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Acute Ptosis in a Middle-aged Man With Hypertension

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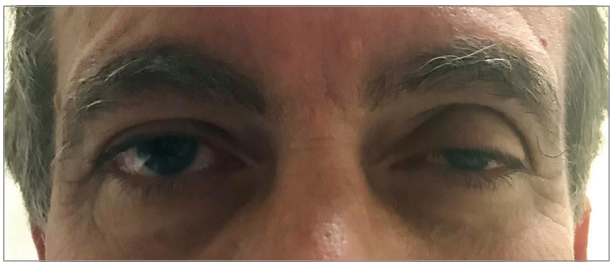

Figure 1. External photograph demonstrating ptosis of the left upper eyelid.

A 56-year-old man with hypertension presented with 2 weeks of acute ptosis in the left upper eyelid. Three weeks prior, the patient had noted severe left-sided facial pain. He denied any diplopia, decreased vision, or recent trauma. The patient had previously been evaluated by his primary care physician and referred to a neuroophthalmologist. During the course of the workup, the patient had undergone computed tomography (CT) of the head and chest, magnetic resonance imaging (MRI) of the brain, and carotid duplex ultrasonography. Following a workup with unremarkable results, the patient presented to the Bascom Palmer emergency department for a second opinion.

On initial examination, his best-corrected visual acuity was 20/20 OU. Intraocular pressure, ocular motility, and confrontational visual fields were normal. There was no axial proptosis or enophthalmos by exophthalmometry. The pupil was 5 mm OD and 4 mm OS. No afferent pupillary defect was observed. External examination demonstrated a left-sided ptosis (**Figure 1**). Anhidrosis was not observed. Anterior and posterior examination, including of the optic nerve head, had normal results in both eyes. The cranial nerve, gross motor, and gross sensory examination had otherwise unremarkable results. The patient's blood pressure was 160/79 mm Hg.

WHAT WOULD YOU DO NEXT?

A. Edrophonium test

B. Oculoplastic referral for ptosis repair

C. Magnetic resonance angiography of the brain and neck

D. Sumatriptan injection

Diagnosis

Horner syndrome

What to Do Next

C. Magnetic resonance angiography of the brain and neck

Discussion

There is a broad differential diagnosis for symptoms of headache, anisocoria, and ptosis. An edrophonium test (choice A) aids in diagnosing myasthenia gravis, while sumatriptan injections (choice D) can treat cluster headache. Although myasthenia gravis and cluster headache may present with acute ptosis, a patient with this constellation of symptoms must first be evaluated for life-threatening causative mechanisms. Myasthenia gravis typically does not present with a headache and anisocoria, while new migraines[1] or cluster headaches[2] in a 56-year-old patient are atypical. Lastly, a diagnosis of blepharoptosis and referral for surgical repair (choice B) should be made only after more lethal diagnoses have been excluded.

This patient with ipsilateral ptosis and miosis prompted the diagnosis of a left-sided Horner syndrome. Horner syndrome, particularly with facial pain and hypertension, must raise suspicion for carotid dissection.[3] An MRA of the head and neck was performed, demonstrating a 12-mm dissection of the left internal carotid artery in the high cervical portion of the neck (**Figure 2**). An intramural hematoma with severe narrowing of the true lumen was present.

The sympathetic innervation of the head and neck consists of a 3-order system.[4] Disruption of the first-order neuron, which runs from the hypothalamus to the cervical spinal cord, occurs secondary to causative mechanisms that include stroke, malignant conditions, and demyelinating lesions. The second-order neuron runs from the spinal cord through the brachial plexus to the superior cervical ganglion. Lesions affecting the thoracic outlet, mediastinum, pulmonary apex, and neck cause second-order lesions. Lastly, the third-order neuron enters the cranium with the internal carotid artery, traveling through the cavernous sinus into the orbit. Lesions affecting the internal carotid artery, including dissections, trauma to the base of the skull, and cavernous sinus lesions, result in third-order lesions.

Topical cocaine, 10%, drops can confirm Horner syndrome by causing dilation of the normal pupil only. Apraclonidine causes di-

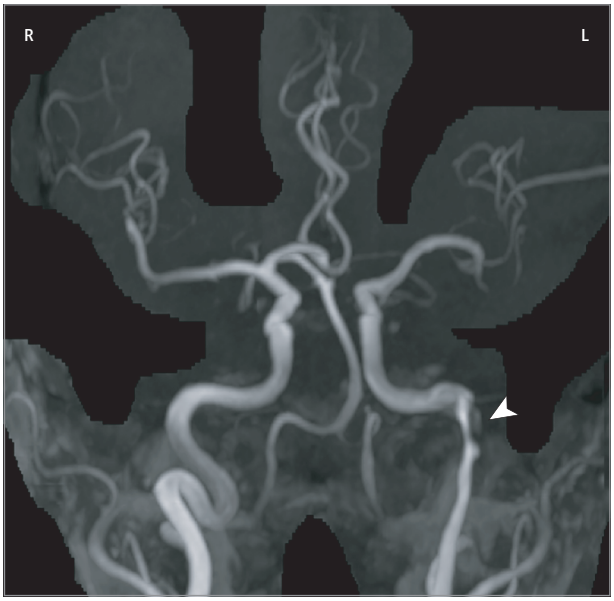

Figure 2. Magnetic resonance imaging and 3-dimensional reconstruction of the internal carotid arteries, showing severe narrowing of the left internal carotid artery in the high cervical region (arrowhead).

lation of the miotic pupil because of denervation sensitivity. Lastly, hydroxymethamphetamine differentiates third-order lesions from first-order and second-order lesions by causing dilation of the abnormal pupil in first-order and second-order neurons only.[5]

While this patient presented with an unremarkable CT of the head, MRI of the brain, and carotid ultrasonographic image, the absence of an angiogram rendered the previous workup incomplete. Carotid ultrasonography is a poor modality for detecting spontaneous carotid artery dissection in patients with isolated Horner syndrome. In 1 study[6] of 88 patients with confirmed spontaneous carotid artery dissection presenting with isolated Horner syndrome, carotid ultrasonography had a false-negative rate of 31%.

Both MRI with magnetic resonance angiography (MRA) and CT angiography are highly sensitive and specific in detecting spontaneous carotid artery dissection. Computed tomography requires radiation and iodinated contrast[7] but is faster and may better identify intraluminal thrombi.[8] With MRA, the sensitivity of MRI for carotid dissection ranged from 50% to 100% across studies, while CT angiography sensitivity ranged from 64% to 100%.[9] Generally, both MRI or MRA and CT angiography are acceptable imaging modalities for the evaluation of carotid dissection. While conventional angiography remains the gold standard test, it is associated with serious complications, including hemorrhage and infection.[10]

Patient Outcome

The patient was transferred to a nearby tertiary hospital. He was medically managed with oral anticoagulation and antihypertensive drugs.

ARTICLE INFORMATION

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