OCCULT TEMPORAL ARTERITIS*† A COMMON CAUSE OF BLINDNESS IN OLD AGE

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HORTON, Magath, and Brown (1932), working at the Mayo Clinic, introduced the term temporal arteritis for the disease which we still know by this somewhat inaccurate name. The condition had been recognized for many years before this and Jonathan Hutchinson described a case which was probably one of temporal arteritis in 1890. It is a disease of the older age groups, affecting both sexes equally, seldom seen before 55 years and of increasing incidence in the seventh and eighth decades.

In classical temporal arteritis there is swelling, tenderness, and pain over the superficial temporal vessels which are thickened, prominent, often pulseless, and occluded. There are associated general symptoms which may precede the signs in the temples, and these consist of headaches, malaise, anorexia, intermittent pyrexia, and loss of weight, and a raised erythrocyte sedimentation rate is often found. The disease is self-limiting, usually lasting about 6 months, but remissions may occur.

The generalized nature of the vascular involvement in this condition has been overlooked in the past but, in recent years, has been re-emphasized, and the so-called complications of the disease—usually ocular and cerebral—are now recognized as part of the widespread arteritic process. Cooke, Cloake, Govan, and Colbeck (1946) were the first to demonstrate this in necropsy material and their cases showed involvement of the aorta and of the coronary, cerebral, subclavian, femoral, mesenteric, and radial arteries. Subsequent reports from elsewhere have confirmed that, although the brunt of the disease is borne by the carotid arteries and their branches, any part of the arterial system can be affected (Heptinstall, Porter, and Barkley, 1954; Harrison, Harrison, and Kopelman, 1955; Lander and Bonnin, 1956; Paulley and Hughes, 1960). Attempts have, therefore, been made to re-name the condition, and such terms as “arteritis of the aged”, “cranial arteritis”, or “giant cell arteritis” have been put forward, but still the original term persists and will no doubt continue to do so, and continue to mislead the unwary, especially the ophthalmologist, who is thus led to believe that he is to look out for an ocular complication of a typical and obvious condition.

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Ocular involvement in temporal arteritis is well documented, and reports of blindness from the disease have appeared sporadically in the literature in recent years. Whitfield, Bateman, and Cooke (1963) reviewed the literature on the incidence of eye involvement, which varied from 33 to 57 per cent. of cases, and they also reported their own observations on 72 cases seen over a period of 15 years, and in 49 of these ocular involvement was recorded. Twelve of their patients suffered total visual loss in both eyes and three lost all vision in one eye. Partial visual loss was recorded in a further 21 cases.

This high incidence of ocular involvement in a relatively uncommon general disease has impressed the writer for some years and several cases of the condition have been reported (Cullen, 1963) under the title of "occult temporal arteritis", where the general involvement was minimal or subclinical and the patients presented with acute visual failure. This syndrome was so designated by Simmons and Cogan (1962) and its main characteristics can be listed as follows:

- (1) The classical symptoms are absent or present only *after* the ocular phase of the disease.
- (2) The temporal artery signs may not be present or may be so minimal as to be of doubtful diagnostic significance.
- (3) There is often only a history of vague headaches, general malaise, loss of weight, and anorexia.
- (4) The disease presents as a quiet ischaemic blindness in one eye often followed by a similar occurrence in the second eye after an interval of days or weeks.
- (5) A raised erythrocyte sedimentation rate may be found.
- (6) Temporal artery biopsy may be positive despite the absence of classical symptoms in this region.

Because of the belief that temporal arteritis, especially in its occult form, is not as uncommon as generally believed, and because of its relatively frequent presentation with acute visual failure, a determined effort has been made to evaluate its incidence and search for it in cases of sudden visual loss in elderly patients. Thus, during the 2½ years from January, 1963 to July, 1965, the investigation reported below has been carried out.

Material and Methods

Two groups of patients presenting with acute visual failure have been investigated:

- (1) Those presenting with *central retinal arterial occlusion* whether partial or complete—the "C.R.A. Occlusion Group".
- (2) Those presenting with presumed vascular occlusion involving the blood supply to the optic nerve, and these have been classified as *ischaemic optic neuritis*—the "I.O.N. Group".

