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Ophthalmic Artery Steal

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Abstract

The optometrist needs to be aware of the potential manifestations of ophthalmic artery steal and its appropriate management, both ocular and systemic. Timely diagnosis by an eyecare specialist can play a crucial role in reducing patient mortality.

Introduction

Vascular "steal" can be defined as the compensatory siphoning or redirecting of blood flow from one area to another. Fisher first employed the term "subclavian steal" in reference to the phenomenon of reversed blood flow through the vertebral artery as a means of compensating for proximal stenosis of the ipsilateral subclavian artery.[1] The distal subclavian artery would "steal" blood from the vertebral circulation, with reverse flow through the vertebral artery, in order to maintain an adequate supply to the upper extremity beyond the point of subclavian occlusion.[1] "Ophthalmic artery steal" has been used in the literature in a slightly different fashion, as a means of describing the reversed blood flow through the ophthalmic artery to compensate for inadequate flow in the internal carotid artery, typically when there is an occlusion of the internal carotid just proximal to the origin of the ophthalmic artery. In this situation, the ophthalmic artery is serving as a conduit for blood from branches of the external carotid to the internal carotid; hence, it is actually the internal carotid that is "stealing" blood by means of reverse flow through the ophthalmic.[2,3,4] A case of a patient with ocular ischemic syndrome from suspected ophthalmic artery steal phenomenon is presented.

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Case Report

A 60-year-old male presented with concerns of a red, painful left eye of one week's duration. Also of note: he reported that for the past few months, he had been experiencing short-lived episodes of graying vision that would last five to ten seconds and then spontaneously resolve. However, on the morning he presented for his eye examination, he reported persistent profound loss of vision in the left eye with the ability to perceive "just shadows." The patient denied symptoms of jaw claudication, headache, scalp tenderness, malaise, fever, and night sweats. He noted intentional weight loss during the last few months due to dietary changes in order to maintain better glycemic control.

The patient's past ocular history was largely unremarkable with good visual acuity noted at his last dilated exam, which had been performed two months prior. His systemic health was significant for type 2 diabetes mellitus, hypertension, hyperlipidemia, and benign prostate hypertrophy for which he was taking lovastatin, glyburide, metformin, and terazosin. Additionally, he reported 40 years of tobacco use (approximately 2.5-3 packs/day) and extensive ethanol consumption (6 pack of beer/day).

On examination his best-corrected visual acuity was 20/20 in his right eye and hand motion at three feet in the left eye. Extraocular motility testing was normal while pupil evaluation revealed a fixed mid-dilated left pupil. Slit lamp biomicroscopy demonstrated age-appropriate findings in the right eye while the left eye was characterized by microcystic edema with Descemet's folds temporally, grade 2+ dilated conjunctival vessels, 1+ flare but no cell in the anterior chamber, as well as neovascularization of the iris root inferotemporally and temporally. Goldmann intraocular pressures (IOP) were 12 mm Hg in the right eye and 30 mm Hg in the left eye. Gonioscopic examination confirmed angle neovascularization in the nasal aspect of the left eye as well as engorgement of the arterial circle. By Spaeth classification, both the right and left eyes were open to D40R.

Fundus evaluation of the right eye was notable for segmental hypoplasia of the optic nerve and a choroidal nevus in the posterior pole. The detail of the left eye was limited by the edematous cornea, so B-scan ultrasound

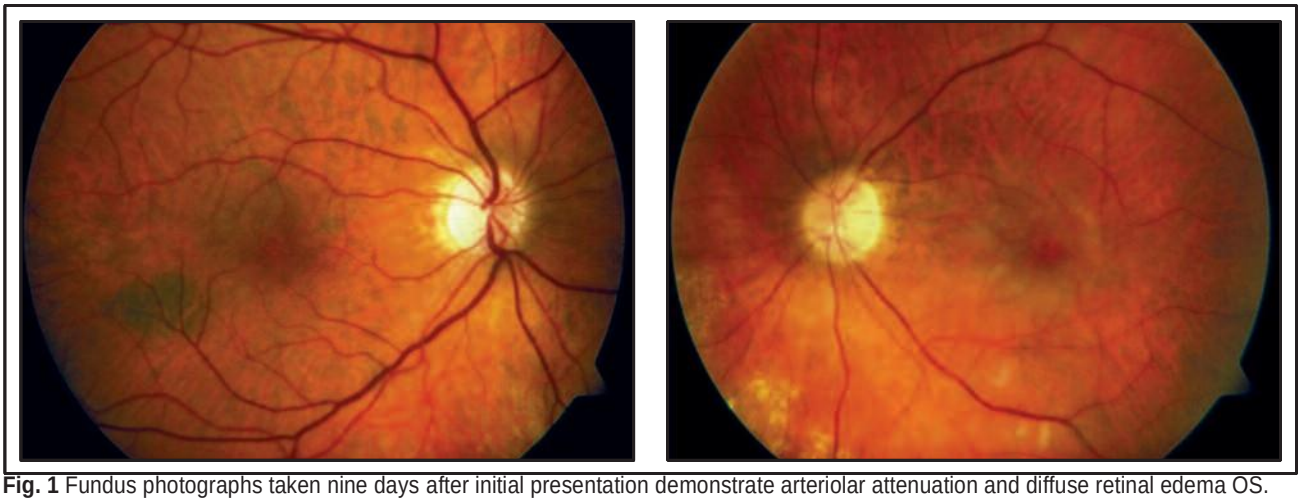

Fig. 1 Fundus photographs taken nine days after initial presentation demonstrate arteriolar attenuation and diffuse retinal edema OS.

