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Headache in Intracranial and Cervical Artery Dissections

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Abstract Dissection refers to a tear in the wall of an artery, with the two main types being intracranial or extracranial. Dissections tend to occur most commonly in the young, sometimes secondary to trauma involving the neck. To confirm a dissection, some type of vessel imaging is necessary, including magnetic resonance angiography (MRA), computed tomography angiography (CTA), or angiography. The most common presentation of a dissection (especially extracranial) is pain, usually head and neck pain along with a Horner's syndrome. Patients may also present with ischemic symptoms, including transient ischemic attack (TIA) or stroke, which may also be a complication of a dissection. Although headache is a common presentation, there is little research into phenotype or long-term outcomes. There are a number of case reports detailing the phenotypes of headaches that may be present in dissection, including a migraine-like or hemicrania-like headache. Dissections are usually treated with some type of anti-platelet or anti-coagulation, although there are only a few randomized controlled trials. In a new acute headache, dissection is an important diagnosis to keep in mind.

Keywords Dissection · Intracranial · Extracranial · Headache · Horner's · Tear · Carotid · Vertebral · Cervical

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Patient Presentation

A healthy 34-year-old man with no prior history of headaches comes into an urgent care center after developing a left-sided temporal headache with some extension behind his ear and into his neck. He reports playing flag football the day prior with one incidence of being pushed to the grass with possible whip lash injury. On exam, his left eye is constricted and does not react as well to light along with some drooping of his left eyelid. He is sent to the local emergency room where CT scan of the head on admission was unremarkable but magnetic resonance angiography of the neck (MRA) reveals a tapering of the left internal carotid artery, consistent with dissection. MRI shows no acute DWI changes. He is started on a low dose of aspirin and monitored closely. A follow-up MRA 6 months later revealed nearly complete resolution of the dissection. Over the next 6 months, he reports gradual improvement of his headache and his neurological exam is non-focal. A thorough workup for collagen vascular disease is unremarkable.

Introduction

Dissection, specifically vertebral dissection, was first described in the 1970s by C. Miller Fisher. Dissection refers to a tear in the wall of an artery, distinguished mainly by location, whether it is extra- vs intracranial. It is referred to as a cervical artery dissection when the tear occurs in the internal carotid or vertebral artery, outside of the skull [1]. When the primary dissection is located above the petrous bone, it is referred to as an intracranial dissection.

The most common presentation of a dissection (especially extracranial) is headache, usually head and neck pain along with a Horner's syndrome [2••]. Other common symptoms

include ischemic symptoms, including transient ischemic attack (TIA) or stroke. Dissections can be complicated by aneurysm formation, stroke, or subarachnoid hemorrhage. Although headache is a very common presenting feature, there is little research into the phenotype and long-term outcomes of the headaches associated with dissections. There is also little known about specific treatment for dissections, as well as the symptomatic treatment of headache specifically secondary to artery dissections.

Epidemiology

Dissection tends to be a disease of the young. Extracranial artery dissections occur mostly in the age range of 35–50 and are rare after age 65, with a slight male predominance, slightly more than half occur in males, across different studies. [1] The annual incidence for spontaneous cervical dissections is about 3–5 cases per 100,000 persons [3]. There is less information about intracranial dissection, since they are thought to occur less often [4]. Extracranial dissections can often be an overlooked cause of stroke, although in recent studies, it has been found to account for up to 2 % of ischemic strokes. In younger patients, those under the age of 55, these rates are much higher, with some reports indicating that they are the cause in about 14–20 % of ischemic strokes in those younger than 50 [1, 5]. Extracranial dissections tend to occur more commonly in the anterior circulation, while the opposite is true for intracranial dissections; with the most common being in the intradural vertebral artery [6]. Intracranial dissections are thought to be much less common although incidence data is not available. One series found a rate of 0.04 % of dissections out of all angiograms completed, accounting for about 10 % of all cervicocephalic dissections [7–9]. However, it is notable that patients presenting with dissections can have both intra- and extracranial locations.

Intracranial dissections tend to be complicated with either ischemic stroke (IS) and/or subarachnoid hemorrhage (SAH) [10]. Headache with ICAD prior to SAH or IS is common, occurring in about 80 % of patients, usually gradual in onset [11]. Isolated headache tends to be a rare finding, although this can be the case with extracranial dissections [4].

Headache Description

Patients with dissections typically present with headache along with other neurological signs and symptoms, including cranial nerve findings, including ipsilateral Horner's syndrome or tinnitus. Head or neck pain is commonly the first and "most frequent" symptom in cervicocephalic arterial dissection [12•].