Only patients over the age of 50 years were included in this study.

The diagnosis of "central retinal artery occlusion" has been straightforward in all cases. The typical picture of arterial spasm or obliteration with oedema of the retina and the "cherry-red spot" at the macula has not always been seen, especially when the visual failure has been of some duration, but still there is always some ophthalmoscopic evidence that an arterial occlusion, whether complete or partial, has taken place.

The diagnosis of "ischaemic optic neuritis" has been made in those who present with acute visual failure, usually complete or nearly so, with inactive or sluggish pupils where a retinal arterial occlusion might have been expected. The fundus signs are, however, minimal or sometimes even a normal fundus picture is seen. More often pale oedema of the disc is present, or pallor without oedema and a few haemorrhages on the disc have been observed in some cases. The retinal

arteries may be attenuated but are never occluded and, in some cases, white patches of exudate or retinal infarcts are present in the posterior fundus.

The ocular signs are, thus, out of proportion to the visual loss and again the diagnosis is easily made and cannot be mistaken once it is borne in mind.

A total of 66 patients aged between 51 and 85 years coming into these two categories was seen, as set out in Tables I and II. All these patients were questioned regarding their general health and especially if they had any symptoms suggestive of temporal or cranial arteritis, and the temporal arteries were palpated in each case.

TABLE I
CENTRAL RETINAL ARTERIAL OCCLUSION IN 46 PATIENTS (4 BILATERAL) = 50 EYES
Age range 51-85 yrs

Complete Central Retinal Arterial occlusion			29
Branch occlusion	Upper division	3	20
	Upper temporal branch	5	
	Lower temporal branch	6	
	Macular vessels	5	
	Lower division	1	
	Lower temporal branch	0	
	Lower nasal branch	0	
C.R.A. and C.R.V. occlusion combined			1
Total No. of Eyes			50

The erythrocyte sedimentation rate (ESR) and a full blood count were carried out and, if the ESR was above 10 mm. in the first hour or if there were any symptoms suggestive of temporal arteritis, a temporal artery biopsy was performed, as set out in Table III.

TABLE II
ISCHAEMIC OPTIC NEURITIS IN 20 PATIENTS
(4 BILATERAL) = 24 EYES
Age range 65-81 yrs

With oedema of disc	21
Without oedema of disc	3
Total No. of Eyes	24

TABLE III
ERYTHROCYTE SEDIMENTATION RATE

Diagnosis	ESR (mm.)	No. of Patients	Biopsy	Side Affected
Central Retinal Arterial Occlusion	1-10	30	Not done	Unilateral
	11-35	10	Negative	1 bilateral
	62-135	6	Positive	3 bilateral
	Total	46		
Ischaemic Optic Neuritis	11-30	8	Negative	Unilateral
	40-131	12	Positive (except one whose ESR was 57 mm.)	4 bilateral
	Total	20		

Results

From this investigation seventeen patients—eleven with ischaemic optic neuritis and six with central retinal arterial occlusions associated with temporal arteritis—were discovered (Table IV). The full details of these patients are listed in Table V.

TABLE IV
SEVENTEEN PATIENTS WITH OCCULT TEMPORAL ARTERITIS
Age range 69–81 yrs (mean 75)

Diagnosis		No. of Cases
Central Retinal Arterial Occlusion	Complete Branch (upper temporal)	5 } 6 (3 bilateral) 1
Ischaemic Optic Neuritis	With oedema of disc (O.N.) Without oedema of disc (R.B.N.)	9 } 11 (4 bilateral) 2

O.N. = Optic neuritis R.B.N. = Retrobulbar neuritis

TABLE V
DETAILS OF SEVENTEEN PATIENTS WITH OCCULT TEMPORAL ARTERITIS
(IN ORDER OF PRESENTATION)

