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

V Purvin, A Kawasaki, S Zeldes

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 scanning 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 in patients with optic neuropathy was nasal 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.

The term dolichoectasia derives from “dolichos” 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.1,2 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.3-6

Visual loss from compression of the anterior visual pathways by dolichoectatic vessels is well recognised.5,7-11 The natural history of this rare condition, however, is not well described. We sought a better definition of the clinical course of this disorder 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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</table>

AF, atrial fibrillation; ARMD, 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.
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.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


References
Paul Broca's thermometric crown

L Cohen, M J Smith and V Leroux-Hugon

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