Spontaneous Internal Jugular Vein Thrombosis and Venous-Stasis Retinopathy

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A case is presented in which the detection of venous-stasis retinopathy in one eye led to investigation of the carotid circulation. There were no neurologic symptoms of carotid insufficiency, and noninvasive tests failed to reveal significant carotid pathology. Digital subtraction angiography and carotid angiography demonstrated a carotid plaque of doubtful significance. At carotid endarterectomy, the venous-stasis retinopathy was found to be associated with venous thrombosis distant from the eye and in the internal jugular vein. This site is beyond the range over which isolated ocular vascular effects would be expected and was thought to be unrelated to the hemodynamically insignificant, nonulcerated carotid artery plaque. The possibility of this association being causal is discussed. (Stroke 1987;18:808-811)

Venous-stasis retinopathy (VSR) is a recognized ocular indicator of arterial insufficiency. Consequently, the detection of this ophthalmoscopic sign implies a threat to the cerebral circulation and should initiate investigation to identify the cause of arterial insufficiency. VSR is commonly the result of significant stenosis or occlusion of the ipsilateral internal carotid artery, although a causative lesion could also be situated in the ophthalmic artery or central retinal artery proximal to the optic nerve head.

Stenosis and occlusion of the internal carotid artery is associated with a wide range of neurologic signs and symptoms. Symptomatic transient ischemic episodes include hemiparesis, hemianesthesia, aphasia, and amaurosis fugax, while signs suggestive of carotid disease include a carotid bruit, retinal emboli, and VSR. When such signs and symptoms occur in combination, the further investigation and management of the patient is a matter of clearly defined clinical protocol. However, the appropriate clinical management is less well established in the presence of an isolated asymptomatic sign such as VSR when other evidence of carotid insufficiency is not forthcoming even after further investigation. The later discovery of a venous, rather than arterial, anomaly raised the question of a causal vs. casual relation between VSR and internal jugular vein thrombosis. If these findings are directly related, it is believed to be the first reported instance.

Report of a Case

A 67-year-old man presented for ophthalmic examination with the complaint of "smoky" vision with the left eye during the last 4–6 weeks. This visual disturbance had remained unchanged during that period. The patient denied symptoms of a transient or episodic nature such as amaurosis fugax, orbital pain, hemiparesis, syncope, or other ocular or neurologic disturbances. The patient reported that the vision of the right eye was entirely normal. Visual acuities were found to be (right) 6/9 and (left) 6/12 = , compared with acuities of 6/9 recorded at a routine eye examination 9 weeks earlier.

The patient's ocular history was remarkable only in that optic disk cupping of a marked degree was present in both eyes, being first noted in 1969. The optic cup/disk ratio was 0.8 in both eyes and had remained unchanged since that time. There had been numerous visual field assessments and estimates of intraocular pressure during that period (including the last examination), but no abnormality had been detected. The anterior chamber angles were wide open. These findings excluded a diagnosis of glaucoma.

The patient was under continuing treatment for rheumatoid arthritis. He suffered maturity-onset asthma and had undergone surgery for aortic aneurysm 10 years previously.

Results

On ophthalmoscopic examination, the right eye was unremarkable except for the disk cupping noted earlier. The left optic disk was edematous, and the disk margins were blurred (Figure 1). The disk edema caused a reduction of the cup/disk ratio to 0.6. There was clouding of the vitreous overlying the disk and in Cloquet's canal. Scattered microaneurysms and small intraretinal hemorrhages were present throughout the left ocular fundus. The retinal veins were mildly turgid in appearance, and there was a minimal degree of macular edema.

Intraocular pressures were 16 mm Hg (applanation tonometry) in both eyes. Both visual fields were nor-
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Figure 1. Fundus photograph of the left eye, showing optic disk edema (reducing the optic cup/disk ratio to 0.6), mildly dilated retinal veins, and scattered retinal hemorrhages.

mal when tested with the Friedmann Visual Field Analyzer Mark 11, although a slight left macular disturbance was reported on the Amsler’s charts. All pupil reactions were normal, and in particular, no Marcus Gunn’s pupil response was elicited. Blood pressure was 140/80 mm Hg (seated), and no bruit could be heard over the cervical course of the carotid arteries.