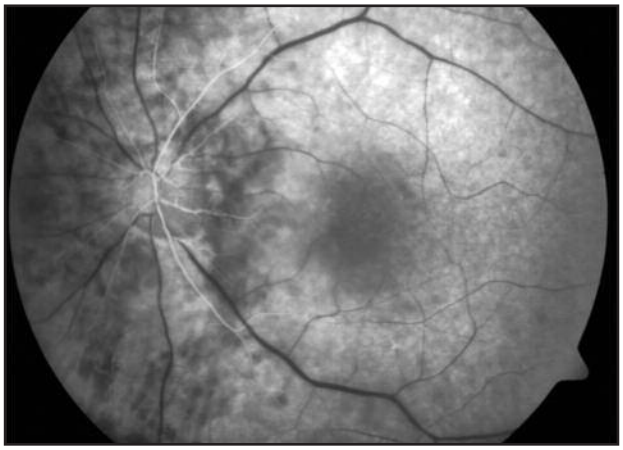

Fig. 2 Fluorescein angiography at 45 seconds demonstrates patchy choroidal filling and delayed retinal arteriolar filling, suggesting a partially reperfused central retinal artery occlusion and reduced ophthalmic artery perfusion.

was utilized. Ultrasonography was grossly unremarkable without evidence of mass or retinal detachment. Despite limited views, it was determined that the left fundus demonstrated attenuated retinal vessels and diffuse edema of the posterior pole; however, the presence of a cherry red spot was equivocal. The diagnosis of neovascular glaucoma was made, and several differential diagnoses for the underlying etiology were considered: central retinal artery occlusion (CRAO), ocular ischemic syndrome, retrobulbar mass, and/or giant cell arteritis in the absence of classic systemic symptoms.

An urgent carotid duplex was ordered and the following labs were obtained: complete blood count (CBC), Westergren erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), finger-stick glucose, glycosylated hemoglobin (HbA1c), prothrombin time (PT), international normalized ratio (INR), liver function tests (LFTs), urinalysis, and a lipid panel. A review of recent vital signs in the record revealed that the patient's systolic blood pressure had varied between 140 and 170 mm Hg and his diastolic blood pressure had ranged from 70 mm Hg to 90 mm Hg. In line with the trend, his blood pressure was 161/85 on the day of his urgent visit. The complete blood count was entirely within normal range. The ESR was 1 mm/hr (normal <30), the CRP level was 0.16 mg/dL (normal <0.744), whole blood glucose measured 180 mg/dL, HbA1c was 7.4%, PT was 12.71 seconds (normal 11.8-13.5) and INR was 1.0. While the alanine aminotransferase level was elevated at 80 mg/dL (normal 7.0-45.0mg/dL), all of the other liver enzymes were normal, and urinalysis was negative for glycosuria or proteinuria. The lipid panel was largely within normal limits with the exception of slightly reduced HDL levels of 27.1 mg/dL (>40 desirable); the total cholesterol level was 141 mg/dL (normal <200), the triglyceride level was 115 mg/dL (normal 40-160), and the LDL was 90mg/dL (<130 desirable).

To address the elevated intraocular pressure, the patient was immediately started on oral acetazolamide 250mg four times a day as well as topical dorzolamide/timolol maleate twice a day and brimonidine solution three times a day in the left eye. Additionally, the patient was prescribed prednisolone acetate 1% (Pred Forte®, Allergan) every hour and atropine 1% twice a day for the left eye.

Fundus photography and fluorescein angiography were ordered immediately but ultimately obtained nine days later (Fig. 1) and a consultation with retina clinic was arranged. On that day, 1508 spots of panretinal photocoagulation were applied to the left eye, followed by 301 shots the following day, by which time the IOP had dropped to 10 mm Hg in the right eye and 13 mm Hg in the left eye. On the third day, an intravitreal bevacizumab injection was administered OS. Five days

later the visual acuity in the left eye had stabilized at hand motion vision, there was full regression of the iris neovascularization, and the IOP remained controlled at 10 mm Hg.

