



Spontaneous Vertebral Artery Dissection

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The patient was a 29-year-old woman with a history of migraine headaches who has otherwise been in good health. She stated that earlier today she was walking in her house and talking on the phone when suddenly she started staggering to her right and fell onto her couch. She stated that when she was lying on the couch and tried to focus on things, the room was spinning and she could not make her eyes focus. She had no nausea, diaphoresis, or vomiting. She noticed no other complaints or deficits. After approximately one to two minutes, the feeling of spinning went away and the patient stated that she has had no recurrence. The only other thing she notes is that since the episode, she has a pain going up the left side of her neck and into the left side of her head. She states that this is not severe. She says that her migraines are usually in the frontal area or behind either eye. She has never had a migraine with similar symptoms in the past.

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Introduction

Past medical history: significant for migraine headaches

Medications: oral contraceptives and Imitrex prn.

Social history: patient doesn't smoke cigarettes or drink alcohol, she is a housewife and mother of two children. Family history is unremarkable.

Physical exam: BP: 137/88 Pulse: 80 Respiratory rate: 14

The pupils were equally round and reactive. Extraocular movements were intact. The neck was supple and non-tender to palpation and she had no pain on range of motion. There were no bruits auscultated in the neck. Cardiac examination was normal with regular rate and rhythm and no murmurs. Neurological examination showed no focal deficits. Sensation was intact and deep tendon reflexes were 2+ and equal bilaterally. Toes were down going and the gait was normal. Heel-toe walking was normal. There was no dysmetria.

Medical decision making: With the history of an unexplained episode of vertigo and left neck pain it was felt that vertebral artery dissection should be ruled out. The patient was given one aspirin while in the emergency department and magnetic resonance angiography was arranged. Magnetic resonance angiography showed a diminutive left vertebral artery with a focal area of absent/decreased flow and abnormal signal hyperintensity from the C1-2 through the C5-6 levels. These findings likely represent dissection of a short segment of vertebral artery.

Etiology

Extracranial cervical cerebral arterial dissections occur when there is a tear in the intima of the blood vessel, allowing blood to dissect in the wall of the artery. Once dissection occurs, neurological sequelae may result from the dissection in one of two ways:

1. The extramural hematoma may compromise the lumen of the blood vessel reducing flow to the affected part of the brain.
2. There may be thrombus formation at the site of the dissection which can either locally occlude the blood vessel or cause an artery to artery embolus to a more distal intracerebral artery.

Cervical arterial dissections occur in all age groups with a peak incidence in the fifth decade. It has been estimated that as many as 2.5% of first strokes may be as a result of artery dissection. The exact incidence, of course, is unknown since most patients presenting with stroke do not have arteriography as part of their initial workup. Although dissections may occur spontaneously as in the patient described, dissections are

frequently associated with trauma and the presence of an arteriopathy. The arteriopathies most often associated with cervical artery dissection are Marfan's Syndrome, Ehlers-Danlos IV, and fibromuscular dysplasia. However, most persons with arterial dissections will not have any associated arteriopathy. Although trauma is frequently associated with the presence of dissection, the history of trauma may be quite trivial. Patients have been reported to have arterial dissections occurring from minimal trauma such as turning the head, looking upward, severe straining or coughing, or chiropractic manipulation. Carotid arterial dissection has been reported with airbag deployment in motor vehicle crashes as well.

Other associations with arterial dissection include migraine and recent respiratory infections. In patients with arterial dissection, the incidence of migraine is reported to be as high as 25-50%. If patients do experience a headache with the dissection, it is typically not described as being migraine-like. There has also been a recently reported association of increased incidence of respiratory infection preceding respiratory infection with development of arterial dissection. It is surmised that systemic inflammatory response associated with infection may contribute to the occurrence of dissection.

Dissections can occur in either the carotid artery or the vertebral artery. The typical location for a dissection in either artery is at the C1-2 level. Although there is a slight predilection for females in vertebral artery dissection, carotid artery dissections occur equally in males and females. Patients with either carotid or vertebral artery dissection may have an underlying arteriopathy.

