Acute Myelopathy With Normal Imaging

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Abstract
A 17-year-old girl presented with rapidly progressive quadriparesis and ventilatory failure. The clinical findings indicated a spinal level, but the diagnosis of myelopathy was not supported by her initial spinal imaging and cerebrospinal fluid studies. She had completed treatment for Guillain-Barré syndrome before a follow-up spinal imaging study showed interval expansion and enhancement of the cervical cord.

Keywords
myelopathy, spinal cord infarction, fibrocartilaginous embolism

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Case Report
This 17-year-old high school athlete woke up with pain between her shoulder blades and a sense of weakness in both arms. During the course of that morning, she developed progressive paraplegia, associated with numbness in her legs up to her waist. There had been no preceding back pain or trauma. She had been in Mexico 2 months prior to admission, and had complained of a self-limited diarrhea during her trip. Otherwise, there was no significant past medical history. Her forced vital capacity was only 1.2 L. She had normal speech but a week cough. Her cranial nerves, including pupils and eye movements, were normal. She had weakness of deltoid and biceps (MRC 3/5), more significant weakness of triceps and the wrists (MRC 2/5), and paraplegia. She had numbness in both legs coming up onto the trunk. Reflexes were reduced in the arms and absent in the legs. A Foley catheter was placed and drained over a liter of urine. Emergent contrast-enhanced magnetic resonance imaging (MRI) of the cervical spine showed only a small annular tear at C5-C6 but was otherwise normal (Figure 1a and b). Cerebrospinal fluid was also unremarkable. She was admitted to the intensive care unit, and within 1 hour underwent elective endotracheal intubation and mechanical ventilation for respiratory distress and low vital capacity. Electrodiagnostic testing showed normal sensory and motor conduction studies, absent F-wave responses, and absent or reduced recruitment of normal motor unit potentials without abnormal spontaneous activity.

The normal spinal MRI, absence of a cerebrospinal fluid pleocytosis and electrodiagnostic findings were felt to be more consistent with an acute inflammatory polyneuritis (Guillain-Barré syndrome) than transverse myelitis, so she was started on a 5-day course of intravenous immunoglobulin 0.4 g/kg/d. By day 3 of her hospitalization, she remained ventilator dependent, and had developed mild bifacial weakness in addition to increased weakness in her arms. After she had completed intravenous immunoglobulin therapy for presumed Guillain-Barré syndrome, she underwent repeat lumbar puncture and electrodiagnostic testing in the hope of confirming the diagnosis. Her repeat cerebrospinal fluid was still normal—the fluid was acellular and there had been no interval increase in protein level. Repeat electrodiagnostic testing showed low-amplitude peroneal motor responses, with some conduction slowing (but no conduction block) across the fibula heads, but was otherwise unchanged. The following investigations were all known to be normal and/or negative: serum Lyme titers, antinuclear antibodies, anti–double-stranded DNA antibodies, anticardiolipin antibodies, anti-GMI antibodies, neuromyelitis optica antibody, serum copper level, urine heavy metal and porphyrin levels, cerebrospinal fluid Viral Reference Laboratory, IgG index and oligoclonal bands, and computed tomographic (CT) angiography of the aorta.

Two days later, repeat spinal MRI showed markedly abnormal signal intensity in the spinal cord (Figures 1C and D). MRI of the brain was normal. She was started on high-dose intravenous steroids. Subsequently, she was transferred to a tertiary care facility at the request of her parents, where she underwent plasma exchange followed by intravenous cyclophosphamide for presumed transverse myelitis. There were numerous...

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medical complications, including aplastic anemia from the chemotherapeutic agents, and she remained hospitalized for many months. She was slowly weaned from the ventilator and then regained some strength in her arms, although she remained paraplegic and confined to a wheelchair.

Discussion
The initial clinical findings indicated a spinal level suggesting acute myelopathy. MRI of the spine with and without contrast is the initial investigation of choice for evaluation of acute myelopathy and is usually abnormal even with noncompressive causes such as transverse myelitis, which has led to the suggestion that a normal study should lead to a reevaluation of the diagnosis of acute myelopathy. A cerebrospinal fluid pleocytosis is present in more than 80% of transverse myelitis cases. The normal MRI and cerebrospinal fluid in this case made a diagnosis of transverse myelitis seem unlikely. The main differential diagnosis of acute quadriplegia and ventilatory failure in a young adult was felt to be Guillain-Barré syndrome. The typical cerebrospinal fluid finding of albuminocytologic dissociation is identified in less than 50% at presentation. Furthermore, there are cases that present with some clinical features of spinal cord disease, and electrodiagnostic testing can be normal in early Guillain-Barré syndrome cases, often necessitating initiation of treatment based on a purely clinical diagnosis.

The lack of evolving albuminocytologic dissociation in the cerebrospinal fluid, the failure of the electrodiagnostic abnormalities to evolve beyond the nonspecific changes that can be seen with Guillain-Barré syndrome or acute myelitis, the lack of clinical response to intravenous immunoglobulin, and the initial clinical presentation with a clear spinal-level therapy all contributed to the decision to obtain a follow-up spinal MRI study. These follow-up images had become strikingly abnormal, showing expansion and enhancement of the cervical cord, findings more commonly associated with transverse myelitis, leading her treating physicians at the tertiary referral center to initiate plasma exchange and then cyclophosphamide infusions as empiric treatment for transverse myelitis, without further investigation.

However, in retrospect, the rapid clinical evolution of symptoms from normal to paraplegic over a few hours, normal cerebrospinal fluid, and normal acute spinal MRI study are more consistent with spinal cord infarction than inflammation. Spinal cord infarction is uncommon and difficult to differentiate from transverse myelitis in children. Fibrocartilaginous embolism, originally felt to be a rare and fatal condition diagnosed at autopsy, is becoming increasingly recognized as a cause of spinal cord infarction, particularly in children, because of a common blood supply between the spinal cord and nucleus pulposus that closes off in later life. Many affected children report a minor traumatic precipitating event, back or neck pain is the most common initial symptom, and the time from symptom onset to maximal weakness is usually less than 4 hours. Spinal MRI done early during the course of the illness is usually normal other than the frequent finding of a disc abnormality at the appropriate level, and the cerebrospinal fluid is noninflammatory. Diffusion-weighted imaging of the spinal cord may be more helpful for demonstrating acute ischemic change. High cervical cord infarction from fibrocartilaginous embolism can present with respiratory failure from involvement of the descending respiratory pathways. There is little response to medical or surgical treatment, and the long-term outcome is persistent moderate to severe disability.

Conclusion
Without tissue pathology, we will never know the etiology of this patient’s acute myelopathy. However, her presentation with acute pain, rapidly progressive weakness, initial MRI showing only a torn annulus at C5-6 but later showing evolving signal abnormality within the cord, and her noninflammatory cerebrospinal fluid all appear more consistent with a diagnosis of spinal cord infarction from fibrocartilaginous embolism than transverse myelitis.
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