Nine days later, the fundus examination was unchanged. The fluorescein angiogram of the right eye was unremarkable; however, the left eye demonstrated not only delayed retinal arteriole filling as had been expected, but also patchy choroidal flush (Fig. 2). Therefore, this was not just a partially-reperfused central retinal artery occlusion given the concurrent obstruction of the choroidal circulation; the collective findings suggested reduced blood flow occurring earlier in the orbital pathway, presumably at the level of the ophthalmic artery. While all the labs returned within expected ranges, the carotid ultrasound showed probable occlusion of the left internal carotid artery starting from the carotid bulb, which was the likely culprit in the ocular findings described earlier (Fig. 3). Subsequent magnetic resonance imaging, including diffusion-weighted images and magnetic resonance angiography (MRA) of the head and neck confirmed 100% obstruction of the left internal carotid artery at its origin (Fig. 4) The right and left external carotid arteries were found to be patent, as well as the right internal carotid artery. The left middle cerebral artery (MCA) showed diminished caliber, flow signal, and cortical branches, suggesting hypoperfusion to the left MCA territory. The left MCA appeared to be "diminutive" and supplied predominantly across the anterior portion of the circle of Willis. There was proximal occlusion of the left vertebral artery with distal reconstitution and focal stenoses in the proximal right vertebral artery with focal flow gaps, indicating the possibility of high-grade stenosis. No large posterior communicating artery could be visualized. Additionally, the MRI of the brain showed no signs of acute infarct or heme. Since the patient was asymptomatic by that time, carotid endarterectomy was not indicated and the patient was scheduled for regular follow-up with vascular clinic. As far as eye care, the patient was monitored frequently while on maximally tolerated medical therapy until the neovascularization, initially responsive to pan-retinal photocoagulation and intravitreal bevacizumab, returned approximately one month after his first evaluation. Gonioscopy was repeated, which revealed an angle classification of AQ30 with nearly 360 degree synechial closure in the left eye. He was arranged for Ahmed valve surgery OS to reduce the dependence on topical ocular anti-hypertensive medications and for the anticipated rise in IOP in the future. The patient was maintained on dorzolamide hydrochloride-timolol maleate (Cosopt®, Merck Frosst) b.i.d. and Pred Forte 1% t.i.d. OS post-operatively, but he subsequently passed away of unknown etiology approximately two months after completion of Ahmed valve surgery.

While certain tests were not utilized during the time of the patient's exam that would confirm the exact mechanism of his clinical outcome, it was concluded, based on the extent and severity of cerebrovascular stenosis, that the patient had experienced ocular ischemic syndrome caused by a phenomenon known as ophthalmic artery steal. The MRA suggested poor perfusion globally throughout his intracranial system, overwhelming the compensatory collateral flow via the circle of Willis to overcome the carotid occlusion. This likely prompted the additional enlistment of secondary collaterals such as the external carotid artery with the ophthalmic artery serving as the conduit for blood transport. The ischemia sustained by the left eye suggests ophthalmic artery steal occurred, in which blood was directed in the retrograde direction, away from the globe and toward the brain, leaving the eye vulnerable to the sequelae of hypoperfusion. Several interesting points regarding the cerebrovascular system and the intertwined role of the ocular blood supply can be learned from this case. Additionally, the mechanism by which an internal carotid artery occlusion results in ocular ischemia is explained in detail.

Discussion

Pathophysiology of Ophthalmic Artery Steal

It must be noted that while most frequently associated with obstruction of the internal carotid arteries, the primary source of the brain's blood supply, ophthalmic artery steal is also occasionally a sequelae of blockage at the level of the aortic arch and can also feasibly occur when there is vascular obstruction anywhere along the pathway from the left ventricle to the brain.[5] Indeed ophthalmic artery steal is a rare clinical entity since the human body can typically compensate for impeded blood flow to the brain through the development of alternative intracranial collateral vessels not derived from the ocular blood supply.[3,6,7,8] However, the system can fail in a patient with very severe vascular disease, when the intracranial mechanisms alone are insufficient in compensating for cerebrovascular hypoperfusion; in this infrequent scenario, ocular blood flow is enlisted and diverted toward the brain to the detriment of the eye and as such, its pres- ence is often an indicator of severely impaired cerebral blood flow.[3,8]

In order to understand the hemodynamic interplay between the eye and the brain, it is imperative to review the basics of the cerebral and orbital vasculature systems, namely how they function in healthy individuals and also how they are altered when vascular health becomes com-promised. Under normal conditions, the cerebrum receives the majority of its blood supply from the internal carotid artery and its branches, while the remaining 20%

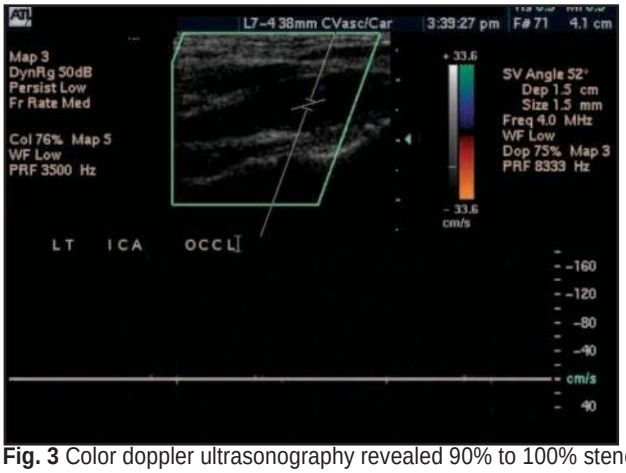

Fig. 3 Color doppler ultrasonography revealed 90% to 100% stenosis o the left internal carotid artery

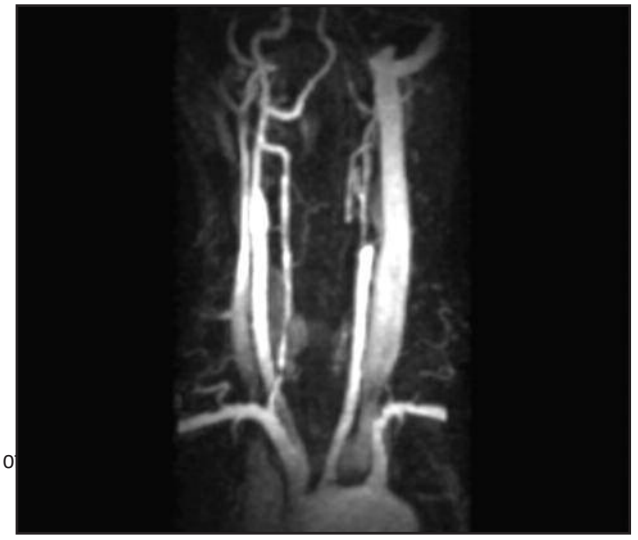

Fig. 4 Complete occlusion of the left internal carotid artery, complete occlusion of the left vertebral artery, and "flow gaps" in the right vertebral artery are demonstrated on magnetic resonance angiography.

