Published in final edited form as:

Am J Med. 2018 February ; 131(2): e39–e42. doi:10.1016/j.amjmed.2017.08.032.

Lights Out: An Unusual Case of Amaurosis Fugax

Mark R. Parsons, OD, FAAO^a, Michael C. Stoner, MD, FACS^b, Adam Doyle, MD^b, Doran Mix, MD^b, and Scott J. Cameron, MD, PhD, FACC^c

^aReed Eye Associates, Rochester, NY, University of Rochester School of Medicine, NY.

^bVascular Surgery, University of Rochester School of Medicine, NY.

^cDivision of Cardiology, Department of Medicine, University of Rochester School of Medicine, NY.

PRESENTATION

A 64-year-old Caucasian woman presented to the office for a routine ocular health assessment with a secondary complaint of occasional episodes of “black vision” occurring in the right eye in the preceding 6 months. She also described occasionally seeing “stars” in her right visual field of the affected eye after blinking, which was coincident with positional vertigo. These episodes would last approximately 5 minutes, with slow recovery to normal vision. Her first symptom occurred 6 months before the initial examination, with 5 recurrences—the last of which was 1 week before the initial examination. The patient’s husband described the patient as having heightened episodes of confusion, decreased alertness, and difficulty staying awake in the preceding several months. The patient endorsed several presyncopal episodes associated with visual disturbances in the right eye. She also endorsed claudication. She denied ever experiencing prior episodes of exertional chest pain, dyspnea on exertion, or dysrhythmia. Past medical history was significant for hypertension, dyslipidemia, and hyperthyroidism. Family history was significant for cardiovascular disease in several first-degree relatives. Vascular risk factors included a current 25 pack-year tobacco history.

ASSESSMENT

On presentation the patient had the following vital signs: blood pressure 88/60 mm Hg (right upper arm); 120/80 mm Hg (left upper arm); heart rate 56 beats per minute; respiratory rate 16 breaths per minute. The external ophthalmic examination was unremarkable, without evidence of ptosis, pupil abnormality, or gross visual field defects on confrontation. Her best corrected visual acuity was 20/25 in the right and 20/25 in the left eye. The patient’s neurologic examination was nonfocal, without gross sensory or motor deficits. The slit lamp revealed areas of anterior basement membrane dystrophy in both eyes, with mild nuclear and cortical cataracts. A dilated fundoscopic examination was unremarkable; there was no evidence of retinal ischemia or retinal hemorrhage, and Hollenhorst plaques were not

Requests for reprints should be addressed to Scott J. Cameron, MD, PhD, FACC, University of Rochester School of Medicine, Department of Medicine, Division of Cardiology, 601 Elmwood Ave, Box 679, Rochester, NY 14642.
Scott_Cameron@urmc.rochester.edu.

Conflict of Interest: None.

appreciated. The peripheral retinal evaluation was similarly unremarkable. There were no carotid bruits upon auscultation, though the right carotid artery was notable for a diminished upstroke by palpitation. Auscultation of the femoral arteries revealed bruit bilaterally. The remainder of the lower extremity examination was notable for spider veins in the anterior tibial regions and mild ankle edema bilaterally. The electrocardiogram revealed normal sinus rhythm without ischemia and no evidence of a past completed transmural infarction.

DIAGNOSIS

When considering the patient's symptoms, cardiovascular history, and testing, in the absence of frank ophthalmic disease, the examining clinician diagnosed the patient with amaurosis fugax, suspecting carotid occlusive disease and atheroembolism. However, with positional vertigo temporally associated with right amaurosis fugax in the context of discrepant bilateral blood pressure recordings by manual sphygmomanometry, proximal subclavian artery stenosis precipitating ipsilateral retrograde flow in the vertebrobasilar system was suspected (the subclavian steal phenomenon). This constellation of symptoms and physical examination findings generally reflects retrograde arterial blood flow and brain hypoperfusion.¹ Arterial duplex ultrasound interrogation of the carotid arteries did reveal a dampened right internal carotid artery signal and retrograde flow in the right vertebral artery. Computed tomography angiography, however, ultimately revealed a significant occlusion in the brachiocephalic artery, with diminished blood flow to the right subclavian artery (Figure 1).

MANAGEMENT

Dual antiplatelet therapy consisting of aspirin 81 mg once daily and clopidogrel 75 mg once daily were started for stroke prophylaxis. Smoking cessation was advised, and a nicotine patch was prescribed. The patient's atorvastatin was increased from 10 mg to 40 mg once daily. The patient was referred to a vascular surgeon, who recommended performing brachiocephalic stenting retrogradely as previously reported² (Figure 2). The patient tolerated the procedure well, without procedure or postoperative complications. Upon follow-up with her cardiologist, she reported "feeling like a new person" and affirmed abstinence from tobacco products. There were no recurrent visual disturbances after brachiocephalic stenting.

DISCUSSION

Amaurosis fugax describes monocular or binocular loss of vision, typically lasting from 4 to 60 minutes.^{3,4} The etiology of this symptom generally is a manifestation of thromboembolic disease in which atherosclerotic debris originating in the carotid artery or the aortic arch temporarily disrupts blood flow in the branch or central retinal arteriolar network. In some cases, appreciating Hollenhorst plaques during the fundoscopic examination on a routine eye examination, even in asymptomatic individuals, reflects significant thromboembolic disease—and especially so if a carotid bruit is appreciated by auscultation.⁵ It is therefore incumbent upon the evaluating clinician to perform auscultation of the carotid artery, but also to consider advanced imaging modalities of the carotid arteries and occasionally the

