Pearls and Oy-sters: Carotid dissection with normal arterial lumen

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PEARLS

- Clinical presentation of carotid artery dissection (CAD) is variable, including acute or subacute headache, cervical pain, or focal neurologic deficit that occurs spontaneously, after a high-speed neck manipulation, or trauma. Isolated lower cranial nerve palsy in carotid dissection is uncommon.
- CAD can be demonstrated by magnetic resonance (MR) angiogram, CT angiogram, or conventional angiography. Crescentic hyperintensity on cervical MR angiogram (T1-weighted with a fat saturation technique) or hyperdensity on CT indicates an intramural hematoma, suggestive of a recent CAD. The patency and normal caliber of the carotid artery filling lumen should not preclude the diagnosis of CAD. A submedial or subadventitial dissection may cause neurologic symptoms by extrinsic compression of the cranial nerves, most commonly cranial nerve (CN) XII.
- Ipsilateral CAD should be suspected in adults presenting with an acute onset of unilateral XII nerve palsy.

OY-STERS

- CT angiogram with normal luminal diameter constitutes a challenge for the diagnosis of CAD in the emergency department. The identification of enlargement or thickening along the wall of the internal carotid artery should be sought in patients with suspected CAD. MRI (axial T1 with a fat saturation technique) is the most sensitive test to identify arterial wall changes and confirm the diagnosis.

CASE REPORT A 60-year-old man presented to the emergency department complaining he could not whistle and had difficulty moving his tongue. The previous day he had developed sudden onset of left-side headache that remained present until his admission. The day of admission, he noticed that his tongue was deviated to the left side (figure, A), causing

swallowing difficulties. He denied any other neurologic symptoms. Past medical history was remarkable for mild hypertension. He denied high-speed neck manipulation. He acknowledged lifting of heavy boxes in the past 2 weeks. Neurologic examination revealed tongue deviated to the left suggestive of left hypoglossal nerve palsy. The remaining neurologic examination was unremarkable. Unenhanced head CT showed crescentic hyperdensity within the upper cervical internal carotid artery (ICA) at the skull base, suggesting dissection with intramural hematoma (figure, B). CT angiogram showed a cuff of abnormal soft tissue along the periphery of the vessel wall (figure, C) with preservation of luminal diameter. Dissection was confirmed with MRI brain (figure, D) which showed T1 hyperintensity (indicating intramural methemoglobin) within the wall of the internal carotid artery. There was no cerebral infarction. The patient was given aspirin 81 mg once daily. He recovered completely within 2 months.

DISCUSSION Cervical artery dissection is a common cause of stroke in young and middle-aged adults.^{1,2} The most frequent vessels involved are the ICA 2 cm above the bifurcation and vertebral artery at V3 segment.¹ Head trauma and high-speed neck manipulation are crucial predisposing factors of CAD.¹ Some conditions affecting the connective tissue (i.e., Ehlers-Danlos syndrome, Marfan syndrome, or osteogenesis imperfecta) have been reported to be associated with CAD.¹ The most common pathologic features include an intimal tear resulting in subintimal dissection. Less commonly, a medial or subadventitial dissection causing an intramural hematoma may occur.² Neurologic symptoms in arterial dissection can result from a cerebral infarction or TIA related to artery-to-artery embolism (due to platelet aggregation or localized luminal thrombus formation at the site of intimal injury) or secondary to a hemodynamic mechanism from a stenotic or occluded artery.³ CAD comprises a wide variety of clinical presentations, including 1) asymptomatic, 2) local signs and symptoms (e.g., headache, cervical pain, cranial nerve palsies, tinnitus,

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(A) Deviation of tongue to the left suggestive of left hypoglossal nerve palsy. (B) Noncontrast CT brain showed crescentic hyperdensity (arrow) indicating intramural hematoma within the upper cervical left internal carotid artery (ICA). (C) Axial enhanced CT angiography image shows normal filling luminal diameter, with "cuff" of tissue thickening (arrow) along the wall of the left ICA. (D) Axial T1 fat suppression MRI brain unenhanced image shows intrinsic crescentic hyperdensity (arrow) in the wall of the left ICA.

