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Brown-Séquard syndrome as a presentation of idiopathic transverse myelitis

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ABSTRACT

Brown-Séquard syndrome refers to a set of signs and symptoms caused by hemisection of the spinal cord from various sources. It may have multiple causes; traumatic injuries are the most frequent ones. The less common causes include inflammation, ischemia, tumors, or infections.

This report is about a 12-year-old boy with an acute and progressive course of right hemisection of the spinal cord, with ipsilateral hypo/areflexic paralysis and contralateral loss of thermalgesic sensation. The MRI of the spinal cord showed inflammation in the right side of the spinal cord at the level of the second and third thoracic vertebrae. The patient was diagnosed with idiopathic transverse myelitis and was started on intravenous high-dose corticosteroids; he showed a favorable clinical course and recovered neurological functions.

Keywords: *transverse myelitis; Brown-Séquard syndrome; demyelinating diseases; pediatrics.*

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INTRODUCTION

The syndrome resulting from hemisection of the spinal cord was first described in 1850 by British physician Charles-Édouard Brown-Séquard (1817–1894).[1] It is caused by an injury to one side of the corticospinal tract and the spinothalamic tract. Its asymmetric clinical manifestations are due to the decussation of the spinothalamic tract, which runs along the same side and decusses one or two levels above the introduction of these fibers into the spinal cord. The involvement of ipsilateral autonomic fibers does not affect the sphincters.[2] Its causes vary; traumatic injuries are the most frequent ones (injuries from firearms or stab wounds).[3,4] Less common causes include infarction, spinal cord tumors, inflammation, or a herniated disk.[2]

CASE REPORT

Our patient was a 12-year-old boy with no relevant medical history, who, following an abdominal trauma referred by the family as minor (he was hit by a ball during a soccer game), started with severe pain of short duration in the epigastric region; 6 hours later he developed progressive weakness of the right lower limb that prevented him from walking. He visited the emergency department 24 hours after the onset of symptoms, where flaccid paralysis of the right lower limb was observed; he was then hospitalized.

The boy lived in an urban area with his mother and his 2 older siblings, in a house with all the necessary utilities; no exposure to environmental contamination was noted during history taking. He attended first year of high school, had a good academic performance and reported not using tobacco, alcohol, or drugs. He had not received any vaccine in the past few months and, as a relevant piece of history, it is worth mentioning that he had had probably viral pharyngitis 10 days before.

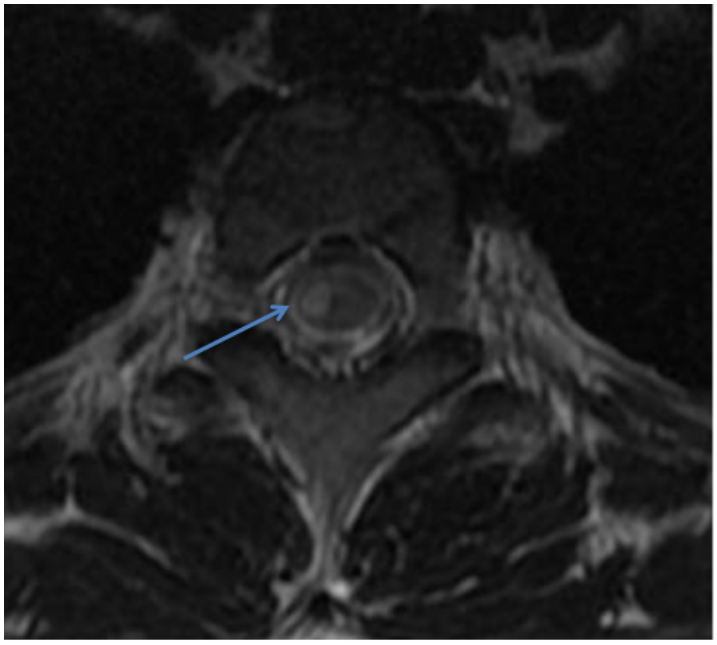
On physical examination, the patient was alert, his Glasgow score was 15/15, and had no alteration in the cranial nerves. He had flaccid paralysis of the right lower limb, from the pelvic girdle, with Achilles tendon areflexia and patellar hyporeflexia that prevented independent gait, without clonus or Babinski sign; he also had loss of thermalgesic sensation and preserved ipsilateral proprioception. He had paresthesia and loss of thermalgesic sensation on the outer side of his left lower limb, which radiated to his navel. Strength, myotatic reflexes, and proprioception were preserved; bilateral skin-abdominal reflexes were absent. There were no signs of dysautonomia or sphincter involvement.

