

# Rapidly Progressive Paraplegia in an 11-Year-Old Girl: A Case of Spinal Cord Infarction and Expected Imaging Findings

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## Abstract

Rapidly progressive non-traumatic paraplegia in a child is uncommonly encountered in clinical practice, but is an important presentation to consider given the potential for significant morbidity. We present the case of an 11-year-old girl who was found to have hyperacute paraplegia due to spinal cord infarction. We discuss the appropriate workup, differential diagnosis in children and how this relates to adults; and describe the prognosis and current state of management options for spinal cord infarction.

## Keywords

paraplegia, infarct, fibrocartilaginous embolism, blood pressure augmentation

## Introduction

Rapidly progressive non-traumatic paraplegia in a child is uncommonly encountered in clinical practice,<sup>1</sup> but is an important presentation to consider given the potential for significant morbidity.<sup>2</sup> The diagnosis highly depends on clinical time course, associated symptoms, and examination findings; which appropriately guides additional clinical management and prognosis.

Here we present the case of an 11-year-old girl who was found to have hyperacute paraplegia due to spinal cord infarction. We also discuss the diagnostic evaluation and therapeutic management for paraplegia, expected outcome and imaging findings.

## Case

An 11-year-old right-handed girl born full-term with history of non-epileptic staring spells presented to the hospital for rapidly progressive back pain and paraplegia. While lifting heavy wooden planks, she experienced acute onset lumbar back discomfort that rapidly radiated to the bilateral legs, with patchy distribution of paresthesias and dysesthesias in the legs. She denied trauma, “popping” sensation, or any preceding infectious symptoms. About 20-30 minutes after the onset of back pain, she noticed weakness throughout the entire distribution of

her bilateral legs, which prompted family to seek care at an outside hospital. She denied arm weakness or sensory changes, headache, vision changes, or difficulty with swallowing, talking, or breathing. She endorsed difficulty urinating.

Her exam was notable for flaccid paraplegia, areflexia throughout the lower extremities, and mute plantar responses bilaterally. She had no abnormalities in mental status, cranial nerves, or motor/sensation/coordination of the upper extremities. There was a lower thoracic sensory level to temperature, light touch, and pinprick. Joint position and vibratory sense was normal in the upper extremities, but reduced in the toes and ankles bilaterally.

MRI lumbar spine without contrast obtained about 6 hours after symptom onset (Figure 1A) was negative for cord signal change, vertebral/disc abnormalities, or masses. Lumbar puncture obtained in the outside emergency department showed

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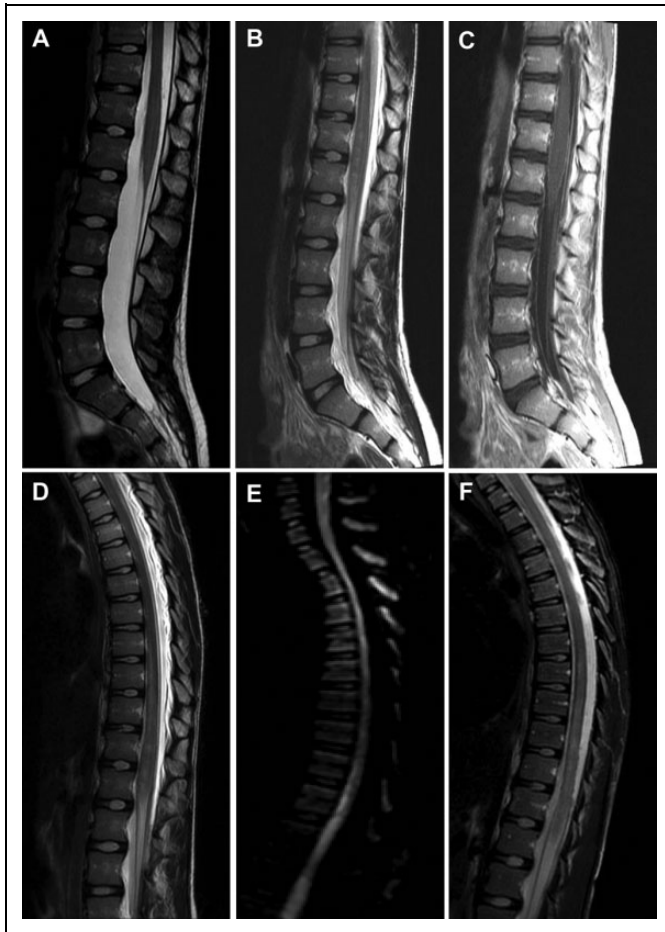
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**Figure 1.** Imaging findings showing progression of spinal cord infarction. A) Sagittal T2-weighted MR image of the lumbar spine from the day of initial presentation is normal. B) Sagittal T2-weighted MR image of the lumbar spine on the day after symptom onset (day 2) demonstrates patchy intramedullary T2 hyperintensity in the anterior cord with parenchymal expansion of the conus. There is notably no abnormal intervertebral disc signal or loss of disc height. C) Sagittal T1-weighted post-gadolinium imaging also from day 2 shows mild vascular prominence without abnormal cord or nerve root enhancement. D) Sagittal T2-weighted MR image of the thoracic spine from the spinal angiogram on day 3 demonstrates progression of cord signal abnormality to the T9 level and remaining ventral. E) Sagittal diffusion weighted image from the same exam showed restricted diffusion corresponding to areas of signal abnormality though slightly more apparent at the T9 level. F) Sagittal T2-weighted MR image of the thoracic spine obtained on day 5 shows signal abnormality is further progressed with extension to the level of T7, again remaining ventral.

