Progressive Tetraparesis in a 57-Year-Old Man With Congenital Absence of an Anterior Spinal Artery: A Case of Anterior Spinal Cord Infarction

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Abstract

A 57-year-old man presented with sudden neck pain radiating down his arms. This pain progressed to bilateral upper and subsequently lower extremity weakness and numbness. His vitals were notable for systolic blood pressures lower than his baseline (down to 90 mm Hg). The patient's neurological examination as well as magnetic resonance imaging of the cervical and thoracic spine localized to a lesion in the anterior spinal cord. The differential diagnosis for such an acute presentation included stroke, demyelination, intramedullary neoplasm, infection, metabolic myelopathy, and a dural arteriovenous fistula. Further imaging with angiography demonstrated that our patient lacked an anterior spinal artery. In its place, collateral flow from cervical artery branches provided sustenance to the anterior spinal cord. In the setting of hemodynamic instability, this variant anatomy likely predisposed the patient to ischemia, leading to the classic presentation of anterior cord syndrome.

Keywords

stroke, spinal cord vascular diseases, neuro-anatomy

Introduction

Anatomically, the subclavian arteries from the aorta give rise to the 2 vertebral arteries, which join to form the anterior spinal artery (ASA). The ASA supplies the ventral two-thirds of the spinal cord. Thus, any aortic insult may circuitously compromise the ventral cord, resulting in archetypal neurologic deficits that map to that territory.

Anterior cord syndrome occurs in 5% to 8% of acute spinal cord injury cases and is usually secondary to ASA ischemia or infarction.¹ In the absence of trauma, sudden development of symptoms referable to the spinal cord strongly suggests infarction or hemorrhage. Symptom onset of ischemia tends to be abrupt, reflecting the sudden impairment of arterial flow to the spinal cord from severe hypotension, for example, during aortic surgery.^{2,3} Because the ASA is an end-artery branch of the vertebral vessels with no anastomoses, the anterior cord may be vulnerable to lack of blood flow in times of hemodynamic instability. Other causes of flow disruption within the ASA include occlusion due to atheroma formation or emboli, aortic dissection, aneurysm, trauma, or syphilitic vasculature.⁴

Affected individuals exhibit a spectrum of findings, including a sensory level with impaired pain and temperature sensation below the level of the lesion.⁵ This deficit reflects disruption of anterior spinothalamic fibers. Weakness or flaccid motor paralysis with diminished deep tendon reflexes occurs because of damage to the descending corticospinal tracts. Para- and quadriplegia are possible outcomes.⁵ Over time, patients may experience progression to upper motor signs like spasticity and hyperactive reflexes. The first symptom of ASA infarction, however, is local or radiating sharp pain at level of the lesion.⁶ Proprioception and vibration remain intact, as the dorsal columns are supplied by the

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Figure 1. Panel A shows longitudinal T2-weighted hyperintensities from C3 to C5 (white arrow) and T1 to T3 (yellow arrow). Panel B shows the same T2 signals on an axial plane, at the C3 level, highlighting the anterior distribution of the lesion.

posterior spinal artery. Further, bowel and bladder autonomic dysfunction are frequently encountered.^{5,7}

Differential diagnosis is key in these cases because an array of pathophysiology can mimic spinal cord infarction. After localizing the disease process to the anterior spinal cord, it is important to rule out transverse myelitis, mass lesions, and infection. Neuroimaging, together with cerebrospinal fluid (CSF) analysis, fills this acute need in clinical practice.⁷ Nevertheless, anterior cord syndrome largely remains a clinical and radiographic diagnosis. Herein, we present a typical case of anterior cord syndrome as a result of a rare, presumably congenital absence of the ASA.

Case Report

A 57-year-old man with a history of chronic low back pain presented with sudden onset neck pain radiating down his arms while lifting a heavy object on a warm summer day. His arms became weak and numb, and he felt he was unable to lift them more than a few inches. A few hours after his arm weakness, the patient's weakness progressed to his legs. His course was complicated by urinary retention that was treated with Foley catheter usage. Review of systems was positive for an episode of diarrhea within 12 hours of hospital admission. His home medications included a pain regimen for his back (baclofen, oxycodone, and cyclobenzaprine). He was a non-smoker and drank 1 to 2 alcoholic beverages per week. He did not have hypertension or diabetes, and his family history was not noncontributory.

His vital signs were notable for a blood pressure of 90/50 mm Hg, considerably lower than his baseline (roughly 120/80 mm Hg). Mental status and cranial nerve examinations were normal. Upper extremity adduction/abduction was 2/5, elbow flexion/extension was 2/5, and his grip strength was 1/5 bilaterally. Hip flexion/extension was 2/5, knee flexion/

extension was 3/5, and plantar flexion/extension was 3/5 bilaterally. His exam was also remarkable for sensory loss to pinprick and temperature at the T3-T4 level. Vibration and proprioception were preserved. Reflexes were diminished to 1+ throughout all 4 limbs.

These examination-localizing features were crucial, as the ASA supplies the ventral two-thirds of the spinal cord, such that an infarction in this territory affects motor, pain, and temperature, but preserves proprioceptive and vibratory sensations otherwise sustained by the posterior spinal artery. In addition to the localizing capacity of the examination, magnetic resonance imaging (MRI) with and without contrast is the radiologic method of choice in evaluating a patient with suspected anterior cord syndrome. Alongside imaging, CSF examination revealed 1 erythrocyte, 1 nucleated cell, glucose 90 mg/dL (serum glucose 154 mg/dL), and protein 76 mg/dL (normal 15-45 mg/dL).