Case No.	Age (yrs)	Sex	Lesion	Eyes Involved	Blood Pressure	ESR (mm.)
1	75	F	O.N.	Both	160/80	109
2	75	F	C.R.A. (Fig. 4)	Both	170/90	108
3	80	F	R.B.N. (R. eye) O.N. (L. eye)	Both	180/80	40
4	69	F	O.N.	Both	156/88	100
5	75	F	R.B.N.	Left	180/90	80
6	77	F	C.R.A.	Left	160/95	102
7	78	F	O.N.	Left	175/80	70
8	76	M	O.N.	Right	150/90	131
9	70	F	C.R.A., C.R.V. (R. eye) C.R.A. (L. eye) (Fig. 5)	Both	130/70	97
10	75	F	O.N. (Figs 1 and 2)	Right	170/70	92
11	71	F	C.R.A. (branch)	Left	180/80	91
12	77	M	C.R.A.	Both	170/95	94
13	81	F	O.N.	Right	130/80	40
14	79	F	O.N. (Fig. 3)	Both	140/80	100
15	72	M	O.N. (Fig. 6)	Right	145/75	50
16	81	M	O.N.	Left	180/80	77
17	71	F	C.R.A.	Left	180/90	135

C.R.V. = Central retinal vein occlusion

C.R.A. = Central retinal artery occlusion
O.N. = Optic neuritis

R.B.N. = Retrobulbar neuritis

In addition, two patients with classical temporal arteritis were seen during this same period in the Eye Department (Table VA). They had been referred because of head pains and headaches which were thought to be due to some ocular condition but, in fact, their eyes were entirely normal and have remained unaffected to date.

TABLE VA
DETAILS OF TWO PATIENTS WITH CLASSICAL TEMPORAL ARTERITIS

Case No.	Age (yrs)	Sex	Eye Involvement	Blood Pressure	ESR (mm.)
18	64	F	None	150/90	105
19	62	F	None	200/110	63

Four Illustrative Case Reports

BILATERAL CENTRAL RETINAL ARTERIAL OCCLUSION

Case 2, a 75-year-old widow, was seen April 27, 1963, because of sudden loss of vision in both eyes the previous day. She admitted on questioning that she had felt unwell for about 10 weeks, had a poor appetite, and had lost one stone in weight. She had shooting pains in the left eye 2 weeks previously. She denied headaches or pain in the temples. She had been a known diabetic for 3 years, controlled by diet, and she had had intermittent claudication since late 1961. She was extremely euphoric and hardly mentioned her blindness. Blood pressure was 170/90. Both superficial temporal arteries were thickened and a faint pulsation was felt in each. The ESR was 108 mm. in the first hour.

Eye Examination.—The visual acuity in the right eye was reduced to counting fingers and, in the left, to hand movements. The pupils were inactive and the fundi showed attenuation of the retinal arterioles with patchy oedema and cherry-red spots at both maculae.

A provisional diagnosis of bilateral central retinal arterial occlusion due to occult temporal arteritis was made and ACTH treatment, 40 units twice daily by intra-muscular injection, was commenced immediately, pending a biopsy report. Within 48 hours the vision in the left eye had dropped to perception of light and eventually this was lost. The vision in the right eye, however, remained at counting fingers, but the peripheral field returned slowly over a period of 4 to 5 months. The biopsy report being positive (Fig. 4, see col. plate overleaf), oral prednisolone was substituted for the ACTH and the ESR fell to 24 mm. after 12 days. The left disc became atrophic, but the right assumed a yellowish, waxy pallor which is typical in this condition. The retinal vessels remained attenuated but not obliterated.

Follow-up.—Corticosteroid treatment has been continued to date for 2½ years. Attempts have been made to stop it, but the patient then feels unwell and complains of extreme lack of energy and the ESR has risen on two occasions from its usual level of below 5 mm. to 18 and 20 mm. in the first hour.

The patient has a full visual field in the right eye with a central scotoma, she lives alone, can manage her daily duties, and gets about unaided.

BILATERAL ISCHAEMIC OPTIC NEURITIS

Case 3, an 80-year-old widow, was first seen in the Eye Department in 1963 because of head pains. No ocular cause was found for these. Visual acuity was full and, apart from arterio-sclerotic retinal vessels, the fundi were normal. On March 1, 1965, she was seen again because of loss of vision in the right eye one week previously. This eye was blind with an inactive pupil and a pale disc. The ESR was 14 mm. in the first hour. A history of myocardial infarction 6 months

previously was obtained. No definite diagnosis was made and observation was arranged; 2 weeks later she returned and stated that her left eye had failed 2 days previously. The visual acuity in the left eye was counting fingers, the pupil was sluggish, and the disc pale and slightly swollen. No temporal artery pulsations were felt and the ESR was now raised to 40 mm. in the first hour. The diagnosis of ischaemic optic neuritis due to occult temporal arteritis was now made and immediate ACTH and prednisolone given but, despite this, the vision in the left eye continued to fail over the next 3 days and finally perception of light was lost. The biopsy was positive. The ESR quickly fell on corticosteroid treatment and this has since been discontinued.

