

# Light-induced Hemiplegia, an Atypical Case of Retinal Steal Syndrome

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**Abstract:** Atherosclerosis affecting the carotid arteries accounts for up to 20% of ischemic strokes. The clinical effects of atherosclerotic occlusive disease vary according to multiple factors, one of which is collateral circulation. In response to a chronic decrease in cerebral perfusion from atherosclerotic occlusion, alternative flow pathways, or collaterals, develop in a variety of patterns. In the setting of carotid occlusion, flow from the Circle of Willis, if complete, often comprises the main cerebral collateral network. In addition, contributions from leptomeningeal and extracranial to intracranial collaterals may develop. In the latter, reversal of flow in the ophthalmic artery can reconstitute to restore supraclinoid internal carotid artery flow. We report a case of the unusual sequela of a collateral flow pattern via the ophthalmic artery that induces a unique retinal steal phenomenon.

**Key Words:** stroke, ICA occlusion, retinal steal

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Atherosclerosis affecting the carotid arteries accounts for up to 20% of ischemic strokes.<sup>1</sup> The clinical effects of atherosclerotic occlusive disease vary according to multiple factors, one of which is collateral circulation. In response to a chronic decrease in cerebral perfusion from atherosclerotic occlusion, alternative flow pathways, or collaterals, develop in a variety of patterns. In the setting of carotid occlusion, flow from the Circle of Willis, if complete, often comprises the main cerebral collateral network. In addition, contributions from leptomeningeal and extracranial to intracranial collaterals may develop. In the latter, reversal of flow in the ophthalmic artery can reconstitute to restore supraclinoid internal carotid artery (ICA) flow. We report a case of the unusual sequela of a collateral flow pattern via the ophthalmic artery that induces a unique retinal steal phenomenon.

## CASE REPORT

A 62-year-old white male with a past medical history of obesity, hypertension, hyperlipidemia, obstructive sleep apnea, chronic kidney disease, pulmonary embolism, and ischemic cerebrovascular accident presented with transient, recurrent episodes of left arm and leg weakness, along with blurry vision in his left eye. These symptoms lasted 10 to 15 minutes with return to baseline thereafter. The patient specifically noted that these episodes were precipitated by light exposure, particularly when driving in bright sunlight.

His work-up included a magnetic resonance imaging (MRI) of the brain, which showed stable encephalomalacia with surrounding

gliosis in the right frontal lobe from a remote infarction of the anterior border zone region (Fig. 1). His magnetic resonance angiogram head and neck with and without contrast showed no appreciable forward flow in the right ICA (Figs. 2, 3). Catheter angiography showed complete occlusion of the right ICA, multiple small (<0.5 mm each) ethmoidal artery collaterals feeding the ophthalmic artery, which, in turn, partially supplied the distal right ICA and part of the right M1 segment of the middle cerebral artery (MCA). However, this collateral pathway was insufficient, with a notable lack of opacification of the right A1 segment of the anterior cerebral artery and MCA branches distal to M1 (Fig. 4). Minimal MCA collateral flow from the right posterior communicating artery and from posterior cerebral artery (PCA) pial collaterals was also identified, but the lack of a discrete, complete capillary phase indicated overall poor flow in the MCA territory. Coupled with the chronic border zone ischemic change, the angiographic findings were suggestive of an underlying diminished cerebrovascular reserve (CVR) due to the chronic ICA occlusive disease. Of note, transcranial Doppler also confirmed retrograde flow in the right ophthalmic artery (Fig. 5).

The MRI included arterial spin labeled (ASL) perfusion imaging. This demonstrated asymmetric arterial transit artifact in the right MCA vasculature and, conversely, little signal in the brain parenchyma itself (Fig. 6). This indicates diminished and delayed flow in the right ICA territory with transit of labeled blood markedly delayed relative to the contralateral hemisphere. In addition, delayed transit was noted in his PCA collateral vasculature. The combination of insufficient native flow in his MCA and delayed flow within recruited pial collateral vessels, coupled with a known CVR impairment, underscores the importance of the ophthalmic collateral flow in maintaining his baseline, asymptomatic status.

