Pearsals & Oy-sters: Ophthalmic artery malperfusion in aortic dissection with common carotid artery involvement

PEARLS
• Acute aortic dissection is associated with cerebral malperfusion in 6%–20% of cases.
• Supra-aortic vessel involvement occurs in 15%–41% of cases with dissection extending into the common carotid or subclavian arteries.
• In some cases, visual symptoms are the only presenting features of common carotid artery involvement.

OY-STERs
• Aortic dissection can present with neurologic symptoms pertaining to vision when the dissection compromises the ophthalmic artery. The most common visual symptom associated with this phenomenon is amaurosis fugax.
• Carotid duplex ultrasound can be a tool for early diagnosis of supra-aortic vessel involvement in aortic dissection.

Aortic dissection presents with a myriad of symptoms, including rare cases of ocular disturbance. Commonly used classification systems for aortic dissection are based on the anatomic involvement and extent of the dissection. The Stanford classification defines type A dissections as those involving the ascending aorta and type B dissections as those that occur distal to the left subclavian artery. Most patients with acute type A aortic dissection present with chest or back pain. However, any branch artery can dissect, causing occlusion or stenosis with distal end-organ ischemia. Carotid artery dissection occurs in up to 24% of cases and is associated with neurologic events on admission.1 In 15%–41% of cases, there is involvement of the supra-aortic vessels, including common carotid or subclavian arteries.2

We present 2 cases of acute type A aortic dissection causing ophthalmic artery insufficiency and monocular blindness.

CASE 1
A 65-year-old man with a history of hypertension, hypercholesterolemia, and a 30-year history of tobacco use presented to the emergency room with acute onset of upper chest pain radiating to his back. The patient reported shortness of breath, right shoulder pain, right neck pain, and right monocular blindness. CT scan revealed an acute type A aortic dissection extending from the ascending aorta with involvement of innominate and right common carotid arteries (CCA) extending through the thoracoabdominal aorta to the level of the renal arteries. The patient was transferred to our hospital for higher level of care.

His right eye vision returned after blood pressure reduction using IV β-blockade. The right neck and shoulder pain also improved. Physical examination revealed equal pupils, round and reactive to light and accommodation, and intact extraocular movements. There was a diminished right carotid pulse when compared to the left. No preoperative carotid imaging was available.

The patient was brought to the operating room urgently and had transesophageal echocardiography, confirming the ascending aortic dissection with severe aortic insufficiency. He was placed on cardiopulmonary bypass and deep hypothermic circulatory arrest with retrograde cerebral perfusion via median sternotomy. A 5-cm dissecting ascending aortic aneurysm was identified and was replaced along with the proximal transverse aortic arch using a 28-mm woven Dacron tube graft. Aortic clamp time was 91 minutes.

Postoperatively, the carotid duplex and CT scans revealed a persistent right common carotid dissection, although the patient did well and had no procedure-related complications.

CASE 2
A 31-year-old man with a history of hypertension and noncompliance with his prescribed β-blockers presented with sudden shortness of breath, acute chest pain, and right eye blindness. CT imaging showed acute type A aortic dissection with extension into right CCA (figure, A), visceral, and renal arteries. He had no history of connective tissue disorders. Upon transfer to our hospital, the patient continued to report chest pain, nausea, and right eye...
blindness, and was found to have severe aortic insufficiency. A carotid duplex scan revealed a right CCA dissection (CCAD) with internal carotid artery thrombosis (figure, B).

At operation, a 3.4-cm aneurysm and a 2-cm tear was discovered on the posterior medial wall of the ascending aorta. The patient underwent replacement of the ascending aorta and proximal transverse arch with a 28-mm woven Dacron graft. Total aortic clamp time was 110 minutes. On postoperative day 2, the patient had a cardiac arrest, pulseless electrical activity, and died, despite resuscitative efforts.

