Ophthalmic masquerades of the atherosclerotic carotids

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Patients with carotid atherosclerosis can present with ophthalmic symptoms. These symptoms and signs can be due to retinal emboli, hypoperfusion of the retina and choroid, opening up of collateral channels, or chronic hypoperfusion of the globe (ocular ischemic syndrome). These pathological mechanisms can produce many interesting signs and a careful history can bring out important past symptoms pointing toward the carotid as the source of the patient's presenting symptom. Such patients are at high risk for an ischemic stroke, especially in the subsequent few days following their first acute symptom. It is important for clinicians to be familiar with these ophthalmic symptoms and signs caused by carotid atherosclerosis for making an early diagnosis and to take appropriate measures to prevent a stroke. This review elaborates the clinical features, importance, and implications of various ophthalmic symptoms and signs resulting from atherosclerotic carotid artery disease.

Key words: Carotid artery disease, ocular ischemic syndrome, retinal emboli

Prevalence of asymptomatic extracranial carotid artery disease (>50% stenosis) ranges from 1.5 to 9% in population-based studies.[14-17] with a higher incidence in the elderly.[13,15] Symptoms in the ipsilateral eye can often herald the presence of asymptomatic atherosclerotic carotid disease. Such patients are at a high risk for stroke. The annual stroke risk ranges between 2.0% and 2.8%,[6-10] and can reach 8.4% in the presence of higher grades of ipsilateral carotid stenosis.[8,11,12] Bruno[13] noted a 10-fold increase in the annual rate of stroke in men with retinal emboli independent of blood pressure and other vascular risk factors. It is also important to note that the risk of stroke following a transient ischemic attack is maximum in the subsequent 48 h.[14-17] The EXPRESS[18] study and the SOS–TIA study[19] have shown that early intervention in such patients is needed to prevent stroke. It is therefore important for the first medical contact to be aware of the importance and implications of the ophthalmic symptoms caused by atherosclerotic carotid artery disease to prevent an impending catastrophic stroke.

Atherosclerosis affects mainly the site of bifurcation of the common carotid artery. The posterior walls of the arteries tend to be more involved. The pathogenesis of atherosclerosis is a decade-long process which results in the formation of the atherosclerotic plaque in the tunica intima.

Falk[20] divided the atherosclerotic plaques into two types, a hard collagen-rich plaque, with barely detectable lipid, and a soft plaque, with a prominent pool of lipid. Soft plaque is more vulnerable because its thin fibrous cap can rupture.

When an atherosclerotic plaque ruptures, the contents of the plaque (collagen, lipids, and smooth muscle cells) protrude into the vascular lumen and come in contact with platelets and blood cells. This leads to activation of platelets and the coagulation cascade, resulting in thrombus formation on the atherosclerotic plaque. Such thrombi may cause distal ischemia by various mechanisms as follows: (1) The newly formed thrombus can cause a sudden marked reduction in the vessel diameter, resulting in decreased distal perfusion. (2) Apart of the thrombus can break off and travel distally (thromboembolism) into the ophthalmic artery or enter the cerebral circulation. (3) The thrombus can grow in size and propagate distally and cause occlusion of a vessel. (4) The thrombus can become organized and get incorporated into the diseased vessel, making its lumen narrow and causing chronic distal ischemia.

When the internal carotid artery (ICA) is obstructed, the blood flow to the brain is compensated by the following: (1) collateral circulation from the circle of Willis which brings blood form the contralateral ICA through the anterior communicating artery (ACom) into the ipsilateral middle carotid artery (MCA) and (2) collateral channels open up between the terminal external carotid artery (ECA) branches and the terminal branches of the ophthalmic artery. Through these channels, there is retrograde flow via the ophthalmic artery to ICA. Thus, ophthalmologic symptoms and signs caused by carotid atherosclerotic disease can occur due to either of the following: (1) retinal emboli arising from the atherosclerotic plaque; (2) hypoperfusion of the retinal and choroid due to the ICA stenosis; (3) opening up of collateral channels; and (4) ocular ischemic syndrome (OIS; venous stasis retinopathy).

In this article, we describe the various ophthalmic symptoms and important physical signs caused by carotid atherosclerosis.

Symptoms and Signs Due to Retinal Emboli

For unclear reasons, an embolus originating in the carotid artery is more likely to enter the ophthalmic artery, which is
The CRA is an end artery with no anastomotic connections. It divides into four major branches supplying non-overlapping sectors of the retina. When an embolus enters and blocks it, the area of retina that they supply infarcts. CRABO are more often attributable to severe ICA disease than CRAOs. The visual loss will be focal and corresponds to the portion of the retina that is ischemic. CRABO may affect only the upper, lower, nasal, or temporal portion and may present as small central or paracentral scotomas. On ophthalmoscopy, the portion of the retina will have a ground glass like appearance due to ischemic edema. Fluorescein angiography can pinpoint the occlusion.

