Clinical Reasoning:

A 54-year-old man with unilateral weakness and vascular risk factors

Vasileios Lioutas, MD
Jose R. Romero, MD
Michael S. Kleinman,
DO
James W. Wang, MD
Michael D. Perloff, MD,
PhD

Correspondence & reprint
requests to Dr. Perloff:
Michael.Perloff@bmc.org

SECTION 1

A 54-year-old man with history of poorly controlled hypertension and hyperlipidemia presented with right arm and leg weakness while exercising. Onset was preceded by a few minutes of acute, right-sided, “stabbing” neck pain. He had no dysarthria, facial weakness, visual disturbance, loss of sensation, chest pain, or palpitations. Blood pressure was 178/78 mm Hg. Cranial nerves were intact, without nystagmus. Comprehension, repetition, and naming were intact. Distal right upper extremity weakness affected extensor more than flexor muscle groups. Right lower extremity weakness was more prominent in flexor than extensor muscle groups. Deep tendon reflexes were symmetric and increased at the ankles. A right Babinski sign was present. There was no dysmetria and screening sensory examination revealed no deficit to

light touch and temperature. His gait was wide-based and unsteady. NIH Stroke Scale score was 2. Cardiovascular examination demonstrated normal rhythm and auscultation of carotid arteries and heart sounds.

Intracranial hemorrhage was excluded with a negative noncontrast head CT, and aspirin 81 mg was administered. EKG revealed normal sinus rhythm. Brain MRI, obtained approximately 6 hours after initiation of symptoms, was negative for stroke. Magnetic resonance angiography (MRA) revealed lack of flow-related signal in the right vertebral artery extracranially extending intracranially.

Questions for consideration:

1. What is the differential diagnosis?
2. What would be the next step in your management of this patient?

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From the Departments of Neurology (V.L., J.R.R., M.S.K., M.D.P.) and Radiology (J.W.W.), Boston University School of Medicine, Boston University Medical Center, Boston, MA.

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SECTION 2

In a patient with cardiovascular risk factors and sudden neurologic deficit, acute stroke must be considered. Cardioembolic stroke typically has cortical signs and symptoms. Absence of cortical involvement, such as language disturbances or visual field deficit, makes a large hemispheric stroke unlikely. Similarly, absence of nystagmus or extraocular movement deficits argue against large brainstem infarction. Chronic, poorly controlled hypertension with pure motor symptoms suggests lacunar stroke, with small vessel disease as underlying mechanism. While lacunes are often located in the internal capsule, they may be seen in the brainstem. Neck pain preceding onset of symptoms during exercise raises concern for carotid or vertebral artery dissection.

The absence of radiographic evidence of acute stroke is perplexing. One would expect ischemia on MRI by 6 hours. However, not all clinical strokes are

evident on MRI and a small stroke might not be seen if imaging is performed too early.^{1,2} In addition, it is well-known that a significant percentage (3%–24%) of clinically evident ischemic strokes do not have MRI correlate.^{1–3} The absence of signal from the right vertebral artery is concerning and suggests arterial dissection, especially given neck pain during exercise. A vertebral artery dissection could cause right-sided weakness via left pontine or medullary stroke (emboli to small basilar perforators that supply the base of the pons). As MRA is a “flow-dependent” study, the reported finding does not exclude slow or retrograde flow.

Questions for consideration:

1. How does absence of ischemia on MRI affect your differential diagnosis?
2. What additional testing would you request?

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SECTION 3

While left hemispheric ischemic stroke sparing the face is still possible, it appears less likely given a negative MRI. CT angiogram revealed occlusion of the right vertebral artery from the origin, extending intracranially (figure 1). Repeat neurologic examination demonstrated no change in right hemiparesis. The patient had no sensory complaints, but a more detailed sensory examination demonstrated a modest decrease to light touch, temperature, and pinprick sensation on the left side with a T3-T4 sensory level. Proprioception was decreased in the right hand but normal in the left. Two-point discrimination was limited to 6 mm on the right palm and fingers, but normal at 3 mm on the left. Vibratory sensation was decreased on the right hand. Proprioception and vibration were normal and equal in the lower extremities.

Taking into account the sensory examination findings, localization moves from the left cerebral hemisphere to the brainstem/spinal cord below the medulla. Absence of cranial nerve involvement with sparing of the face further suggests a spinal cord lesion. The constellation of unilateral motor findings in a pyramidal pattern with sensory findings that suggest ipsilateral posterior column involvement (vibratory sense, proprioception) and contralateral spinothalamic tract involvement (light touch, temperature, and pinprick) is consistent with the Brown-Sequard (hemicord) syndrome. Vertebral artery

Figure 1 Coronal thick slab MIP image shows lack of contrast filling within the right vertebral artery (arrows), consistent with occlusion

occlusion with a Brown-Sequard syndrome raises concern for a spinal cord infarction, as vertebral artery occlusion can lead to occlusion of the small cervical segmental artery branches.

Questions for consideration:

1. With the information present thus far, can you localize the lesion within the spinal cord?
2. What tests will validate the Brown-Sequard syndrome diagnosis and reveal its etiology?

