Infarction in the optic nerve

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Infarction in the optic nerve is a well-known complication in cases of giant-cell arteritis, but may not be diagnosed when it is related to atherosclerosis. The three patients to be reported were referred during the same year with papilloedema and a diagnosis of suspected brain tumour. Two underwent extensive radiological procedures to exclude an intracranial tumour. The three patients illustrate the syndrome of infarction in the optic nerve and provide a basis for the suggestion that, when this diagnosis is suspected, a short period of observation appropriately may replace some of these procedures. In addition, studies were made of the vasculature of selected optic nerves at necropsy.

CASE 1

A 52-year-old man noted sudden, painless onset of right inferior altitudinal hemianopsia. The defect remained unchanged. A year later, he consulted an ophthalmologist immediately after an identical defect occurred on the left. On admission to the hospital, vision was 20/30 on the right, 20/20 on the left. Intraocular tension was normal. The right optic disc was pale and the arterioles narrowed. The left disc was slightly elevated, had indistinct margins, but was not hyperaemic. Retinal arterioles on the left were narrowed in segments, and a few small haemorrhages radiated from the nerve head. Evidence of retinal vascular occlusion was not seen. Inferior altitudinal fibre bundle defects were present bilaterally (Fig. 1). Blood pressure 160/100, but the remainder of the physical examination was normal. Fasting blood sugar, VDRL, erythrocyte sedimentation rate (ESR), cerebrospinal fluid (CSF), radiographs of the skull with views of the optic foramen, electroencephalogram (EEG), bilateral carotid arteriograms, and pneumoencephalogram were normal. Biopsies of temporal artery, skin and muscle, and examination of bone marrow were also normal. One month, and again two years later, bilateral pallor of the discs was noted, but visual acuity and fields were unchanged.

COMMENT A hypertensive, middle-aged man presented with a Foster Kennedy syndrome. Extensive studies did not give evidence for an intracranial lesion. Sudden onset of symptoms, permanent bilateral altitudinal field loss, and prompt disappearance of disc swelling are important features suggesting infarction in the optic nerve.

CASE 2

A 29-year-old woman suffered sudden decrease of vision in the left eye and was seen by an ophthalmologist who diagnosed 'papillitis'. Acuity improved over a few months. At the age of 41, she noted episodic blurring of vision for three days, and then sudden painless onset of a superior altitudinal field defect on the right. Another ophthalmologist diagnosed papilloedema and recommended neurological evaluation.

On admission, no history of other previous neurological deficit was obtained. Blood pressure and general examination were normal. Vision with glasses was 20/40 in both eyes. Intraocular tension was normal. The right optic disc was elevated two dioptres, had indistinct margins, but was not hyperaemic. A small linear haemorrhage radiated from the disc. Retinal veins were dilated, tortuous, and segmented, and arterioles extremely narrowed. The left disc was pale, had sharp margins, and the left retinal arterioles were small in caliber. Superior altitudinal fibre bundle defects were noted in both fields (Fig. 2).

Moderate iron deficiency anaemia and an abnormal glucose tolerance curve were found. Fluorescent treponeme antibody (FTA-ABS) test was negative. ESR was normal. Electrocardiogram demonstrated changes of ischaemia, and biopsy of the temporal artery revealed mild atherosclerosis. Radiographs of the skull with views of the optic foramen, EEG, CSF, and brain scan were normal. Papilloedema disappeared within a


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ESR, FTA-ABS test, radiographs of the skull with views of the optic foramen, and tomograms of the sella turcica, EEG, brain scan, and CSF were normal. No abnormalities were seen in the left carotid arteriogram. This procedure was followed by dysphasia and dyslexia, improving slowly over two months. The patient refused pneumoencephalography. Two weeks and again two months after discharge from the hospital, the left disc was flat, had distinct margins, and was slightly pale. Acuity and fields remained unchanged.