**Anterior Ischemic Optic Neuropathy**

Unlike the above conditions, AION is caused by hypoperfusion of the optic nerve head due to ischemia to the anterior part of the optic nerve. Rarely, an ICA occlusion with thrombus extending to the carotid siphon can block the ophthalmic artery causing AION. Painless monocular visual loss is the usual symptom. If the ischemia involves the anterior part of the optic nerve, the examination will show edema of the optic disc and small splinter hemorrhages at the disc margins. If the ischemia affects the posterior part of the optic nerve, then the fundus examination can be normal.

**Symptoms and Signs Due to Hypoperfusion of the Retina and Choroid**

ICA narrowing can cause hypoperfusion of the retina and choroid. This can result in some interesting symptoms. Patients can experience an iris diaphragm type of TMVL where the constriction of the visual fields start from the periphery and move towards the center. This TMVL of hypoperfusion can occur in many specific clinical settings.

1. **Bright light amaurosis**
   - The TMVL is brought on after exposure to bright light (retinal claudication). The symptom can persist for seconds to hours after the light exposure.
   - The mechanism is severe retinal ischemia causing a delay in a regeneration of visual pigments in the pigment epithelial layer of the retina. With bilateral carotid disease, this can present as asymmetrical visual loss occurring with bright light exposure.

2. **Position-induced TMVL**
   - Episodes of TMVL may be induced by changes in posture or neck position that temporarily reduce blood flow through the already stenosed artery.

3. **Postprandial transient visual loss**
   - The TMVL following a heavy meal can be secondary to shunting of blood to the mesenteric arteries.

4. **Exercise-induced TMVL**
   - The probable mechanism in this is thought to be diversion of blood to a dilated external carotid vascular bed.

5. **TMVL following facial heating**
   - In such suspected cases of ICA narrowing, a carotid or ocular bruit may be heard on auscultation. A carotid bruit due to carotid stenosis may be heard in the following locations: (1) common carotid region – base of neck; (2) carotid bifurcation region – level of the superior thyroid cartilage; and (3) ICA region – angle of jaw. It should be recalled that cardiac murmurs
can radiate to the neck vessels. So, a careful cardiac auscultation should be done, and if any cardiac murmurs are heard, they should be followed up into the neck. If there is an associated carotid bruit, then a change in pitch and/or intensity can be noted.

A carotid bruit is usually heard at the origin of the ICA when the stenosis is ≥70% of the lumen diameter and/or the residual lumen diameter is ≤1.5 mm.[44]

It should be noted that a carotid bruit may be heard often in asymptomatic patients, but if heard in the setting of a suspected carotid stenosis, it warrants further investigation.

Occasionally, a carotid bruit may be absent in a patient with a “tight” stenosis. This may be due to a “slow-flow” state.[45]

The orbit can be auscultated with the bell of the stethoscope to look for ocular bruits. The patient should be either sitting or lying. Patients should be instructed to open the opposite eye to reduce blinking noise. The opposite eye should also be fixated to reduce the movement of the globe during auscultation. Both the sides should be auscultated. In carotid stenosis, an ocular bruit may be heard on the contralateral side. The increased flow through the patent contralateral ICA may be the cause.[46,47]

It should be borne in mind that just palpation of the carotid arteries may not be adequate. On palpation, only the common carotid artery pulse can be felt and an ICA occlusion which is posteriorly placed after the bifurcation cannot be ruled out. The left carotid artery is usually posterior and deeper than the right, so there may be a mild asymmetry, normally the right being more prominent than the left.

**Symptoms and Signs Due to Opening Up of Collateral Channels**

The opening up of ECA–ICA collateral channels via the ophthalmic artery can cause many signs and symptoms which can be picked up by a careful vascular examination [Fig. 1].

1. **Delay/asymmetry of ECA pulses:** If the common carotid artery is blocked, then the branches of the ipsilateral ECA, the facial artery (felt along the angle of the lower jaw), the preauricular artery (felt anterior to the ear), and the superficial temporal artery (felt in the temporal region) will be diminished/delayed. The face on that side of the block will feel cool to touch. Both the sides should be felt simultaneously to detect this asymmetry.

