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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-Barre syndrome before a follow-up spinal imaging study showed interval expansion and enhancement of the cervical cord.

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 waste. 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 liters. 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 (MR) imaging of the cervical spine, showed only a small annular tear at C56, but was otherwise normal (figure 1a and b) and cerebrospinal fluid (CSF) was also unremarkable. She was admitted to the intensive care unit, and within one 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 MR imaging, absence of a CSF pleocytosis and electrodiagnostic findings were felt to be more consistent with an acute inflammatory polyneuritis (Guillain-Barre syndrome, GBS) than transverse myelitis, so she was started on a five day course of intravenous immunoglobulin 0.4g/Kg/day. By day three of her hospitalization, she remained ventilator dependent, and had developed mild bifacial weakness in addition to increased

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weakness in her arms. After she had completed IVIG therapy for presumed GBS, she underwent repeat lumbar puncture and electrodiagnostic testing in the hope of confirming the diagnosis. Her repeat CSF was still normal – the fluid was acellular 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, anti-nuclear antibodies, anti-double stranded DNA antibodies, anti-cardiolipin antibodies, anti-GM1 antibodies, neuromyelitis optica (NMO) antibody, serum copper level, urine heavy metal and porphyrin levels, CSF VRL, IgG index and oligoclonal bands, and computed tomographic angiography of the aorta.

Two days later repeat spinal MR imaging showed markedly abnormal signal intensity in the spinal cord (figures 1c and d). MR imaging of the brain was normal. She was started on high dose intravenous steroids. 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 medical complications, including aplastic anemia from the chemotherapeutic agents, and she remained hospitalized for many months. She was slowly weaned from the ventilator, and 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. MR imaging of the spine with and without contrast is the initial investigation of choice for evaluation of acute myelopathy¹, and is usually abnormal even with non-compressive causes such as transverse myelitis^{2,3,4}, which has led to the suggestion that a normal study should lead to a reevaluation of the diagnosis of acute myelopathy^{1,5}. A CSF pleocytosis is present in more than 80% of transverse myelitis cases⁶. The normal MRI and CSF 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 GBS. The typical CSF finding of albuminocytologic dissociation is identified in less than 50% at presentation⁷, there are cases that present with some clinical features of spinal cord disease⁸, and electrodiagnostic testing can be normal in early GBS cases⁹, often necessitating initiation of treatment based on a purely clinical diagnosis¹⁰.

The lack of evolving albuminocytologic dissociation in the CSF, the failure of the electrodiagnostic abnormalities to evolve beyond the nonspecific changes that can be seen with GBS or acute myelitis¹¹, the lack of clinical response to IVIG, and the initial clinical presentation with a clear spinal level therapy all contributed to the decision to obtain a follow-up spinal MR imaging study.

These follow-up images had become strikingly abnormal, showing expansion and enhancement of the cervical cord, findings more commonly associated with transverse myelitis², leading her treating physicians at the tertiary referral center to initiate plasma exchange 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 CSF and normal acute spinal MR imaging study, are more consistent with spinal cord infarction than inflammation^{2,12,13,14,15,16}. Spinal cord infarction is uncommon and difficult to differentiate from transverse myelitis in children^{16, 17,18}. Fibrocartilagenous embolism, originally felt to be a rare and fatal condition

diagnosed at autopsy¹⁹, is becoming increasingly recognized as a cause of spinal cord infarction²⁰, particularly in children¹⁷, because of a common blood supply between the spinal cord and nucleus pulposus that closes off in later life²¹. 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 hours²⁰. Spinal MR imaging done early during the course of the illness is usually normal other than the frequent finding of a disc abnormality at the appropriate level^{16,21} and the cerebrospinal fluid is non-inflammatory²⁰. Diffusion weighted imaging (DWI) of the spinal cord may be more helpful for demonstrating acute ischemic change⁴. High cervical cord infarction from fibrocartilagenous embolism can present with respiratory failure from involvement of the descending respiratory pathways²². There is little response to medical or surgical treatment, and the long term outcome is persistent moderate to severe disability²⁰.

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 C56, but later showing evolving signal abnormality within the cord, and her non-inflammatory CSF all appear more consistent with a diagnosis of spinal cord infarction from fibrocartilagenous embolism than transverse myelitis.

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Figure 1.

Sagittal T2 (1a) and contrast enhanced T1 (1b) weighted images of the spine done acutely at the time of admission to the hospital, normal aside from a small annular tear at C56 (arrow). Repeat sagittal spinal imaging done 7 days after presentation, now showing diffuse cervical cord expansion with abnormal signal on T2 images (1c), with associated nodular enhancement on contrast enhanced T1 images (1d).