CSF glucose 62 mg/dL (serum glucose 111 mg/dL), protein 17 mg/dL, total nucleated cell count 0, RBC count 1, and xanthochromia negative. She was directly admitted to our hospital for further management.

Although acute spinal cord infarction seemed most likely, we worked to rule out other etiologies to ensure we were not missing a treatable condition. EMG was obtained to assess for inflammatory polyradiculoneuropathy and returned with normal results. MRI thoracic/lumbar spine with contrast obtained

1 day after symptom onset showed new development of T2 hyperintensity within the anterior portion (anterior horns) of the distal thoracic cord (T10) to the conus medullaris (Figure 1B-C). Importantly, there were no signal abnormalities in the vertebrae or discs, and the brain MRI exam was normal. Her MRI findings were most consistent with acute spinal cord infarction. MR spine angiogram with diffusion weighted sequences were obtained 2 days after symptom onset and showed extension of T2 hyperintensity rostrally to T9 with diffusion-weighted imaging showing restricted diffusion most notable at level T9 (Figure 1D-E). No overt vascular abnormalities were visualized. MRI cervical spine was obtained 4 days after symptom onset (day 5) to assess the remainder of the spine, which showed extension of T2 hyperintensity rostrally to T7 (Figure 1F).

The expansion of T2 hyperintensity in the anterior cord from T10 on day 2 to T7 on day 5 prompted further testing, given possibility of inflammatory process, such as anti-MOG disease that can involve the conus medullaris.<sup>3</sup> Anti-MOG and AQP4 antibodies in the serum and autoimmune encephalitis panel in the CSF were negative.

To provide our patient the best opportunity for clinical recovery, she was brought to the intensive care unit 2 days after symptom onset for 12 hours of pressure augmentation using phenylephrine from 0.1-0.4 mcg/kg/min to sustain mean arterial pressure >85 mmHg, without clinical improvement. Upon hospital discharge, she was wheelchair dependent for ambulation and working to become independent in activities of daily living. After being discharged from inpatient care, she underwent intense inpatient rehabilitation. Her examination about 3 weeks after infarct showed no activation and persistent areflexia of the bilateral legs, sensory level to T7 on the right and T9 on the left.

## Discussion

Our patient presented with hyperacute onset non-traumatic paraplegia. Her physical examination best localized to the anterior spinal cord given paraplegia, lower thoracic sensory level, early areflexia, and sparing of vibratory sensation. Thus, spinal cord infarct was high on the differential diagnosis, along with spinal cord compression and trauma.

