

Posterior Spinal Cord Infarction Due to Fibrocartilaginous Embolization in a 16-Year-Old Athlete

AUTHORS: Seema Bansal, MD, Wendy Brown, MD, Anuradha Dayal, MD, and Jessica L. Carpenter, MD

Children's National Medical Center, Washington, District of Columbia

KEY WORDS

spinal cord, stroke

ABBREVIATIONS

CSF—cerebrospinal fluid

DWI—diffuse-weighted imaging

FGE—fibrocartilaginous embolization

IV—intravenous

Dr Bansal cared for the patient, performed the associated literature search, and wrote the manuscript and approved the final manuscript as submitted; Dr Brown reviewed the patients imaging, contributed to the differential diagnosis, described the radiographic findings in the manuscript, and prepared the figures and captions for the manuscript and approved the final manuscript as submitted; Dr Dayal cared for the patient and reviewed and revised the manuscript and approved the final manuscript as submitted; and Dr Carpenter cared for the patient, critically reviewed the manuscript, and approved the final manuscript as submitted.

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Address correspondence to Seema Bansal, MD, Epilepsy Center, Ann and Robert H. Lurie Children's Hospital of Chicago, 225 East Chicago Ave, Box 29, Chicago, IL 60611-2605. E-mail address: sbansal@luriechildrens.org

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abstract

Spinal cord infarction is extremely rare in children, and, similar to cerebrovascular infarcts, the pathogenesis is different from adults. Spinal cord infarcts are most commonly reported in adults in the context of aortic surgery; in children, the etiology is frequently unknown. Fibrocartilaginous embolization is a potential cause of spinal cord infarct in both populations. It is a process that occurs when spinal injury has resulted in disc disease, and subsequently disc fragments embolize to the cord, resulting in ischemia and/or infarction. In this report, we present a 16-year-old athlete who presented with symptoms of acute myelopathy after a period of intense exercise. Our original concern was for an inflammatory process of the spinal cord; however, given her history of competitive tumbling and degenerative disc changes on her initial spine magnetic resonance imaging scan, diffusion-weighted imaging was performed, which demonstrated acute spinal cord infarction. Unlike many cases of spinal cord infarction, our patient was fortunate to make a near-complete recovery. This case highlights the importance of recognizing rare causes of spinal cord pathology and considering infarction in the differential diagnosis of acute myelopathy because management and prognosis varies. *Pediatrics* 2014;134:e289–e292

Spinal cord infarction is a rare but well-established diagnosis in adults; however, in pediatrics, knowledge is limited to a few case reports. Clinical presentation is similar to other causes of acute myelopathy, yet diagnosis, management, and prognosis are considerably different. Symptoms vary based on the involved arterial territory. Anterior spinal artery ischemia causes motor deficits with spinothalamic tract symptoms, whereas posterior spinal artery ischemia causes lemniscal sensory deficits.¹ Common etiologies include prolonged arterial hypotension, cardioembolic disease, chronic spinal disease, tumor embolism, dissection, and systemic arteriopathy (ie, syphilitic arteritis). An increasingly recognized etiology is fibrocartilaginous embolization (FCE).^{2,3} Care providers should be aware of this entity and include it in the differential diagnosis when symptoms localize to the spinal cord.

Prognosis after spinal cord infarction varies based on the spinal level and territory involved, but permanent disability is common. Naess et al showed that 1 week after symptom onset, functional scores of patients with spinal cord infarctions were lower compared with those with cerebrovascular infarcts.⁴ FCE is thought to carry a particularly poor prognosis. After FCE was first described in 1961,⁵ diagnoses were exclusively made postmortem. Although a presumed diagnosis can now be made earlier, the degree of recovery varies, and therapeutic options remain limited.

PATIENT PRESENTATION

A 16-year-old girl presented with acute-onset numbness and tingling of the right leg soon after a period of intense exercise. She was a highly competitive athlete who was being actively recruited to compete for placement on the Olympic aerial ski jumping team. Her past medical history was negative for significant spinal trauma. Over the next 2 days,

symptoms progressed to the opposite leg and she developed gait disturbances, attributed to abnormal sensation. In the emergency department, creatine phosphokinase was found to be slightly elevated at 581 mcg/L, and she was diagnosed with rhabdomyolysis. She was discharged from the hospital with medications for pain control and instructions to aggressively hydrate. Two days later, despite compliance, there had been no improvement; she returned to the emergency department and was admitted for inpatient management.

