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Case report

Spinal cord infarction in a patient with multiple sclerosis

David N. Abarbanel, Nicholas L Zalewski, B. Mark Keegan*

Department of Neurology, Mayo Clinic, 200 1st Street SW, Rochester, MN 55905, USA

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ABSTRACT

We describe a 49 year old woman with relapsing-remitting multiple sclerosis (MS) with a suspected severe recurrent attack of myelitis that was ultimately diagnosed as a spinal cord infarction (SCI). This case of SCI in a patient with an established diagnosis of MS highlights the clinical, laboratory, and radiographic characteristics that help distinguish SCI from inflammatory myelitis due to MS.

1. Introduction

Spontaneous spinal cord infarction (SCI) is an uncommon cause of myelopathy that is frequently misdiagnosed (Zalewski et al., 2018). Symptom onset is often hyperacute with a severe deficit reaching nadir within 12 h in the majority of cases, commonly in the presence of vascular risk factors (Zalewski et al., 2019). Multiple sclerosis (MS), however, is a rather common cause of myelopathy, manifesting as either an attack of inflammatory myelitis or as a progressive myelopathy over years in the progressive phase of the disease. Contrary to the hyperacute, severe onset of SCI, most attacks of inflammatory myelitis in MS are subacute in onset over several days to weeks, sensory predominant, and mild to moderate in severity. We present a case of SCI in a patient with MS that highlights the contrasting features of SCI and inflammatory myelitis due to MS. This case serves as an example of the importance of critically assessing new disease activity in MS, and whether the suspected attack has typical features of MS or warrants consideration for an alternative etiology requiring different evaluation and management.

2. Case report

A 49 year old woman with a history of relapsing-remitting MS presented with hyperacute onset of severe paraplegia. Her past medical history includes hypertension, type 2 diabetes, hyperlipidemia, and obstructive sleep apnea.

The patient had a clearly delineated 7 year history of multiple typical relapses of MS, with attacks manifesting as subacute, focal, mild sensory and motor deficits and typical corresponding MS lesions on magnetic resonance imaging (MRI) brain and spinal cord. At baseline, she had ambulated without assistive devices and had been clinically and radiographically stable on teriflunomide for approximately 2 years. Twelve months before referral to our facility, she had sudden onset right lower back pain and loss of sensation in the entire right lower extremity immediately following horseback riding. She then had rapid, severe, right lower extremity weakness followed within 30 min by severe left lower extremity weakness and pain throughout both lower extremities. Her neurological exam within hours revealed severe flaccid paraparesis, sensory level at T7, and depressed deep tendon reflexes. Magnetic resonance imaging (MRI) of the brain and entire spine were obtained on the day of symptom onset, revealing faint new T2-hyperintense signal from T6 through T8 without contrast enhancement, and stable T2-hyperintense non gadolinium enhancing demyelinating lesions at T8-9 and T11-12 (Fig. 1a). Diffusion weighted imaging (DWI) and apparent diffusion coefficient (ADC) sequences were not performed. Serum aquaporin-4 (AQP4)-IgG and myelin oligodendrocyte glycoprotein (MOG)-IgG autoantibodies were negative. Cerebrospinal fluid (CSF) studies were not performed. The findings at the time were felt consistent with a recurrent severe attack of inflammatory myelitis secondary to the established diagnosis of MS.

She was treated with 1000 mg of intravenous methylprednisolone daily for 5 days without improvement. Given the suspicion of a refractory inflammatory MS attack, she was treated with plasma exchange for seven total treatments over the course of 2 weeks with minimal initial improvement followed by symptomatic worsening after the fourth exchange. Completion of plasma exchange and a repeated course of intravenous methylprednisolone after the clinical worsening led to no improvement.

MRI performed 10 days after symptom onset showed significant change in imaging from initial MRI, with an extensive T2-hyperintense lesion extending from the T6 to the T10-T11 level along with a linear craniocaudal strip of contrast enhancement (Fig. 1b and c). Her MS disease modifying therapy was escalated from teriflunomide to rituximab. On hospital discharge she had severe paraparesis and bladder

* Corresponding author.