The timing of symptoms in patients with myelopathy has been shown to be one of the best predictors of etiology.<sup>4</sup> Almost all patients with a vascular cause of myelopathy have symptom onset in the hyperacute (<6 hours) to acute (6-48 hours) window. While inflammatory myelopathies generally present subacute (2-21 days) to chronic (>21 days).<sup>4</sup> An inflammatory myelopathy (i.e. anti-MOG) or polyradiculoneuropathy (i.e. Guillain Barré) are less likely to have such an acute onset, but are more common than stroke in children and should be ruled out, which was done in the diagnostic evaluation of our patient.

A key learning point is that spinal cord infarcts are generally diagnosed later in pediatric patients than adults, due to vague symptoms, inability to communicate symptoms, or healthcare

providers not recognizing the possibility of infarct. Another important aspect about this case is the imaging findings (Figure 1A-F), which display the expected evolution of spinal cord infarct that is rarely documented clinically.<sup>5</sup> This is not necessary or routine in clinical practice, but when there is a question of diagnosis, repeat images may help guide further management. Importantly, an acute spinal cord infarct may initially show no changes on imaging. If there is concern for spinal cord infarct, diffusion-weighted sequences and spinal angiography should be considered. In the acute phases, enhancement within the cord is unlikely, and if present may suggest inflammatory, infectious, or neoplastic process.<sup>5-7</sup>

Patient demographics and risk factors must always be considered when assessing the etiology of spinal cord infarct, which also differs between the pediatric and adult populations. In pediatrics, one should consider fibrocartilaginous embolism, congenital AV malformations, thrombophilia, congenital cardiovascular abnormalities, or vasospasm. Vasospasm can arise from hyperflexion injuries, which may be complicated by poor spinal cord pressure autoregulation leading to ischemia.<sup>2,6</sup> In adults, the mechanism of infarct is often secondary to traditional vascular/atherosclerotic risk factors, aortic injury, systemic hypotension, or cardioembolic event.<sup>7-9</sup> Our patient had a thorough workup to rule out vascular risk factors for spinal cord infarct, which was unrevealing. Although there is no definitive confirmatory test, the etiology of our patient's infarct was most likely cryptogenic fibrocartilaginous embolism, based on absence of vascular risk factors, temporal relationship to heavy lifting, and imaging features consistent with evolving infarction. Fibrocartilaginous embolism is the embolization of nucleus pulposis material from the intervertebral disc after a pressure related injury. The embolized nucleus pulposis enters arteries in the anterior spinal artery distribution leading to ischemia.<sup>10</sup>

Unfortunately, there are limited interventions in the acute to subacute period of spinal cord infarction, unless it is secondary to dural arteriovenous fistula, where liquid embolic endovascular procedure or surgical occlusion of the intradural vein may be considered.<sup>11</sup> Pressure augmentation or the pharmacologic support of blood pressure to meet an elevated pressure goal<sup>12</sup> and CSF drainage may be considered, but there is limited data in patients with spontaneous infarction.<sup>8</sup> Pressure augmentation was attempted for our patient without functional improvement. While tissue plasminogen activator (tPA) may be considered for acute infarct.<sup>13</sup> Its accepted use is within a maximum of 4.5 hours after symptoms onset. By the time our patient presented to our care, she was outside of the window for tPA administration.

The long-term recovery in spinal cord infarct is not necessarily bleak. Previous case series in patients with spinal cord infarct suggest gradual improvement may be seen in the years following infarct. The primary factors affecting long term functional status are severity of symptoms at nadir, extent of spinal cord involvement, early clinical improvement, age, and comorbid vascular risk factors.<sup>6,9,14</sup>

## Author Contributions

Bryan Neth: case concept and primary authorship. Angela L. Hewitt: case concept and critical revision of manuscript for intellectual content. Wendy S. Edlund: case concept and critical revision of manuscript for intellectual content. Julie B. Guerin: contribution of neuroimaging expertise and critical revision of manuscript for intellectual content. Marc C. Patterson: case concept and critical revision of manuscript for intellectual content. Gesina F. Keating: case concept and critical revision of manuscript for intellectual content.


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