ED Diagnosis

The major presenting features will vary somewhat with the artery involved. The most common presenting feature with either vertebral or carotid artery dissection is stroke or transient ischemic attack associated with pain in the ipsilateral neck, face or head. Neck pain is more common with vertebral artery dissection occurring in approximately 46% as opposed to 26% with carotid dissection. Headache occurs equally in about 70% of patients. It has been reported that the development of symptoms may be delayed for days after the onset of carotid dissection vs. less than one day in patients with vertebral dissection.

In carotid dissection, an ipsilateral Horner syndrome may occur in 40% of patients. The head pain associated with carotid dissection is frequently retro-orbital, temporal or elsewhere on the ipsilateral side of the face. Pulsatile tinnitus has also been reported with carotid dissection. Various cranial nerve palsies may present with carotid dissection including palsies of cranial nerve 2, 4, 5, 6, 7, 9, 10 and 12. Amaurosis fugax may also be a presenting complaint for carotid dissection.

Patients with vertebral artery dissection typically will present with strokes in the posterior circulation and will often have bilateral neurological findings. Neck pain as mentioned is more frequently associated with vertebral artery dissection.

ED Management

The key to diagnosing arterial dissection is suspecting the disease. Evaluation should be undertaken in any patient who presents with neurologic signs or symptoms of stroke/TIA with associated neck pain or headache. Patients with a history of arteriopathy (Marfan's, Ehlers-Danlos, etc.) should be highly suspect. Also, a history of neck trauma, even relatively trivial, should raise the index of suspicion.

The best non-invasive test for diagnosing arterial dissection is magnetic resonance angiography. If the diagnosis is suspect on MRA/MRI, angiography may be needed to confirm the diagnosis. In many cases MRI/MRA alone is sufficient to establish the diagnosis. If MRA/MRI or angiography is not immediately available, carotid duplex scanning may be useful in diagnosing a carotid artery dissection. Duplex scanning may be valuable in diagnosing vertebral artery dissection with experienced operators. In patients who present with ischemic stroke or TIA symptoms, one should obtain non-contrast cranial CT as the initial test.

There are no randomized controlled trials of the management of extracranial artery dissection. Because the symptoms or sequelae of dissection are related to thrombosis and/or embolus, anticoagulation is the main stay for treatment. A patient with a documented vertebral or carotid artery dissection should be given intravenous heparin and then switched to warfarin anticoagulation. Some physicians might opt for anti-platelet therapy alone which may be quite adequate. As mentioned, there are no randomized-controlled trials to establish the superiority of one treatment over another.

Re-imaging is usually done in three to six months. After the artery appears normal on MRA or angiography, anticoagulation or anti-platelet therapy can be discontinued. There is an incidence of recurrent dissection which is approximately 3% for carotids and 5% for vertebral artery dissections.

In patients with acute stroke secondary to extracranial arterial dissection, intra-arterial or intravenous thrombolysis have been used with good results. While this has not been extensively studied, there is no evidence that patients with stroke resulting from extracranial carotid or vertebral artery dissections should have thrombolytic therapy withheld if they otherwise meet other criteria for IV thrombolysis.

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Case Outcome

The patient was admitted to the Neurology inpatient service and started on intravenous heparin. On hospital day two, the patient was begun on Coumadin to achieve an INR of 2-3. This was achieved after several days and the patient was discharged from the hospital. As an outpatient she underwent evaluation by Medical Genetics for Ehler-Danlos Type 4 syndrome, which was negative. The Medical Genetics service felt that there was no indication that the patient had any underlying arteriopathy or connective tissue disorder. Six months later, the patient had repeat magnetic resonance angiography which showed normalization of the MR angiographic appearance of the left vertebral artery since the prior examination. The MR was read as normal. The patient was discontinued from the Coumadin and has remained well.

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Annotated Bibliography

1. **Silbert et al: “Headache and Neck Pain in Spontaneous Carotid and Vertebral Artery Dissections,” Neurology 45:1517-1522, 1995.**

Documents signs and symptoms in 161 patients with dissection.

