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Relative Afferent Pupillary Defect: An Unusual Manifestation of Carotid Artery Dissection

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ABSTRACT

We report the case of a man with a carotid dissection who had relative afferent pupillary defect (RAPD) that persisted throughout his hospital course. Other causes for RAPD were quickly ruled out. Carotid dissections often have ophthalmic manifestations—including RAPD, which, although uncommon as a presenting sign, can be tested for with the swinging flashlight test.

CASE REPORT

A 42-year-old man with a past medical history of hypertension, hypercholesterolemia, and hypothyroidism presented to the emergency department with a blood pressure of 221/117 mmHg, episodic transient blurry vision in his left eye, right upper extremity weakness, and expressive aphasia. The next day the patient developed a left facial droop, a right hemiplegia, and total aphasia. Carotid ultrasound revealed no flow in the proximal left internal carotid artery (ICA), and magnetic resonance angiography revealed a common carotid dissection leading to the occlusion of the left internal carotid from the skull base to the carotid terminus (Figure 1). Computed tomography and magnetic resonance imagining revealed an acute infarction of more than 75% of the left middle carotid artery and hemorrhage in the left basal ganglia (Figure 2).

On physical examination, the patient spontaneously opened his eyes and responded to pain but not to verbal commands. He had 0/4 strength on his right side but moved his left side spontaneously. He also had a positive Babinski sign on the right. Significantly, the left pupil was minimally reactive. During a swinging flashlight test, the right pupil constricted from 4 mm to 2 mm, while the left remained at 5 mm. The patient did not blink to threat on testing of his left eye. This left relative afferent pupillary defect (RAPD) persisted throughout his hospitalization.

The patient remained aphasic, hemiplegic, and minimally responsive. Because he would require intensive nursing care and probable percutaneous endoscopic gastrostomy tube placement for dysphagia, the family decided the patient would not desire such aggressive care. Palliative care was consulted, and soon after the patient was discharged to inpatient hospice.

DISCUSSION

Because the ophthalmic artery is a branch of the ICA, ophthalmic signs and symptoms are a common feature of carotid dissections. In one study, two-thirds of extracranial ICA dissections had ophthalmic symptoms or signs, and in half the cases these were presenting symptoms.[1] These disturbances tend to be monocular in nature and ipsilateral to the lesion.[2] The most common symptom, present in up to 58% of patients with dissections, is a partial painful Horner syndrome consisting of ptosis and miosis, secondary to damage to the sympathetic pathway from either ischemia or compression of sympathetic fibers running alongside the carotid. Transient episodic blindness, known as amaurosis fugax, occurs in up to 30% of patients, secondary to decreased blood flow from either hemodynamic compromise or emboli to the retina. Permanent visual loss and cranial nerve palsies are less common symptoms, occurring in less than 5% of patients with dissection. A carotid dissection can lead to an RAPD on the ipsilateral side of the

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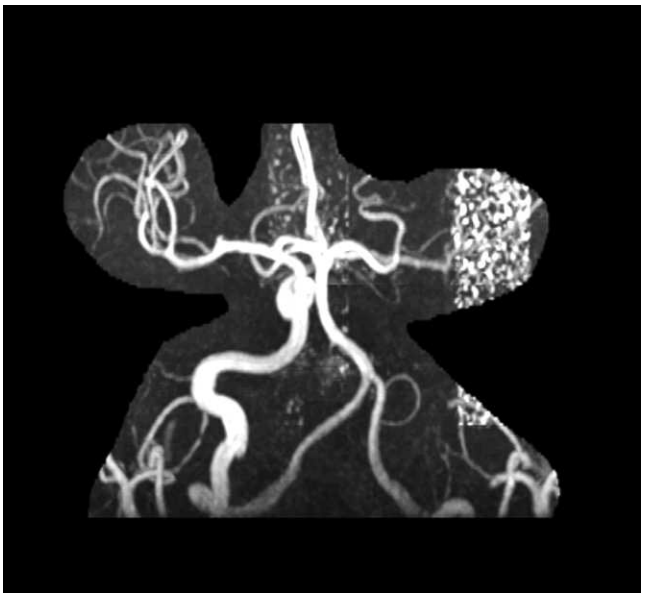

