Chapter 42

The Man with Neck Pain and Arm Numbness

Judit Áfra, MD, PhD

Case History

A 41-year-old man presented to the Neurology Department one morning accompanied by his wife. His previous medical history revealed he had slight hypertension treated for several weeks, and he was a smoker (20 cigarettes per day). He said that he had had a terrible headache the evening before. He was working at his desk, and to rest, he leaned back on his chair. He then felt numbness in his right arm and had the impression he could not type as fast. This symptom lasted 10 to 15 minutes, and then he had a sharp, strong pain behind his left eye. He had the feeling someone wanted to "push his eye from the inside." The pain radiated to the occipital region and the upper neck on the same side. His wife noted his left eye was somewhat smaller, the eyelid a bit swollen, and the left pupil was smaller than the right one. There was some tearing and redness of the left eye. He tried to lie down, but finally, he stood up and walked around. This pain lasted 2 hours, then stopped, and he went to bed. The next morning his wife noted his left pupil was still smaller, and he felt his right arm was numb and clumsy, but he had no more pain.

On neurologic examination, a left Horner's syndrome, a slight right-sided central facial palsy, and a weakness of the right upper extremity were found.

Questions on the Case

Please read the questions, try to answer them, and reflect on your answers before reading the author's discussion.

- What investigation should be done?
- What kind of primary headache could the patient have?
- What elements do not fit with this diagnosis in the case history?
- What kind of treatment would you suggest?
- Should hypertension and smoking be regarded as risk factors?

Case Discussion

Initially, this appeared to be a case of left-sided cluster headache attack in this patient. According to the 2004 classification of the International Headache Society, the left-sided, predominantly periorbital strong pain accompanied by ipsilateral conjunctival injection, lacrimation, miosis, ptosis, and eyelid edema with the duration of 2 hours would fulfill the diagnosis of a cluster attack. Age, sex, and the smoking history of the patient were further clinical data supporting the diagnosis. To make the diagnosis of cluster headache, five attacks are needed (criterion A) and history, physical, and neurologic examination should not suggest any other illness (criterion E). Reviewing the history of this patient, he said that before his headache started, he had transient numbness and clumsiness in his right arm after leaning back on his chair. Moreover, on neurologic examination the day after his attack, a persistent Horner's syndrome on the side of the pain and a slight contralateral faciobrachial palsy were noted. Taken together, the criterion listed under E is not fulfilled, and thus the diagnosis of cluster headache cannot be made.

In light of the permanent focal neurologic symptoms, computed tomography was performed to exclude bleeding and/or ischemic lesions, and this was normal. A second, noninvasive test, duplex sonography of the carotid and vertebral arteries, was used and this showed dissec-

tion of the extracranial part of the left internal carotid artery with a significant stenosis. This diagnosis was confirmed by angiography.

Management Strategies

The therapy of arterial dissection is controversial. Most authors suggest anticoagulation for 3 to 6 months (or until complete recanalization), with or without antiplatelet therapy afterwards. No controlled, randomized trials have been performed so far. The rationale for anticoagulation is to prevent cerebral embolization from the dissected artery and development of a permanent occlusion. The main risk of this therapy is hemorrhagic transformation and progression of the dissection. Antiplatelet therapy could be an alternative as a first choice in patients who cannot receive anticoagulation. Traditional vascular surgery is not recommended in arterial dissection as a first-choice therapy. In some patients with recurrent symptoms and cerebral ischemia despite anticoagulation therapy, who are not suitable for endovascular treatment, it is still an option. In case of adequate collateral circulation, ligation of the internal carotid artery is possible. Endovascular treatment by stenting or percutaneous balloon angioplasty is another treatment option based on some clinical experience.

In our patient, anticoagulation therapy was started and continued for 3 months. Neurologic signs disappeared during the next 2 days, and headache did not recur. Three months later, magnetic resonance imaging (MRI) detected no infarction, and duplex sonography showed complete recanalization of the left internal carotid artery.

Case Summary

- The 41-year old man had a minor neck trauma (when leaning back on his chair).
- He had transient numbness and clumsiness of his right hand, followed by a headache mimicking a cluster attack. A Horner's syndrome on the left side persisted, and he developed a slight faciobrachial palsy on the right.
- The triad of unilateral head and neck pain with ipsilateral Horner's syndrome and ischemic signs are typical for carotid artery dissection. Minor neck trauma before the onset of symptoms also is often reported.
- Duplex sonography suggested dissection of the left carotid artery, which was later confirmed by angiography.
- In this case, 3-month anticoagulation therapy was administered, his symptoms and neurologic signs disappeared, and recanalization of the artery could be demonstrated.