Magnetic resonance imaging spine revealed T2 hyperintense signals in the anterior spinal cord, spanning C3 to C5 and T1 and T3 in the sagittal plane (Figure 1A). From an axial view, T2-weighted imaging revealed the same confluent, abnormal signal within the anterior cord (Figure 1B). Because of the discrete vascular distribution of the lesion, it was felt that the patient's anterior cord syndrome resulted from an ASA stroke. Vertebral angiography was consequently pursued to investigate stroke etiology.

Conventional angiogram of the posterior circulation showed penetrating vertebral branches supplying the cord (Figure 2). There was no filling of the proximal anterior and posterior spinal arteries from either vertebral artery. There were a few small muscular branches of the cervical vertebral arteries bilaterally that sent branches to the cervical spinal cord, but they did not reconstitute the anterior and posterior spinal arteries. Thus, in close consultation with neuroradiology and the vascular neurologists, it was felt that there was a

Figure 2. Conventional angiogram of the posterior circulation showing (A) left and (B) right vertebral arterial circulation without an anterior spinal artery (sagittal oblique view). In both panels, arrows point to small muscular branches of the cervical vertebral arteries sending additional branches to the cervical spinal cord. These branches did not reconstitute anterior or posterior spinal arteries.

congenitally absent ASA. The artery of Adamkiewicz was not visualized.

Discussion

This case stresses the importance of clinical presentation in tandem with the neurologic examination. Though our patient's clinical syndrome was an expected presentation of an ASA stroke, his spinal angiogram revealed that there was no filling of the proximal ASA from either vertebral artery. Instead, there were a few small muscular branches of both cervical vertebral arteries sending branches to the cervical cord but failing to reconstitute the anterior and posterior spinal arteries. This variant collateral anatomy was likely the reason he was asymptomatic prior to his ischemic event. This anatomy also explains his increased vulnerability to hemodynamic instability, as likely occurred during an episode of hypotension (systolic pressures down to 90 mm Hg), which was thought to be secondary to the episode of diarrhea he experienced the morning of his admission.

Anterior spinal artery stroke represents approximately 1.2% of all stroke syndromes.⁸ While anterior cord syndrome is a clinical diagnosis, MRI is recommended in the pursuit of a definitive diagnosis and etiology. The characteristic feature of the ASA stroke syndrome is high signal intensity on T2-weighted sequences in the central and anterior aspect of the cord. Restricted diffusion in the ventral two-thirds of the cord would confirm the diagnosis, although diffusion-weighted imaging was technically challenging and difficult to visualize, as it can be in most spinal cases.^{9,10} Based on our patient's CSF studies alone, the clinical picture may have been confused with an inflammatory process, rather than primary ischemia, which has also been shown to produce elevated cell count and protein in the CSF.^{11,12} Without considering the

localization of his symptoms, alongside the anatomy of the spinal cord, this may have been confused with transverse myelitis.

Furthermore, to our knowledge, this case represents the first reported documentation of a congenitally absent ASA. Anatomic variants within the circle of Willis are well recognized, but we did not find any similar cases of an absent ASA in the literature. Savica et al reported a cerebellar stroke in a 76-year-old female patient with pure basilar artery agenesis. As in our case, the authors speculate that their patient remained asymptomatic until an advanced age because of sufficient collateral flow.¹³ However, in the context of vascular aging, her progressive glycemic burden and history of hypertension likely predisposed her collateral circulation to fail, leading to stroke at 76 years of age. While our patient did not possess any vascular risk factors, his hypotension on the day of presentation, superimposed on vulnerable variant anatomy, was the likely underlying etiology of his ischemia.

Nevertheless, it is important to note that our case stands alone, and so we can only infer etiology based on our patient's presentation and angiogram. It is possible that variant anatomy such as an absent ASA does not come to clinical attention because it remains asymptomatic and adequately compensated by collateral vasculature. While our patient's anatomy laid unstable brickwork for cord infarction during hemodynamic instability, this situation may not be the case for others in the population with unknowingly absent ASA anatomy.

Unfortunately, there is no specific treatment for spinal cord infarction. Antiplatelet, supportive care, and extensive physical rehabilitation are recommended.^{1,2,8} In this patient's case, midodrine was administered to augment blood pressure, thereby improving collateralization and perfusion to the cord. Regarding prognosis, the trajectory toward recovery is

variable.¹⁴ A retrospective, longitudinal 3-year study of 115 patients with spinal cord stroke showed that 41% of patients using a wheelchair at discharge were walking, and 33% of patients with a long-term catheter at discharge were catheter free.¹⁵ This study emphasizes that gradual improvement is not uncommon in this patient population, and recovery may continue well after hospital discharge. Six months after the stroke, our patient is catheter free and doing well with the aid of a rolling walker for ambulation.

Authors' Note

Informed patient consent was received using standardized forms at Yale-New Haven Hospital. The patient consented to the manuscript preparation and submission as well as to non-identifiable imaging and angiography of his spinal cord and cerebrovascular tree, respectively.

Declaration of Conflicting Interests

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