Spontaneous bilateral internal carotid and vertebral artery dissections with dominant-hemisphere circulation maintained by external carotid artery–ophthalmic artery anastomoses

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Spontaneous cervical artery dissection (sCAD) is a major cause of stroke in young adults. Multiple sCAD is a rarer, more poorly understood presentation of sCAD that has been increasingly attributed to cervical trauma such as spinal manipulation or genetic polymorphisms in extracellular matrix components. The authors present the case of a 49-year-old, otherwise healthy woman, who over the course of 2 weeks developed progressive, hemodynamically significant, bilateral internal carotid artery and vertebral artery dissections. Collateral response involved extensive external carotid artery–internal carotid artery anastomoses via the ophthalmic artery, which were instrumental in maintaining perfusion because circle of Willis and leptomeningeal anastomotic responses were hampered by the dissection burden in the corresponding collateral vessels. Endovascular intervention by placement of Pipeline embolization devices and Atlas stents in bilateral internal carotid arteries was successfully performed. No syndromic or systemic etiology was discovered during a thorough workup.

https://thejns.org/doi/abs/10.3171/2018.11.FOCUS18443

KEYWORDS spontaneous cervical artery dissection; carotid dissection; vertebral dissection; flow diverter; meningolacrimal artery
FIG. 1. A–D: 3D centerline reconstructions of cervical vessels on admission CT angiogram. Left ICA (A) showing critical stenosis starting distal to the carotid bulb through to the petrous segment and persistent irregularity in the cavernous segment; right ICA (B) showing irregular vessel caliber at the cervical segment, but with good contrast filling; left VA (C) showing a critical stenosis at the V3 segment; right VA (D) with no abnormality. E: Diffusion-weighted MRI sequence obtained on admission showing no diffusion restriction or evidence of ischemia. F: T2-weighted MRI showing multivessel simultaneous dissection as diminished caliber of flow voids accompanied by surrounding intramural hematoma in the right ICA (arrow), left ICA (arrowhead), and left VA (asterisk).
cal function attributable to a robust congenital anastomosis between the external carotid artery (ECA) and ophthalmic artery (OphA) systems.

Case Report

History

A 49-year-old woman presented to the emergency department with 3 hours of mild right-hand weakness and mild headache. She had a history of migraine and a remote history of C6–7 anterior cervical discectomy and fusion. She intermittently participated in hot yoga and Pilates, but denied recent trauma, chiropractic manipulations, or family history of stroke or vasculopathy. She insisted that her headache did not resemble her typical migraines, nor had she ever had migraines with auras. She had reported similar symptoms 5 days prior; CT angiography (CTA) performed at another institution was suggestive of a left internal carotid artery (ICA) dissection, for which she was medically managed with aspirin and clopidogrel.

Examination and Initial Workup

When the patient was initially triaged, a stroke code was called. Examination revealed only a slight slowing in rapid right-hand finger movements (National Institutes of Health Stroke Scale [NIHSS] score = 0). Emergency head CT scans obtained without contrast showed no acute intracranial pathology. However, given the mild asymmetry in her finger movements in the context of mild headache and recent history, vascular imaging was pursued.

Admission CTA of the head and neck revealed abrupt tapering of the proximal left ICA at the level of the C3 vertebral body, followed distally by a severe > 90% stenosis with persistent irregularity through the cavernous segment—consistent with severe dissection (Fig. 1A). Also noted was new irregular appearance of the right ICA at the cervical segment with approximately 50% stenosis (Fig. 1B). Additionally present was a new critical stenosis in the left vertebral artery (VA) at the V3 segment (Fig. 1C). The right VA was unremarkable (Fig. 1D). The aortic arch and origins of the great vessels were normal.

MRI was obtained without contrast to rule out infarction, and showed no acute ischemia. Again seen were dissections of the bilateral ICAs and left VA on T2-weighted imaging (Fig. 1E and F).

Management

The patient was admitted to the stroke unit. She was medically managed with aspirin, clopidogrel, and atorvastatin and closely monitored. Given the critical nature and unclear etiology for her multiple sSCADs, she consented to an urgent diagnostic angiogram with a possibility for intervention.

