior ciliary arteries. Connexions between the retinal and ciliary arterial systems in the region of the lamina cribrosa have been described and may have some physiological significance, but it is unlikely that they contribute much if one or other system is occluded alone. Meadows (1954) suggested that involvement of the short ciliary arteries concerned in the formation of the arterial circle of Zinn around the optic nerve head might be responsible for severe visual loss unaccompanied by fundus changes other than slight optic disc oedema. Thrombosis is not a constant histological feature in this condition and it is likely that intimal proliferation is the main cause of reduction in size of the arterial lumen resulting in ischaemia of the optic nerve.

Although blindness is common in giant cell arteritis, there are few published accounts of *post mortem* findings in cases with visual pathway involvement. This may be because giant cell arteritis is rarely fatal and removal of an eye is unlikely to be required. Cardell and Hanley (1951) described a case with giant cell arteritis involving the ophthalmic, left central retinal, and right posterior ciliary arteries. Greenfield (1951) described a case with intense arteritis affecting the central retinal and ciliary arteries of both eyes spreading into the intrascleral arteries and causing extreme reduction of lumen. Both ophthalmic arteries were involved in one of the cases described by Heptinstall and others (1954). Crompton (1959) examined the whole visual pathway in a man with bilateral blindness due to giant cell arteritis. There was involvement of the ophthalmic, posterior ciliary, central retinal, chiasmatic,

vertebral, and other small arteries by severe giant cell arteritis. The nasal branch of the right central retinal artery at the disc was involved by arteritis with complete occlusion of the lumen by organizing fibrinoid material, but the choroidal arteries were not involved. In one of the cases described by Cooke and others (1946), both central retinal arteries were equally affected with obliteration of the lumen by cellular fibrous tissue, and arteritis could be traced forwards into the main branches of the central retinal arteries, but it is not stated how far the disease extended into the retinal arteries within the globe. Seitz (1961) illustrated his case report with a microphotograph showing giant cell arteritis affecting the central retinal artery and one of its branches on the optic disc, but the intraretinal branches were not involved. This is important in view of the fact that the internal elastic lamina ceases as the artery emerges from the disc and the theory of Kimmelstiel, Gilmour, and Hodges (1952) that giant cell arteritis may have its origin in a specific lesion of elastic membranes.

In a fatal case reported by Spencer and Hoyt (1960), typical lesions of giant cell arteritis were found in the ophthalmic and short ciliary arteries and there was a clear zone of ischaemic necrosis immediately behind the lamina cribrosa of the optic nerve.

Secondary haemorrhagic glaucoma following massive necrosis of the retina and optic nerve was described by Wolter and Phillips (1965).

A 77-year-old woman had a 6-month history of severe headache before sudden loss of sight in the affected eye. The temporal and occipital arteries were clinically normal but the erythrocyte sedimentation rate was raised. The blind eye was removed because of severe pain and the ophthalmic and posterior ciliary arteries were seen to be severely affected by giant cell arteritis. The optic nerve was surrounded by dense scar tissue containing the short posterior ciliary arteries, which showed marked thickening of the walls, narrowing of the lumen, granulomatous inflammation, and many giant cells. Similar changes could be traced into the intrascleral vessels but not into the choroidal vessels.

Although Crompton (1959) showed that small arteries can be affected in this disease, choroidal involvement has not been described and Wolter and Phillips (1965) stated that giant cell arteritis is found only outside the eye and in the sclera and not in the choroidal arteries. In the present case arteritis can be traced right through the sclera in the short posterior ciliary arteries, but no giant cells or elastic degeneration were seen in the choroidal vessels.

Summary

The *post mortem* findings are described in a case of bilateral blindness associated with giant cell arteritis of the ophthalmic, central retinal, and posterior ciliary arteries.

The retinal arteries in the globe are not directly involved and the classical picture of central retinal artery occlusion is not present. Blindness probably resulted from failure of the circulation in the optic nerve due to extensive involvement of the short posterior ciliary arteries around the optic nerve head. Absence of giant cell arteritis in the choroidal vessels is emphasized.

Ischaemia of the optic nerve is the commonest cause of blindness in giant cell

arteritis and this condition should be considered in the differential diagnosis of optic nerve ischaemia in elderly people especially when other signs of the disease are not present.

I should like to thank Professor Norman Ashton for his help and kindness in allowing me to report this case which was originally under the care of Mr. J. M. L. Howat. I am also grateful to Mrs. H. Mehra for secretarial help and to Mr. V. J. Elwood for technical assistance.

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