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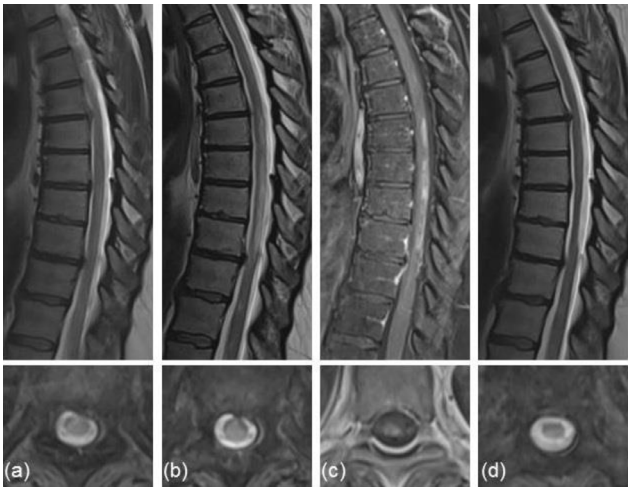

Fig. 1. Evolution of MRI findings of spinal cord infarction in a patient with multiple sclerosis Fig. 1. Thoracic spinal cord magnetic resonance imaging (MRI) of the patient obtained on day 0, day 10, and 1 year following symptom onset. (a) Day 0. Faint T2-hyperintense signal from T6 through T8. (b,c) Day 10. Extensive T2-hyperintense lesion extending from T6 to the T10-T11 level (image b) with a linear craniocaudal strip of contrast enhancement (image c). (d) Residual atrophy of the spinal cord at the site of the lesion.

dysfunction requiring self-catheterization.

Due to lack of improvement over time, she sought a second opinion at our facility one year after her hospitalization. She could take small steps with a walker but otherwise required a motorized wheelchair for mobility. She continued to experience pain in the right lower back and hip. Her neurological exam showed severe bilateral sensory loss and bilateral lower extremity weakness in an upper motor neuron pattern. Repeat spinal cord MRI revealed thoracic cord atrophy at the site of the recent longitudinally extensive lesion (Fig. 1d).

Based on the myelopathy features of hyperacute severe deficits within an hour, associated severe pain, activity at onset, spinal cord lesion spanning 5 vertebral segments with subsequent severe atrophy, and the initial gadolinium enhancement pattern of a linear craniocaudal strip, she was diagnosed with spontaneous spinal cord infarction (SCI) separate from her relapsing remitting MS. The etiology of the SCI was uncertain (common in SCI) but presumed to be either related to her known vascular risk factors or from a fibrocartilaginous embolism in the setting of physical activity. Her vascular risk factors were optimized and it was recommended that she could return to her initial MS disease modifying therapy if desired as breakthrough MS disease activity was not the cause of the severe impairment.

3. Discussion

This case highlights the under-recognized features of SCI, which were required to discriminate her presentation from inflammatory myelitis of MS.

The important discriminating features included: severe paraparesis within an hour of onset, severe pain with onset, and a longitudinally extensive lesion ($\geq$3 vertebral segments), all quite typical of a spontaneous SCI. Such important discriminating clinical and radiological features have recently been highlighted with a large case series and proposed clinical/research criteria (Zalewski et al., 2019).

SCIs have a number of typical clinical, radiographic, and laboratory characteristics that can be used to aid in diagnosis. Key clinical features include severe nadir of myelopathy deficits within 12 h (77%), associated pain with onset (72%), and the presence of vascular risk factors (76%). Typical radiographic features as highlighted by this case include a longitudinally extensive lesion (60%), variable axial T2-hyperintensity patterns, craniocaudal strip of gadolinium enhancement, and follow-up imaging demonstrating atrophy at the site of infarction (Zalewski et al., 2019). A cerebrospinal fluid analysis is generally recommended in the diagnosis of spontaneous SCI to help exclude inflammation and specific alternative etiologies, however in this case oligoclonal bands and/or an elevated IgG index would have been expected in the context of the patient's established diagnosis of MS.

The typical presentation of myelitis in MS that was not seen in this case includes sensory predominant deficits evolving subacutely over days to weeks, which are usually mild to moderate in severity (Zalewski and Flanagan, 2018). A mildly inflammatory CSF profile is expected, with possible mild elevations in total nucleated cell count, supernumerary oligoclonal bands in ~85%, and elevated IgG index in ~70% (Gajofatto et al., 2010). Spinal cord lesions in MS are usually well-delineated ovoid lesions, involve the periphery of the cord (most commonly dorsal), typically involve less than 1/3 the cross sectional area of the cord, and are almost always less than 3 vertebral segments long in adults. Acute lesions usually enhance with gadolinium, either homogeneously or with ring-like enhancement. After an episode of MS-related myelitis, a well-defined focal T2-hyperintense lesion remains (Zalewski and Flanagan, 2018). Patients generally demonstrate minimal residual disability in one functional system (Expanded Disability Status Scale score of 2.0) at 1 year (Presas-Rodriguez et al., 2016), although more disabling attacks can be seen.

Establishing the correct diagnosis of SCI as opposed to an attack of myelitis is important. It can help avoid unnecessary alterations in disease modifying therapy and use of acute rescue treatments such as plasma exchange, which could lead to worsening of ischemia from treatment related systemic hypotension. It is also important for appropriate long term management, including optimizing vascular risk factors.

4. Conclusion

Our case highlights the clinical, radiological and therapeutic differences between SCI and MS in a unique patient with both present.

Declaration of Competing Interest

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