GO TO SECTION 4

SECTION 4

Occlusion of the origin of the vertebral artery, in the setting of poorly controlled hypertension, warrants imaging to rule out aortic dissection. There is also a possibility of traumatic injury of the cord during exercise, necessitating a cervical MRI. Blood pressure was equal in both arms, and a CT angiogram of the aorta was negative for dissection. After detailed review of the imaging, with special attention to the origin of the vertebral arteries and to potential evidence of dissection (intramural hematoma), it was concluded that the vertebral artery occlusion was most likely chronic. At times difficult to visualize with imaging,⁴ dural fistula was considered with transient amplification of myelopathic symptoms during activity (from elevated venous pressure). Cervical spine MRI was negative for infarction, but revealed moderate to severe spinal canal stenosis from C3-C4 through C5-C6. Spinal stenosis was greatest

at C3–4, with disc protrusion on the right causing more pronounced focal narrowing, and chronic spinal cord injury (myelomalacia) was noted at C5–C6 (figure 2). On questioning, the patient related that he was doing his daily sit-ups (typically 100–200) on the day his symptoms started; he decided to “push limits” and had done approximately 1,500. He also revealed that he felt an “electric-like” tingling sensation down his spine upon turning his head to the right. The most likely scenario supported by history, examination, and imaging is that of acute mechanical trauma to the cervical spinal cord caused by vigorous exercise, in the setting of pre-existing cervical stenosis, resulting in traumatic partial Brown-Sequard syndrome. The underlying mechanism was likely contusion without infarction of the spinal cord, maximal at the site of the more pronounced focal C3–C4 canal stenosis on the right side. The presence of Lhermitte phenomenon with head turning further supports the hypothesis. Brown-Sequard syndrome can be a manifestation of a variety of insults to the spinal cord that result in damage to the ascending and descending tracts on one side. Spinal cord anatomy accounts for the clinical presentation: the corticospinal tracts cross at the junction of medulla and spinal cord. The spinothalamic tracts cross 2–3 levels above their entrance to the spinal cord. The ascending dorsal columns decussate in the medulla. In our patient, corticospinal tract deficits affected cervical through lumbar innervated muscles on the right, while left-sided spinothalamic deficits began in high thoracic segments, and posterior column proprioception and vibration only had deficits in the right upper extremity. Based on long tract somatotopic organization, localization suggested a more lateral partial hemicord syndrome (figure 2D).

Any space-occupying lesion compressing the spinal cord can manifest as Brown-Sequard syndrome (true incidence unknown), most commonly trauma and neoplasm.^{5,6} Inflammatory or infectious processes such as transverse myelitis, multiple sclerosis, herpes zoster, and acute bacterial meningitis have been implicated as well.^{5,6} Ischemia of the spinal cord typically involves the anterior spinal artery, affecting the ventral two-thirds of the spinal cord. This results in bilateral motor impairment, pain, and temperature loss below the level of the lesion, but with relative preservation of posterior column function.^{7,8} Ischemia can result in a cord hemisection syndrome with occlusion of small segmental branches that arise from the vertebral artery and provide lateral blood supply to the cervical spinal cord. The majority (50%–80%) of spinal cord infarctions are painful, an unexplained difference from cerebral infarction, likely related to radicular sensory nerve ischemia.^{7,8}

Figure 2 Cervical MRI and lesion localization

Sagittal T2 turbo spin-echo (TSE) image of the cervical spine (A) shows focal high signal within the cervical spinal cord at the C5–C6 level with associated atrophy, suggestive of myelomalacia (chronic injury). Note moderate to severe canal narrowing, most prominent at C3–C4. Axial T2 TSE image of the cervical spine at the C5–C6 level (B) confirms high signal within the gray matter of the cervical spinal cord. Note degenerative changes with a disc osteophyte complex causing moderate canal and bilateral neural foraminal narrowing. CSF surrounding the cord is effaced, but still present. Axial T2 TSE image of the cervical spine at the C3–C4 level (C) shows severe canal narrowing due to degenerative disc osteophyte complex (right greater than left). Note complete effacement of the CSF surrounding the cord, with deformity of the cord with trefoil appearance. In theoretical depiction of the cervical lesion (D), note that both the spinothalamic tracts and the posterior columns are partially interrupted, while all levels of the corticospinal tracts are affected at the level of the compression injury.

Fibrocartilaginous embolism can also cause an ischemic cord syndrome. This is a rare etiology with embolism of intervertebral disc fragments, usually traumatic in nature, almost invariably with poor outcome.⁹ Dissections of the vertebral (and carotid) arteries are usually provoked, commonly associated with hyperextension or rotation of the neck.¹⁰ Ischemic symptoms manifest rapidly, but separated from the initial painful episode by days to weeks.^{11,12} This is a distinguishing feature from other diagnoses, such as acute myelopathy from a disc herniation that presents acutely (with pain symptoms), but evolves slowly. Hence, in our patient, a thorough history suggested compressive etiology over ischemia. MRI is useful in differentiating spinal cord infarction from compressive myelopathy, but there is no gold standard method.¹³

Discogenic causes of Brown-Sequard syndrome are rare. A 2009 review identified 37 cases in the English language literature.¹⁴ Pre-existence of central spinal canal stenosis “limits” the available space, making a patient more prone to development of a clinically significant lesion. Often, the outcome of acute discogenic Brown-Sequard syndrome is favorable.¹⁴ This is predicated on early diagnosis and initiation of treatment including decompressive surgery or IV steroids with supportive care. In our patient, the neurosurgical consultant recommended conservative management with steroids and physical therapy, as the patient was not agreeable to surgery in the acute setting. After dexamethasone treatment and acute rehabilitation, the patient was discharged with no neurologic deficits 15 days after admission.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. **Go to Neurology.org for full disclosures.**

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