On admission, vital signs and general medical exam were unremarkable. The patient was a well-appearing teenager in no acute distress. Review of systems revealed 1 episode of urinary incontinence on the day of admission. She denied back pain. On neurologic examination, mental status and cranial nerves were normal. Manual motor testing of the upper extremities was unremarkable. In the lower extremities, strength was 4+/5 in the bilateral iliopsoas and quadriceps, 5/5 in all other muscle groups. Muscle tone was normal. Deep tendon reflexes were 2+ in the bilateral upper extremities and 3+ in the bilateral patella with 2 to 3 beats of clonus at the Achilles. Plantar response was extensor on the right and flexor on the left. Digital rectal examination revealed poor sphincter tone and absence of anal wink. Sensory examination revealed intact vibration but diminished proprioception, temperature, and pinprick distal to T12, worse on the right. Heel-shin-knee testing disclosed dysmetria bilaterally. Gait was wide-based and Romberg sign was positive.

Initial spine MRI showed several T2 hyperintense lesions in the dorsal spinal cord at T11 and T12 (Fig 1). Lesions did not enhance with gadolinium. Degenerative disc disease, with disc desiccation and bulge, was present at T11/T12. The initial impression was myelitis, and therefore a lumbar puncture was performed. Cerebrospinal fluid (CSF) analysis showed

normal leukocytes, erythrocytes, glucose, and protein. Myelin basic protein was weakly positive, immunoglobulin G synthesis rate was normal, and oligoclonal bands were negative. Fluid culture and Gram stain were negative. Serum and CSF neuromyelitis optica antibody were negative.

The degenerative disc disease with negative inflammatory laboratories raised the possibility of spinal cord infarction. The next day, diffuse-weighted imaging (DWI) of the spine was performed, and focal areas of restricted diffusion were seen (Fig 2), corresponding to the T2 hyperintense lesions noted on initial imaging. Given the adjacent disc changes, FCE was proposed as the likely etiology.

A limited hypercoagulability panel and echocardiogram were done as part of the workup for infarction, and all studies were negative. The patient was treated with anticoagulation while prothrombotic laboratories were pending. She received a brief period of inpatient rehabilitation. Two months later, neurologic examination showed only mild sensory changes in the right leg. Four months later, MRI demonstrated resolution of the signal abnormalities within the spinal cord.

DISCUSSION

Spinal cord infarctions comprise 1% of all strokes in adults and are even less common in children.¹ It can be difficult to differentiate spinal infarct from other, more common, causes of acute myelopathy such as transverse myelitis, demyelinating diseases, and external compression. Diagnosis is often delayed,⁶ in 1 case by 2 years.⁷ Typically, spinal cord infarction is characterized by onset of paraplegia and/or paresthesias over minutes to hours associated with sudden back pain in the setting of trauma. In contrast, inflammatory causes of myelopathy may have a more insidious presentation. Our patient was atypical in that her deficit was initially unilateral and then



FIGURE 1

Axial (A) and sagittal (B) T2-weighted images demonstrate multiple foci of hyperintense signal at the dorsal aspect of the spinal cord, at the levels of T11 and T12. Adjacent disc desiccation and non-compressive disc bulge noted at T11/T12 (B). Lesions did not enhance after contrast administration (C).

became bilateral; however this 2-stage presentation has been previously described.⁸

CSF may distinguish spinal infarct from an inflammatory process. Typically, CSF is unremarkable, although leukocytosis² and elevated protein⁵ have been reported independently. Thus, the diagnosis can be difficult to make without imaging.

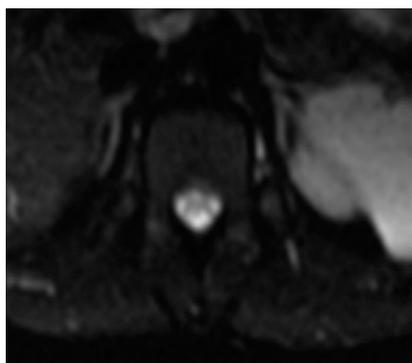


FIGURE 2

Axial diffusion DWI of the spine demonstrates focal areas of restricted diffusion that correspond to the T2 lesions within the spinal cord.

