**Dolichoectatic arterial compression of the anterior visual pathways: neuro-ophthalmic features and clinical course**

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**Aim:** To characterise the clinical findings and natural history of anterior visual pathway compression by dolichoectatic intracranial vessels.

**Methods:** A retrospective case review of patients evaluated in an outpatient neuro-ophthalmology clinic.

**Results:** 10 patients with this condition were identified. Dolichoectatic compression was confirmed by magnetic resonance imaging. In all patients, the average age at presentation was 70.6 years, and eight of the 10 were female. The carotid artery was involved in seven patients and the basilar in three. Patterns of visual loss varied depending on the site of compression. The most common pattern was optic neuropathy followed by visual field loss. In most patients, visual loss showed little progression over time. Over an average follow-up interval of 2.8 years, progressive visual loss was documented in only three cases. In one of these, neurosurgical intervention was undertaken with subsequent improvement of vision.

**Conclusions:** Visual loss resulting from compression of the visual pathways by dolichoectatic arteries is usually mild and only slowly progressive. Most patients are elderly, with other forms of vascular disease. Conservative management is thus usually appropriate in this disorder. In occasional cases with more rapid progression, surgical intervention may be beneficial.

**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,
<table>
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<th>Clinical features</th>
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<tr>
<td>Age/sex</td>
<td>73/F</td>
<td>81/F</td>
<td>68/M</td>
<td>48/M</td>
<td>59/M</td>
<td>68/F</td>
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<td>Diagnosis</td>
<td>Optic neuropathy, right-left</td>
<td>Right optic neuropathy, left NAION</td>
<td>Left optic neuropathy, Left optic neuropathy</td>
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<td>Left junctional scotoma, left Horner's syndrome</td>
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<td>Left optic tract compression</td>
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<td>Duration of symptoms</td>
<td>Many years</td>
<td>Uncertain OD, sudden OS</td>
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<td>Uncertain</td>
<td>3 Years</td>
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<td>5 Years</td>
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<td>1 Year</td>
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<tr>
<td>Medical history</td>
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<td>COPD, ARMD</td>
<td>Hypothyroid</td>
<td>HTN, CAD, AF, TIA, ARMD OS</td>
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<td>HTN, CAD, stroke</td>
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<td>HTN, CAD, aortic aneurysm, hypercholesterolemia</td>
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<td>Visual acuity at presentation</td>
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<td>OS</td>
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<td>20/50</td>
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<td>Visual fields at presentation</td>
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<td>OD</td>
<td>Inferior nasal defect</td>
<td>Inferior nasal defect</td>
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<td>OS</td>
<td>Central depression</td>
<td>Central depression</td>
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<td>MRI</td>
<td>Ectatic carotid arteries; elevation of optic nerves (OD&gt;OS)</td>
<td>Ectatic right carotid artery elevating chiasm and right optic nerve</td>
<td>Ectatic left carotid artery elevating chiasm and right optic nerve</td>
<td>Ectatic right carotid artery elevating chiasm and right optic nerve</td>
<td>Ectatic right carotid artery elevating chiasm and right optic nerve</td>
<td>Ectatic right carotid artery elevating chiasm and right optic nerve</td>
<td>Ectatic right carotid artery elevating chiasm and right optic nerve</td>
<td>Ectatic basilar artery compressing right optic tract</td>
<td>Ectatic basilar artery compressing right optic tract</td>
<td>Ectatic basilar artery compressing right optic tract</td>
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<td>Clinical course</td>
<td>Stable until CRAO OS 5 years later</td>
<td>Improved to 20/25 OD following carotid surgery 3 years later</td>
<td>Subjectively stable 4 years later</td>
<td>Decreased VA to no light perception</td>
<td>Stable right optic neuropathy</td>
<td>Initial progression then improved optic neuropathy (20/25 OS) following craniotomy 3 Years</td>
<td>Some progression of right optic neuropathy (20/200 OD) stable OS</td>
<td>Progressive ataxia</td>
<td>Stable exam at 7 month follow-up died of stroke 2 years later</td>
<td>Stable</td>
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<td>Follow up interval (last examination)</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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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 superior temporal 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.
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).

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.19

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.20

**References**

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 mechanisms of arterial lesions of the limbs, and to guide amputation whenever necessary.

In order to apply this approach to the field of neurology, 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 a 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