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The EM Educator Series

The EM Educator Series: Cervical Vessel Dissection

Author: Alex Koyfman, MD (@EMHighAK) // Edited by: Brit Long, MD (@long_brit) and Manpreet Singh, MD (@MprizzleER)

Case 1: A 32-year-old male presents with headache and neck pain. This is his second visit in the last week. 10 days ago he was in a motor vehicle accident and had a negative non contrast head CT and C spine CT at that time.

Case 2: A 38-year-old female presents with anterior neck pain. You notice ptosis and miosis on exam, but no anhydrosis.

Case 3: A 31-year-old female presents with focal motor weakness in the left arm and leg. Her husband states she was complaining of a headache and neck pain several days ago. A "Code Stroke" is activated.

Questions for Learners:

1. What's the pathophysiology behind cervical vessel dissection?
2. Who is at risk for the condition?
3. How can patient's present, and what should be on your differential?
4. What is the ED evaluation?
5. What is the ED management?
6. What are the complications if the condition is left untreated?

Suggested Resources:

- Articles
  - emDOCs – Cervical Artery Dissection: The Elusive Diagnosis
  - Emergency Medicine Cases
  - EM@3AM
- Journal Articles
  - Emergency Medicine Clinics of NA
  - JEM – Thunderclap Headache

Answers for Learners:

1. What's the pathophysiology behind cervical vessel dissection?

The arterial wall is made of the intima layer, media layer and adventitia layer. The vaso vasorum are a set of blood vessels within the middle media layer that feeds the vessel. Cervical artery dissections are created by a tear in one of the walls leading to separation between these layers. One mechanism proposed suggests a rupture in the vaso vasorum of the media (middle muscular layer of the artery) leading to the creation of an intramural hematoma and a false lumen. Another mechanism proposed is the tearing of the intimal layer leading to the creation of an intramural hematoma after blood is forced in between the arterial wall layers by strong arterial pressures. In some postmortem and surgical specimens analyzed, there were no communications noted between the layers suggesting that some dissections may also be caused by primary intramural hematomas. The exact pathogenesis is not fully understood.

Patients presenting with cervical artery dissection can also develop brain ischemia leading to transient ischemic attacks or cerebrovascular accidents. The ischemia can develop from emboli formation and/or narrowing of the arterial lumen. In one retrospective study, the authors used brain imaging to examine the causes of ischemic stroke (hemodynamic vs thromboembolic etiologies) in 141 patients. They concluded that the majority of patients developed ischemic strokes due to a thrombo-embolic mechanism based on the pattern of infarct on imaging.

2. Who is at risk for the condition?

Risk factors include trauma (chiropractic manipulation of the neck, coughing, sneezing, vomiting, yoga, recent motor vehicle collision, any mechanism leading to sudden hyperextension and/or rotation of neck), genetic pre-disposition (fibromuscular dysplasia, Marfan's syndrome, osteogenesis imperfecta type 1, autosomal dominant polycystic kidney disease, Ehlers-Danlos syndrome), recent infection and migraines.

3. How can patient's present, and what should be on your differential?

Cervical artery dissections can have different clinical pictures depending on where the dissection occurs. Common presentations include headache, neck pain, pulsatile tinnitus, cranial neuropathies and partial Horner's syndrome (ipsilateral miosis, ptosis). Anhidrosis does not occur because the fibers involved in facial sweating runs along the external carotid artery. Pulsatile tinnitus has been reported in about 16-27% of patients. Headache has been reported in up to 65-68% of patients presenting with dissection. Patients can present with sharp headache, severe non-throbbing bilateral frontal headache and/or thunderclap headache. Cranial nerve palsies were reported in around 8-16% of patients. Some patients are also known to present with signs of ischemic stroke including visual changes, weakness of face or extremity muscles and/or numbness in these muscle groups. In Arnold et al., a multi-center prospective cohort study analyzing 169 patients at three tertiary care centers (Paris, Zurich, Berne) who were found to have vertebral artery dissections, 77% of the patients showed signs of brain ischemia (predominantly posterior circulation). 114 patients had signs of ischemic stroke and 17 patients had transient ischemic attacks. Surprisingly, 13 patients (8%) in the study were asymptomatic and there was 1 case where the patient only had cervical radiculopathy as a presenting symptom.