Comment.—This case exemplifies the tragedy of not making the diagnosis at the onset of the condition in the first eye. If this had been done there is no doubt that treatment would have prevented the involvement of the second eye and total blindness would have been prevented.

UNILATERAL CENTRAL RETINAL ARTERIAL OCCLUSION

Case 6, a 77-year-old spinster, presented at the Eye Department on March 12, 1964, because of loss of vision in the left eye the previous day. Her previous history was unremarkable except that, in 1963, she was operated upon for a gastric ulcer which showed evidence of malignancy. She had not felt unwell and did not complain of headaches or head pains. Both temples were somewhat tender on palpation but the temporal arterial pulses were present.

Eye Examination.—The visual acuity in the the right eye was 6/6 and N5. That in the left was reduced to perception of hand movements, the pupil reaction being sluggish, the retinal vessels attenuated, and the disc pale. There was no obvious retinal oedema but a diagnosis of central retinal arterial occlusion was made. The ESR was 102 mm. in the first hour and, therefore, a temporal artery biopsy was performed, which showed typical lesions of temporal arteritis. Oral prednisolone therapy was given, 40 mg. daily at first, and in reducing doses until the ESR had fallen to 22 mm. 10 days later. The left optic disc became atrophic and the vessels remained attenuated. The visual acuity remained at perception of hand movements.

Follow-up.—The right eye has remained normal for 18 months; the patient is symptomless and oral prednisolone is still being taken (7.5 mg. daily). An attempt to stop the treatment has resulted in the ESR rising to 25 mm. and the patient has felt unwell. On the above dosage the ESR remains below 10 mm.

UNILATERAL ISCHAEMIC OPTIC NEURITIS

Case 10, a 75-year-old housewife, was seen on September 18, 1964, because of loss of vision in the right eye 3 days previously. The eye was blind with no perception of light, and the disc was described as "pale and fuzzy" with some haemorrhages at its margin (Fig. 1). The other eye was normal. She claimed to be in good health but, on questioning, admitted to some recent weight loss and vague headaches for the previous month. Her temporal arteries were thought to be normal on palpation. Occult temporal arteritis was suspected. The ESR was 92 mm. in the first hour and the blood pressure 170/70. A biopsy was positive (Fig. 2). In the meantime, treatment with systemic prednisolone was started, the ESR quickly fell to 16 mm. after 11 days, and the patient felt better. She has since been maintained on oral prednisolone (7.5 mg. daily) and after one year the other eye remains unaffected.

Discussion

Incidence

During the 2½ years January, 1963 to July, 1965, seventeen cases of occult temporal arteritis were seen in our Eye Department. During this same period two cases of classical temporal arteritis without ocular involvement were also seen. In the general pathological departments of the hospitals serving the same areas from which our patients are drawn only three positive temporal artery biopsies were examined over the same period. One is thus led to suspect that ocular involvement may be the commonest mode of presentation of this disease in present-day practice.

OCCULT TEMPORAL ARTERITIS

1
(Case 10)

2
(Case 10)

3
(Case 14)

4
(Case 2)

5
(Case 9)

6
(Case 15)

FIG. 1.—Case 10, right eye, showing ischaemic optic neuritis in acute phase. Visual acuity = No perception of light.

FIG. 2.—Case 10, superficial temporal artery biopsy, showing typical temporal arteritis. $\times 40$.

FIG. 3.—Case 14, right eye, 2 weeks after acute ischaemic optic neuritis. Visual acuity = Perception of light.

FIG. 4.—Case 2, section of superficial temporal artery, showing healing arteritis. $\times 45$.

FIG. 5.—Case 9, left eye, showing central retinal artery occlusion. Visual acuity = No perception of light.