Overview of Internal Carotid Artery Dissection

Epidemiology

Jentzer reported the first case of an internal carotid artery dissection in 1954, and dissection is now considered as one of the most frequent causes of ischemic stroke in young adults. The annual incidence is 2.6 in 100,000, and it was the cause of stroke in 20% of patients in a population younger than 30 years of age in a study by Bogusslavsky and Regli, but the incidence still seems to be underestimated.

Dissections of the internal carotid artery are more frequent than other vessels, and in 10% of cases, intracranial portions of arteries are involved. In 9 to 21%, dissection is bilateral, and affects multiple arteries in 28%.

Pathophysiologic Mechanisms

In the dissecting arteries, blood penetrates through the intimal layer into the outer layers of the media of the arterial wall. The subintimal hematoma may partly or completely occlude the arterial lumen. Dilation of the vessel wall may result in aneurysm formation and bleeding. Brain infarcts may be of embolic origin or may result from hemodynamic changes. Connective tissue disorders and arterial dissections show strong associations that may enhance the risk of arterial dissections in certain individuals. Among risk factors for stroke, hypertension and hypercholesterolemia were found more frequently in patients with arterial dissection. Preceding minor neck or head trauma is present in about one-third of patients. A positive family history was found in some studies in 2 to 18% of patients. Preexisting migraine was present in 26 to 49% of patients with arterial dissection in some studies, whereas others failed to demonstrate migraine comorbidity.

Symptoms and Signs

By far the most common clinical sign both in carotid and vertebral artery dissection is headache (68% and 69%, respectively). The pain is typically ipsilateral on the anterior head and is the first symptom in 47% of patients with internal carotid artery dissection. In vertebral artery dissection, headache is ipsilateral or bilateral, located in the posterior region of the head, and is a first symptom in 33% of cases. Neck pain is more frequent in vertebral artery dissection. According to Silbert and colleagues, the quality of pain is mainly pressing, aching, or sharp, in most cases constant, and not pulsating. Other signs of dissection are those of cerebral or retinal ischemia, including Horner's syndrome, ipsilateral pulsatile tinnitus, syncope, scalp tenderness, cranial nerve palsies (most commonly the XIIth), swelling of the neck, dysgeusia, retinal blindness, and symptoms of

orbital ischemia. The most characteristic triad of dissection is 1) the unilateral headache with 2) ipsilateral Horner's syndrome and 3) signs of focal cerebral ischemia.

Diagnostic Procedures

Among noninvasive studies, duplex sonography can detect stenosis or occlusion, double lumen if present. Transcranial Doppler is suitable for following changes of intracranial hemodynamics and detecting cerebral microemboli. Using MRI and magnetic resonance angiography (MRA), size and age of the hematoma in the vessel wall and double lumen can be shown. The most sensitive test for dissection is three-dimensional time-of-flight (3D TOF) MRA, which is now regarded as the imaging technique of choice in these cases. Typical angiographic signs are the irregular stenosis with a "string sign." Aneurysmal dilatation and embolization are less frequent, and Pozzati and colleagues found double lumen in only 4% of cases.

Prognosis

Unilateral dissection of the extracranial carotid artery has the best prognosis. When the intracranial part of the vessel is involved or the dissection is bilateral, the outcome is generally worse. Dissections of vertebral arteries have less favorable prognosis, especially if subarachnoid hemorrhage or bilateral involvement is present. Recurrence rate is usually low, under 5%.

Selected Readings

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Editorial Comments

There are few neurologic conditions that are more important to diagnose than cranial artery dissection, as the sequelae can be significant to the patient. Neck pain should be a significant clue and should not be overlooked if there are associated neurologic symptoms and signs. This review is comprehensive in its approach, and details the nuances of diagnosis and treatment. Therapy of dissection is controversial in the absence of high-level evidence as how to proceed in any individual case. In some cases, anticoagulation may be considered unless there are contraindications such as intracranial extension of the dissection (Schievink WI. Spontaneous dissection of the carotid and vertebral arteries. N Engl J Med 2001;344:898–906). Some stroke specialists treat spontaneous dissections with aspirin alone, in the absence of clear evidence.

Final diagnosis:

Arterial dissection