The patient was initially treated with intensive medical therapy, which included aspirin 325 mg daily and atorvastatin 80 mg daily. However, as his symptoms persisted, and given the relatively poor collateral flow to the MCA territory, the patient underwent a right-sided craniotomy for indirect external carotid artery to ICA bypass, in the form of an encephaloduroarteriomyosynangiosis. Postoperatively, the patient's course was complicated by incisional infection, and transient dizziness in the setting of hypotension. He also experienced left upper extremity and lower extremity limb shaking with preservation of consciousness observed at an outside hospital 1 month after the procedure. An electroencephalogram was performed at the time, which did not show any epileptiform activity, but he was started on an antiepileptic (levetiracetam).

Several months later, the patient developed similar episodes that began with tingling in his left leg that travels up to his hip and left hand followed by weakness on the left side, then shaking of the left arm and leg that was sometimes subtle, sometime violent. He denied loss of consciousness, tongue biting, bowel incontinence or bladder incontinence, and vision change during these spells. The episodes last for a few minutes and occur 2 to 3 times per week and are always associated with a decrease in his hemoglobin to <9 gm/dL due to chemotherapy (required for newly diagnosed malignancy). Repeat MRI did not show any evidence of new cerebrovascular accident. His episodes of left-sided weakness provoked by bright light resolved.

## DISCUSSION

Although it is common for patients with internal carotid occlusion to develop collateral pathways through the

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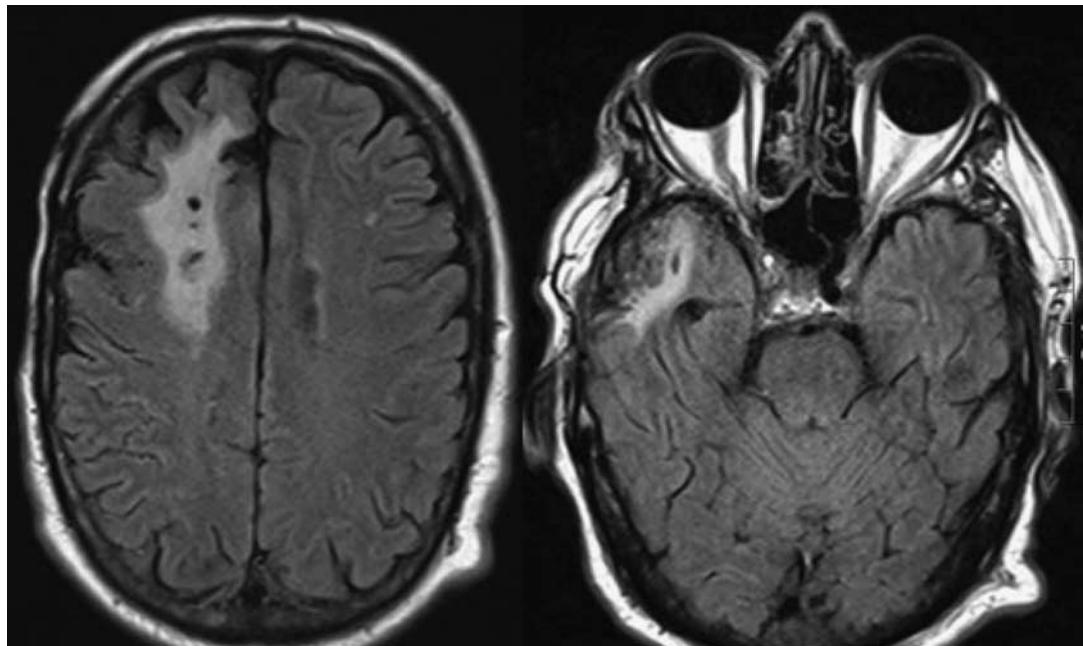
The authors declare no conflict of interest.

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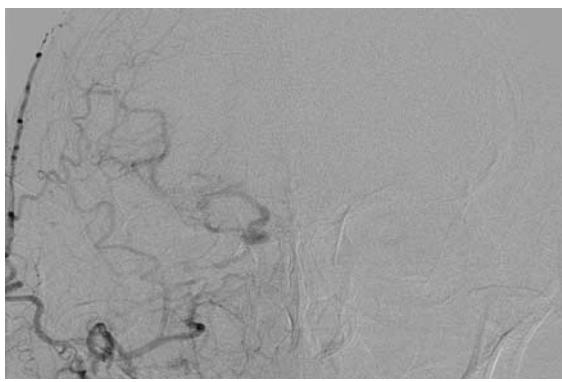


**FIGURE 1.** Magnetic resonance imaging of brain, FLAIR sequence showing classic “border zone” chronic ischemic change (long-standing and present on old studies). FLAIR indicates fluid-attenuated inversion recovery.

