The term dolichoectasia derives from “dolicho” meaning elongation and “ectasia” meaning distension.

Dolichoectasia of the intracerebral vessels is a rare disorder that affects the large arteries at the base of the brain. The vertebro-basilar system is more often affected than the internal carotid arteries. Neurological deficits may occur secondary to local embolisation, thrombotic occlusion, compression, or rarely rupture. Neuro-ophthalmic manifestations are most often related to compression of neighbouring structures and include cranial nerve palsies, optic neuropathy, chiasmal syndromes, nystagmus, hemifacial spasm, and ocular tilt.

Visual loss from compression of the anterior visual pathways by dolichoectatic vessels is well recognised. The natural history of this rare condition, however, is not well described. We sought a better definition of the clinical course to assist the clinician in managing these patients.

METHODS

A retrospective review of the clinical records was undertaken for all patients with dolichoectatic compression of the anterior visual pathways evaluated in an outpatient neuro-ophthalmology clinic from 1984 to 1999. All patients were examined by one of us (VP or AK). Ten such patients were identified. All patients underwent a complete neuro-ophthalmic examination including visual field testing by Goldmann perimetry. For each patient the age, sex, presenting symptoms, time course of visual loss, past medical history, visual acuity, visual fields, radiographic studies, and clinical course were reviewed. In each case the clinical findings and radiographic investigations were carefully studied and correlated, both to exclude other potential causes of visual loss and to ensure that the visual deficits were explainable on the basis of vascular compression. In each case there was clearly demonstrable distortion of the optic nerve, chiasm, or tract by the involved artery. Patients were excluded if the optic disc(s) showed excavation rather than pallor, or if intraocular pressures were raised (>21 mm Hg). Intervention (craniotomy) was undertaken in one patient (case 6). In the remainder, the clinical course represents the natural history of this disorder.

RESULTS

Ten patients were studied (table 1). The average age at presentation was 70.6 years (range 48 to 88). Women were more commonly affected than men (seven women, three men). Vascular risk factors included hypertension in eight and diabetes in three. Five patients had a history of coronary artery disease. One had a history of previous transient ischaemic attacks. One (case 8) had suffered a lacunar stroke two years before neuro-ophthalmological examination. Four patients presented with a unilateral optic neuropathy, one with bilateral optic neuropathy, three had a chiasmal syndrome, and two an optic tract syndrome. One eye was affected in four patients, both eyes in six (total 16 eyes). In addition, vision was abnormal in three eyes because of unrelated ocular conditions (non-arteritic ischaemic optic neuropathy in one, age related macular degeneration in two). One patient experienced additional loss of vision in one eye because of central retinal artery occlusion five years after the initial presentation.

Visual acuities in the 16 involved eyes at presentation ranged from 20/20 to hand motion. In nine eyes acuity was better than 20/40, six ranged from 20/40 to 20/100, and in one vision was worse than 20/100 (hand motion). Three patients had a unilateral nasal visual field defect, one had a unilateral temporal defect, and one had severe diffuse loss of field. Three patients had variants of a chiasmal syndrome: one had a central scotoma plus a small superior temporal defect in the fellow eye (fig 1A), one had a bitemporal defect plus a nasal defect in one eye (fig 2), and one had a bitemporal hemianopia (fig 3A). Two patients had a homonymous hemianopia (fig 4A).

All patients underwent magnetic resonance imaging (MRI); seven also had magnetic resonance angiography (MRA) and two had conventional cerebral arteriography. Seven patients had a dolichoectatic internal carotid artery,
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<tr>
<td><strong>Clinical course</strong></td>
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<td>Improved to 20/20 OD following cataract surgery 3 years later</td>
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<td>Some progression of right optic neuropathy (20/200 OD) stable OS</td>
<td>Progressive ataxia</td>
<td>Stable at 7 month follow-up died of stroke 2 years later</td>
<td>Stable</td>
</tr>
<tr>
<td><strong>Follow up interval (last examination)</strong></td>
<td>5 Years</td>
<td>6 Years</td>
<td>2 Years</td>
<td>1 Year</td>
<td>3 Years</td>
<td>2 Years</td>
<td>NA</td>
<td>7 Months</td>
<td>2 Years</td>
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</table>