The patient was diagnosed with Brown-Séquard syndrome, and the following causes were proposed: inflammation, ischemia (vascular territory of the anterior spinal artery), or trauma. The following supplementary tests were indicated: magnetic resonance imaging (MRI) of the spinal cord (SC) with gadolinium, which showed lateral inflammation on the right side of the spinal cord, at the level of the second and third thoracic vertebrae, which measured 2–3 cm, hyperintense in T2, gadolinium enhancement (*Figures 1 and 2*); normal MRI of the brain and optic nerves; normal visual evoked potentials; cerebrospinal fluid (CSF) with a mononuclear cell, a CSF protein level of 61 mg/dL, a CSF glucose level of 61 mg/dL, and lactate of 1.8 mmol/L; oligoclonal bands (OCBs) without intrathecal production of IgG antibodies (by kinetic nephelometry), and negative enterovirus. Anti-aquaporin-4 IgG (AQP4 IgG), anti-myelin oligodendrocyte glycoprotein (MOG)-IgG were tested in serum by indirect immunofluorescence in cells transfected with aquaporin 4 and MOG (cell-based assay), together with serologies for Epstein-Barr virus, cytomegalovirus, herpes simplex 1 and 2, and human immunodeficiency virus; all tests were negative.

The patient was diagnosed with acute transverse myelitis; treatment was started with daily intravenous methylprednisolone pulses at a dose of 1 g for 5 days; upon partial improvement, a second cycle was indicated in order to obtain the maximum possible recovery. He then continued treatment with oral meprednisone at 1 mg/kg/day, tapered down over 4 weeks. He continued with physiological doses of hydrocortisone until his cortisol levels were back to a normal range. He started physical therapy early. The patient continued with favorable course and progressive recovery of muscle strength. He was able to walk independently upon discharge and the only sequelae he had was, 6 months later, a slight decrease in right foot extensor strength with hyperreflexia in the right leg, with no Babinski sign, and with occasional paresthesia in the contralateral leg. The rest of the neurological exam was within normal ranges.

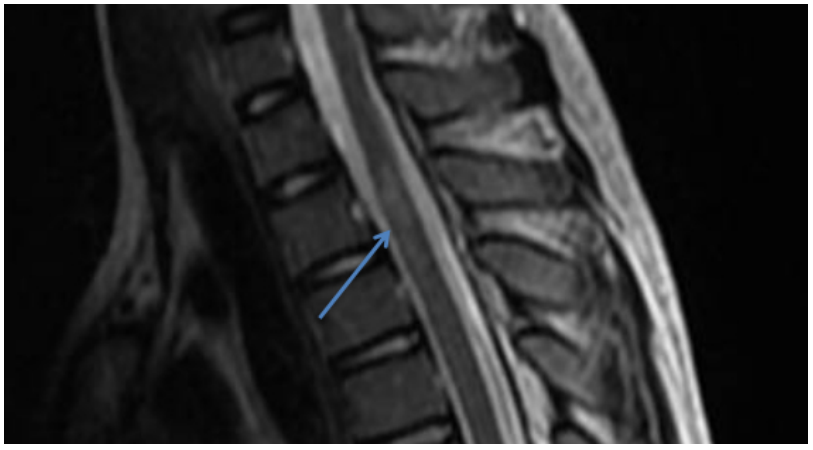
The patient continued receiving neurological follow-up every 6 months for the following 2 years, with no new clinical events.

Figure 1. Transverse section of the magnetic resonance imaging of the spinal cord



Hyperintense image in T2 in the right side of the spinal cord.

Figure 2. Sagittal section of the magnetic resonance imaging of the spinal cord



Hyperintense, 2-cm image in the second and third thoracic vertebrae, with intravenous contrast enhancement.