FIG. 6.—Case 15, section of superficial temporal artery, showing giant cell systems. $\times 130$.

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Age and Sex

The preponderance of females (13 out of 17) is striking. The age range, 69–81, is that expected in this disease.

Clinical Symptoms and Signs

The relative absence of clinical symptoms, apart from the sudden loss of vision, was characteristic in these seventeen cases. No patient specifically complained of symptoms in the temples or head but, when directly questioned, six admitted to a degree of malaise, anorexia, and/or weight loss over a period of 1 or 2 months. Headaches or vague head pains were admitted by four patients and three mentioned pains in the temples and behind the eyes. In the majority, however, no subjective complaints were made, yet nearly all patients admitted to feeling much better after a period of treatment when the erythrocyte sedimentation rate had fallen to normal. Two patients admitted to some tenderness over the superficial temporal arteries and pulsation was noted to be diminished or absent in a few cases. Euphoria, as mentioned by Meadows (1954), was a prominent feature in most of our cases and, even after the occurrence of blindness in the second eye, they still seemed quite unconcerned and cheerful.

Other General Disease

Another feature of these patients was the relative absence of other general disease. Two patients were known diabetics, but insulin-independent. One had had a myocardial infarct in the previous year, and one had had blackouts during the previous year. One had had an operation for gastric neoplasm. Some degree of general arteriosclerosis was present in all patients, which was not unexpected in this age group, yet only one showed any significant degree of arteriosclerotic retinopathy. Table V shows that several were mildly hypertensive, but in no case was the diastolic pressure above 100, and none was receiving any formal hypotensive therapy.

Erythrocyte Sedimentation Rate

A raised ESR is a characteristic feature of the disease, despite the absence of clinical symptoms in most cases. In the present series the ESR range in the central retinal arterial occlusion group was 62 to 135 mm. (mean 98), and in the ischaemic optic neuritis group 40–131 mm. (mean 85). We were somewhat surprised to find such a clearcut demarcation between the ESR levels in the positive and negative cases, because positive cases have been reported in the past where the ESR was within normal limits even at the time of onset of visual loss. One of the cases previously reported by the present author (Cullen, 1963) had an ESR of 12 mm. in the first hour, yet he showed generalized arteritic involvement at *post mortem* and typical lesions of giant cell arteritis were found in the blood vessels supplying the eye and optic nerve. We feel that a raised ESR is a great help in establishing the diagnosis but, in the presence of the typical ocular picture, especially that of an ischaemic optic neuritis, occult temporal arteritis should be suspected and a biopsy carried out despite a normal ESR reading. This is particularly so if both eyes have been involved and, in these circumstances, treatment with corticosteroids should be commenced pending pathological confirmation. Only one case of our series with an ESR above 40 mm. had a negative biopsy. This patient had a pale swollen disc following acute complete

visual failure and occult temporal arteritis was provisionally diagnosed. This type of patient presents a real problem in diagnosis because it is difficult to see how arteriosclerosis, thrombosis, or embolism could produce this clinical picture which, in the proven cases, as will be seen later, is associated with involvement both of the ophthalmic and posterior ciliary vessels in a generalized arteritis. Palm (1958) assumed that these were also examples of temporal arteritis and included six such patients in his series of 31 cases. It is impossible to say that they have not got an arteritis in other branches of the carotid system, and the temporal arteries have escaped. Such patients with negative biopsies and ischaemic optic neuritis, especially if the ESR is elevated, should be carefully observed because, if they are examples of an occult temporal arteritis, the second eye is undoubtedly in danger, but in the nine such patients with negative biopsies in our series involvement of the second eye has not occurred in the follow-up period.

Biopsy

Superficial temporal artery biopsy should be performed in all suspected cases and is the only way of proving the diagnosis. The biopsy should be taken from the same side as the involved eye in unilateral cases. We have had experience in one case in which the biopsy was taken initially from the side opposite to that of the blind eye and the pathological picture was equivocal. When repeated on the other side typical lesions of temporal arteritis were reported. The biopsy has been done in all cases by the author or by one of the junior staff of the Eye Department. It is a simple procedure which can be performed in the out-patient department and under local anaesthesia.