On conventional MRI, inflammatory and ischemic spinal cord lesions may both show T2 hyperintensity; however, with gadolinium, acute infarction will not enhance. DWI, which confirms the presence of ischemia, is unfortunately not routinely performed on spinal MRIs. Therefore, when clinical suspicion for infarct is high in a patient with acute myelopathy, care providers should specifically ask for a spinal MRI with DWI to confirm the diagnosis.²

The etiology of spinal infarct varies; in 1 study of 164 spinal infarcts, FCE was the cause in 9 patients (5.5%).³ Various activities predispose to FCE, including repetitive trauma from heavy-weight-bearing and intense exercise. FCE may occur more frequently in females and in the cervical cord.⁹ Age distribution is bimodal, peaking in adolescence and late middle age.¹⁰ In pediatrics, it is most often reported in teenagers,^{7,10–12} but notable cases of presumed FCE in young children include a 6-year-old gymnast¹³

and an 8-month-old infant who fell from a changing table.¹⁴

Postmortem biopsies have showed fibrocartilaginous material in spinal cord vasculature (arterioles and venules); however, the exact mechanism through which emboli enter remains unclear. Several mechanisms have been proposed. One theory is that the intervertebral disc ruptures laterally, causing disc fragments to enter arterial circulation via radicular arteries. A second hypothesis is that after high axial loading, intradiscal pressure is increased, resulting in injection of semifluid nucleus pulposus into small intradiscal arteries, which persist as embryological remnants in children. This may explain the increased incidence of FCE in adolescence. Another theory suggests that herniated disk material traverses the vertebral body endplate (forming Schmorl nodes), entering arterial circulation directly via segmental arteries.

The majority of spinal cord infarcts involve the anterior territory.^{3,8} In FCE, it seems logical that emboli would enter anterior microcirculation, which is closer to the intervertebral disc compared with posterior vessels. Our patient's MRI showed a posterior infarction; however, her symptoms suggested involvement of the anterolateral and corticospinal systems. Furthermore, her weakness was mild and may have been due to involvement of the posteriorly located spinocerebellar tracts.¹

Traditionally FCE was diagnosed histopathologically; however, increasing case reports led to diagnostic criteria that include acute back pain after trauma or Valsalva maneuver, absence of evidence of inflammatory CSF fluid or systemic thrombi, and MRI findings of intervertebral disc disease and hyperintense lesions on T2-weighted images.² A spinal cord lesion with restricted diffusion on DWI MRI, when adjacent to areas of disc disease, further supports FCE as the likely etiology.

Currently no treatment guidelines exist for spinal cord infarct due to FCE. Most patients receive supportive care including physiotherapy. Anticoagulation has been used, but no trials exist to support its use for this indication. In a series examining spinal cord infarct due to FCE at the Mayo Clinic in Rochester, Minnesota, 9 patients were identified who received a variety of treatment including intravenous (IV) steroids, IV immunoglobulin, plasma exchange, and IV heparin; none had a significant clinical improvement after treatment.³ Another series of 11 patients treated with heparin or corticosteroids failed to show significant clinical improvement in any patient.¹⁵

Not surprisingly, clinical outcome is related to location of the infarct. Full re-

covery has been seen in a child with a midthoracic lesion,² whereas high-cervical lesions can result in tetraparesis¹¹; death due to respiratory failure has also been reported.⁵ Age seems to be influential as well, with older patients consistently having poorer outcomes.³ Overall, some degree of initial recovery is observed in the majority of patients regardless of etiology, although the maximal recovery period appears to be soon after infarct.¹⁶ Furthermore, there are no data regarding recurrence risk, making it difficult to counsel patients. Given the potential for permanent neurologic disability, however, it is not unreasonable to advise patients to limit activities that load the spinal column.

Two features make this case unique. First, our patient's infarct localized to

the posterior cord, and second, her outcome was excellent. Although the initial examination and imaging were thought to be consistent with transverse myelitis by several pediatric neurologists in our institution, the presence of disc disease on MRI gave rise to the possibility of spinal cord infarction and prompted repeat imaging with DWI sequencing, which was positive. The time course of symptoms, history of trauma, and evidence of disc disease supports the diagnosis of spinal cord infarct due to FCE. Awareness of this entity may help care providers reach this diagnosis earlier and therefore limit unnecessary testing and therapies, as well as provide more specific prognostic information to patients and parents.

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