2. **ABC (angular, brow, cheek) pulses:** When the ICA is occluded before its ophthalmic artery branch, there will be increased flow in the ECA branches to open up collateral vessels around the orbit. This augmented flow can be felt by palpation at the inner angle of the eye, the brow, and cheek.[47]

3. **Frontal artery sign:** Normally, there is antegrade flow of blood from the ICA to the ophthalmic artery and then to its supratrochlear and supraorbital (frontal) branches. In severe carotid stenosis, blood forms superficial temporal artery (ECA) flows retrograde through supratrochlear and supraorbital branches into the ophthalmic artery and enters the ICA system.[48,49] Normally, if the supratrochlear and supraorbital (frontal) pulses are seen on the forehead, they can be blocked by applying pressure at the brow (as they are being filled antegrade from the ophthalmic artery). In cases of severe carotid stenosis where these arteries have retrograde flow into the ophthalmic artery, applying pressure on the brow will not diminish them, but pressure on the superficial temporal artery on the forehead will obliterate them. This is called the frontal artery sign.[50]

4. **Dilated episcleral arteries** may be seen with ipsilateral carotid artery occlusion.[51] This is caused by ECA orbital collateral channels which cause retrograde filling of the ophthalmic arteries.

5. **Olivarius’ external carotid sign:** Increased prominence and pulsation of the branches of the superficial temporal artery, especially the frontal, in occlusion of the ipsilateral ICA.[52] Rarely, a subjective bruit over the ipsilateral ear may be the complaint. Occlusion of ipsilateral ICA can cause compensatory hyperperfusion of the ECA. This can cause a subjective bruit over the ipsilateral ear (due to increased flow across the temporal artery). Digital compression on the temporal artery can eliminate this bruit.[53]

**Symptoms and Signs Due to Chronic OIS (Venous Stasis Retinopathy, Hypotensive Retinopathy)**

Initially, this condition was called “venous stasis retinopathy.” Since mild central retinal vein obstruction was also designated as “venous stasis retinopathy,” the term “ocular ischemic syndrome” is now used for changes in the eyes caused by chronic ipsilateral carotid stenosis.[54,55]

It is postulated that the decreased vascular perfusion caused by carotid stenosis/occlusion in tissue hypoxia and increased ocular ischemia, leading to neovascularization.[56,57] Reported frequencies range between 5% and 21% in series of patients with carotid artery stenosis or occlusion.[58,59] The patient with OIS can present with pain or visual loss. In 90% of cases, visual loss occurs gradually over a period of weeks.
to months; in rare instances, it may be abrupt. Sudden visual loss may occur when the intraocular pressure (IOP) suddenly exceeds the perfusion pressure within the CRA, causing a CRAO. The possible mechanisms for pain in OIS are ischemia to the globe, increased intraocular pressure secondary to neovascular glaucoma, or dural ischemia. It is usually a dull ache over the brow and eyes. This pain usually is relieved on assuming a supine position.

An ophthalmic examination may show rubeosis iridis (iris neovascularization) in two-thirds of patients. It is interesting to note that in chronic OIS, a raised IOP is rare even if the anterior chamber angle is closed because of ciliary body hypoperfusion and decreased aqueous production. Other findings in this condition could be mature cataracts, especially if the ICA occlusion is longstanding. Retinal examination can show pale, narrow, and thread-like retinal arteries, and the retinal veins can appear dilated with occasional beading.

Retinal hemorrhages (dot and blot) and microaneurysms can be seen mainly in the midperiphery, and vitreous or subhyaloid hemorrhage can be seen following rupture of the new vessels. Varying degrees of optic nerve and retinal revascularization and retinal cherry-red spot can be seen.

It should be noted that most of the changes caused by OIS can be seen with diabetes and hypertension. Marked asymmetry of findings (more marked in the side of the occluded carotid) and disproportion in the extent of anterior and posterior segment pathology can be a clue that the changes are more likely due to carotid narrowing.

**Signs of Stroke**

It is very important not to miss out signs and symptoms of a stroke in patients presenting with visual symptoms and known or suspected carotid stenosis. Very often being preoccupied with his/her visual symptoms, the patient may not have noted subtle symptoms of the stroke.

In a busy clinical setting, the FAST approach is an easy way to recognize and remember the most common signs of a stroke. Face – Check the patient’s face for any facial droop or drooping of the eyes. Arms – Ask the patient to lift both his arms and to keep it horizontally for 10 seconds, with the palms facing the ceiling and eyes closed. The arm on the affected side will start to drift down in 10 seconds if there is subtle weakness. Speech – Look for any slurring of speech and whether the patient is able to understand your speech. Time – If any of these signs are present, do not waste time. The patient may be having a stroke and should be rushed to a hospital.

It is important to pick up these early signs of stroke since the patient may be a candidate for thrombolytic therapy if still within the window period.

**Conclusion**

Often ophthalmologic symptoms and findings may be the only manifestation in patients with carotid atherosclerotic disease. It is important for ophthalmologists to be familiar with these symptoms and signs. So, high-risk patients for an ischemic stroke can be picked up and referred for early intervention.

**References**