is supplied by the vertebral artery system.[9] After yielding its first branch to the eye as the ophthalmic artery, the internal carotid artery subsequently divides into the anterior and middle cerebral arteries, which collectively supply the parietal lobe, the medial aspect of the frontal lobe, and the lateral portion of the temporal lobes.[9] The vertebral arteries, in contrast, carry the burden of the posterior cerebral vascular supply. The left and right vertebral arteries join to form the solitary basilar artery, which supplies the brainstem and cerebellum; the basilar artery then splits into two posterior cerebral arteries that then nourish the occipital lobes and the medial portions of the temporal lobes.[9] The internal carotids and the vertebral branches in turn join together via communicating arteries to form the circle of Willis,[9] thereby unifying the anterior and posterior networks as well as the right and left hemispheric systems.[3] This Willisian network of vessels typically compensates for any stenotic or occlusive event by redirecting blood flow from patent pathways to areas of diminished cerebral perfusion[3,6,7,9] (Fig. 5).

Ordinarily the orbital blood supply exists relatively independently of the cerebral system. The eye is largely supplied by the ophthalmic artery with minor contributions from the external carotid artery (ECA) via the infraorbital artery and the middle meningeal artery branches.[10,11] Distally, the ophthalmic artery typically divides into three branches, the first of which is the central retinal artery (CRA), which supplies the inner retina from the nerve fiber layer at the most anterior aspect to the inner portion of the inner nuclear layer at the most posterior aspect. The other two branches from the ophthalmic artery are the medial and lateral posterior ciliary arteries.[12] These give rise to long posterior ciliary arteries, which supply the anterior aspect of the globe, and between seven to ten short posterior ciliary arteries, which compose the bulk of the choroidal/choriocapillaris system.[12] While the central retinal artery feeds the inner portion of the retina, the choroidal system supplies the outer layers, which traditionally includes the outer portion of the inner nuclear layer through to the retinal pigment epithelium.[12]

These two systems, the orbital and cerebral, tend to run parallel and independently of one another unless poor cerebral perfusion necessitates cooperation. In the event of carotid stenosis or occlusive disease, the human body can elicit several compensatory mechanisms to overcome focal blockage, the success of which can determine a myriad of systemic manifestations ranging from clinically "normal" when effective collaterization takes hold to the unfortunate other end — debilitating cerebral infarct — presumably when adequate ancillary blood supply cannot be established.[4,13]

In most patients with severe ipsilateral carotid disease, the circle of Willis is the primary collateral system;[3,7,8,14,15,16] this network can be recruited almost instantaneously,[14] and when functional, can re-perfuse ischemic areas very effectively since blood supplied by the patent contralateral internal carotid can travel in any necessary direction either via the anterior communicating arteries or by the vertebral system via the posterior communicating arteries.[3,15,16] In this situation there is no intervention of the ophthalmic artery and hence no disruption of ocular blood flow. The ophthalmic artery ipsilateral to the occlusion continues to maintain blood flow in the normal and undisturbed anterograde direction

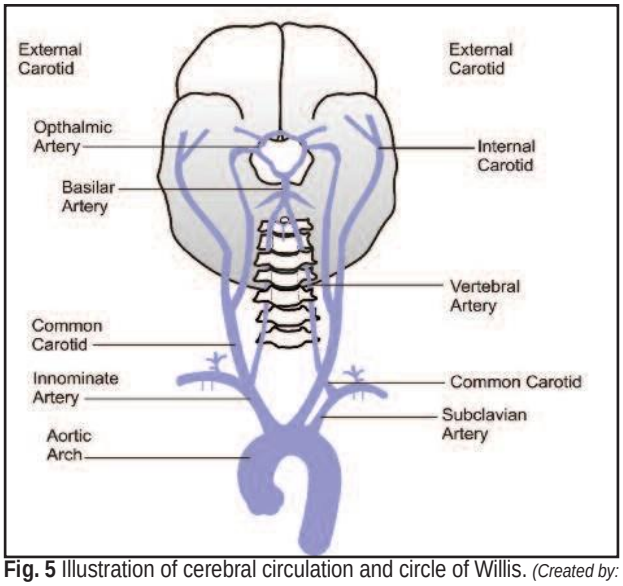

Fig. 5 Illustration of cerebral circulation and circle of Willis. *(Created by: Mr. Terrence M. Washington, Medical Illustrator)*

as it is perfused by the external carotid artery rather than the now occluded internal carotid artery.[7]

The demand for collateralization can exceed the capacity of the circle of Willis in cases of system-wide stenosis, bilateral internal carotid artery disease, or anatomical limitations such as congenitally hypoplastic or absent vessels.[2,3,7,14,15,17] In such scenarios, secondary collaterals such as the leptomeningeal anastomoses[3,4,14,17] or collaterals with the ipsilateral external carotid [2,3,6,7,14,17,18] become necessary, which then use the ophthalmic artery as a conduit to supply the occluded ipsilateral internal carotid artery.[7,8,19] Kluytmans et al reported that patients with a normal volume of anterograde flow through the ophthalmic artery essentially had patent collaterization via the circle of Willis in that secondary collaterals such as the ophthalmic artery were not recruited.[15] In their study, the presence of the ophthalmic artery collateral, usually a mechanism of second resort, was indicative of more severely impaired hemodynamic perfusion to the brain[15] (Fig. 6).

