

Spontaneous Vertebral Artery Dissection with Thunderclap Headache: A Case Report and Review of the Literature

Yung-Chu Hsu, Sheng-Feng Sung

Abstract

Purpose: Vertebral artery dissection (VAD) is an important and under-recognized etiology of stroke in young patients. No clinical symptoms or signs appear to be specific for VAD. This report describes a representative patient and reviews the headache pattern and imaging findings commonly noted in VAD to help with the early diagnosis of VAD.

Case Report: A 44-year-old female presented with severe right posterior neck pain and vertigo followed closely by thunderclap headache that was confirmed as right VAD (V4 segment) with delayed right dorsal medullary infarction two days later. Her headache, vertigo, and truncal ataxia were completely improved one week later.

Conclusion: The most common neuroimaging findings of VAD were vertebral artery stenosis, followed by the string and pearls sign, arterial dilation, arterial occlusion, and the less common but most characteristic features of pseudoaneurysm formation, double lumen, and intimal flap. Pain in VAD mostly occurred in the ipsilateral posterior occipitocervical region, with throbbing features in 50-60% of the patients. Only one fifth of patients with VAD present with a thunderclap pattern. In stroke among young patients or stroke with pain in the head and neck, angiography study of the craniocervical artery is highly recommended.

Key Words: vertebral artery, cervical artery dissection, thunderclap headache, neck pain

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INTRODUCTION

Spontaneous cervical artery dissection (CAD), although rare with an incidence of only 3 per 100,000

per year⁽¹⁻³⁾, is a definite etiology for stroke among young patients and a potentially disabling disease. It accounts for 13-20% of stroke among young patients worldwide⁽⁴⁾ and 9.1% of stroke among young patients in Taiwan⁽⁵⁾.

CAD can be classified as either vertebral artery

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dissection (VAD) or internal carotid artery dissection (ICAD). The triad of ICAD symptoms are pain, focal sign, and delayed ischemic event, and the presence of any two components warrants clinical attention regarding this diagnosis⁽¹⁾. The classic presentations of VAD, which are less distinct than those in ICAD, are headache and neck pain followed by delayed ischemic event⁽¹⁻³⁾. As a result, early stage VAD is usually misinterpreted as musculoskeletal disease, especially in cases with isolated neck pain which worsens after spinal manipulation⁽⁶⁾. Despite the development of neurovascular imaging, VAD remains a clinically elusive diagnosis owing to the broad spectrum of clinical presentations. For example, some patients present with isolated headache or neck pain, and there have even been reports of asymptomatic patients with proven VAD^(1,2).

Headache and pain are the most common, initial, and even isolated symptoms in VAD. The knowledge and insight of pain symptoms facilitates the ability of physicians to make early and accurate diagnosis of VAD. We herein report a young woman presenting with thunderclap headache who was diagnosed with spontaneous VAD.

CASE REPORT

A 44-year-old woman presented with sudden severe right side nuchal pain and vertigo which developed while quietly sitting on a chair at midnight. The pain reached

the peak intensity in seconds first then radiated to the right occipital region 2-3 minutes later. The occipital pain, which also reached maximum severity in seconds, exhibited throbbing feature followed by nausea and vomiting. Before this event, she did not have any head motion. She denied having visual problems, tinnitus, facial pain, dysarthria, or focal limb weakness. She did not have past history of headache or vertigo and had not been massaged recently.

On examination, her initial blood pressure was 156/88 mmHg, and her pulse rate was 78 beats per minute. Her right occipitocervical region was exquisitely tender to palpation. Furthermore, she was quite uneasy in bed and displayed general discomfort. The other physical and neurological examinations were unremarkable. Fundoscopy showed normal results. The laboratory tests, including autoimmune tests, were all within normal limits. We gave her diphenidol, metoclopramide, intravenous non-steroid anti-inflammatory drugs and lorazepam. However, her general disposition did not improve during the first night.