COMMENT A 37-year-old diabetic woman presented with unilateral papilloedema. The sudden onset, sectoral visual field deficit, and prompt disappearance of disc swelling are consistent with infarction in the optic nerve. Evidence for an intracranial mass was not found. Preservation of visual acuity and permanent field defect make demyelinative disease unlikely.

DISCUSSION

Infarction in the optic nerve, also called ischaemic optic neuritis (Hollenhorst, Brown, Wagener, and Shick, 1960), ischaemic optic neuropathy (Miller and Smith, 1966) or vascular pseudopapillitis (François, Verriest, Neetens, De Rouck, and Hanssens, 1962) has been reported frequently in the French and German medical literature for more than a decade, but has only recently been considered as a nosological entity in the English literature (Miller and Smith, 1966). Involvement of the eye in many systemic vascular diseases is generally recognized, but infarction in the optic nerve on the basis of arteriolo- or atherosclerosis is seldom diagnosed. The resulting choked disc may be mistaken as evidence of increased intracranial pressure, and, when the process later occurs on the other side, the finding of a Foster Kennedy syndrome may suggest a mass in the anterior fossa.

Infarction in the optic nerve probably occurs more frequently than generally realized, and may account for some cases of 'low tension glaucoma', optic neuritis of unknown origin and 'chiasmatic arachnoiditis'. It is a frequent result of giant-cell arteritis, whether typical (Hollenhorst et al., 1960) or 'occult' (Cullen, 1967), and has occurred in association with polyarteritis nodosa (Kimbrall and Wheliss, 1967), lupus erythematosus (Lasco, 1961), syphilitic arteritis (Smith, Israel, and Harner, 1967), acute blood loss (Piper and Unger, 1957), and polymyalgia rheumatica (Fessell and Pearson, 1967). We have observed one case after subarachnoid haemorrhage (University of Virginia Hospital, Case No. 54-05-45). Nevertheless, infarction in the optic nerve is probably most common with atherosclerosis (François et al., 1962).

The syndrome, as recently described also by Miller and Smith (1966), begins with sudden onset...
of a visual field defect which may include central vision. Pain is usually not present when athero-
sclerosis is the cause. Permanent visual deficit may 
occur after several attacks of transient ischaemia. 
Initially, the process is often unilateral, but it may 
eventually occur in the other eye within days to 
years. Twenty-four to 36 hours after the onset, the 
appearance of the optic disc may be indistinguishable 
from papilloedema resulting from raised intracranial 
pressure. More commonly, however, the disc, 
although oedematous, is not hyperaemic, and may 
even be pale. Frequently, only a segment of the 
disc is elevated, and occasionally it is normal. Small 
linear haemorrhages may radiate outward from the 
disc, and the pattern of nerve fibre bundles be 
accentuated by oedema. Elevation gradually disap-
ppears in two or three weeks, leaving a flat, pale disc.

Evidence for retinal arteriosclerosis is usually 
present. In addition, in the acute stage, retinal 
veins may be dilated, tortuous and segmented, and 
arterioles narrowed, sometimes to thread-like 
calibre.

An important distinguishing sign of infarction in 
the optic nerve is the abnormality of the visual field. 
Sudden onset of a large fibre bundle defect, particu-
larly an altitudinal hemianopsia, should suggest a 
vascular mechanism for the elevated disc. Fibre 
bundle defects are not characteristic of this entity 
alone; they are common in glaucoma, for example. 
Total loss of vision, sectoral defects, and scotomas 
also occur frequently. Visual acuity may not be 
diminished.