AF, atrial fibrillation; ARM, age related macular degeneration; CAD, coronary artery disease; COPD, chronic obstructive pulmonary disease; CRAO, central retinal artery occlusion; HTN, hypertension; MRI, magnetic resonance imaging; NA, not available; NAION, non-arteritic ischaemic optic neuropathy; OD, right eye; OS, left eye; OU, both eyes; TIA, transient ischaemic attack; VA, visual acuity.
causing compression of one optic nerve in four, of both optic nerves in one, and of the chiasm in two. Three patients had a dolichoectatic basilar artery, compressing the optic chiasm in one (fig 3B and 3C) and the optic tract in two (fig 4B). The pattern of visual loss correlated well with the radiographic findings in all patients.

In all 10 patients initial management was observational. One patient (case 6) experienced progressive visual loss that prompted surgical intervention. One patient was lost to follow up. In the remainder, follow up intervals ranged from 7 months to 10 years (mean 3 years, median 2.8 years). During the follow up period six patients remained stable and three showed some progression. Of the three with progression, the change was small in one (case 4, who had already experienced profound visual loss at the time of presentation) and mild to moderate in one (case 7). In the third patient (case 6), surgical intervention was undertaken with subsequent improvement of vision (see case report below).

CASE REPORTS
Patient No 6
This man was 59 years old when he presented with a two month history of progressive visual loss in the left eye. There was no associated head or eye pain and no other focal neurological deficits or systemic symptoms. His past medical history was positive for non-insulin-dependent diabetes and hypertension.

Initial examination revealed visual acuity of 20/20 in the right eye and 20/40 in the left eye. Goldmann perimetry in the right eye showed a mild temporal hemianopic defect affecting the superior field more than inferior; in the left eye there was a relative central scotoma and mild supertemporal defect (fig 1A). Pupils measured 5 mm OD (right eye), 3.5 mm OS in dim illumination, and 3 mm OD, 2.5 mm OS in light, with dilatation lag and a 2+ relative afferent defect OS (left eye). There was mild left upper lid ptosis with reverse lower lid ptosis. The right disc had a healthy appearance, the left was mildly pale.
MRI showed elevation of the left prechiasmal optic nerve and the left side of the chiasm by a markedly ectatic left internal carotid artery (fig 1B). A conventional arteriogram confirmed the fusiform nature of the arterial dilatation (fig 1C). The patient’s history of recent and progressive visual loss prompted an attempt to decompress the left optic nerve surgically through a left frontotemporal craniotomy. Exploration of the chiasmatic and carotid cisterns confirmed compression of the left optic nerve by the internal carotid artery. Areas of arteriosclerotic plaque were identified in the lateral and superior portions of the artery. Although the optic nerve was elevated by the artery, it did not appear pale or particularly flattened. There was a moderate amount of thickened arachnoid between the nerve and the artery. The optic canal was unroofed and the dura opened. The nerve was then dissected from the carotid and a fenestrated special angled small Sugita clip was placed around the artery at the point where it appeared to be compressing the nerve. The clip was then sutured to the dura laterally, decompressing the optic nerve.

Postoperatively the patient noted subjective visual improvement. Examination two months later revealed definite improvement of the visual field, although visual acuity and colour vision were unchanged. At re-examination four months later there was some additional improvement of the field, and visual acuity in the left eye had improved to 20/25. Subsequent yearly examinations over the next three years were stable. His left Horner’s syndrome has persisted.
Patient No 7
This 68 year old woman was found to have decreased vision on routine eye examination. In retrospect she had noticed gradually decreasing vision, more in the right eye than the left, for the preceding five years. Her past medical history was significant for diabetes, hypertension, and coronary artery disease. She was on chronic anticoagulant treatment for atrial fibrillation.

