
SHORT COMMUNICATION

Case report

Isolated ocular ischemic syndrome with no cerebral involvement in common carotid artery occlusion

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PURPOSE. *The ocular ischemic syndrome (OIS) is sometimes a complication of common carotid artery (CCA) occlusion causing complete interruption of blood flow through both the internal and external carotid arteries we investigated a single case of an isolated OIS that remained undiagnosed for two years, because the underlying CCA pathology caused no cerebral involvement.*

CASE REPORT. *A 57-year-old man presented with subacute painful visual loss in the right eye in a setting of hypertension, smoking and coronary artery disease.*

RESULTS. *Neurological examination, a brain CT and MRI scan were all normal. Extensive laboratory work-up excluded small artery disease, inflammatory arteritis or cardiac causes of retinal embolism. Ophthalmologic evaluation and fluorescein angiography gave findings consistent with OIS, while vascular ultrasound evaluation and aortic arch angiography verified right CCA occlusion accompanied by an extensive collateral network.*

CONCLUSIONS. *Had this patient been referred sooner for a simple carotid artery work-up, both the CCA occlusion and the OIS could probably had been prevented. (Eur J Ophthalmol 2001; 11: 97-101)*

KEY WORDS. *Common carotid occlusion, Ocular ischemic syndrome*

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INTRODUCTION

Common carotid artery (CCA) occlusion may produce cerebral and/or ocular ischemic symptoms in relation to reduced blood flow or embolism in the internal carotid system (1). Concomitant impairment of the external carotid collateral network in CCA occlusion may explain the higher frequency of cerebral transient ischemic attacks, stroke and amaurosis fugax than with isolated internal carotid artery (ICA) occlusion (1), although 25% of cases remain asymptomatic (2).

The ocular ischemic syndrome (OIS) (3) is frequently the only or earliest clinical manifestation of severe

carotid artery disease (CAD) (4) but its recognition is hampered by its diverse and sometimes subtle presentation (3, 4), the similarity of visual phenomena to other ocular conditions (3, 4) and the frequent absence of neurological symptoms or signs (4, 5).

These patients are at risk for ischemic cerebrovascular, coronary or peripheral artery disease and the prognosis for life, as well as for vision, is threatened (6-8). Thus, early diagnosis (3, 8) requires alert and cooperative ophthalmologists and neurologists.

We present a documented case of isolated severe ocular ischemia that remained undiagnosed for two years, probably because the underlying CCA pathology caused no cerebral involvement.

Case report

A 57-year-old white man with a history of hypertension and cigarettes smoking for ten years was admitted for evaluation of a right-sided monocular visual loss, accompanied by periorbital pain ipsilaterally. His medical history was significant for coronary artery disease that had been managed with transluminal angioplasty in another hospital, two years ago. Since then, he had experienced several episodes of transient monocular blurring of vision in the right eye, that resolved in a few minutes. Treated with acetylsalicylic acid 100 mg/d these episodes did not recur. One month before admission he had noticed gradual visual impairment in the right eye, followed three weeks later by almost complete visual loss and periorbital pain.

On admission, the best corrected visual acuity was 20/20 for the left eye, but only hand motion vision in the temporal hemifield for the right eye. Under daylight conditions, the left pupil was 3 mm with normal reac-

tion to direct light, to dark and near, but no consensual light reaction. The right pupil was 7 mm and fixed to either direct or consensual light stimulation, to dark, near and to the instillation of 1% pilocarpine solution. Extraocular movements were full, there was no ptosis and corneal sensation was normal bilaterally. The intraocular pressure in repeated measurements, at different times of day did not exceed 18 mmHg, with no more than 4 mmHg difference between the two eyes, the right eye having the higher pressure. Visual fields with automatic perimetry only disclosed an island in the temporal hemifield of the right eye. Ipsilateral funduscopy showed marked arterial stenoses and straightening, with irregular venous dilation, dot and blot retinal hemorrhages and microaneurysms, most marked in the superior nasal arcade, with no edema, cherry red spot or embolic material (Fig. 1a). The right optic nerve head was pale with clear borders and stagnation of the blood column at the exit of vessels from the optic disc. Slit lamp examination and gonioscopy detected con-

