A 47-year-old man presented to the emergency department with a drooping right eye. He also complained of a constant right-sided headache of 1 week's duration; the pain involved the temporal region. Another physician had diagnosed new-onset migraine and prescribed sumatriptan, which failed to alleviate the pain. The patient had no weakness, vomiting, or double vision. Both his father and his son had Marfan syndrome.

Analysis of cerebrospinal fluid from a lumbar puncture showed no white blood cells, 3 to 6 red blood cells per microliter, a glucose level of 55 mg/dL, and a protein level of 53 mg/dL. These findings ruled out subarachnoid hemorrhage. During the patient's hospital stay, a CT scan of the chest was negative for Pancoast tumor of the lung. An MRI scan/magnetic resonance angiogram of the brain showed caliber irregularity and slight course derangement of the high cervical, petrous portion of the right internal carotid artery. Long-segment atherosclerosis was considered unlikely because there was no other site of atherosclerotic change. Fibromuscular dysplasia was also considered. Carotid dissection was strongly suspected. An angiogram of the brain performed a month later showed a 2.5-cm-long segment of mild irregularity and focal ulceration in the middle right internal carotid artery, with no definite evidence of a focal arterial dissection in this area. Horner syndrome—a functional sympathectomy of the ipsilateral eye—is caused by injury or disruption of the neural plexus that runs from the sympathetic chain, past the apex of the lung, and up the carotid artery to the eye. Ptosis may be subtle. Miosis is more marked in dim light; it may be difficult to notice in bright light. Potential causes of Horner syndrome include carotid or vertebral artery dissection, aortic dissection, traumatic carotid injury, deep neck infections, cerebrovascular accident, cerebellar bleed, cluster headache, and Pancoast tumor of the lung. Initial symptoms of carotid dissection usually involve pain that affects one side of the neck, face, or head; the pain may start abruptly but usually the onset is gradual. Pulsatile tinnitus or a bruit is present in about 25% of affected patients. Early neurologic findings may involve the sympathetic plexus (Horner syndrome is present in about 50% of patients), cranial nerve XII, or cerebellar function. Eventually, transient ischemic attack or thrombotic stroke may occur. The mean time between onset of pain and onset of neurologic symptoms is 4 days. Carotid dissection is an important cause of stroke in young adults and accounts for up to 25% of cases. If carotid dissection is suspected, magnetic resonance angiography (MRA) with fat suppression is the "gold standard" test. If MRA is not available, alternative tests include carotid duplex ultrasonography and CT angiography (which is almost as sensitive as MRA). Treatment of suspected carotid dissection involves consultation with a neurologist and a neurosurgeon. In contrast to aortic dissection, thrombosis—not rupture—causes complications, and treatment with standard-dose heparin is started in suspicious cases, following a negative head CT scan. Anticoagulation is usually maintained for 3 months. Surgery is rarely required. This patient almost certainly had a carotid dissection related to subclinical Marfan syndrome. Although his imaging studies were nondiagnostic, they were highly suggestive of these disorders. He probably should have received anticoagulant therapy for 3 months. Nevertheless, he did well and did not have a stroke. His Horner syndrome resolved after 3 months, although he complained of occasional mild right temporal headaches.

(Case and photograph courtesy of D. Brady Pregerson, MD. Dr Pregerson is the author of 2 medical...
References:


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