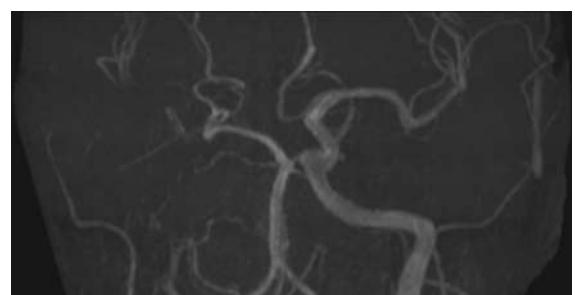
ophthalmic artery, our case is unique in that the patient’s presenting symptoms were provoked by a retinal “steal phenomenon,” wherein the increased retinal demand in bright light resulted in reduced collateral blood flow to the brain. In patients with carotid occlusion and inadequate intracranial collaterals (via the circle of Willis), external carotid artery vessels can be recruited to maintain flow to the ICA territory. The ophthalmic artery is frequently recruited due to extensive connections with the facial, internal maxillary, and superficial temporal arteries. In such cases, the blood flow in the ophthalmic artery can reverse to supply the ipsilateral brain.<sup>2</sup> As a result, a “steal phenomenon” can occur, wherein the increased metabolic demand of the brain causes decreased blood flow to the eye, resulting in acute or chronic ocular ischemia.<sup>2-5</sup> In our case, however, the “steal” seemed to be reversed, with

increased retinal demand inducing return of ophthalmic artery anterograde flow and producing brain ischemia, evidenced by symptomatic hemiparesis provoked by bright light.

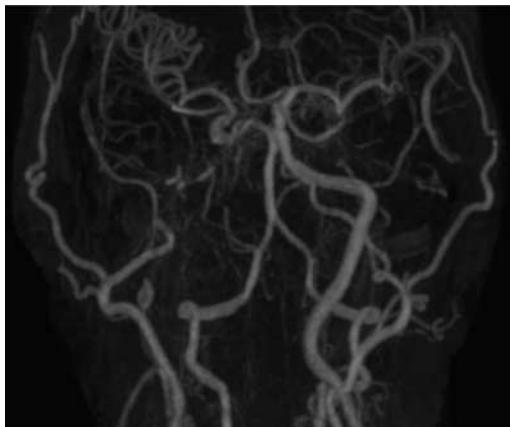
In vivo studies have shown that there is a sustained retinal hyperemia when the eye is exposed to photic stimulation. It is hypothesized that light dilates the primary arterioles on the retinal surface leading to an increase in blood flow in arterioles, capillaries, and venules of the retina.<sup>6</sup> We theorize that our patient was experiencing ischemic brain symptomatology as a direct result of increased retinal blood flow requirements in response to bright light. This in turn, resulted in decreased retrograde ophthalmic artery flow to the right MCA and right PCA territories. As this comprised the bulk of the patient’s collateral flow at baseline, the retinal steal produced significant symptomatology including left-sided weakness and blurred vision. The left-sided weakness can be explained from the decreased flow to the motor centers supplied by the right MCA and the visual symptoms attributed to either to decreased flow



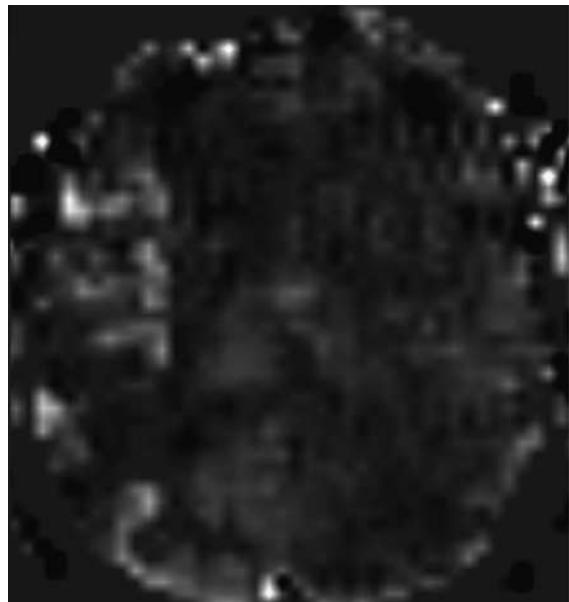
**FIGURE 2.** Catheter angiography showing collaterals from the facial and maxillary arteries were feeding the ophthalmic artery in retrograde manner, which in turn is feeding the internal carotid artery and middle cerebral artery.