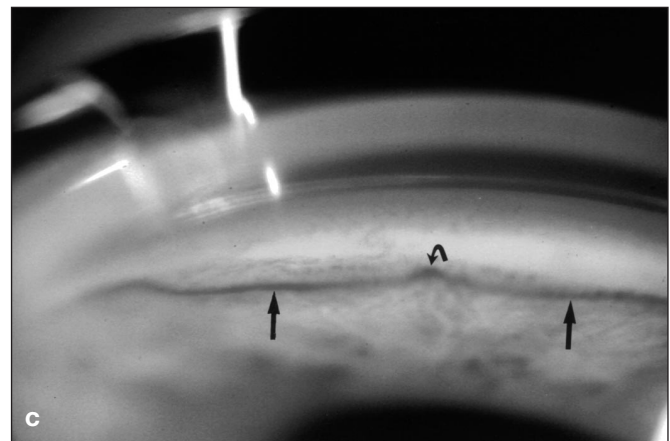
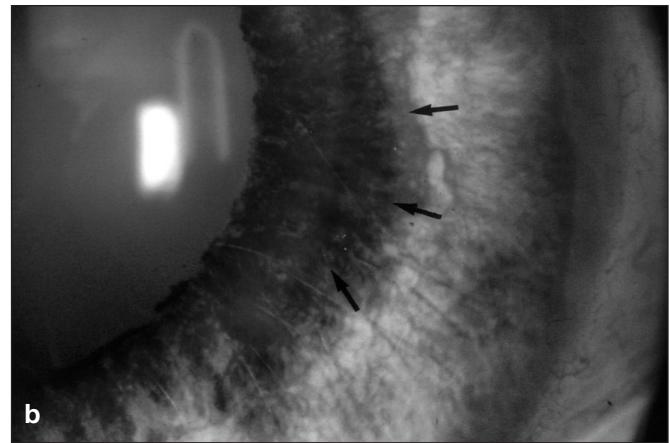


Fig. 1a - Right funduscopy showed marked arterial stenoses and straightening (small arrow), with venous dilations (large arrow), dot and blot retinal hemorrhages and microaneurysms (curved arrow).

b - Confluent right iris neovascularization throughout the surface (arrows).

c - Neovascularization included the angle (arrows) with the anterior chamber angle partially closed, due to anterior synechiae (curved arrow).