Figure 1. Magnetic resonance angiography of the circle of Willis showing the absence of flow from the left distal internal carotid artery from the skull base to the carotid terminus.

lesion via occlusion or hemodynamic compromise of the ophthalmic artery, preferentially affecting the retinotectal tract, similar to the etiology of amaurosis fugax. However, fewer than 1% of dissections present with an RAPD, making this finding fairly unusual.[3]

An RAPD, or Marcus-Gunn pupil, presents with pupillary asymmetry during the swinging flashlight test, in which the pupils constrict less when a flashlight is swung from the unaffected eye to the affected eye. A flashlight swung to the normal eye produces normal constriction. Afferent pupillomotor signals originate in the photosensitive ganglionic cells of the retina and terminate in the pretectal olivary nuclei in the midbrain. The efferent signal then travels to the Edinger-Westphal nuclei to synapse on the ciliary ganglion via the occulomotor nerves, finally innervating the constrictor muscle of the pupil. Because RAPDs result from an asymmetry in afferent signals, they can result from any unilateral lesion between the retina and the midbrain.[4] As such, the finding is fairly nonspecific, although the most common causes are disorders of the optic nerve. For example, an RAPD is pathognomonic for optic neuritis of multiple sclerosis, with more than 90% of patients with a new diagnosis presenting with an RAPD.[5] Furthermore, up to one-third of patients with open angle glaucoma will display an RAPD, which has led some to consider this presentation as a screening tool for glaucoma.[6] Ischemic optic neuropathies such as giant cell arteritis and optic nerve tumors such as gliomas are less common causes of

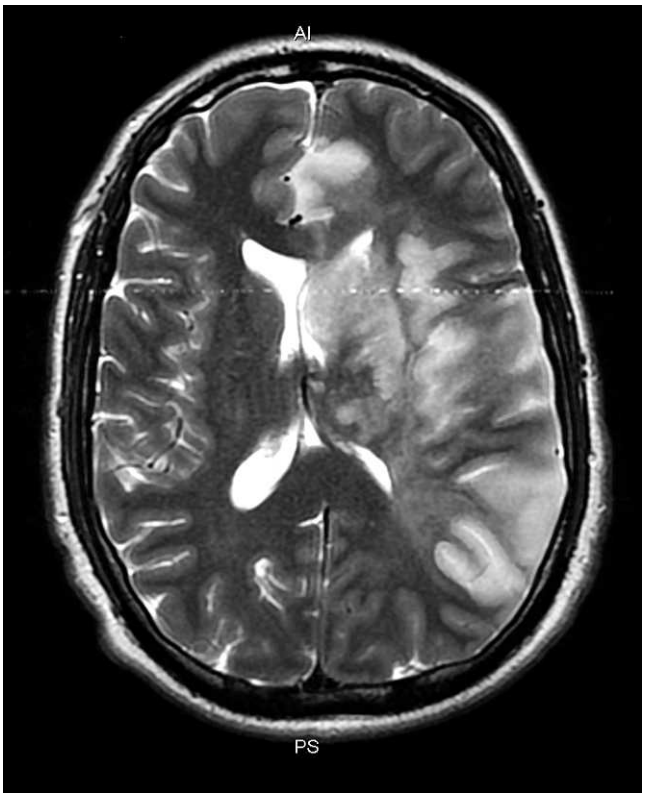

Figure 2. Magnetic resonance imaging fluid-attenuated inversion recovery showing a large area of restricted diffusion involving the left middle carotid artery and left anterior cerebral artery distributions, compatible with infarction. AI, anterior; PS, posterior.

RAPDs. Damage to the retina is a considerably less common cause of an afferent pupillary defect. Such damage is usually caused through ischemia, although severe macular degeneration, intraocular tumor, or retinal infection can also be the cause.

CONCLUSIONS

Because ophthalmic signs and symptoms are common presentations for dissections, especially among younger adults, clinicians should perform thorough ophthalmic examinations on patients with suspected dissections because early intervention, namely anticoagulation therapy, may help prevent the development of a thrombosis in the dissected vessel.[7] Physicians should furthermore include carotid dissection on their differential diagnosis when identifying new-onset RAPD, especially if it is associated with other ophthalmologic phenomena.

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