Method of Biopsy.—The superficial temporal artery is first palpated within the hair line in front of and above the ear. The artery is marked with dye and the local anaesthetic injected. A transverse incision is made boldly through the scalp across the line of the vessel and, when the scalp is fully divided, the fascia overlying the temporalis muscle is seen. The vessel lies on the fascia surrounded by fine fibrofatty tissue and is easily dissected out. The artery is clipped and divided and a piece about one inch long is removed. The ends are tied off with catgut, the scalp is sutured with silk and a dry dressing applied. The sutures are removed in 5 or 6 days. No complications have occurred following this procedure and bilateral biopsies have been performed in some cases.

Ocular Picture

In previously reported series of eye involvement in temporal arteritis retinal vascular occlusions have not been common. Meadows (1954) saw one out of twelve cases; Wagener and Hollenhorst (1958) three in one hundred eyes of 58 patients; Palm (1958) four in 31 cases, and Greaves (1961) found central retinal arterial occlusion in four out of thirty cases. On the other hand, Ellis, Hamer, Hunt, Lever, Lever, Peart, and Walker (1964), in a medical investigation of retinal vascular occlusions, found four instances of temporal arteritis in forty cases of central retinal arterial occlusions, both complete and segmental (*i.e.* a 10 per cent. incidence). In our series, the incidence of vascular occlusion has been high, six out of seventeen cases (35 per cent.). This is probably because we have suspected the condition and

looked for it. Of all our cases of central retinal arterial occlusion, the incidence of temporal arteritis is similar to that of Ellis and others (1964), *i.e.* 11 per cent., and where the involvement is bilateral temporal arteritis is much more likely to be the cause.

The arterial occlusion is usually complete, but branch involvement may occur as in Case 11, and we have observed one case with occlusion of the central retinal artery and vein (Case 9).

Bilateral Involvement.—In the ischaemic optic neuritis group the incidence was four out of eleven and no bilateral case was found in those with negative biopsies. Ischaemic optic neuritis is the typical lesion to be expected and, if bilateral, the diagnosis is even more certain. Oedema of the disc was present in all but two proven cases. In these there had been intervals of 5 days and 2 weeks before they were seen. It is probable that oedema had been present but had subsided by the time they were examined. The involvement is usually severe, and complete or substantial visual loss is the rule, although less severe involvement has been seen in two patients. In both groups the interval between the involvement of the first and second eye varied from simultaneous onset in one case to 3 weeks (Table VI). All the bilateral cases presented after involvement of the second eye and no unilateral case so far treated had incurred a similar involvement of the fellow eye. No patient had different lesions in the two eyes. If a central retinal arterial occlusion occurred in one, this was repeated in the other, and likewise with ischaemic optic neuritis.

TABLE VI
INTERVAL BETWEEN INVOLVEMENT OF FIRST AND SECOND EYES
IN SEVEN CASES OF OCCULT TEMPORAL ARTERITIS

C.R.A. Occlusion Group (3)	I.O.N. Group (4)
Simultaneous	2 wks
1 wk	2 wks
3 wks	1 wk
	3 days

Premonitory Symptoms.—These have been reported by other authors and, in a number of patients of this series, the acute visual failure was preceded by transient visual obscurations, usually within 24 hours of the final visual failure.

Pathology and Aetiology

There is a cellular infiltration of the vessel wall, usually most marked in the media and internal lamina elastica. Lymphocytes and mononuclear cells predominate, especially in the early stage of the disease, and giant cells are found later but are by no means a constant feature. There is, in addition, a patchy necrosis of the media and the internal elastic lamina is broken up or destroyed. Fibroblastic proliferation of the intima occurs, reducing the lumen or even occluding the vessel, but thrombosis is not an essential feature. Secondary fibrosis occurs in the adventitia and may involve the nerves, thus accounting, some say, for the pain in the temples and for the relief which sometimes occurs after biopsy. The giant cells are often found around the broken-up internal lamina elastica and it is held by some workers that the disease is, in fact, an allergic or auto-immune reaction to elastic tissue.

The aetiology of temporal arteritis is obscure and, in the patients comprising this series, no further clue in this regard is forthcoming. Some allergic or auto-immune process is suspected, probably for want of a better explanation, but none of the seventeen patients here studied has shown any other evidence or history of allergic conditions, none was taking or had taken in the past any drugs which might have set off this peculiar response, and none was suffering from any other obvious predisposing condition.