On the second day, her headache and vertigo were much better, but her neck pain persisted. Her trunk was deviated to the right side while sitting, which was not observed initially. Repeated neurological examination did not show Horner's sign, dysarthria, tongue deviation, motor or sensory deficits. Finger-nose-finger and heel-knee-shin tests were all normal, but she was unable to stand up, hinting at truncal ataxia. Neck duplex showed

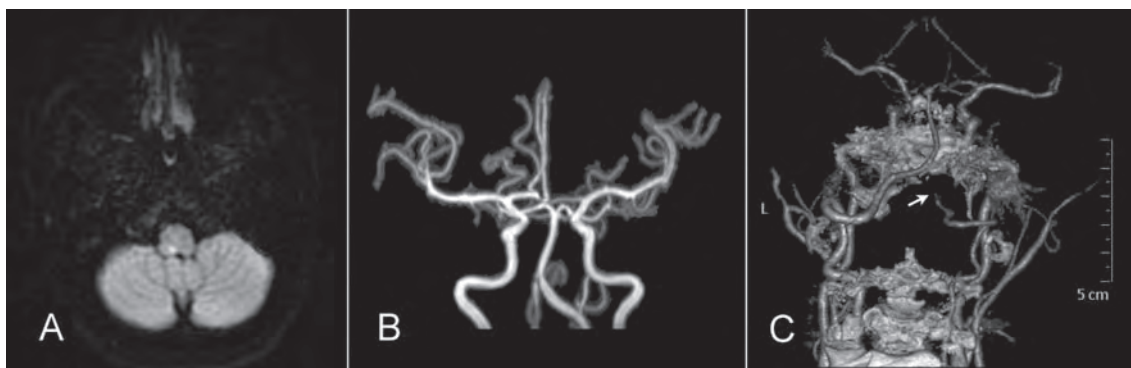


Figure 1. (A) Brain magnetic resonance imaging on the second day after pain revealed a faint hyperintensity in the right upper dorsal medulla on diffuse weighted imaging. (B) Brain magnetic resonance angiography showed normal intracranial vasculature. (C) Computed tomography angiography on the second day (rotated 180° to demonstrate the dissected vessel) disclosed a string sign (arrow) in V4 segment of the right vertebral artery.

relatively decreased flow in the right vertebral artery (peak systolic velocity: right 53 cm/sec, left 78 cm/sec; end diastolic velocity: right 12 cm/sec, left 32 cm/sec; flow: right 35 ml/min, left 188 ml/min, measured with Siemens Acuson™ Sequoia® C256). Brain magnetic resonance imaging (MRI) showed a faint hyperintensity in the right upper dorsal medulla on diffuse weighted imaging (Fig. 1A), and brain magnetic resonance angiography (MRA) revealed normal intracranial vasculature (Fig. 1B). Further cervical computed tomography angiography on the same day (CTA) showed a string sign in the V4 segment of the right vertebral artery, confirming the diagnosis of right vertebral dissection (Fig. 1C). Aspirin 100mg per day was prescribed, and her truncal ataxia quickly improved 2-3 days later. Her headache, neck pain, and vertigo completely disappeared at day 7. Because of good recovery of all symptoms, she refused to undergo follow-up CTA three months later. We discontinued aspirin therapy 6 months later, and her headache and neck pain did not recur in the following two years.

DISCUSSION

Our patient presented with severe right posterior neck pain and vertigo followed closely by right occipital thunderclap headache which was then confirmed as right VAD with delayed right dorsal medullary infarction two days later. Besides CAD, initial differential diagnosis for thunderclap headache includes subarachnoid hemorrhage, cerebral venous thrombosis, and reversible cerebrovascular constriction syndrome⁽⁷⁾. Furthermore, when facing a patient with abrupt headache associated with severe nuchal pain, cervical spinal epidural hemorrhage⁽⁸⁾ should also be considered.

The severe pain rendered our patient unable to be cooperative for further MRI study of the cervical region. Even though the initial brain MRI demonstrated the right dorsal medullary lesion (Fig. 1A), we could not recognize the dissection site clearly by intracranial MRA (Fig. 1B). Therefore we performed CTA and discovered well-demonstrated right vertebral artery dissection in the V4 segment (Fig. 1C). Both CTA and MRI with MRA have replaced conventional angiography as the golden standard because of accessibility and play an important role in the consideration of the above comprehensive

differential diagnoses⁽¹⁰⁻¹³⁾. CTA has benefits in offering prompt imaging in an emergency condition and requires less patient cooperation, but iodinated contrast and the cumulative radiation dose are its main limits. In addition to the ability to detect posterior fossa lesions, MRI provides better visualization of subacute hematoma on cross-section view of T1-weighted imaging with fat saturation. These hematomas classically appear as a crescent-shaped hyperintense area surrounded by an eccentric flow void zone in the lumen⁽¹¹⁻¹³⁾.