The CADISP study found that patients with ICAD presented more often with a headache when compared with VAD (OR 1.36 [1.01-1.84]). The presence of cerebral ischemia (OR 0.32 [0.21-0.49]) and cervical pain (OR 0.36 [0.27-0.48] was less frequently seen in patients with ICAD when compared to VAD. [16] In Arnold et al, the authors noted that women in the study more likely presented with tinnitus (16 vs 8%; $p = 0.001$) and migraines (47 vs 20%; $p < 0.0001$).

In one case series, pain (head and/or neck) was noted to be the only presenting symptom even in patients who were found to have multiple dissected arteries. 20 out of the 245 patients (8%) presented with pain as their only initial symptom. The pain included headache and neck pain (different onsets, quality and locality of pain for all of the patients) Of these 20 patients, 5 had multiple dissections, 12 had VA dissections, and 3 had ICA dissections. In one case report, a patient with vertebral artery dissection presenting with radiculopathic pain in the C5 region.

→ Carotid artery dissection presents as thunderclap headache or subacute headache with unilateral facial, neck or head pain with a partial Horner's syndrome (myosis and ptosis, but not anhydrosis), and 1/3rd of patients will have retinal or cerebral TIA within one week (neurological symptoms lag behind because it takes time to have a thrombus formed and thrown from the site of dissection)

→ Vertebral artery dissection presents with posterior neck or occiput pain and posterior circulation symptoms – ataxia, vertigo, dysarthria, diplopia and dysphagia

4. What is the ED evaluation?

Although conventional angiography is the gold standard, both CTA and MRA can be used for the diagnosis of CAD. Consider using CTA over MRA if concern for vertebral artery dissection.

If your patient has concerning findings for CAD with negative CTA and/or MRA results, DO NOT stop your work-up. Proceed to conventional angiography (gold standard).

5. What is the ED management?

Treatment includes antiplatelet or anticoagulation therapy, except in the presence of large infarct with mass effect, hemorrhagic transformation of an infarct, or intracranial extension of the dissection, but consultants should weigh in before treatment is initiated.

- Consult vascular surgery to discuss management:
    - Patients with extra-cranial carotid artery dissection, evidence of acute ischemic stroke, and symptom onset < 4.5 hours prior to arrival => consider thrombolysis if no contraindications.
        - Note: thrombolysis carries the potential risks of extending the intramural hematoma, dislocating an intraluminal thrombus with subsequent embolization, development of a subarachnoid hemorrhage due to leakage, or formation of a pseudoaneurysm.
    - Patients with extra-cranial carotid artery dissection and evidence of ischemic stroke, presenting > 4.5 hours following symptom onset or thrombolysis contraindicated => antithrombotic therapy.

        - A 2012 randomized controlled trial (CADISS – Cervical Artery Dissection In Stroke Study) demonstrated no evidence of superiority in treatment with anticoagulation vs. antiplatelet therapy.
        - Anticoagulation is preferred if severe stenosis, vessel occlusion, or pseudoaneurysm are present (assumed that anticoagulation more effectively prevents thromboembolic complications).
        - Antiplatelet therapy is preferred if the patient is likely to have a poor prognosis (large infarct) or in individuals with a medical history pre-disposing to increased risk of bleeding (blood dycrasias, etc.).
    - Patients with intra-cranial extension of the carotid artery dissection or dissection involving the aorta:
        - Thrombolysis is contraindicated (risk of SAH or aortic rupture) => requires antithrombotic therapy.
    - For all patients, endovascular or surgical repair may be utilized in the setting of severe stenosis, expanding dissecting aneurysm, or persistent ischemia despite antithrombotic therapy.

6. What are the complications if the condition is left untreated?

CAD increases the risk of thromboembolic events.