It is believed that much like primary collaterals, secondary collaterals develop in the prenatal period but the maximum functionality of such pathways takes time to develop.[3] Some studies regard the ophthalmic artery to be a less than ideal collateral pathway given the relative limits to its blood volume capacity in supplying the brain,[4,8,14] while van Laar et al indicate collateralization with the ECA via the ophthalmic artery is substantial enough to maintain sufficient cerebral perfusion to preserve patient life.[18] In either scenario, recruitment of

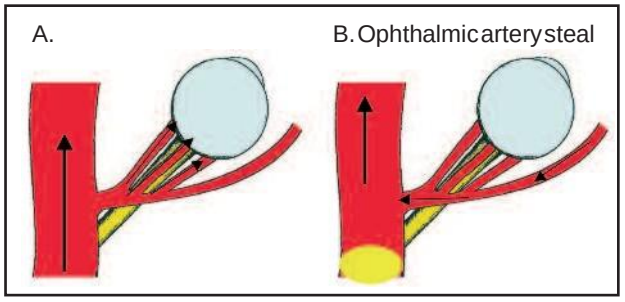

Fig. 6 (A) Diagram of normal circulation. **(B)** Reverse flow through the ophthalmic artery (ophthalmic artery "steal") from the external carotid.

the ophthalmic artery as a collateral is suggestive of more severe and/or extensive occlusive disease[8,15] since this indicates inadequate perfusion though the primary collateral system alone.

In secondary collateral development, blood derived from the branches of the ECA, the superficial temporal, facial, supraorbital, maxillary, and middle meningeal arteries,[3,10,11] travel through the ophthalmic artery, reversing the direction of flow away from the eye toward the brain.[2,3,4,7,19,20,21] (Fig. 6). In this process, the ophthalmic artery is the conduit that diverts blood from the patent extracranial circulation to the impaired intracranial system.[7] Given the small caliber of the ocular blood vessels prohibiting adequate visualization through traditional angiography, a non-invasive technology referred to as color Doppler imaging (CDI) is often utilized to image the retrobulbar vessels and their internal patterns of flow.[2,7,16,19,20,22] CDI yields color schemes depicting the pattern of blood flow superimposed upon a B-scan ultrasound of the vascular anatomy.[19] By using this technology, Yamamoto et al were able to shed light on the role of the retrobulbar vascular system and to explain the variable clinical presentation in the eye when internal carotid artery (ICA) occlusion occurs. They identified various groups of patients with an ICA occlusion based on four different patterns of orbital blood flow.[2] In situations where the occluded ICA received collateral support only from the circle of Willis, the ophthalmic artery (OA) and likewise as expected, downstream at the level of the central retinal artery (CRA) and the choroidal system, demonstrated undisturbed blood flow.[2] The second category involved collateralization via a branch of the ECA by way of the OA with continued but limited perfusion to the ophthalmic artery's branches, the central retinal artery and the short posterior ciliary arteries.[2] Under this model, given perfusion to the ICA as well as the ocular end arteries (the CRA and the choroidal circulation), it was assumed that some of the blood flow from the OA was not directed exclusively to the cerebral

Table I Ocular manifestations of ophthalmic artery steal

Spectrum of Clinical Findings		
Normal	**Acute**	**Chronic**
Asymptomatic, clinically "normal"	Moderate retinal opacification	Rubeosis irides
	Variable cherry red spot	Retinal arterial attenuation
	Vascular box-carring	Dilated, non-tortuous veins
	Retinal artery attenuation	Mid-peripheral intraretinal hemorrhages
	No retinal hemes/exudates	Optic disc collaterals
	Intraretinal gray lesions (choroid)	Optic nerve edema
	Late optic atrophy	Retinal arteriolar pulsation
	Delayed RPE hyperplasia	Neovascularization NVG
	Primary sx: profound vision loss	Primary sx: amaurosis fugax

tissues in the retrograde direction but also a portion managed to reach the eye in the normal anterograde fashion.[2] In the third group, secondary collaterals from the ECA were recruited but none of the retrograde flow within the OA was directed toward the eye, fully exemplifying the phenomenon of ophthalmic artery steal.[2,7] This form of collateralization shunts blood to the lower resistance vessels of the intracranial system with the outcome of terminated retrobulbar flow.[7] The final category Yamamoto et al identified was one in which no effective collateralization developed, leaving the eye and the cerebral tissues devoid of any measurable vascular flow, causing cerebral infarct and ocular ischemia.[2] Such differences in blood flow patterns presumably explains why some eyes become ischemic after an ICA occlusion (when retrobulbar flow is diminished or absent) and why other eyes appear to have undisturbed ocular blood flow after a carotid occlusive event.