**FIGURE 3.** Noncontrast time of flight brain magnetic resonance angiogram showing no appreciable flow-related enhancement in the right ICA (compare with normal signal in the left ICA). This indicates that there is no real forward flow in the ICA that can be detected without contrast. ICA indicates internal carotid artery.



**FIGURE 4.** Postcontrast neck magnetic resonance angiogram (looking at the intracranial portion of the study) showing enhancement of a prominent right ophthalmic artery and resultant collateral-induced filling of the right internal carotid artery.

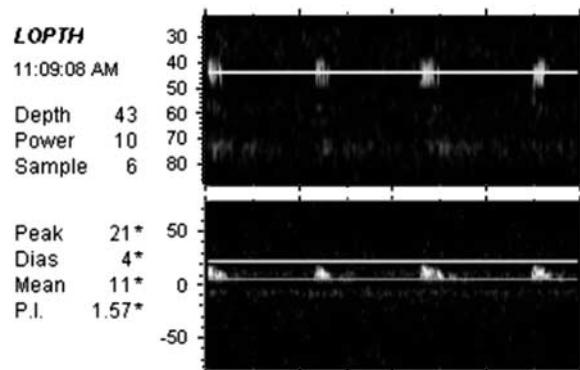
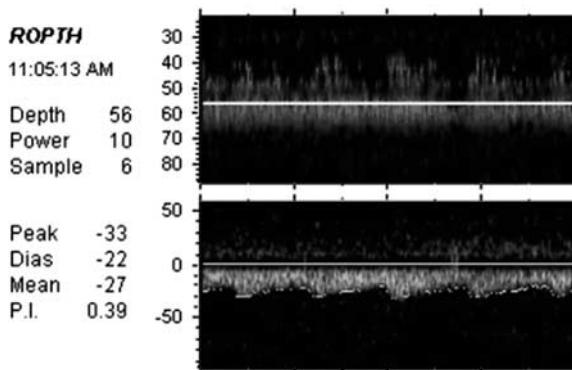


**FIGURE 6.** Arterial spin labeled perfusion magnetic resonance imaging demonstrates asymmetric signal within the right middle cerebral artery branch vessels in the Sylvian fissure. This phenomenon is termed “arterial transit artifact,” and coupled with chronic border zone ischemic change seen in Figure 1, indicates delayed flow to brain parenchyma with impaired cerebrovascular reserve.”

to the optic radiations (MCA or PCA territory) or to the visual cortex (PCA territory) as there were both ophthalmic artery and PCA collaterals involved in forming the collateral blood supply. Following indirect MCA territory revascularization and the induction of additional collateral supply, the effect of retinal steal on parenchymal perfusion was mitigated. However, the patient still suffers from intermittent left-sided weakness, tingling, and variable left limb shaking which occur with decreases in hemoglobin suggesting that his brain is still susceptible to ischemia in the setting of low flow and hypoxia.

Our hypothesis of a reversed “steal” phenomenon is supported by multimodal imaging studies suggestive of chronically diminished CVR in the right MCA territory, including preexisting border zone ischemic changes and findings of delayed blood flow on ASL perfusion. The latter is a novel MR perfusion sequence that relies on detecting signal generated from endogenously labeled blood. No contrast is required and the imaging allows direct detection of the flow of blood. In the normal setting, this signal rapidly progresses from the intracranial vasculature into the brain parenchyma where perfusion parameters can be measured. In our patient, the

signal on ASL imaging was delayed in the right MCA territory with labeled blood “caught up” in the arteries themselves. In addition, a similar finding was seen in his native PCA collaterals. This is further evidence of chronically impaired and delayed perfusion and of the importance of the baseline retrograde ophthalmic flow to maintaining normal neurologic status. In the setting of light-induced retinal steal, delayed flow in his MCA and PCA collaterals was no longer sufficient to maintain function, and symptomatic ischemia developed. We can speculate that surgical revascularization provided sufficient perfusion to the hemisphere, providing his oxygen carrying capacity was adequate.



**FIGURE 5.** Transcranial Doppler of the left and right ophthalmic artery confirming retrograde flow in the right ophthalmic artery. ROPTH indicates right ophthalmic artery; LOPTH, left ophthalmic artery; Dias, diastolic; PI, pulsatility index.

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