Initial examination showed visual acuities of 20/70 OD and 20/40 OS. There was a moderate right relative afferent pupillary defect. Goldmann perimetry revealed mild bitemporal loss plus generalised depression in the right eye. The right disc showed mild diffuse pallor.

MRI showed elevation of the right optic nerve and the right side of the chiasm by an ectatic right internal carotid artery (fig 2). No treatment was undertaken. At follow up three months later acuity in the right eye had decreased to 20/80. Other tests of optic nerve function, including Goldmann perimetry, were unchanged. Examination findings four months later were unchanged. Six months later visual acuity in the right eye decreased to 20/200, vision in the left eye was unchanged. Nine months later the examination remained stable.

DISCUSSION
Visual loss associated with ectatic intracranial arteries was described in 1932 by Caramazza.12 A review of this subject by Mitsi and McQueen in 1964 characterised the clinical findings in four cases plus 10 more culled from the literature.13 Patients ranged in age from 38 to 66 years and typically presented with progressive visual loss in one or both eyes. The most common pattern of visual field loss was a superior defect that was bilateral in three patients and unilateral in four. Other defects described were bitemporal loss (2), unilateral constriction (2), and bilateral constriction (1). In two patients the pattern of field loss was not reported.

In seven, the diagnosis was established by exploratory craniotomy. Unroofing of the optic canal with ligation of the internal carotid artery was undertaken in four patients. In one of the surgically treated patients vision improved postoperatively.

In the past, dolichoectatic compression was diagnosed by surgical exploration in most cases. Our ability to diagnose this disorder has been greatly aided by recent advances in radiographic techniques.14–16 Specifically, the combination of MRI and MRA has been particularly helpful in this regard. MRI allows simultaneous visualisation of both neural and vascular structures, with the potential for high spatial resolution. MRA affords imaging of intracranial vessels without the risk of an invasive procedure. This increased ability to diagnose this condition radiographically should be matched with a heightened awareness of the disorder by the clinician.

We describe 10 patients with compression of the anterior visual pathways by dolichoectatic intracranial arteries, all of whom were diagnosed by MRI. In only one case was neurosurgical intervention undertaken and thus our description largely represents the natural history of the disorder.

The majority of our patients were women, usually aged over 70 at time of presentation. Visual acuity was usually only mildly diminished, nine of 16 eyes having better than 20/40 acuity. Patterns of visual field loss depended on the site of compression. The most common pattern in patients with optic nerve compression was a nasal defect. The most commonly involved artery was the internal carotid, which usually caused unilateral optic neuropathy. In one patient (case 1), both carotid arteries were ectatic, causing bilateral optic neuropathy.

In most patients visual loss showed little change over time. In one patient (case 7), progression was documented but was not severe and the fellow eye remained stable. In another (case 4), further progression led to complete visual loss, but the level of vision at presentation was so diminished that intervention was not considered. In a third, progression was documented over several months, prompting neurosurgical intervention which reversed much of the visual loss (case 6).

Of interest, these latter two patients, whose clinical course differed from the majority by being more rapidly progressive, also differed demographically, being younger and male.

It should be noted that our patient group may have been selected for being at the more severe end of the spectrum of this disorder. We intentionally chose cases in which the MRI findings were sufficiently marked that there could be no doubt as to the compressive aetiology of the visual loss. This was done in an effort to exclude cases in which the causal relation between the radiographic findings and visual loss might be ambiguous. Controversy over this causal relation has arisen because varying degrees of contact between the carotid artery and ipsilateral optic nerve have been noted as an incidental finding in some patients without visual complaints.

For most patients with this disorder conservative management is appropriate. Most are elderly and have other forms of vascular disease, making them poor candidates for surgery. These individuals can be reassured that in most cases progression is extremely slow and the degree of visual loss is not disabling. In occasional patients with more rapidly progressive visual loss, surgical intervention may be effective in reversing the visual loss and preventing further deterioration.