It is believed that any reverse flow through the OA occurs in approximately 38.5% to 76% of patients with internal carotid artery occlusion[4,7,8] and with lower frequency in patients with significant stenosis.[19] Reverse flow through the OA is indicative of secondary collateralization via the ECA, usually as a result of an ICA occlusion in the setting of overall poor intracranial arterial status.[3,7,19] This agrees with Costa et al's finding that reverse flow through the OA is much more common in patients with severe bilateral disease as the intracranial collaterals (i.e., the circle of Willis) are generally the first and only line of collaterization if patent and sufficient.[3,6,16] One critical finding of Yamamoto et al's study was that ocular ischemia is caused by reverse OA flow, but retrograde ocular blood flow did not always result in ocular ischemia.[2] Only those with high velocity reverse OA flow seemed to be susceptible to the ravages of ocular ischemia as the rapid retrograde redirection of blood resulted in severely diminished or absent perfusion through the ocular end arteries[2] as measured by reduced peak systolic velocities in both the central retinal and short posterior ciliary arteries.[16,19,20,22] Specifically, it was found that further downstream, reduced perfusion at the level of the posterior ciliary artery moreso than at the central retinal artery was particularly devastating as far as patient susceptibility to ocular ischemic syndrome with vision loss.[19] This is presumably due to the numerous ocular structures dependent on perfusion by the posterior ciliary arteries, namely the optic nerve, choroid, RPE, and the outer retinal layers.[19] The variety of collateral pathways and flow patterns of the retrobulbar vasculature in ICA occlusion seems to explain why the clinical outcome can be so variable — ranging from a complete absence of clinical signs to devastating ocular ischemia with profound vision loss and deterioration.[19]

Clinical Manifestations of Ophthalmic Artery Steal

In internal carotid occlusion, the clinical manifestations within the eye can be quite variable.[7,16,19,20,23] Depending on the mode of collaterization (primary vs. secondary), the direction of blood flow through the OA (anterograde vs. retrograde), as well as the magnitude of the flow (high velocity vs. low velocity), the eye can be potentially spared of ischemic changes, as in the case of a patient with 100% effective intracranial collaterization through the circle of Willis.[2] However, it is possible that collaterization with the extracranial blood supply, which requires retrograde flow through the OA to supply the brain, leaves the eyes potentially vulnerable to ocular ischemia.[2,7,8,19,20,22] According to Fogelholm and Vuolio whose study depended on archaic x-ray technology only, there is no time required for the OA to develop into a collateral; rather, they believed the OA to be fully patent and functional at the moment of ICA occlusion.[4] A more current researcher, Liebeskind, suggested that while anatomically capable of collateral formation at birth, the OA it is not necessarily at peak capacity as a collateral circuit until cerebral hypoperfusion necessitates it.[3]

Table II Clinical features of acute ophthalmic artery hypoperfusion vs. central retinal artery occlusion	
OAH	**CRAO**
VA: HM to NLP	VA: CF to HM (better if (+) cilioretinal artery)
Cherry red spot: variable (+) to (-)	Cherry red spot: (+)
Retinal whitening: moderate to severe	Retinal whitening: mild to moderate
Pigment disturbance: (+)	Pigment disturbance: (-)
FA: delayed retinal and choroidal flows; late choroidal stain	FA: delayed retinal flow only
ERG: reduced/absent a- and b-waves	ERG: reduced b-wave
Decreased IOP (early?)	

Therefore, the exact onset of ophthalmic artery steal after an ICA occlusion is debatable though more likely to be of delayed onset.[3]

A spectrum of clinical presentations (Table I) in the eye can occur in the setting of internal carotid artery occlusion.[19,22] On the most benign end and under the most ideal conditions, this may result in a clinically normal appearing eye and a systemically asymptomatic patient.[19,22] This would occur under circumstances in which, despite the presence of reverse OA flow due to a diversion of blood from the ECA to the brain, a portion of the extracranial blood supply is still sent to the eye, providing it with enough blood to sustain function.[7]

Should ophthalmic artery hypoperfusion from the steal phenomenon occur, it can manifest differently in the eye depending on the rapidity with which blood flow is re-directed away from ocular structures.[2,20] If done rapidly, this may present itself similar to a direct and acute occlusion of the ophthalmic artery, which would basically appear as a concomitant central retinal artery occlusion superimposed on an acute choroidal infarct.[21] One would find moderate retinal opacification that extends into most layers of the retina, possibly as deep as the outer retinal layers and even the retinal pigment epithelium.[21,24] A less likely finding would be the faint hint of a cherry red spot depending on the contrast between the retinal opacification and the underlying level of choroidal perfusion – this is much less likely than in a CRAO where choroidal perfusion is intact and a cherry-red spot is prominent.[25,26] Box-carring of the vessels, retinal arterial attenuation, and a likely absence of hemorrhages or exudates are also possible findings of acute ophthalmic artery hypoperfusion,[25] which can occur either via direct occlusion as in the case of a retinal embolus or indirectly by the ophthalmic artery steal phenomenon.[21,24] In acute ophthalmic artery hypoperfusion, occasionally gray intraretinal lesions can also appear as focal manifestations of retinal ischemia deep to the retinal vessels,[21,24] postulated to be in the vicinity of the outer nuclear and outer plexiform layers.[24] In late stages, the posterior segment can be characterized by optic atrophy and prominent retinal pigment epithelium hyperplasia within weeks to months of the event given the extensive retinal involvement.[25] It is believed that the poor choroidal perfusion results in ischemia and permanent damage to the RPE that results in changes including hyperplasia, dropout, and metaplasia, localized mostly in the macula.[25] This is a distinction from the fundus appearance of a pure central retinal artery occlusion, which is typically not marked by late pigmentary changes, as the ischemia is limited to the inner retinal layers.[27]