An expanding intracranial or intraorbital lesion 
compressing one optic nerve is most important in 
the differential diagnosis of infarction in the optic 
nerve. Some expanding lesions, particularly 
aeurysms, may rarely cause sudden loss of vision, 
and even altitudinal hemianopsia (Mitts and 
McQueen, 1965). Thrombosis of a retinal branch of 
the central retinal artery may cause similar abnor-
malities of the visual field, particularly 
sectoral or fibre bundle defects. These defects, 
however, are more likely to radiate from the blind 
spot rather than from the centre of fixation (as in 
Case 3), and also are usually more sharply 
demarcated at the horizontal line because the 
superior and inferior halves of the retina are supplied 
by separate branches of the central artery. Similar 
field defects may result from trauma to the optic 
nerve (Turner, 1943), probably because the same 
vessels, the pial arterioles of their branches, are 
involved. These arterioles, being on the outer 
surface, may be more vulnerable than the central 
vessels.

Preservation of visual acuity frequently aids in 
distinguishing infarction in the optic nerve from 
demyelinating diseases in which severe loss of vision 
is common.

A helpful diagnostic point illustrated by the 
present cases is the resolution of disc elevation 
within two or three weeks. After recognition of a 
static visual deficit, adequate reason exists to 
postpone arteriography and pneumoencephalo-
graphy for the short time necessary to observe the 
disc.

Occlusion of a 'central artery of the optic nerve' 
has been a frequent explanation of this syndrome 
(Piper and Unger, 1957; François et al., 1962; 
François and Neetens, 1965). François and Neetens 
(1965) have cited the occurrence of lower altitudinal 
hemianopsia as evidence for existence of this artery 
thought to supply the upper half of the optic nerve 
by these authors (François et al., 1962), macular 
fibres by others (Wolff, 1954).

The pattern of arterial supply of the optic nerve 
is a controversial subject. The concept of a central 
artery of the optic nerve has been strongly supported 
by François and Neetens (1954, 1963, 1965), on 
the basis of latex injection and microradiographic 
study, and has become widely accepted. These 
authors stated that nutritive branches of the central 
retinal artery supplying the optic nerve do not exist, 
and maintained that the nerve is supplied by 
perforating arterioles from the pia, and a separate 
axial system that enters the nerve to supply the upper 
half (Fig. 4a). In 10 of 31 specimens, François et al. 
found the axial system to be a discrete central artery 
of the optic nerve; in the remainder, more than one 
axial artery was present.

Hayreh (1963) stressed the variability of the blood 
supply to the optic nerve. Using latex injection, he 
demonstrated nutritive branches from the intraneural 
part of the central retinal artery in 75% of 64 
specimens, but noted absence of these branches 
from the distal third of the artery (Fig. 4b). The 
section of optic nerve just behind the lamina 
cribrosa was supplied only by branches from the 
circle of Zinn. Although Hayreh found abundant 
anastomoses between the central retinal artery and 
pial plexus, he found none between this artery and 
branches of the circle of Zinn immediately behind 
the lamina cribrosa. This may be a relatively 
vulnerable location for infarction. Several other 
authors have found intraneural branches of the 
central retinal artery and have denied the existence 
of a central optic nerve artery (Wybar, 1956; Steele 
and Blunt, 1956; Blunt, 1963). Present clinical and 
pathological evidence does not support the concept 
of occlusion of a central optic nerve artery as being 
the cause of infarction in the optic nerve in all cases.
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The variety of visual field defects resulting from infarction in the optic nerve suggest that more than one small blood vessel may be involved.

Several pathological studies of ocular lesions in giant-cell arteritis are available (Cardell and Hanley, 1951; Kreibig, 1953; Crompton, 1959; Spencer and Hoyt, 1960; Rodenhäuser, 1964; Wolter and Phillips, 1965) but optic nerves have not been studied in cases of infarction resulting from arteriolo- and atherosclerosis. The location of the lesions in both instances may be similar, however. Narrowing of arterial lumens by cellular infiltration and intimal proliferation is frequently seen in the ophthalmic and central retinal arteries, mainly in their most proximal parts, and, almost invariably, in the long and short posterior ciliary arteries, and their pial branches. The central retinal artery is usually patent in these cases. A localized region of opticomulacalacia immediately posterior to the lamina cribrosa has been noted in a few cases of temporal arteritis (Kreibig, 1953; Spencer and Hoyt, 1960) but other reports describe total destruction of nerve fibres just behind the disc (Rodenhäuser, 1964; Wolter and Phillips, 1965), ‘gross degeneration’ (Cardell and Hanley, 1951), or a normal nerve.