The ocular involvement in temporal arteritis is similar as regards its pathology to that mentioned above, the vessels involved being the branches of the ophthalmic artery which supply the optic nerve and eyeball.

Histopathological studies on the eyes in this condition have been reported in only ten cases in nine papers (Cooke and others, 1946; Cardell and Hanley, 1951; Kreibitz, 1953; Heptinstall and others, 1954; Crompton, 1959; Spencer and Hoyt, 1960; Cullen, 1963; Manschot, 1965; Wolter and Phillips, 1965). Material is scarce because patients may live for many years after the ocular involvement and also because enucleation is rarely indicated. Wolter and Phillips (1965) obtained their specimen after the occurrence of haemorrhagic secondary glaucoma which followed occlusion of the central retinal artery, and this case exhibited total necrosis of the whole retina, optic nerve head, and distal optic nerve caused by severe combined involvement of the ophthalmic and posterior ciliary arteries. In this specimen the central retinal artery was found patent on the disc and inside the optic nerve. The specimens mentioned by the present author (Cullen, 1963) showed typical lesions of giant cell arteritis in the ophthalmic arteries, in the central retinal arteries at their origins, and in the ciliary vessels. Again, the retinal vessels within the globes were not involved, nor were the choroidal vessels, and the central retinal arteries within the optic nerves were patent. The other reported cases all showed a similar picture of involvement of the posterior ciliary arteries and of the central retinal artery outside the globe and outside the nerve, and all authors agree that combined involvement of these vessels must occur before true infarction can take place in the optic nerve head.

Cases of Central Retinal Arterial Occlusion not Biopsied

It is noted that thirty patients in the central retinal arterial occlusion group were not biopsied, as their ESRs ranged from 1 to 10 mm. These were all unilateral and have remained so over the period of observation. It might be argued that they could also have occult temporal arteritis but this has not been proven. It would appear to be unlikely, however, in view of the observations made in the positive cases, and it was felt to be unjustified to subject all patients to biopsy, even though this is a simple and harmless procedure. It was also hoped that the ESR level could be established as a guide as to whether biopsy should be done or not, and we feel that this study supports our contention that, if the ESR is normal, temporal arteritis is unlikely in patients with central retinal arterial occlusion, especially if the lesion is unilateral. We feel, however, that all bilateral cases and all patients with ischaemic optic neuritis should have a biopsy done, even if the ESR is below 10 mm. Furthermore, careful questioning may reveal other suspicious evidence of arteritis which will also lead one to a decision as to whether this type of investigation should be pursued.

Treatment

This disease can be controlled by adequate corticosteroid therapy. Thus, early diagnosis is of prime importance so that treatment can at least be given in time to prevent involvement of the second eye. Our system of treatment is as follows:

In emergency cases, where the patient presents after involvement of the second eye and provided no longer than 24 to 48 hours has elapsed since this occurrence, we give ACTH by intramuscular injection (40 units twice a day) pending confirmation of the diagnosis by biopsy. If this is positive, the ACTH is replaced by prednisolone by mouth 40 mg. daily to start and in reducing doses as the ESR begins to fall. This regime obviates the danger of giving large and unnecessary doses of corticosteroids to elderly patients, especially when one has not made a definite diagnosis. The only patient who recovered vision in our bilateral series was treated in this fashion. If both eyes are blind and if more than 48 hours have elapsed, we give oral prednisolone until the ESR returns to normal. In all our seventeen cases the ESR has fallen to normal levels within 7 to 10 days and has remained down in all cases, but the majority have had to continue with maintenance treatment up to the present time. This is in contrast to the response in classical temporal arteritis as reported by Whitfield and others (1964), where the ESR may remain raised despite adequate corticosteroid therapy.

Those patients who present with involvement of one eye only are treated with oral prednisolone as above until the ESR returns to normal and remains down after withdrawal of the drug. Treatment is commenced at once without waiting for the biopsy report because of the potential danger to the second eye. To date no patient so treated has developed trouble in the second eye, but the corticosteroids may have to be continued for years or even indefinitely. A case mentioned by Miller (1964) developed ocular involvement 3 years after the diagnosis of temporal arteritis was made, and a case reported by the present author (Cullen, 1963) developed ocular involvement 6 weeks after cessation of treatment.