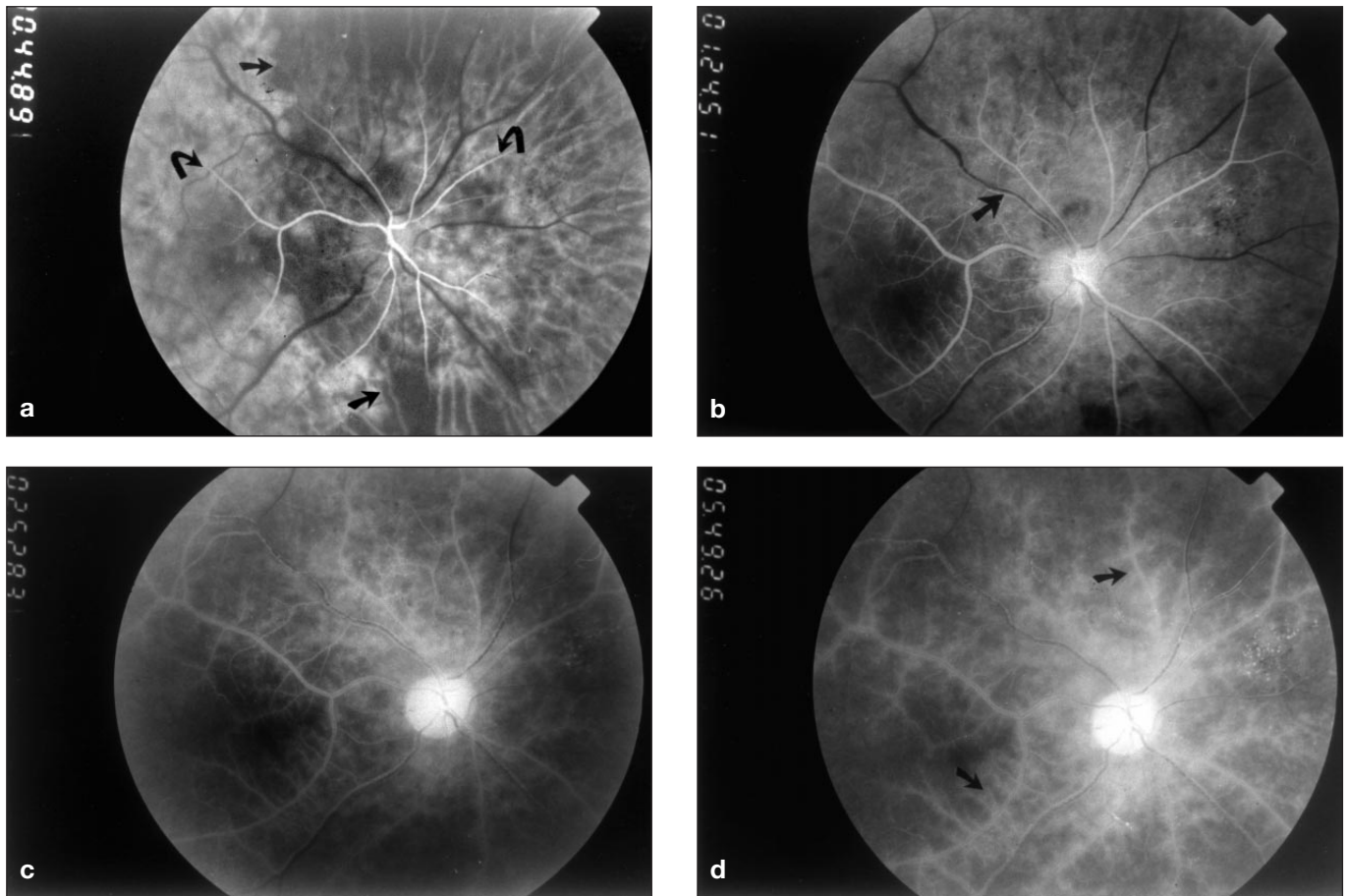


Fig. 2 - Right fluorescein angiography.

a - 45 sec after injection. Patchy choroidal filling (straight arrows) and delayed arterial staining (curved arrow).

b - 84 sec Choroidal filling is partially incomplete and the veins remain empty (arrow).

c - 173 sec. Choroidal filling is completed.

d - 350 sec. Dye leakage is obvious (arrows). The venous circulation remains abnormal.

fluent right iris neovascularization throughout the iris surface (Fig. 1b), including the angle, with the anterior chamber angle partially closed due to anterior synechiae (Fig. 1c). The conjunctiva, cornea, lens and anterior vitreous were clear. Fluorescein fundus angiography showed markedly delayed and patchy choroidal filling (over 60 sec), prolonged arteriovenous transit time (over 120 sec) and late staining of the retinal arteries and veins in the right eye (Fig. 2).

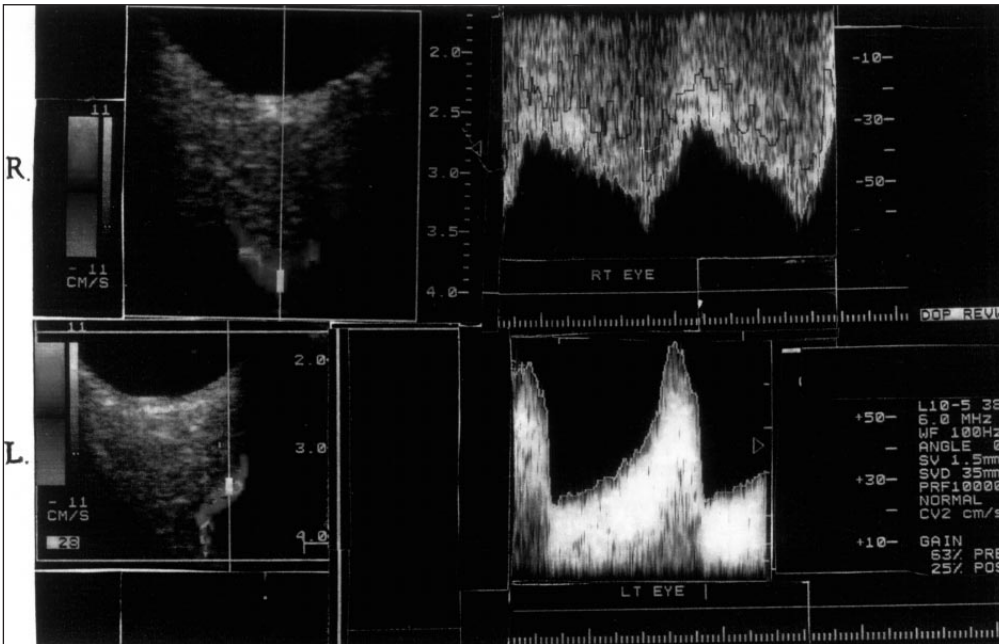
The left eye and the remainder of the general and neurological examination, a brain CT and MRI scan were all normal. Extensive hematological, biochemical and immunological tests were either negative or unrevealing. Vascular ultrasound evaluation with color-coded Duplex imaging indicated a right CCA occlusion, with retrograde flow in the right oph-

thalmic artery (Fig. 3). Transcranial Doppler detected intracranial left-to-right hemisphere collateral circulation through the contralateral ICA and middle cerebral artery, as well as from the posterior circulation towards both carotid systems.