The headache in dissections can be helpful in determining the location of the dissection, although there can be much variation. In extracranial dissections, carotid dissections are thought to cause ipsilateral frontotemporal headaches while vertebrobasilar dissections are typically more associated with neck pain in addition to a posterior headache [9, 13]. Neck pain occurs in about a fourth of internal carotid artery (ICA) dissections [13]. Dissection is also in the differential for a "thunderclap headache," as some can present with an acute, severe headache, especially if there is associated subarachnoid hemorrhage.

There are case reports which describe the range of headache presentations for patients with extra- and intracranial dissections. Ashkenazi et al. described a 51-year-old woman who presented with a continuous side-locked headache with autonomic symptoms, including eye tearing and ptosis after a car accident. Her headache description was consistent with a phenotype of hemicrania continua that continued for a year after onset, with initially significant relief after starting indomethacin [14]. Another case report described a 26-year-old woman who presented with a migraine-like headache, unilateral throbbing pain with nausea, and photophobia, after cervical manipulation. She was found to have a vertebral artery dissection on CT angiography [15].

Other presentations of a dissection are determined by the location and what other structures are nearby. Horner's syndrome, with ptosis and miosis of the ipsilateral eye, is mostly commonly involved in carotid dissections as the pericarotid sympathetic fibers are affected. It can also occur with vertebrobasilar dissections if there is brainstem ischemia [16]. Vertigo and nausea are frequently present with dissections in the posterior circulation [17]. A group in Japan described a case of a patient who presented with trigeminal neuralgia-like facial pain with an intracranial vertebral artery dissection [18].

In addition to head/neck pain and a Horner's syndrome, the third most common presentation is cerebral ischemia, including transient ischemic attacks (TIA) or ischemic stroke. This occurs in up to two thirds of patients with cervical artery dissections [6]. Symptoms are consistent with the area of the ischemia, including vertebral artery dissections that can present with brainstem signs and symptoms. Less common presentations include transient monocular visual loss or ischemic optic neuropathy, as well as retinal infarctions [19, 20].

Evaluation

To confirm a dissection, some type of vessel imaging is necessary. Digital subtraction angiography is considered the gold standard, although MRA and computed tomography angiography (CTA) are usually also acceptable alternatives [9]. Dissection on imaging is seen as an "eccentric, smooth or

irregularly tapered stenosis," sometimes referred to as a "flame-shaped occlusion" [21]. Magnetic resonance angiography (MRA) is very sensitive, in some studies up to 99 %, in detecting dissection, especially if it is done with a fat-suppression protocol. The most common finding on MRA is a "crescent-shaped intramural hematoma" on axial T1-weighted images, while fat-suppressed T1 images allow the intramural hematoma to be distinguished from the fat tissue around the vessel [22]. Computed tomography angiography (CTA) is also very sensitive in picking up a dissection, usually done if someone is unable to tolerate MRI. The typical finding is an "eccentric arterial lumen combined with mural thickening" [23].

Usually along with vessel imaging, MRI is done to evaluate for other possible causes or complication from a dissection, especially if there are signs or symptoms of ischemia. MRI is useful in looking for subarachnoid hemorrhage or infarcts that may be a result of the dissection.

Pathophysiology

Arterial dissections are due to a tear in the layers of the arterial wall, which is usually complicated by blood entering the wall leading to thrombus [9]. Dissections can occur from major or minor trauma, including whip lash or sports injuries. They are referred to as spontaneous when no preceding traumatic event is identified. Risk factors for dissection including high blood pressure and those with connective tissue disorders tend to have a higher rate of dissections, including but not limited to Ehlers-Danlos syndrome type IV, Marfan syndrome, polycystic kidney disease, and fibromuscular dysplasia [9].

In intracranial dissections, aneurysmal formation and extravascular hemorrhage tend to be more common than in extracranial dissections [16]. In intradural arteries, spontaneous dissections arise most commonly from the internal elastic lamina, while extracranial arteries can have tear in one or more of the possible layers in the vessel [9].

The headache or neck pain likely results from the direct tear in the blood vessel wall. Blood vessels, like arteries, are known to have many nerves surrounding them. Irritation of the wall likely leads to a cascade of events, including pro-inflammatory neurotransmitters from the nerve terminals that surround the blood vessel. This may cause pain away from the actual site of the dissection [24].