Because the patient had impending overseas travel, referral for vascular work-up was arranged without delay. Further investigation by oculoplethysmography and ultrasonic arteriography (Hokanson) was noncontributory. An intravenous digital subtraction angiogram (DSA) was performed and demonstrated a plaque at the origin of the left internal carotid artery. This plaque appeared to be irregular and possibly ulcerated, and this finding was confirmed by selective carotid angiography. Since the plaque may have been ulcerated and a possible source of emboli, surgical removal was recommended, and the patient subsequently underwent left carotid endarterectomy.

At surgery, arteriotomy confirmed the presence of a plaque that did not significantly encroach on the arterial lumen. There was no evidence of ulceration of the surface of the plaque. However, the internal jugular vein was found to be occluded between the occipital tributary and the common facial tributary for 1.5 cm. Both the occipital vein and the common facial vein were very much enlarged. The surgeon had not observed this pathologic circumstance in over 1,000 carotid endarterectomies.

Carotid endarterectomy was performed. The operation was technically uncomplicated, with the atheroma feathering off very nicely above and below the plaque. A routine postoperative angiogram confirmed a smooth reconstruction, and the patient made an uneventful postoperative recovery.

Over the following months, there was gradual resolution of the optic disk edema, microaneurysms, intraretinal hemorrhages, and macular edema, accompanied by improvement of visual acuity to 6/9. However, 4 months after surgery the patient was still aware of slight blurring of the vision of the left eye. Carotid endarterectomy did not appear to hasten the resolution of the retinopathy. Further oculoplethysmography indicated normal carotid artery competence. Visual acuity has remained 6/9 since that time.

Discussion

Venous-stasis retinopathy is not an uncommon accompaniment of carotid artery disease. Kearns et al observed VSR in 18% of patients having unilateral carotid artery occlusion, while Samples et al suggest that approximately 10% of subjects with angiographically proven stenosis of >90% of the cross-sectional area of the vessel have VSR. An earlier estimate of 5% of cases with occlusion or significant stenosis may reflect the current increased awareness of ocular correlations of carotid disease and more extensive investigations for systemic causes of ischemic ocular signs. An increase in one such important sign was found by one of the authors (J.P. Royle, unpublished data) following careful inspection of “at risk” patients. In a prospective study of patients about to undergo carotid endarterectomy, 18% were observed during routine ophthalmoscopy to have retinal emboli. However, this proportion increased to 35% when these patients were again examined ophthalmoscopically while their pupils were dilated during surgery.

VSR appears to be the result of chronic arterial insufficiency rather than the more acute disturbances that give rise to amaurosis fugax. Indeed, Kearns, who initially coined the term VSR, recently suggested that the name be changed to “chronic ischemic retinopathy” to contrast with those acute symptoms that are more typical of carotid disease. Such chronic or progressive deficiencies constitute a serious threat, perhaps greater than that of embolic stroke, when available collateral cerebral supply is inadequate. The theory of chronic insufficiency as a cause of VSR is supported by a report that describes the rapid resolution of VSR in a patient following restoration of normal arterial supply with successful endarterectomy. The present case also appears to support this view since the retinopathy gradually disappeared, presumably as collateral vessels restored venous drainage to that side of the head.

In the case reported here, it was necessary to exclude local ocular causes of the patient’s retinopathy and disk edema. It is well documented that central retinal vein occlusion frequently occurs in association with elevated intraocular pressure or frank glaucoma. Approximately 30% of patients with central retinal vein thrombosis have preexisting open angle glaucoma in that eye. Elevated intraocular pressure reduces the vascular perfusion pressure in the eye, thus contribu-
ing to the development of venous stasis.\textsuperscript{7,8} Because the patient described here had markedly excavated optic disks, he had been under regular and frequent review for glaucoma for several years. However, at no time were intraocular pressures found to be abnormal, and no glaucomatous visual field defects were elicited.

Papilledema (secondary to increased intracranial pressure), optic neuritis, and anterior ischemic optic neuropathy were eliminated as possible diagnoses since the signs, symptoms, or subsequent course of the condition were not consistent with these diseases. In particular, visual field results were negative, disk edema was unilateral, and no neurologic signs or symptoms were detected.