Endovascular Procedure and Findings

Under general anesthesia, right femoral artery access was obtained in the usual fashion via a 5-Fr catheter. Initial imaging of the descending aorta, renal vasculature, aortic arch, and proximal great vessels showed no abnormality. Digital subtraction angiography of the right VA showed leptomeningeal anastomoses between the right posterior cerebral artery (PCA) and middle cerebral artery (MCA) and retrograde flow through a fetal PCoA in the presence of a hemodynamic stress from the stenosis in the anterior circulation (A). PCoA also visualized in the injection of the left VA (B). Acute right ICA critical stenosis seen on right CCA injection (C) with corresponding loss of perfusion of the right hemisphere (D). Known left ICA critical stenosis seen on left CCA injection (E) with distal perfusion preserved, probably from retrograde collateral flow, given the early appearance of an ECA-ICA anastomosis between the MMA and OphA (F).
critical stenosis of distal left ICA (Fig. 2E). Cranial views showed early retrograde flow through the left OphA from the ECA circulation reconstituting the left ICA territory (Fig. 2F). Irregular vessel caliber with a 30% stenosis was seen in the left VA at the level of C1 (Fig. 2B).

Given rapid progression of the right ICA dissection and after discussion with the patient's husband, himself a physician, emergency revascularization was undertaken. Balloon angioplasty of distal and middle segments of the right ICA was performed using a Maverick 4 × 15-mm balloon, with partial restoration of vessel caliber (Fig. 3A). Two Atlas stents (Stryker) measuring 4.5 × 30 mm and 4.5 × 20 mm were sequentially deployed from the horizontal petrous to midcervical segments, restoring full luminal patency and cerebral perfusion (Fig. 3B–D).

The left CCA was then similarly catheterized, followed by placement of a 4.5 × 30-mm Atlas stent from the horizontal petrous to upper cervical segments of the left ICA with minimal improvement in vessel patency (Fig. 3E). Balloon angioplasty was then performed, followed by the sequential deployment of two 5 × 25-mm Pipeline embolization devices (PEDs) spanning the vertical cavernous segment to the proximal cervical segment (Fig. 3F and G). Restoration of luminal caliber and normalization of cerebral perfusion were observed (Fig. 3F and H). The left VA dissection did not have hemodynamic significance and was not treated.

After hemostasis was achieved, the patient emerged from anesthesia without neurological deficits. Right-hand rapid finger movements were restored to baseline. She was discharged to the stroke unit in stable condition.

**Postoperative Course**

Postintervention CTA showed improved flow through both anterior and posterior circulations despite persistent intramural hematomas in the cervical segments of both

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ICAs and the left VA. Other workup excluded inflammatory, autoimmune, or infectious vasculitis and hyperhomocysteinemia, and fibromuscular dysplasia was unlikely given normal results on the aortogram. A genetic polymorphism combined with increased risk from her history of anterior cervical discectomy and fusion was the most likely mechanism for her 3-vessel sCAD. She was discharged with an NIHSS score of 0 on hospital day 3. Close follow-up imaging 2 weeks later showed asymptomatic, persistent, irregular narrowing of the midcervical right ICA, which was treated by placement of a single 5 × 25–mm PED to cover the previously placed Atlas stents and the residual narrowed segment (Fig. 4A and B). Also found was yet another new, mild, right VA dissection that was managed conservatively.

MR angiography (MRA) sequences obtained at 3 and 6 months showed progressive healing of all dissections with only mild residual stenosis of the distal cervical left ICA (Fig. 4C and D). The patient remains without neurological deficit and is being maintained on dual-antiplatelet therapy given the dissection burden and lengths of the stented segments. A lifelong ban on cervical neck stress, including yoga, was advised.

**Discussion**

Multiple sCAD is a rare, life-threatening pathology for which the etiology is frequently unknown. Approximately 20% of patients with sCAD have multivessel involvement, but only a handful of these cases are explained by syndromic connective tissue disorders. The diagnosis of multivessel sCAD continues to be made most reliably by catheter angiography because MRA and CTA have relatively worse sensitivity. The elusiveness of this diagnosis is exaggerated by its wildly variable presentation. Treating these patients is further complicated by the inherent—probably genetic—instability of the involved vasculature, compounded by the plurality of the insult.

The goals in managing dissection are to prevent progression, limit thromboembolic events, and preserve distal organ perfusion. Stabilizing the dissection and preventing the formation of thrombus with antiplatelet, anticoagulation, or antifibrinolytic agents is routine. The recent CADISS (Cervical Artery Dissection in Stroke Study) trial suggested that antiplatelet and anticoagulant drugs have equal efficacy in preventing stroke or death in cervical dissections, but intracranial hemorrhage was more likely with anticoagulants. In cases with intracranial extension, a high rate of associated subarachnoid hemorrhage has been observed, making dual-antiplatelet therapy a better choice for our patient over intravenous fibrinolytics or anticoagulation. In critical cases, endovascular intervention is highly favored over open surgical repair due to its exceedingly lower perioperative stroke risk (0.02% vs 10%) and mortality rate (nearly 0% vs 2%). Our patient’s critical stenoses made her a candidate for acute endovascular intervention especially since she, a young, active woman, was experiencing recurring dominant-hemisphere symptoms at rest (despite collateral circulation), putting her at high risk for future hypoperfusion and possible stroke.