The efficacy of corticosteroid treatment in preventing involvement of the second eye is emphasized by the high incidence of bilateral visual loss in untreated cases. In these, if the diagnosis had been made after the occurrence in the first eye (and in all but one case in this series there was plenty of time for this to have been done), there is little doubt that bilateral blindness would have been prevented.

Prognosis (Table VII)

Of the seventeen patients reported, seven incurred involvement of both eyes and of these five became completely blind, one retained counting fingers in one eye with a good field, and one (Case 4) made a good recovery. This last was a case of bilateral

TABLE VII
VISUAL RESULT IN SEVENTEEN CASES OF OCCULT TEMPORAL ARTERITIS

Visual Loss	Bilateral	Unilateral
Total	5	5
Substantial (C.F.-H.M.)	1 (complete loss other eye)	3
Partial (Better than C.F.)	1	2

ischaemic optic neuritis, and the initial visual loss was only partial—6/18, 6/18—and she was not seen for one week after the onset of symptoms. None of the five who became blind in both eyes presented within 24 hours of the onset of blindness in the second eye, and in all of them the vision was reduced to no perception of light or counting fingers at best by this time. The only patient (Case 2) who recovered sight in the bilateral group presented within 24 hours and received immediate treatment; she had bilateral central retinal arterial occlusions.

In the unilateral group, if one excludes the partial central retinal arterial occlusion case, the prognosis seems better with ischaemic optic neuritis but again, in this group, those who retained some vision were those who had not incurred complete loss at the outset and who were treated within 24 hours. The final outcome, therefore, seems to depend on the type of involvement, its severity, and the time at which treatment is instituted.

The prognosis for life is good. Only two patients have died within the period of observation which, for the whole group, is 1 year and 2 months (range: $2\frac{1}{2}$ years to 6 months); 4 months after the visual involvement Case 9 sustained a cerebral vascular accident which, at *post mortem*, was found to be associated with carotid occlusion in the neck, but there was no evidence of arteritis either in the carotid at the site of the thrombosis or in any of the intracranial arteries. Case 12 sustained an acute fatal myocardial infarction 8 months after he became blind. The remaining fifteen patients are all well and treatment with corticosteroids is being continued in twelve cases.

Conclusions

- (1) Temporal arteritis in its occult form is a common cause of blindness in elderly people.
- (2) The occult form of the disease appears to be more common than the classical variety.
- (3) Patients with acute visual failure of presumed vascular origin should be investigated with this condition in mind.
- (4) A raised erythrocyte sedimentation rate is a good guide to the diagnosis, although not absolute.
- (5) All patients with ischaemic optic neuritis should be presumed to be suffering from occult temporal arteritis until proved otherwise.
- (6) In 10 per cent. of patients with central retinal arterial occlusion the cause is occult temporal arteritis.
- (7) Biopsy of the superficial temporal artery is the only way to prove or exclude the condition. This should be done by the ophthalmologist as soon as possible in all suspected cases.
- (8) Bilateral involvement resulting in total blindness may occur if adequate treatment is not given.
- (9) Involvement of the fellow eye can be prevented if treatment is instituted in time.
- (10) Treatment should be continued until all symptoms subside and the erythrocyte sedimentation rate remains down after withdrawal of therapy.

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ADDENDUM

Since this paper was submitted for publication we have encountered a further three cases of central retinal artery occlusion (all unilateral) and fourteen cases of ischaemic optic neuritis (five bilateral), associated with occult temporal arteritis. The erythrocyte sedimentation rate range in these additional seventeen patients was 46–122 mm./hr (mean 82). Three further patients with classical temporal arteritis and no ocular involvement have also been seen.

CORRIGENDUM

The following list of references was omitted from the paper by J. NOLAN and J. F. CULLEN which appeared in the June issue (*Brit. J. Ophthal.*, 1967, **51**, 361):

- BOYD, W. (1961). "Textbook of Pathology", 7th ed., p. 37. Lea and Febiger, Philadelphia.
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