Illustrative Teaching Case

Young Patient With Headache and Amaurosis Fugax

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Case Description
A 48-year-old male farmer with a past medical history of non-specific headache experienced acute onset of throbbing headache centered over his forehead. About an hour later he acutely lost sight in his right eye, lasting for ≈5 minutes. When the severe headache continued, the patient’s wife called an ambulance. In the ambulance, he received fentanyl for the headache but was otherwise normal. At the primary hospital an internist investigated the patient and the vision was found to be normal and no neurological deficit was detected. The patient received some more painkillers for presumed migraine. An hour later, the patient developed left-sided hemiparesis and he became slightly drowsy. Computed tomographic (CT) scan of the head revealed a hyperdense middle cerebral artery sign on the right. The patient was then transferred to a tertiary care center where a neurologist obtained a National Institutes of Health Stroke Scale of 11 (age wrong, left hand and foot drifted into the bed during 10/5 seconds, visual and tactile neglect, partial gaze deviation, sensory deficit in the left hand, and deficit in the left visual field). There were no contraindications for thrombolysis and the patient received the bolus of thrombolysis treatment 2 hours and 50 minutes from the development of hemiparesis. After the bolus administration, the patient underwent perfusion CT and CT angiography. The latter showed occlusion of the right middle cerebral artery corresponding to the hypodense sign on the original CT. In addition, dissection of the right internal carotid artery was detected. Findings on perfusion CT indicated penumbra within the middle cerebral artery territory. Follow-up CT scan the next day showed brain infarction of a size slightly smaller than the lesion seen in CT perfusion imaging. The patient was eventually discharged to the rehabilitation ward, where his 30-day modified Rankin Scale was 4. The patient has a visual and tactile neglect, anosognosia, and he walks with assistance. He continues with physiotherapy, occupational therapy, and neuropsychologic rehabilitation.

Discussion
Transitory blindness or blurred vision in 1 eye (amaurosis fugax) is a form of transient ischemic attack (TIA) localized within the eye. The symptom is caused by interruption of the ocular arterial circulation, usually lasting a few minutes. Classically amaurosis is caused by thromboembolism originating from an atherosclerotic plaque of the internal carotid artery; however, it can also be caused by embolization from the aorta, heart, local thrombosis of the vasculature of the optic nerve or the retina, or thromboembolism from carotid artery because of nonatherosclerotic causes including dissection. Amaurosis fugax is an emergency that requires a similar diagnostic workup as other TIAs.

Carotid artery dissection (CAD) is common cause of stroke in young adults (<50 years) and should be considered when previously healthy young individuals experience an ischemic stroke or TIA. Local signs and symptoms of CAD include head, facial, or neck pain, partial Horner syndrome (ptosis, miosis), pulsatile tinnitus, and cranial nerve palsies. The most common is head, facial, or neck pain occurring in 64% to 74% and is the initial symptom in 60%, and the only symptom in 2% to 5% of patients. In particular, headache is observed in 65% to 68%, facial pain in 34% to 53%, and neck pain in 9% to 26% of patients. Partial Horner syndrome is observed in 28% to 41%; the pupil and eyelids are innervated by the sympathetic pathway, postganglionic axons of which ascend as a plexus intimately associated with internal carotid artery.

It is partial because sudomotor fibers (sweating) follow the external carotid artery and are thus not affected in internal CAD. Cranial nerve palsies are reported in 8% to 16%, and cranial nerves IX to XII are most commonly affected; most commonly the hypoglossal nerve, which descends from the hypoglossal canal to the angle of mandible right next to the lateral side of extracranial part of internal carotid artery. Also, the facial nerve, ocular motor, and trigeminal nerves may be affected. About 75% of CADs lead to ischemic events making it imperative that they be recognized and treated as soon as possible. Ischemic events include cerebral infarction (80% to 84%), TIA (15% to 16%), amaurosis fugax (3%), ischemic optic neuropathy (4%), and retinal infarct (1%). We want to stress that CAD is not a contraindication for intravenous thrombolysis, which is as safe as thrombolysis of acute ischemic stroke because of other causes. Anticoagulation and antiplatelets are both used for prevention of recurrent stroke after CAD. There is currently no evidence for superiority of either therapy.

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Study (CADISS) is a randomized prospective multicenter study comparing antiplatelet therapy with anticoagulation for patients with carotid and vertebral artery dissection and it may provide some answers in the near future. Surgical and endovascular intervention have also been used in the acute management of the CAD. Surgical approaches use (1) ligation or clipping of the carotid artery, (2) thromboendarterectomy with patch angioplasty, or (3) extracranial to intracranial bypass. Endovascular techniques using percutaneous balloon angioplasty and placement of a self-expanding stent have largely replaced surgical interventions. However, no randomized trial has been completed to compare the treatment options and hence the indications, efficacy, and necessity of endovascular intervention remain unclear.

In contrast, migraine with aura is defined as a recurrent disorder manifesting in attacks of reversible focal neurological symptoms that usually develop gradually for 5 to 20 minutes and last for <60 minutes. Headache with the features of migraine usually follows the aura symptoms. Less commonly, headache may occur without aura or aura may be present without headache. Typical auras include visual, sensory, and language symptoms, which can be a mix of positive and negative phenomena. Symptoms usually, but not always, follow one another in succession beginning with visual, then sensory, and language symptoms (International Headache Society’s International Classification of Headache Disorders, 2nd edition [IHS ICHD-II]). In our case, the headache preceded the visual symptoms which is unusual for migraine and should alert the clinician to the possibility of other pathogenesis.

**TAKE-HOME POINTS**

- Migraine with aura follows a typical pattern where the aura is followed by headache. If the symptom order is reversed, other causes should be ruled out, particularly TIA.
- Throbbing or sharp headache (including neck and facial pain) is a typical symptom of carotid artery dissection. Although not specific, other signs of ischemia including transient blindness (amaurosis fugax) should alert the clinician to the possibility of carotid dissection.
- Other symptoms of carotid dissection include paresis, miosis, migraine-like symptoms (scintillating scotoma), and possibly arm/limb weakness. Less common symptoms include pulsatile tinnitus, neck swelling, and cranial nerve palsies, most commonly hypogeusia (decreased sensation of taste).
- CAD is not a contraindication for thrombolysis.
- Amaurosis fugax is an emergency and carotid pathology should be suspected and investigated urgently.

**Disclosures**

None.

**References**


Key Words: amaurosis fugax • carotid artery, internal, dissection • headache
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