A key diagnostic tool to distinguish between acute ophthalmic artery hypoperfusion and a CRAO would be fluorescein angiography: in early phases, ophthalmic artery hypoperfusion produces delayed central retinal artery filling as well as delayed choroidal flush.[21,24,25,26] In a "classic" central retinal artery occlusion, only the retinal system is delayed and choroidal filling occurs within the normal time frame. Later frames of the angiogram may show several areas of leakage at the level of the retinal pigment epithelium in ophthalmic artery hypoperfusion due to compromised outer retinal blood flow and subsequent breakdown of the blood-retina barrier;[21,25,28] such late angiographic findings are not seen in CRAO. Additionally, while unlikely to be used in diagnosis, electroretinography (ERG) would show extinguished a- and b-wave responses due to the loss of functionality of both the inner and outer retina due to simultaneous inner and outer retinal ischemia in ophthalmic artery hypo-perfusion.[25] In contrast, in central retinal artery occlusion, typically, only the b-wave would be extinguished while the a-wave, which represents the outer retina, is preserved given the uncompromised choroidal blood supply.[25] Furthermore, the vision loss in CRAO tends to dwell on the order of count fingers to hand motion whereas vision loss with ophthalmic artery hypoperfusion is usually more profound since the ischemia is more extensive.[25] Acute pain-less vision loss is the most common symptom of patients presenting with acute ophthalmic artery hypoperfusion.[25,29]

More concerning and more widely known on the spectrum of ocular manifestations of the ophthalmic artery steal phenomenon is ocular ischemic syndrome, which occurs when the ocular bloodflow is reduced on a more delayed and chronic scale.[23] This seems more in line with the studies of Liebeskind et al as ophthalmic artery steal occurs secondary to extracranial collateralization,

Table III Suggested laboratory and imaging studies in suspected ophthalmic artery steal		
Ancillary Ophthalmic Tests	**Labs and Vitals**	**Imaging Studies**
Fluorescein angiography (FA)	Blood pressure	Carotid duplex ultrasound
Electroretinography (ERG)	Lipid panel	Brain magnetic resonance imaging (MRI)
B-scan ultrasonography	Erythrocyte sedimentation rate (ESR)	Magnetic resonance angiography (MRA)/ Computed tomography angiography (CTA)
	C-reactive protein (CRP)	Orbital Color Doppler Imaging (CDI)
	Fasting plasma glucose (FPG)	
	Hemoglobin A1c (HbA1C)	
	Prothrombin time (PT)	
	Partial thromboplastin time (PTT)	
	International normalized ratio (INR)	

which is typically recruited later than at the time of ICA occlusion when the intracranial mechanisms have been exhausted.[3] In some studies, it has been found that 4% to 18% of patients with ocular ischemic syndrome have underlying carotid occlusion.[19,23] In its earliest stages, ocular ischemic syndrome is often referred to as venous stasis retinopathy, when the funduscopic changes are consistent with chronic low perfusion pressure to the eye.[30] It is believed to occur with higher frequency in patients with compromised cerebral perfusion, given the necessity to recruit the ophthalmic artery as a collateral rather than the usually more effective intracranial Willisian system.[16,30] Clinically, this can manifest with rubeosis irides, arterial attenuation, dilated, non-tortuous veins, mid-peripheral intraretinal hemorrhages, optic disc collaterals, optic nerve edema,[21] and retinal arteriolar pulsation.[19,25,30] Classically, the patient experiences amaurosis fugax,[21] presumably due to fluctuating perfusion to the eye that can result in permanent loss of vision.[19] Other patients may be completely asymptomatic.[19]

Left untreated, anterior neovascularization caused by belatedly detected ocular ischemic syndrome can result in neovascular glaucoma.[30] Initially, reduced blood flow to the ciliary body can cause a reduction in intraocular pressure secondary to reduced production of aqueous humor from an ischemic ciliary body.[5,23] However, as the new vessel growth progresses, obstruction of the trabecular meshwork can cause a steady imbalance between the outflow and the production of aqueous, resulting in elevated IOP.[23] Symptoms in such a patient will not only include preceding episodes of amaurosis fugax, but also blur, photophobia, and pain from the corneal edema that can result from an acute rise in IOP.[5,30] Cataract and uveitis can also occur in such instances.[16,23]

Ophthalmic Differential Diagnoses

There is a wide range of potential conditions that must be considered when faced with what appears to be ophthalmic artery hypoperfusion. In the acute presentation, higher on the list of differentials would be central retinal artery occlusion from embolic disease,[32] vasculitis including giant cell arteritis, hypercoagulability disorders,[33,34] compression, laceration, and vasospasm. Concurrent central retinal artery occlusion and choroidal infarct, commotio retinae with a history of recent trauma, or an inheritied metabolic lysosomal storage disease such as Tay-Sachs are also possibilities for the clinical picture involving diffuse retinal opacification with a variable cherry-red spot.[31,32] In contrast, advanced diabetic retinopathy and central retinal vein occlusion would be more consistent with a chronic presentation of ocular ischemia as demonstrated by the presence of neovascularization. While ocular ischemic syndrome usually is a result of an ICA occlusion, this entity can feasibly occur as a result of any mechanism that drives retrobulbar blood flow rapidly away from the eye such as in the case of high-flow carotid-cavernous fistula.[24]