aorta and its branches for culprit atherosclerotic lesions as noted in this patient. Immediately treating a patient with amaurosis fugax in whom aortic or carotid atherosclerosis is detected with an antiplatelet agent and a statin are consistent with established guidelines.⁴ Amaurosis fugax is most often associated with significant internal carotid artery atherosclerosis, though some suspicion for atheroembolism from the aorta or its branches, including the subclavian or brachiocephalic artery, should remain. The patient in question showed significant atherosclerosis in the aortic branches and the carotid artery, and so an atheroembolic etiology behind the patient's symptoms is reasonable—despite an unrevealing fundoscopic examination. Whenever stenosis is observed in the brachiocephalic artery, the vertebral artery, or the subclavian artery, it is critical that the treating clinician immediately consider giant cell arteritis or Takayasu arteritis. Both giant cell arteritis and Takayasu arteritis constitute vascular emergencies and should prompt immediate referral to a specialist in vascular medicine or rheumatology. Although giant cell arteritis is more common in patients older than 50 years, Takayasu arteritis is more common in patients younger than 40 years and particularly prevalent in women of child-bearing age.^{6,7} Recognizing a potential case of giant cell arteritis or Takayasu arteritis is of utmost importance because treatment with anti-inflammatory and sometimes immunologic agents takes priority over immediate revascularization procedures, according to established guidelines.^{8,9} In our patient, however, the constellation of clinical variables associated with large vessel vasculitis was absent.

Together, the imaging finding of retrograde vertebral artery blood flow with symptoms of vertebrobasilar insufficiency and presyncope are sometimes resolved by revascularization.¹⁰ In patients with severe subclavian stenosis—as was initially suspected after the discovery of marked discrepancy in bilateral blood pressures—increased blood flow to the ipsilateral arm with exertion can decrease and sometimes reverse blood flow in the vertebral artery immediately distal to the stenotic lesion. These symptoms should prompt the treating clinician to request arterial duplex imaging to interrogate the subclavian and carotid arteries. A more unusual feature in this patient revealed only by computed tomography angiography was stenosis of the brachiocephalic artery rather than severe carotid or subclavian stenosis.

The decision was made to stent the brachiocephalic artery in a retrograde fashion through the carotid artery using the ENROUTE transcarotid device (Silk Road Medical, Sunnyvale, Calif) as described in the Investigation of Transcarotid CAS with Dynamic Flow Reversal in Subjects with Significant Extracranial Carotid Stenosis, carrying a risk of adverse events of less than 2%.² The patient's visual symptoms disappeared immediately after this procedure, and she was found to be in good health in clinic 1 month and then 5 months after the procedure.

Acknowledgments

Funding: This study was supported by National Institutes of Health grants HL120200 and K08HL128856 (to SJC).

References

1. Potter BJ, Pinto DS. Subclavian steal syndrome. *Circulation*. 2014;129:2320–2323. [PubMed: 24891625]

2. Kwolek CJ, Jaff MR, Leal JJ, et al. Results of the ROADSTER multicenter trial of transcatheter stenting with dynamic flow reversal. *J Vasc Surg.* 2015;62:1227–1234. [PubMed: 26506270]
3. Kvickstrom P, Lindblom B, Bergstrom G, Zetterberg M. Amaurosis fugax—delay between symptoms and surgery by specialty. *Clin Ophthalmol.* 2016;10:2291–2296. [PubMed: 27895459]
4. Pula JH, Kwan K, Yuen CA, Kattah JC. Update on the evaluation of transient vision loss. *Clin Ophthalmol.* 2016;10:297–303. [PubMed: 26929593]
5. Bakri SJ, Luqman A, Pathik B, Chandrasekaran K. Is carotid ultrasound necessary in the clinical evaluation of the asymptomatic Hollenhorst plaque? (An American Ophthalmological Society thesis). *Trans Am Ophthalmol Soc.* 2013;111:17–23. [PubMed: 24072943]
6. Steel L, Khan A, Dasgupta B. Giant cell arteritis: beyond corticosteroids. *Drugs Aging.* 2015;32:591–599. [PubMed: 26239828]
7. Brunner J, Feldman BM, Tyrrell PN, et al. Takayasu arteritis in children and adolescents. *Rheumatology (Oxford).* 2010;49:1806–1814. [PubMed: 20562196]
8. Delecoeuilliere G, Joly P, Cohen de Lara A, Paolaggi JB. Polymyalgia rheumatica and temporal arteritis: a retrospective analysis of prognostic features and different corticosteroid regimens (11 year survey of 210 patients). *Ann Rheum Dis.* 1988;47:733–739. [PubMed: 3178314]
9. Kyle V, Hazleman BL. Treatment of polymyalgia rheumatica and giant cell arteritis. II. Relation between steroid dose and steroid associated side effects. *Ann Rheum Dis.* 1989;48:662–666. [PubMed: 2782976]
10. Wrotniak L, Kablak-Ziembicka A, Roslawiecka A, et al. Resolution of ischemic symptoms after percutaneous angioplasty for a symptomatic subclavian artery stenosis. *J Vasc Surg.* 2016;64:684–691. [PubMed: 27565589]

Figure 1. Computed tomography angiography. Sagittal section (left) and coronal section (right) showing severe stenotic disease at the confluence between the brachiocephalic artery (BCA) and aortic origin of the left common carotid artery (CC [red arrow]) and left subclavian artery (SC). Calcific atherosclerosis is indicated by the black arrow. AA = aortic arch; LV = left ventricle.

Figure 2. Digital subtraction angiography. Contrast dye injection before (left) and after (right) stent deployment in the brachiocephalic artery (BCA). Severe stenosis of the BCA is indicated by a filling defect in the vessel lumen after dye injection into the aorta using a pigtail catheter. The region of BCA stenosis and stent deployment is indicated by the white arrow. AA = aortic arch; BC = brachiocephalic artery; CC = common carotid artery; SC = subclavian artery.