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HISTORICAL NOTE

Paul Broca’s thermometric crown

The fame of Paul Broca as a precursor of cognitive neuroscience is mainly due to his groundbreaking work in describing brain lesions, most notably patient Leborgne’s speech disorder, and the correlation he drew with the left inferior frontal lesion that caused it, in a region known ever since as Broca’s area. Based on further cases of aphasia, he was also able to elucidate the left hemisphere’s systematic specialisation for language. Less recognised is his pioneering interest for in vivo brain imaging. He used measures of scalp temperature for the early diagnosis of brain lesions, as well as for functional imaging in normal subjects, which are today the two main applications of techniques such as magnetic resonance imaging. His imaging ideas were inspired from studies at the time that measured skin temperature to infer the localisation and mechanism of arterial lesions of the limbs, and to guide amputation whenever necessary.

In order to apply this approach to the field of neurolology, Broca devised a “thermometric crown, allowing the simultaneous application of six thermometers around the head. It is made of a series of small identical cotton pockets, tied together with a circular band of elastic material, with thermometers placed inside the pockets.” He later improved this apparatus by means of two additional thermometers critically located over the inferior frontal gyrus “which is assigned to language”.

Broca was well aware that “the dura mater, the skull, the scalp constitute an obstacle to heat radiations”, and acknowledged that it was “an open question to what extent thermometric exploration could be used for the general diagnosis of brain diseases”. He nevertheless gained some optimism from the obstinate attempts he made in patients and normal subjects.

Thus, “when the sylvian artery is occluded [by an embolism], the temporal thermometer should show a lower temperature than on the healthy side”. Despite its obvious limitations, Broca tried to make the most of the coarse spatial resolution of the technique, observing that following sylvian infarcts “blood is provided by collateral pathways, ie..., branches of the anterior and posterior cerebral arteries... which then become the site of an exaggerated blood flow. Hence one observes a higher temperature on the occipital and even more on the frontal thermometers, relative to the healthy side.”

Broca also tried to infer underlying pathophysiological mechanisms, and surmised that progressive thrombosis, contrary to embolism, leads to increased rather than to reduced temperatures. “One of the first patients in which I made this exploration was a doctor from the provinces who was introduced to me by his son, himself a doctor. Speech was not abolished, but it was already severely impaired; there was no disorder of sensation or motility. I easily recognized with my hand that the temperature of the left temporal region was notably increased, and I observed, using two thermometers applied on the temporal regions, that this increase was of 3 degrees, an excessive figure which I never observed again.” He predicted that the condition should worsen as a consequence of an extension of a “congestive softening” to the whole hemisphere. Indeed, the patient’s son soon informed Broca that “speech was entirely abolished, that intelligence was deeply impaired, that the patient was bed-ridden”, and three months later that his father eventually died.

Broca also hypothesised that brain temperature should increase with the execution of cognitive tasks by normal subjects. Importantly, he thought that those variations should be local, mostly affecting frontal regions dedicated to higher brain functions. He observed that “following intellectual work, the frontal temperature increases more than the temporal temperature, and the latter more than the occipital temperature. One may confirm this by placing the thermometric crown on resting subject. After about 20 minutes, when all thermometers have reached equilibrium, one asks the subject to perform a mental task. If he is only half-literate, if he does not read very fluently, one asks him to read a text aloud, and after a few minutes, one sees the thermometers rise, mainly the frontal ones.”

Stressing the influence of cultural acquisitions on brain function, Broca noted that “the result is nil or extremely minuscule if the subject can read without difficulty. To make the cerebral temperature rise [in medical students], I had to give them a more arduous work, generally consisting in adding thirty five digit numbers. Then the thermometers, mainly the frontal ones, rose substantially”.

Because of a regrettable lack of confidence in this venerable thermometric technique, we did not endeavour to replicate Broca’s findings. Nevertheless, we deemed it justified to acknowledge his open-mindedness and curiosity. There is no doubt Broca would have felt both perfectly comfortable and immensely excited with the present plethora of brain imaging techniques.

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References