2. **Biousse et al: “Head Pain in Non-Traumatic Carotid Artery Dissection,” Cephalgia 14:33-36, 1994.**

Discusses presenting signs and symptoms of carotid artery dissection.

3. **Schievink et al: “Heritable Connective Tissue Disorders in Cervical Artery Dissections,” Neurology 50:1166-1169, 1998.**

Documents that connective tissue disorders are common in dissection patients although they don't meet the classic criteria for diagnosis.

4. **Jeret JS: “Stroke following chiropractic manipulation: Report of 3 cases and a review of the literature” J. Neuroimaging 10:52, 2000.**

This is an abstract describing 3 cases of dissection associated with manipulation of the neck.

5. **Schievink W: “Spontaneous dissection of the Carotid and Vertebral arteries” NEJM 344(12): 898-906, 2001.**

This is a great review of the etiology, presentations and treatment of patients with arterial dissection. If you're going to read one article, this is it.

6. **Lyrer P et al: “Antithrombotic drugs for Carotid Artery dissection” Cochrane Database of Systematic Reviews (4): CD000255, 2001.**

This is a review of all the articles describing treatment for dissections. They conclude that there is not even good data to support the use of antithrombotic treatment as no randomized trials have been done. There is no evidence to support the use of heparin(s) over antiplatelet agents.

7. **Saeed AB et al: Vertebral Artery dissection: Warning symptoms, clinical features and prognosis in 26 patients” Canadian Journal of Neurological Sciences 27(4): 292-296, 2000.**

This article summarizes the findings in 26 patients with vertebral dissections over a 10 year period.

8. **Sampagnaro G et al: “Intra-arterial thrombolysis in a patient presenting with ischemic stroke due to spontaneous internal carotid artery dissection” *Catheterization and Cardiovascular Interventions* 48(3): 312-315, 1999.**

This article describes intra-arterial thrombolysis in a patient with an evolving stroke from carotid dissection.

9. **Wityk: “Stroke in a Healthy 46 year old man,” *JAMA* 285(21):2757-2762, 2001.**

Case presentation of spontaneous carotid dissection with a literature review.

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Questions

1. The most appropriate acute treatment for extracranial artery dissection is:

- a. Angioplasty and stenting
- b. Endarterectomy
- c. IV Heparin
- d. Arterial ligation
- e. Beta blockers and antihypertensives

2. Which of the following is most often associated with arterial dissections?

- a. Atherosclerosis
- b. History of MI
- c. Prior intracranial hemorrhage
- d. Migraine
- e. Aortic dissection

3. Which of the following is NOT a common presenting complaint with dissection?

- a. Neck pain
- b. Facial pain
- c. Headache
- d. Nuchal rigidity
- e. Neurological deficit

4. The diagnostic test of choice for diagnosis of arterial dissection is:

- a. Contrast angiography
- b. Magnetic resonance angiography
- c. Duplex ultrasound
- d. CT
- e. PET imaging

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Answers

1. Answer c.

IV heparin.

Because the dissection is between the media and the muscular layer of the vessel, rupture is highly unlikely. The morbidity from dissection is typically a result of thrombosis or embolus and heparin can prevent these from occurring.

2. Answer d.

Migraine.

Patients with migraine are often the victims of arterial dissection for unknown reasons. Migraine has been reported in up to 50% of patients with dissection. The other disorders have little or no association with dissection.

3. Answer d.

Nuchal rigidity

Neck pain, facial pain, headache and stroke syndromes are the common presenting complaints in patients with dissection. Nuchal rigidity is not present as there is no associated meningeal irritation with extracranial artery dissection.

4. Answer b.

Magnetic resonance angiography

While contrast angiography is quite accurate for dissection, it is invasive and carries a higher morbidity than MRA. Duplex ultrasound is acceptable for carotid dissections but will not have the same reliability as MRA, and the absence of dissection on carotid duplex does not rule out dissection. CT and PET imaging are not appropriate or accurate studies for assessing dissection.