Pearls & Oy-sters: Unusual manifestations of bilateral carotid artery dissection

Deep monocular pains

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PEARLS

- Internal carotid artery dissection can present with isolated ocular pain
- MRI and angiography may be indicated in new-onset, unexplained eye, neck, or facial pain or headache

OYSTER

- Therapy for internal carotid artery dissection is not evidence-based and generally includes either antiplatelet or anticoagulant drugs

CASE REPORT A 44-year-old man in good general health presented with a 1-week history of deep-seated, moderate right eye pain without exacerbation on eye movement. Ophthalmologic examination showed visual acuity of 20/20 in both eyes, equal pupils with normal pupillary reflexes, mild right upper lid ptosis known to the patient for more than 5 years, and normal fundus examination in both eyes.

Neurologic examination was normal. There was no history of head or neck trauma in the past weeks. Cocaine drop test for Horner syndrome was negative.

Fluorescein and indocyanine green angiography were normal without delayed arterial filling.

Cranial MRI with fat-saturated sequences and magnetic resonance angiography (MRA) revealed bilateral acute internal carotid artery (ICA) dissections without fibromuscular dysplasia (figure 1, A–C). Aspirin 100 mg a day was prescribed.

Pain progressively disappeared after 2 months. No further neurologic events were reported after a follow-up of 5 months and MRI and angiography showed regression of the mural hematoma in both internal carotid arteries (figure 2, A and B). On the left side, a 4 × 3 mm pseudoaneurysm had appeared (figure 2C).

DISCUSSION Spontaneous ICA dissection has an annual incidence of around 2/100,000.¹ Bilateral ICA dissection represents 4% to 16% of all dissections.^{1,2} Extracranial ICA dissection is more common than intracranial. Main risk factors are blunt or penetrating trauma including cervical manipulation or strain, hyperhomocysteinemia, hereditary connective tissue disorders, and other genetic diseases.³

ICA dissection most commonly presents with head or neck pain (65%–80%), cerebral ischemia (60%–75%), and Horner syndrome (25%–50%).⁴ Pain is likely caused by stretching of the arterial wall, which contains pain-sensitive nerve fibers.⁵ Orbital pain may correspond to referred pain in the territory of the ophthalmic artery or to direct involvement of this artery by the dissection in the case of an intracranial ICA dissection.

Presentation with deep monocular pain as the only symptom of ICA dissection is very uncommon.⁶ To our knowledge, this is the first report of bilateral carotid artery dissection with monocular pain as the only symptom.⁷ Other conditions may present with deep ocular pain, including primary headaches such

Figure 1 Imaging

(A) Axial MRI, T1-weighted fat-saturated image, shows wall hematomas of the right (arrow) and left (arrowhead) internal carotid arteries at the level of their entry in the carotid canal. (B) Magnetic resonance angiography reconstruction, right internal carotid artery, shows the slight impact of the wall hematoma on the vessel lumen (arrow). (C) Magnetic resonance angiography reconstruction, left internal carotid artery, shows a 50% stenosis of the vessel lumen (arrow) by mural hematoma.

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Figure 2 Five-month follow-up imaging

(A) Five-month follow-up axial MRI, T1-weighted fat-saturated image, shows regression of the wall hematomas in the right (arrow) and left (arrowhead) internal carotid arteries. (B) Magnetic resonance angiography reconstruction, right internal carotid artery, shows the regression of the stenosis (arrow points to previous location of stenosis). (C) Magnetic resonance angiography reconstruction, left internal carotid artery, shows the regression of the stenosis and appearance of a small pseudoaneurysm (arrow).

as migraine and cluster headaches, orbital pathology such as inflammation, infection, tumor, thyroid orbitopathy, optic neuritis, and cavernous sinus pathology such as thrombosis or Tolosa-Hunt syndrome.

The diagnosis of Horner syndrome can be confirmed by topical application of cocaine to the eye. Cocaine blocks the reuptake of norepinephrine at the synapse between the postsynaptic sympathetic neuron and the iris dilator muscle, causing pupillary dilation in eyes with normal sympathetic innervation. In Horner syndrome, the oculosympathetic pathway is interrupted, and cocaine does not cause pupil dilation (a positive cocaine test). A normal pupillary dilation response (negative cocaine test) effectively rules out Horner syndrome.⁸

Why was there pain only on the right side, despite the presence of bilateral ICA dissection? One might speculate that the right-sided dissection was more of a subadventitial type causing outward dilation of the vessel wall without any significant impact on the vessel lumen, stimulating the nociceptive fibers in the wall of the ICA as well as those of adjacent tissues. In con-

trast, the left-sided dissection was more of the subintimal type, causing a greater reduction in the vessel lumen, but a lesser outward expansion of its wall.

It must be noted that a previous episode of ICA dissection might have explained the preexisting ptosis in our patient, although the initial MRI/MRA evaluation did not disclose sequelae of ICA dissection, such as vessel wall irregularity, ectasia, or pseudoaneurysm.

ICA unilateral or bilateral dissection can be asymptomatic. Why symptomatology from ICA is so variable is not understood; it was recently suggested that the number and distribution of activated intracranial collateral vessels may predict the gravity of ICA.⁹ A prospective study analyzing predisposing factors that predict ICA poor outcome would be interesting.

AUTHOR CONTRIBUTIONS

Dr. Olivier: writing the article. Dr. Schutz: critical revision of the article. Dr. Mégéand: writing the article, critical revision of the article.

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