Involvement of vessels supplying the optic nerve by atherosclerosis is a reasonable possibility, because this disease is so generalized. However, the nature and extent of vascular disease in the optic nerve has received little attention, this nerve being relatively inaccessible in the routine necropsy.

Battastini and Caffi (1959) examined optic nerves from 14 elderly patients and found thickening of arterial walls due to invasion of media and adventitia by connective tissue and ‘sclerosis’. The intima was also thickened, causing narrow lumens. Atheromata and other degenerative changes were not found. Brooser, Börzövi, and Åhi (1967) noted irregular regions of fibre destruction in optic nerves of diabetic patients, and PAS-positive material in the walls of vessels in and around the nerve.

To extend our own observations, 40 optic nerves were removed from 20 cadavers after injection of embalming fluid into the cranial arteries. All patients were older than 45 years. The entire length of the nerves was obtained by removing the roofs of the orbits and optic canals with a chisel. Nerves were fixed in 10% formalin, embedded in paraffin, and sectioned in either sagittal or transverse planes. Sections were stained with haematoxylin and eosin, phosphotungstic acid, and by Weigert’s method for elastic tissue.

Three changes were present in various degrees of severity in almost all optic nerves examined. Intimal thickening, usually diffuse and minimal, was present in many of the posterior ciliary arteries, but was less marked in the central retinal artery. A more frequent finding was the thickening of the walls of arterioles lying in the pia (Fig. 5) and within the substance of the nerves. In addition, increase in the amount of fibrous connective tissue was observed in the septae of many optic nerves, structures that consist only of capillary walls in the newborn. In some cases, this increase of fibrous tissue was excessive, even though present in patients without visual complaints (Fig. 6). Focal opticomalacia was not found in our specimens.

The results of the present pathological study

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FIG. 4. (a) Scheme of the arterial supply of the optic nerve, according to François and Neetens (1954). (b) The scheme of Hayreh (1963). Note central artery of the optic nerve (CAO) in (a) only, intraneural branches of central retinal artery (CAR) in (b) only.

suggest that the arterial supply of the optic nerves is subject to athero- and arteriosclerosis no less than other vessels of similar size. These changes provide a limited pathological basis for the syndrome of infarction in the optic nerve. Although not found by us, emboli lodged in small arterioles may have an effect similar to thrombotic occlusion. Routine examination of optic nerves at necropsy would yield more definite data.

The name ‘infarction of the optic nerve’ is more specific and descriptive than ‘ischaemic optic neuropathy’ or ‘vascular pseudopapillitis’, and is in accord with current nomenclature. ‘Neuropathy’ designates primary disease of neural tissues rather than of its blood supply. Only infarction, not ischaemia, can account for permanent visual loss. ‘Pseudopapillitis’ is one more addition to a long list of confusing terms describing the appearance of the optic disc in various conditions, and has been used, as has ‘pseudopapilloedema’, to designate congenital anomalies of the disc. Infarction may occur in the optic nerve without causing abnormalities of the disc.

SUMMARY

Three cases are presented to illustrate a syndrome,
Infarction in the optic nerve, which most probably results from arteriolo- and atherosclerosis of vessels supplying the nerve. Sudden onset of a static visual field defect, particularly of the altitudinal fibre bundle type, and transient, pale swelling of the optic disc are salient features of this syndrome. When infarction in the optic nerve is suspected, the patient should be observed for a short time with particular reference to optic disc and visual fields. Such innocuous studies may replace more hazardous diagnostic procedures for exclusion of an intracranial mass. Atherosclerosis and arteriosclerosis of the arterial supply and thickening of connective tissue septae of the optic nerve were demonstrated in a small series of necropsy specimens.

REFERENCES