Although not secondary to increased intraocular pressure, the patient's retinopathy may have been a form of central retinal vein occlusion rather than VSR. In fact, the terminology used to describe these two conditions is confusing. The former is characterized by dramatic and florid retinal hemorrhaging; the latter has much less obvious features, although there is some dispute as to whether optic disk edema is found in VSR.\textsuperscript{1,2,3} It is possible that the two conditions are the extremes of a spectrum, with intermediate presentations also occurring. Despite this uncertainty, all types of occlusion or stasis are generally believed to be the result of some form of arterial insufficiency.

In this case, principal suspicion fell on the carotid circulation. Further investigations of the patient were based on this assumption; however, all noninvasive tests results were negative. Of course, it is possible that these results were falsely negative. False-negative results of ophthalmodynamometry have been reported\textsuperscript{10} in a small percent of patients with angiographically proven occlusion or high-grade stenosis and presumably result from the establishment of adequate collateral or retrograde flow. Carotid auscultation and phonoangiography\textsuperscript{11} and oculoplethysmography\textsuperscript{12,13} also give significant false-negative rates.

The use of a battery of noninvasive tests of arterial competency, as advocated by Ackerman,\textsuperscript{14} reduces the probability of false-negative and false-positive results. However, when suspicion is strong, the patient should be submitted to angiography. Intravenous DSA, while carrying less risk than the more invasive form of angiography, suffers from poorer resolution capabilities and may not adequately define the extent of a lesion.\textsuperscript{15,16} In this case, the suspicious findings seen on DSA were confirmed by carotid angiography before proceeding to carotid endarterectomy. At surgery, the carotid lesion proved to be occluding approximately 20\% of the vessel lumen and could not explain the ocular signs and symptoms. The occlusion of the left jugular vein just above its junction with the common facial vein became obvious during surgery. The resulting venous backpressure extending to the level of the orbit would explain the VSR observed in the left eye.

There are a number of interesting aspects of this case with its presumed relation of VSR with internal jugular vein thrombosis. Firstly, the ocular signs were unilateral. The venous drainage of the eye is such that one might expect bilateral, albeit asymmetric, involvement of the other eye. Secondly, there was no history or other clinical evidence elsewhere of venous stasis on either side. Thirdly, spontaneous occlusion of the internal jugular vein appears to be a very rare occurrence, most occlusions being secondary to central venous catheterization.\textsuperscript{17} McNeill\textsuperscript{17} also commented that many occlusions may go unrecognized due to the deep location of the vessel, the adequacy of collateral circulation, and the high venous flow rate. In fact, this case might have remained undetected without direct visualization of the internal jugular at surgery. Although catheterization via the left internal jugular vein may have been carried out during the patient's previous aortic surgery, it is unlikely that thrombotic complication would occur after the lapse of 10 years.

This report is believed to be the first case in which a spontaneous internal jugular vein thrombosis has been detected as a result of investigation of VSR as a presenting sign, although the possibility exists that the association was fortuitous rather than causal. A possible mechanism would be that the increase in venous backpressure limited arterial flow through the ophthalmic vascular tree. A similar mechanism was proposed in a case involving carotid-cavernous sinus fistula where central retinal vein occlusion was seen to follow 3 months after VSR.\textsuperscript{18} (In this report, the VSR was accompanied by other evidence of distant venous obstruction.) It could be predicted that retinal signs might also be seen following secondary jugular vein thrombosis or ligation. Although this association has not previously been reported, the possibility of internal jugular vein or related venous thrombosis may require consideration. This is especially relevant if other causes of arterial insufficiency are equivocal or absent, or where there has been no instrumentation involving the internal jugular vein. Any additional cases of this or similar venous thrombosis associated with VSR, yet to be reported, may support the contention of a causal relation discussed in this case.

**Acknowledgment**

We would like to acknowledge the assistance of Dr. Geoffrey A. Gutteridge, FFARACS, Austin Hospital, Melbourne, Australia.

**References**


Key Words • venous-stasis retinopathy • internal jugular vein thrombosis • carotid artery
Spontaneous internal jugular vein thrombosis and venous-stasis retinopathy.
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Stroke. 1987;18:808-811
doi: 10.1161/01.STR.18.4.808

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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