Systemic Considerations

Systemically, the patient may be completely asymptomatic after an ICA occlusion provided that collateral systems are recruited quickly and effectively to overcome cerebral hypoperfusion. However, if the collateralization is poor on a systemic level, the patient may experience transient ischemic attacks, with symptoms such as unilateral weakness, speech disturbances, confusion, and temporary monocular vision loss.[35,36,37,38,39]

Typically, the patient with ophthalmic artery hypoperfusion secondary to the steal phenomenon has stenosis or occlusion of the ipsilateral internal carotid artery along with extensive stenosis throughout the cerebrovascular system.[3,7,16] This is a rare clinical entity, and the signs and symptoms can be quite variable, because intracranial collateralization is generally a very effective compensatory mechanism.[3] Generally patients with this clinical picture of ophthalmic artery hypoperfusion, however, have ipsilateral carotid disease with several comorbidities, including hyper- tension, diabetes mellitus, and atherosclerosis.[19]

Ophthalmic and Systemic Diagnostic Testing

As far as ocular testing is concerned, clinical history-taking is critical first and foremost. Inquiring about patient ocular and systemic symptoms suggestive of transient ischemia, laterality of the symptoms, rapidity of onset, and quality of vascular health are examples of questions to be posed. Visual acuity measurements, pupil testing, IOP readings, gonioscopy, funduscopic exam, and fluorescein angiography are critical elements of ocular testing. ERG could be of interest as well, though not necessarily practical, accessible, or essential to diagnosis.[25,40]

Systemic diagnostic investigation is imperative as is cooperation with other disciplines, potentially including the patient's primary care provider, as well as specialists in neurology, cardiology, and vascular surgery. Carotid duplex ultrasound is the initial means for determining if carotid stenosis is present.[3] Further investigation of the cerebral vasculature typically would involve magnetic resonance or computed tomography angiography.[3] Color Doppler imaging (CDI) of the orbit would be helpful to determine the type of flow through the ophthalmic artery and its branches, which are all considerably smaller caliber vessels not readily imaged by traditional ultrasound techniques alone.[19,20,22] How carotid occlusive disease will impact the patient's ocular circulation is best understood with the evaluation of blood flow in the ocular end arteries with regard to direction of flow, systolic velocity, as well as the amount of vessel resistance (as measured by the pulsatility index).[19,20,22]

In addition to these imaging techniques, certain laboratory tests are critical in ruling out other systemic etiologies for the patient's ocular condition, and these include Westergren erythrocyte sedimentation rate (ESR), C-reactive protein, lipid panel, blood glucose, and complete blood count (CBC). Blood pressure should also be measured.[21,24,31]

Management

As far as the ocular management of patients manifesting clinical signs and symptoms of ophthalmic artery steal phenomenon, there is no accepted or proven means for attempting to restore vision loss from ischemia.[23,25,41,42] The treatment strategies are largely aimed at preventing progression to more serious ocular sequelae. Frequent follow-up with dilated fundus exams and gonioscopy to monitor for neovascularization is critical.[31] In the event that new vessel growth occurs in the angle or on the iris, pan-retinal photocoagulation should be performed to reduce vascular demand and hence the release of vaso-proliferative factors.[31] However, if the neovascular- ization progresses to glaucoma, topical intraocular pressure lowering medications should be implemented and surgical referral is indicated if maximally tolerated medical therapy cannot achieve adequate pressure control.

One study has shown that systemic intervention for occlusive disease may arrest the progression of neovascularization as the internal carotid artery and subsequently the ophthalmic artery regain perfusion.[2,21] It has also been found that following carotid endarterectomy, return of blood flow may cause some reversal of vision loss in some patients.[21] Such decisions regarding carotid endarterectomy should be left in the hands of the patient and their vascular specialist as the decision is usually based principally on the risks of cerebral infarction.[19,41] While some researchers suggest carotid endarterectomy may be indicated in patients with ocular manifestations of ophthalmic artery steal as a means to improve visual status, other authors believe that insufficient evidence exists to conclude that this procedure is indicated for patients with these clinical findings.[19,41] As an alternative, Yamamoto et al found that patients suffering from ophthalmic artery steal may benefit from STA-MCA bypass (anastomosis of the superficial temporal and middle cerebral arteries) by creating an alternate collateral with the ECA and in the process, reducing the impact on the ophthalmic circulation.[2,23] The idea is to restore cerebral perfusion surgically, but there is no widespread consensus on the utility of this method with regard to the preservation of the ocular circulation.[2]

It should be noted that while possible retention of vision can occur with rapid anterograde reperfusion of the ophthalmic artery,[24,43] usually the visual prognosis is relatively grim once ischemia to the ocular structures is so vast.[42] However, patient management should not be delayed given the underlying risks to patient mortality. With timely examination and diagnosis, the patient may be spared further systemic and visual loss despite already sustained ocular damage.

Conclusion

Ophthalmic artery steal is an infrequently discussed phenomenon, which may have potentially devastating effects on the eye. Optometrists should have an understanding of the role that the ophthalmic artery can play in patients with severe carotid occlusive disease and be aware that this can present itself clinically in a myriad of manifestations. Timely recognition and intervention may help to prevent or mitigate sight- and life-threatening ischemic vascular consequences. ❑

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