Finally, an aortic arch angiography verified the right CCA occlusion and also showed irregular atheromatous stenoses of the right vertebral artery and insignificant (<50%) left bifurcation change (Fig. 4).

DISCUSSION

Hedges (9) first described chronic ocular ischemia secondary to ICA occlusion and later reports included CCA occlusion as well (1, 2, 4). Although the true in-

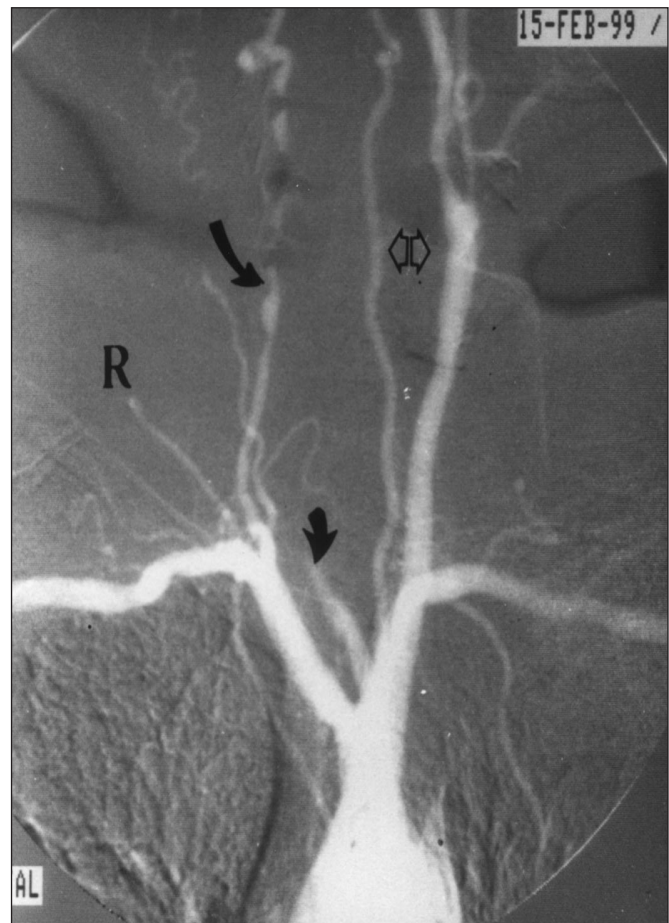


◀ **Fig. 3** - Color-coded Duplex imaging with the corresponding Doppler spectra of the right (R) and left (L) ophthalmic artery. Note the retrograde flow, with a downward spectrum, in the right ophthalmic artery, in contrast with the normal left ophthalmic artery, with an upward spectrum.

idence of OIS is not known, it is estimated that 5% of patients with marked carotid artery stenosis present with the syndrome (7). The OIS is presumably caused by reduction of blood flow to the eyeball producing anterior or posterior segment ischemia or both, sometimes accompanied by periorbital pain (3, 10). However not all patients with severe CAD develop OIS (7), suggesting an ocular steal mechanism (11). Recent hemodynamic studies support this, and also indicate that in certain patients with bilateral ICA (12, 13) or ipsilateral CCA occlusion (14), flow reversal in the ophthalmic artery acts as collateral, supporting the cerebral circulation, stealing blood away from the eye (12-14).

Our patient presented with subacute painful visual loss in the right eye, associated with ipsilateral CCA atherothrombotic occlusion, in a setting of hypertension, smoking and coronary artery disease. There was no evidence of associated small-artery disease, inflammatory arteritis or cardiac causes of potential retinal embolism. Ophthalmologic evaluation and fluorescein angiography gave findings consistent with both anterior and posterior segment ocular ischemia indicative of OIS (3, 6, 13), in the absence of diabetes, central retinal vein occlusion or uveitis, that could masquerade as OIS (3). The clinical characteristics of the areactive right mydriasis in our patient possibly reflected iris or cil-

▼ **Fig. 4** - Aortic arch angiography (oblique view) confined in the right CCA occlusion (short arrow) and also showed irregular stenoses of the right vertebral artery (long arrow) and insignificant left carotid bifurcation and left vertebral artery changes (double arrow).



iliary ganglion ischemia due to prolonged hypoperfusion of the eye as has been suggested in CCA occlusion in patients with the OIS (15) or in ICA occlusion with ischemic optic neuropathy (16) and central retinal artery occlusion (17). However ipsilateral mydriasis due to oculomotor nerve paresis, has been reported in CCA occlusion, accompanied with visual loss (18) or not (19), as well as in ICA occlusion in association with the OIS (20).