According to a systematic review collecting 75 studies and 1972 VAD patients⁽¹⁰⁾, the most common neuroimaging findings were vertebral artery stenosis (51%), followed by the string and pearls sign (48%), arterial dilation (37%), arterial occlusion (36%), and the less common but most characteristic features of pseudoaneurysm formation, double lumen, and intimal flap (22% each). Traditionally, the V3 and V1 segments were regarded as the most common sites of dissection⁽¹⁾, probably due to the presence of the most tortuous portion of the artery and the lack of vertebral bone protection respectively. However, according to the aforementioned review⁽¹⁰⁾, dissections in the V1 segment were found less frequently (27.5%) than in the other dissection sites (33.6-35.8%). Neck ultrasonographic techniques are non-invasive and helpful for the initial assessment of CAD patients, especially in daily outpatient practice. This, however, should be only regarded as a screening tool because it is highly technician dependent and has a poor diagnostic rate for tearing vessels hidden beneath bony structures^(1,2,11).

Clinical suspicion is always the key to the diagnosis of CAD. Our patient demonstrated characteristic thunderclap headache and neck pain, which alerted us to the accurate clinical diagnosis. Headache and pain are the hallmarks of VAD, but the features of the headache can sometimes be misleading, mimicking migraine, primary thunderclap headache, cervicogenic headache, or even musculoskeletal pain. Up to now, there have been only a few studies focused on the details of the features of headache and pain in CAD.

In the largest prospective study on headache in CAD in 1995⁽¹⁴⁾, Silbert et al. analyzed 26 patients with VAD, with 23% of them having past history of migraine. The headache was the initial symptom in approximately one third of the patients, mostly ipsilateral to the dissected

artery. Among the patients with headache, 44% had a pulsatile headache, while the rest described their symptoms as steady pain. Furthermore, 83% of the headaches were located posteriorly. Half of the patients described their headache as unique. Interestingly, thunderclap headache was not as common as we thought. Only 22% of patients reported “abrupt” onset, and the majority of patients (72%) suffered from gradual onset of headache. The mean time interval from headache to delayed stroke was 3.7 days (median 14.5 hours), while the mean duration of headache was 8.3 days (median 3 days), which was quite similar to our patient’s clinical course. No patients developed chronic headache later. Neck pain was observed in nearly half of the patients, mostly developing gradually and occurring with headache. All cases of neck pain were located in the posterior region, and were unilateral in two thirds of patients and bilateral in the rest. Neck pain preceded other symptoms in 5 patients by a mean time interval of 12 days and median of 14 days.

Arnold et al. collected 15 patients with spontaneous VAD, and only four of them (27%) experienced thunderclap headache⁽¹⁵⁾. Among patients with VAD, 7 (46.7%) had headache located in the occipital region, and 9 (60%) presented with pulsatile feature. All patients were pain free at 3 months. Another recent study from Japan described a higher rate of acute headache in dissection patients. They reported 6 VAD patients with isolated pain, with 5 of them having acute headache⁽¹⁶⁾. All suffered from occipital headache. Overall, 5 of the 7 (71%) patients had pulsatile pain.

Ipsilateral occipitocervical pain followed by delayed stroke is the outstanding clinical presentation of VAD. The main explanation of the pain is due to artery distension by the mural hematoma resulting in the stimulation of pain-sensitive receptors. The predominantly occipital location of headache in VAD may be explained by the referred pain associated with innervation of this region by the upper cervical nerves, quite similar to that of cervicogenic headache^(15,16). Dissection usually occurs in the subintimal level, and the consequential hematoma causes a long, irregular stenosis or even gradually evolves to an occlusion. Moreover, the distal thromboembolism is another possible mechanism of delayed ischemic stroke. Medullary stroke is the most common site of ischemic event, but other posterior strokes and cervical

cord ischemias have been previously reported^(1,2,4). If the dissection extends intracranially, a poor prognosis is usually indicated because subarachnoid hemorrhage or lower brain stem compression may occur^(1,2). Factors associated with better outcome were younger onset and a lower initial National Institutes of Health Stroke Scale score⁽¹⁷⁾.

VAD can present with isolated thunderclap headache as initial manifestation and with delayed stroke. Pain mostly occurs in the ipsilateral posterior neck region and is associated with occipital headache. Most evidence^(14,15) showed that only 22-27% of VAD patients present with thunderclap headache, and about 50-60% of patients had throbbing features. Excruciating neck pain could occur in a case of dissection. In stroke among young patients or stroke with pain, MRA or CTA of the craniocervical artery is highly recommended.

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