or Horner syndrome), or 3) TIA or focal neurologic deficits due to stroke.^{1,2} Cranial nerve palsies, as a presenting sign in CAD, are rare. In a large study including 190 patients with spontaneous CAD, the authors reported that 12% presented with cranial nerve palsy, and 5.2% of them had a lower cranial palsy.⁴ The hypoglossal nerve (CN XII) is most commonly affected.^{4,5} Less commonly, the V, VII, IX, X, and XI cranial nerves may also be involved. Ocular cranial nerve involvement has been described in intracranial CAD or progression of extracranial CAD (e.g., III, V, VI, affecting the cavernous segment of the ICA).^{4,5} Cranial nerve palsies from CAD are usually explained by 2 mechanisms: 1) mechanical compression or stretching of the cranial nerves below the jugular foramen by the expanded artery or aneurysmal dilation at the origin of the dissection,⁶ or 2) impairment, transient or permanent, of the blood supply to the cranial nerves.^{4,5} CN XII has only a somatic motor

(general somatic efferent) component innervating all the intrinsic and 3 of the 4 extrinsic muscles of the tongue, including genioglossus, styloglossus, and hyoglossus. After passing through the hypoglossal canal, CN XII descends in the neck behind the vagus nerve and passes between ICA and internal jugular vein lying on the carotid sheath. Then, it courses over the internal and external carotid arteries and the hyoglossus muscle to supply tongue. CN XII compression or palsy results in tongue deviation to the ipsilateral side due to weakness of the innervated muscles and unopposed contraction of the contralateral genioglossus muscle. The close anatomic proximity to high cervical ICA explains its involvement in CAD.

The main diagnostic tools for CAD include MR angiogram, CT angiogram, and conventional angiogram. Classic findings include a luminal flap, false lumen, long tapered narrowing, or occlusion.^{1,2} On cross-sectional imaging (especially MR), a characteristic sign of CAD is an enlarged artery with crescent-shaped rim of hyperintense signal surrounding a lumen, which may or may not be reduced in size. These signs can be demonstrated by T1-weighted axial cervical MRI with a fat saturation technique.^{1,2} In our patient, the CT head also showed intramural hematoma within the upper cervical ICA at the skull base, suggesting dissection, and CT angiogram showed a cuff of abnormal soft tissue along the periphery of the vessel wall with preservation of luminal diameter. This case emphasizes the importance of looking at the vessel wall and not just the (filling) luminal contour, which can be normal.

The treatment of cervical artery dissection remains controversial. Anticoagulation or antiplatelet therapy is the mainstay. Currently, there is an ongoing randomized trial aimed at comparing the efficacy of antiplatelet vs anticoagulation in acute cervical dissections.⁷ In a meta-analysis including 34 nonrandomized studies (n = 762 patients), there was no difference in the risk of death (antiplatelet 5/268 [1.8%], anticoagulation 9/494 [1.8%], $p = 0.88$), stroke (antiplatelet 5/268 [1.9%], anticoagulant 10/494 [2.0%], $p = 0.66$), or stroke and death between antiplatelet and anticoagulation therapy.⁸ Overall, the prognosis in cervical artery dissection is good. Excellent outcome (defined as modified Rankin Scale score of 0–2) was reported in 92%.⁹ Recurrence rate is usually low (0.9%) after long-term follow-up.¹⁰

AUTHOR CONTRIBUTIONS

Dr. Jitphapa Pongmoragot, Dr. Aditya Bharatha, and Dr. Gustavo Saposnik contributed to the study concept and design, acquisition of data, and analysis and interpretation of results, and made critical revision of the manuscript for important intellectual content. Dr. Gustavo Saposnik supervised the study.

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DISCLOSURE

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