**DISCUSSION** We reviewed the literature to find other cases addressing supra-aortic extension of type A aortic dissections involving the CCAs that manifested as ophthalmologic symptoms. We found 11 additional cases of retinal compromise due to aortic dissection involving the CCAs. The most common visual complaint among this patient group was noted to be amaurosis fugax. Similar to our case, the visual symptoms in 5 of these cases were present on admission, while in the rest they did not appear until after the aortic repair, probably related to persistent CCA malperfusion. Additional CCA intervention was required in 2 patients because of recurrent retinal TIAs. Table e-1 on the Neurology® Web site at Neurology.org summarizes the findings of these cases.

Sudden monocular blindness is a rare manifestation of acute aortic dissection. A high index of suspicion is imperative for recognition, timely diagnosis, and early repair of proximal aortic dissection to improve the prognosis of these high-risk patients. Carotid duplex ultrasound is a sensitive modality useful to diagnose the supra-aortic extension of dissection, intimal tears, and dynamic flaps, and to assess patency of the false and true channels. The mechanism of ophthalmic artery insufficiency involves either thromboembolism or hypoperfusion due to dissection flap obstruction of the internal carotid artery. Obstruction of branch arteries due to a dissection flap can be static or dynamic. Static obstructions are caused by extension of the dissection into the branch artery that causes thrombosis. Dynamic obstructions cause intermittent occlusion of the branch artery origin. The dissection flap prolapses into the branch artery origin depending on the false lumen pressure. Thus, dynamic obstruction is intermittent and lowering systemic pressures leads to improved perfusion by lessening the size of the false channel and increasing the area available for flow through the true channel. Thus, unlike most approaches to management of ischemia in the brain, where lowering systemic pressures is avoided, lower systemic pressure in the setting of dynamic obstruction due to dissection is critical to improving perfusion. This explains the improvement in symptoms in case 1 after IV β-blockade.

Based on our experience in management of CCAD of aortic origin and from the traumatic or spontaneous carotid dissections literature, we recommend medical therapy with aspirin alone (81–325 mg daily), clopidogrel alone (75 mg daily), or full anticoagulation for 6 months after proximal aortic repair. Carotid revascularization should be reserved for patients with refractory ischemic symptoms referable to CCAD following adequate medical therapy. Both open and endovascular approaches are feasible options depending on patient’s anatomy and extent of aortic dissection.

**AUTHOR CONTRIBUTIONS**

Drafting/revising the manuscript for content, including medical writing for content: V.K., H.K.S., A.-C.L.M., A.A., A.E., H.J.S., K.M.C.-O. Study concept or design: V.K., H.K.S., K.M.C.-O. Analysis or interpretation of data: H.K.S., K.M.C.-O.
STUDY FUNDING
No targeted funding reported.

DISCLOSURE
V. Kumar and H. Sandhu report no disclosures relevant to the manuscript. A. Meyer has received support from the Fogarty International Center of the NIH (1K01TW008764-01A1) and National Institute of Neurological Diseases and Stroke (1R21NS077858-01), the Global Health Equity Scholars Program of Yale University, the University of California San Francisco Hellman Fellows Program, and the World Federation of Neurology. She has received a drug donation from Valeant Pharmaceuticals. A. Azizzadeh is a consultant with Medtronic and W.L. Gore et al. A. Estrera, H. Safi, and K. Charlton-Ouw report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES
Pearls & Oy-sters: Ophthalmic artery malperfusion in aortic dissection with common carotid artery involvement
Neurology 2015;84:e27-e29
DOI 10.1212/WNL.0000000000001208

This information is current as of February 2, 2015

Updated Information & Services
including high resolution figures, can be found at:
http://n.neurology.org/content/84/5/e27.full

Supplementary Material
Supplementary material can be found at:
http://n.neurology.org/content/suppl/2015/02/01/WNL.0000000000001208.DC1

References
This article cites 5 articles, 2 of which you can access for free at:
http://n.neurology.org/content/84/5/e27.full#ref-list-1

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
Carotid artery dissection
http://n.neurology.org/cgi/collection/carotid_artery_dissection
Visual loss
http://n.neurology.org/cgi/collection/visual_loss

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints
Information about ordering reprints can be found online:
http://n.neurology.org/subscribers/advertise

Neurology © is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2015 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.