The right ICA stenosis was prioritized given the over-night change in vessel caliber and was treated with 2 Neuroform Atlas stents, which were approved by the FDA in 2017. The Atlas is a self-expanding, nitinol, laser-cut mini-stent with an open cell design developed for stent-assisted aneurysm coiling. The Atlas has several technical advantages over previous models, principally a lower delivery profile. However, it has a relatively low outward radial force, favoring balloon predilatation prior to deployment. Our patient additionally received 2 PEDs for her longer left cervical and petrous ICA dissection. PEDs, also well described in aneurysm management, are flexible mesh devices that can conform to vessels despite high degrees of regional tortuosity, and are well suited for (off-label) treatment of complex dissections particularly with intracranial extension. However, this device is substantially more thrombogenic than the Atlas stent family, and therefore greater attention to the antiplatelet regimen is required.

It is possible for patients with a critical ICA stenosis to initially present without neurological deficits in the setting of robust collateral circulation, which includes, in general descending order of efficiency, the circle of Willis, multiple ECA-ICA anastomoses (principally via the OphA),
FIG. 5. Upper: Magnified image from interventional angiogram (Fig. 2F) showing early reconstitution of left anterior circulation from MMA (white arrows) to the lacrimal artery (white arrowheads) and OphA (black arrowheads) via a fetal neomorphic remnant, the sphenoidal artery (black arrows). Lower: Anatomical illustration in the anterior view of potential MMA-OphA anastomoses—including the sphenoidal and meningolacrimal arteries—and their associated embryologic origins. Pr DOA = primitive dorsal OphA; Pr VOA = primitive ventral OphA.
and leptomeningeal collateral vessels. Reconstitution of the OphA can proceed via multiple collaterals, which include the middle meningeal artery (MMA) and accessory meningeal, superficial temporal, deep temporal, infraorbital, and facial arteries. 

In the context of transarterial embolization, it is critical to identify these anastomoses to prevent unintended embolization into the OphA territory. 

In our case, the patient’s anastomosis served a protective role: a robust ECA-ICA anastomosis allowed for sufficient collateral flow to her dominant hemisphere despite a > 90% left ICA stenosis and limited efficiency of the circle of Willis and leptomeningeal collaterals due to dissections in the contralateral carotid and VA systems. The principal ICA reconstitution in this case proceeded via an MMA-OphA connection along the greater sphenoid wing, occasionally named the sphenoidal artery (Fig. 5). The sphenoidal artery enters the superior orbital fissure to Anastomose with the proximal OphA. The full persistence of this anastomosis—with complete MMA supply of the orbit and no identifiable origin of the OphA from the ICA—represents the meningo-ophthalmic variant. 

The same sphenoidal artery also participates in more limited MMA supply of the lacrimal gland, known as the meningo-lacrimal artery variant, recognized as a short, straight vessel entering the orbit through its own foramen of Hyrtl. Review of the relevant ontogenetics suggests that the sphenoidal artery may be a vestige of a later-developing neomorph. This embryological phenomenon is of particular interest to interventionalists because variants involving partial or complete OphA origin of the MMA are more heavily associated with the presence of a sphenoidal artery. This sphenoidal artery variant complicates surgical planning for cases in which an MMA derivating its circulation from the OphA is supplying pathologic processes such as meningiomas, dural arteriovenous fistulas, moyamoya disease, aneuysms, and chronic subdural hematomas. 

Conclusions  
In sCAD, clinical presentation can be misleading. The presence of robust fetal anastomoses between ECA and ICA circulations can mask the severity of disease and is only visualized on high-resolution intravascular imaging. Identification of potentially dangerous ECA-ICA anastomoses is critical to ensuring safe embolization procedures; however, the same anastomoses can be useful in maintaining cerebral perfusion. Off-label use of the PED and the Atlas stent for revascularization of cervical dissections deserves further study. 

Acknowledgments  
Employees at the NYU Radiology 3D Imaging Lab; NYU School of Medicine: Dr. Aaron Lord, Dr. Eytan Raz, Dr. Miguel Litao, Dr. Breehan Chancellor, and Dr. Asya Wallach. 

References  

Disclosures
Dr. Maksim Shapiro discloses a consultant and Pipeline device proctor relationship with Medtronic, Inc.

Author Contributions
Conception and design: Golub, Torres, Shapiro. Acquisition of data: Golub, Hu. Analysis and interpretation of data: Golub, Torres. Drafting the article: Golub, Hu, Dogra. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Golub. Administrative/technical/material support: Golub, Dogra, Torres, Shapiro. Study supervision: Golub, Torres, Shapiro.

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