The complete absence of ipsilateral hemisphere involvement detectable either clinically or by neuroimaging studies (CT and MRI), might be due to the development of an adequate collateral supply through the circle of Willis from the contralateral carotid and verte-brobasilar system (14, 18), as well as from the ipsi-

lateral ophthalmic artery (8, 11, 13), as was indicated by the vascular ultrasound findings.

We believe that the fact that this patient had had no neurological symptoms, delayed the diagnosis of the underlying CCA pathology for two years, and his well-developed collateral supply protected the cerebral circulation at the expense of his ocular function and vision (12-14). Had this patient been referred sooner for a simple carotid artery work-up (8, 13, 16), both the CCA occlusion and the OIS could probably had been prevented (3, 6, 21, 22).

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REFERENCES

1. Levine SR, Welch KMA, Common carotid artery occlusion. *Neurology* 1989; 39: 178-86.
2. Podore PC, Rob CG, DeWeese JA, Green RM. Chronic common carotid occlusion. *Stroke* 1981; 12: 98-100.
3. Mizener JB, Podhajsky P, Hayreh SS. Ocular Ischemic syndrome. *Ophthalmology* 1997; 104: 859-64.
4. Ross Russell RW, Page NGR. Critical perfusion of brain and retina. *Brain* 1983; 106: 419-34.
5. Kearns TP. Differential diagnosis of central retinal vein occlusion. *Ophthalmology* 1983; 90: 475-80.
6. Sivalingam A, Brown GC, Magargal LE. The ocular ischemic syndrome. Visual prognosis and the effect of treatment. *Int Ophthalmol* 1991; 15: 15-20.
7. Kearns TP. Ophthalmology and the carotid artery. *Am J Ophthalmol* 1979; 88: 714-22.
8. Riordan-Eva P, Restori M, Hamilton AMP, Levy IS. Orbital ultrasound in the ocular ischemic syndrome. *Eye* 1994; 8: 93-6.
9. Hedges TR. Ophthalmoscopic findings in internal carotid artery occlusion. *Bull Hopkins Hosp* 1962; 111: 89-97.
10. Carte JE. Chronic ocular ischemia and carotid vascular disease. *Stroke* 1985; 16: 721.
11. Huckman MS, Haas J. Reversed flow through the ophthalmic artery as a causes of rubeosis iridis. *Am J Ophthalmol* 1972; 74: 1094.
12. Kerty E, Horven O. Ocular hemodynamic changes in patients with high-grade carotid occlusive disease and development of chronic ocular ischemia. Doppler and dynamic tonometry findings. *Acta Ophthalmol Scand* 1995; 73: 66-71.
13. Costa VP, Kuzniec S, Molnar LJ, et al. Clinical findings band hemodynamic changes associated with severe occlusive carotid artery disease. *Ophthalmology* 1997; 104: 1994-2002.
14. Brown GC, Magargal LE, Simeone FA, Goldberg RE, Federman JL, Benson WE. Arterial obstruction and ocular neovascularization. *Ophthalmology* 1982;17: 139.
15. Fisher CM. Some neuro-ophthalmological observatins. *J Neurol Neurosurg Psych* 1967; 30: 383-93.
16. Biousse V, Schaison M, Touboul PJ, D'Anglejan-Chatillon J, Bousser MG. Ischemic optic neuropathy associated with internal carotid artery dissection. *Arch Neurol* 1998; 55: 715-9.
17. Newman NJ, Kline LB, Leifer D, Lessel S. Ocular stroke and carotid artery dissection. *Neurology* 1989; 39: 1452-64.
18. Bogousslavsky J, Pedrazzi PL, Borruat FX, Regil F. Isolated complete orbital infarction: A common carotid artery syndrome. *Eur Neurol* 1991; 31: 72-6.
19. Koennecke HC, Seyfert S. Mydriatic pupil as the presenting sign of common carotid artery dissection. *Stroke* 1998; 29: 2653-55.
20. Galetta SL, Leahey A, Nichols CW, Raps EC. Orbital ischemia ophthalmoparesis and carotid dissection. *J Clin Neuro-Ophthalmol* 1991; 11: 284 -7.
21. Thompson JE. Carotid endarterectomy for asymptomatic stenosis: benefits confirmed by randomized trial. *J Vasc Surg* 1993; 17: 798-800.
22. Johnston ME, Gonder GC, Canny CL. Successful treatment of the ocular ischemic syndrome with panretinal photocoagulation and cerebrovascular surgery. *Can J Ophthalmol* 1988; 23: 114-19.