# What's Your Diagnosis?

## **Doctor!** I Can't Stand The Noise in my Ear!

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A 46-year-old lady presented with a history of troubling whistling sound in her left ear for the past week. She noted the sound increased with bending. The sound has become more noticeable and was interfering with her sleep. There was no nausea. She had no ataxia.

She had some neck pain six weeks earlier and went for chiropractic therapy. She did not have any acute neck pain following the visit to the chiropractor.



Figure 1. Left carotid oblique view.

## What's your diagnosis?

### **Patient Stats**

- Well and comfortable.
- Machinery type cranial bruit more prominent over the left mastoid.
- No Horner syndrome.
- Ear examination normal.
- No signs of vestibular dysfunction.
- No lower cranial nerve signs.
- Remainder of neurologic exam normal.
- Blood pressure was 120/180 in the right arm sitting.
- Pulse rate was 88.

### **Medical History**

- Thyroidectomy a number of years ago.
- Tuberculin reactor.
- Taking replacement thyroid.
- Had a tubal ligation.
- Noted in 1992 to have mildly elevated cholesterol.
- Three pregnancies.
- Non-smoker.
- No known drug allergies.
- Her father had his first of four strokes at the age of 60.
- Her mother, in her early 60s, is having some transient ischemic attacks (TIAs).
- Second sibling of four; all the other siblings are in good health.

### **Clinical Investigations**

- The carotid ultrasound study did not reveal any definite abnormality, but suggested dual anteriovenous malformations (AVM) on the left side.
- The carotid angiogram showed a significantly long dissection of the left internal carotid artery as it entered the skull, with the lumen ranging from 70% to 80% compromised.
- At the proximal point, there was a small ulcer.
- There was occlusion of the left anterior cerebral artery distal to its first segment; this was likely due to embolism from the dissected part of the artery. Fortunately, there was a collateral supply from the right side to the left side.
- The other vessels looked normal (Figures 1 and 2).
- Blood investigations, including complete blood cell count, electrolytes, glucose and coagulation profile, were normal.

### Answer: Long Dissection of the Left Internal Carotid Artery Due to Chiropractic Neck Manipulation

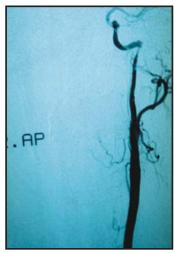


Figure 2. Carotid angiogram-left carotid anteroposterior view.

Given that her vessels were otherwise normal, and the lack of other risk factors for dissection, namely arteritis, the cause is most likely chiropractic neck manipulation, which she had six weeks earlier.

# What is the treatment?

She was anticoagulated with both heparin and warfarin. Her international normalised rate was monitored closely, and it was advised that the rate be maintained between 2 and 3. She will be anticoagulated for approximately six weeks. A carotid ultrasound study will be repeated, and if

more than 30% improvement is noted in the narrowed carotid artery, anticoagulation will be discontinued, or else, may continue for another month further or so. The patient has been advised about strenuous activity and sudden neck movements.

In a recent Canadian survey, dissection of the cervical arteries was one of the most common cause of stroke in patients under 45 years of age.<sup>1</sup>

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Dissections can occur in either the carotid artery or the vertebral artery. The typical location for dissection in either artery is the C1-2 level.<sup>2,3</sup> Although there is a slight predilection for females in vertebral artery dissection, carotid artery dissection occurs equally in males and females. Patients with either carotid or vertebral artery dissection may have an underlying arteriopathy.<sup>3</sup>

Cervical arterial dissections occur in all age groups with a peak incidence in the fifth decade.

## What's Your Diagnosis ?

#### Table 1

#### Neurologic Sequelae Resulting From the Dissection

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, neurologic 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 embolus to a more distal intracerebral artery.

It has been estimated that as many as 2.5% of first strokes may be a result of artery dissection. The exact incidence, of course is unknown as most patients presenting with stroke do not have arteriography as part of their initial workup. Although dissections may occur spontaneously, they 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 syndrome, and fibromuscular dysplasia. Most persons with arterial dissections, however, will not have any associated arteri-

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



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### What are the symptoms?

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.<sup>1,3</sup> Neck pain is more common with vertebral artery dissection, accruing in approximately 46% of patients as opposed to 26% of patients 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, as opposed to patients

The best non-invasive test for diagnosing arterial dissection is magnetic resonance angiography (MRA). with vertebral dissection where symptoms appear in less than one day.<sup>1,2,3</sup>

In carotid dissection, 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, 3, 4, 5, 6, 7, 9, 10 and 12.<sup>1-3</sup>

Patients with vertebral artery dissection will normally present with strokes in the posterior circulation and will often have bilateral neurologic findings (Table 1).

# How do I manage arterial dissection?

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 or transient ischemic attack with associated neck pains or headaches. Patients with a history of arteriopathy (Marfan's, Ehlers-Danlos, *etc.*) should be highly suspected. A history of neck trauma, even if relatively trivial, should also raise the index of suspicion.

The best non-invasive test for diagnosing arterial dissection is magnetic resonance angiography (MRA). If the diagnosis is suspect on MRA/magnetic resonance imaging (MRI), angiography may be needed to confirm the diagnosis.

## What's Your Diagnosis ?

In many cases MRI/MRA alone is sufficient to establish the diagnosis. If MRI/MRA or angiography is not immediately available, carotid duplex scanning may be useful in diagnosing vertebral artery dissection with experienced operators. In patients who present with ischemic stroke or transient ischemic attack symptoms, a non-contrast cranial computed tomography scan should be obtained as the initial test.<sup>3</sup> The symptoms or sequelae of dissection are related to thrombosis and/or embolus, therefore, anticoagulation is the mainstay for treatment. A patient with 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. There are no randomised, 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. The incidence of recurrent dissection is approximately 3% for carotids and 5% for vertebral artery dissections.<sup>3</sup>  $D_{k}$ 

#### References

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