Complications

Ischemic events, including transient ischemic attacks and stroke are known and common complication of extracranial dissections. One study evaluated the time course of dissections and found that ischemic events were more likely to occur early in the course of the disease, as part of the initial presentation [25]. In the study of 80 extracranial ICA dissections, 88 % of those with strokes occurred in the first week of presentation [25]. This study did not find any differences in those who did have ischemic symptoms compared to those who did not as part of their dissection. In another study, dissections were found to be the cause in about 1.5 % of acute ischemic strokes or TIAs and 10.4 % of strokes/TIAs in the young [26]. The group found that vertebral artery dissections were the most common type followed by dissection in the internal carotid artery and almost two thirds of these were intracranial. It was found that artery to artery embolism was the most common cause of ischemic stroke, as a complication of the dissection [26]. Intracranial dissections are commonly associated with intramural hemorrhage, formation of aneurysms, and subarachnoid hemorrhage [27].

Treatment

There are limited trials for treatment of dissections. For the initial dissection, the treatment options include using anti-platelet therapy versus surgical treatment options to stabilize the dissections. The most important treatment acutely is focused on treating the dissection in order to prevent possible complications, including stroke or SAH. Although there has been controversy over the exact treatment, it centers around preventing thromboemboli from the area of dissection [9]. Studies using Dopplers suggest that thrombus formation with emboli traveling distally is the most common cause for stroke after dissection [28]. Current treatment options include anti-platelet therapy, i.e., aspirin versus clopidogrel, either with or without a loading dose or anti-coagulation using warfarin or heparin. Due to the lack of sufficient trials, there is controversy whether one is superior to the other. Recently, a trial was reported that "compared the effectiveness of anti-platelet drugs with anticoagulant drugs for the prevention of recurrent stroke in patients with carotid and vertebral dissection," called the Cervical Artery Dissection in Stroke Study (CADISS) [29]. The randomized trial evaluated 250 patients with either carotid or vertebral dissections, most presenting with either stroke or TIA symptoms, who received either an anti-platelet including aspirin, dipyridamole, or clopidogrel alone or in combination versus heparin followed by warfarin. At presentation, 67 % of patients had headache and half had neck pain. Overall, there was "no difference in efficacy" between the two groups in preventing stroke [29].

A 2010 systematic review for stenting of extracranial carotid and vertebral artery dissections showed high rate of success and low complication rates. The authors reviewed 140 cases of stenting with low rates of new neurological events [30]. An updated review in 2013 of only internal carotid artery dissections treated with stenting revealed similar rates of

success and complications [31]. Intracranial dissections, which can often have subarachnoid hemorrhage (SAH) as a complication, are more likely to require endovascular procedures since they are prone to re-bleeding [32]. However, most patients with intracranial dissections and no evidence of SAH are initially treated medically. Endovascular treatments are initiated if there are recurring or worsening ischemic symptoms or complications like SAH [1]. In most cases, follow-up imaging, usually with a less invasive technique, is done to assure that the dissection has healed. This is usually done 6 months or so after the initial event.

Prognosis

A study published in *Stroke* which studied almost 100 cases of intracranial artery dissections (IADs) showed that IADs not associated with aneurysms had a better prognosis [33]. Dissections with aneurysms tend to be characterized by SAH and poorer prognosis [33]. Another study revealed a recurrence rate of about 1 % during at least 1 year of follow-up. When there were recurrences, patients presented mostly with ischemic symptoms [34]. Overall, it is thought that the prognosis for neurological outcome is good to excellent in more than three fourths of cases [9], with low mortality. Overtime, the dissection should heal and the wall will repair itself, usually over the course of 3–6 months [35], with low recurrence rates.

In terms of the headache, there are only a few cases that describe the long-term outcomes for headaches after a dissection. A 20-year retrospective study on the nature history of carotid dissections found that 70 % of the 20 patients followed up had resolution of all their symptoms; about half of patient presented with either facial or head pain [36••]. A few case studies report long-standing headaches after dissections, one resolved after treating with steroids [12•].

Conclusion

Dissection is an important differential to keep in mind for someone who presents with ischemic symptoms. This is especially true for patients who are younger or have had recent neck trauma or manipulation. Although there is little evidence regarding the best type of treatment, most patients have a favorable prognosis. However, dissection can have complications, including subarachnoid hemorrhage or stroke. Therefore, it is important to monitor these patients closely, especially if they are having evolving symptoms. Long-term headaches may be a complication although most studies indicate that dissection is usually a one-time occurrence.

Compliance with Ethical Standards

Conflict of Interest Huma Sheikh declares personal fees from Neurology Advisor for submission articles unrelated to this article.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by the author.

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