DISCUSSION

Acute transverse myelitis may have an idiopathic onset or become the onset of multiple sclerosis (MS), neuromyelitis optica (NMO), or develop in the context of systemic autoimmune diseases or post-infectious conditions. Its incidence worldwide is low: 1 to 4 cases per million/year.[5,6] According to a case series observed at the Johns Hopkins Transverse Myelitis Center, 28% of cases occur in the pediatric age group, 40% report a history of non-specific febrile symptoms, and 30% received a vaccine in the 3 weeks prior to the onset of symptoms. The neurological deficit has an acute onset (more than 4 hours and less than 4 weeks) and involves the motor, sensory, and autonomic nervous systems at the level of the spinal cord inflammation. Pain in the dorsal region, abdomen,

or extremities is a frequent symptom at onset.[3]

This patient was reported as having a rare case of Brown-Séquard syndrome as a presentation of transverse myelitis. It had an acute onset, with abdominal pain after a minor traumatic injury. No associated infections or immune-related causes were observed, except for a mechanism that triggered the previous respiratory symptoms, so it is defined as idiopathic transverse myelitis, as per the international diagnostic criteria proposed by the Transverse Myelitis Consortium Working Group, published in 2002.[6]

There have been few reports of pediatric and adult patients with this form of presentation in the bibliography.[7–10] One of them referred to an 11-year-old girl;[8] in the adult population, no triggering events were observed, except for a case triggered after the administration of the flu vaccine.[7]

The supplementary test of choice for diagnosis is the MRI of the spinal cord with gadolinium, which allows to enhance spinal cord inflammation and, at the same time, to rule out traumatic, ischemic, or compression injuries. It should be complemented with images of the central nervous system (CNS) to assess for demyelinating lesions at this level. Serological and CSF studies may be used to assess triggering causes and associated infections or immune factors. In 75–90% of cases, it is an isolated episode; however, 10–25% of patients have a possibility of recurrence or progression to MS or NMO spectrum disorder. *Table 1* describes their predictors. The presence of positive serological markers (anti-AQP4 or anti-MOG IgG) may help to predict the course of disease. The persistence of positive serology for anti-MOG antibodies implies a risk for recurrence of up to 75% (taking into account that, in 60% of pediatric patients, titers tend to decrease after the acute onset); in cases of anti-AQP4 IgG, the possibility of recurrence is more than 50% in the following 12 months. The presence of intrathecal OCBs (positive in CSF and absent in serum) is the typical finding in MS and implies the presence of a chronic immune response restricted to the CNS.[10] CNS lesions and female sex have also

Table 1. Predictors of recurrence or progression

Predictors of recurrence after transverse myelitis	
MRI	Diffuse injuries Fusiform lesion involving more than 3 spinal cord segments Demyelinating injuries in the CNS
CSF	Positive oligoclonal bands
Other	Optic nerve involvement Positive autoantibodies (ANA, dsDNA, c-ANCA, SNCA, and NMO-IgG)
Predictors of progression to MS-NMO	
Clinical	Prior demyelinating disease Asymmetry, prevalent sensory symptoms
MRI	Demyelinating injuries in the CNS Involving more than 2 segments and less than 50% of the SC diameter (for MS) Involving more than 3 segments (for NMO)
CSF	Positive oligoclonal bands
Other	Optic nerve involvement Positive autoantibodies (anti-AQP4 and anti-MOG)

Modified from reference 5.
MRI: magnetic resonance imaging.
CNS: central nervous system.
CSF: cerebrospinal fluid.
ANA: antinuclear antibody.
dsDNA: double-stranded DNA.
c-ANCA: anti-neutrophil cytoplasmic antibody.
SNCA: alpha-synuclein.
IgG: immunoglobulin G.
NMO: neuromyelitis optica.
MS: multiple sclerosis.
SC: spinal cord.

been described as risk factors for recurrence.[11,12]

Advances in diagnostic methods, particularly neuroimaging, serological tests, and CSF studies, have decreased the proportion of patients diagnosed with idiopathic transverse myelitis.[13]

Treatment begins with high doses of intravenous corticosteroids due to the immune mechanism of the lesion. The purpose is to accelerate recovery and improve patients' prognosis. Although no adequately designed studies have been conducted due to the low frequency of this disease, this is considered the first line of treatment. In patients with little response to corticosteroids, the next therapeutic step is therapeutic plasma exchange[13–15] and, in refractory or recurrent cases, different immunosuppressive therapies or immunomodulators are indicated, including cyclophosphamide, azathioprine, mycophenolate, among others.[5]

Another pillar of treatment is physical therapy, which should be started early to prevent secondary complications, such as immobility, contractures, and bedsores. The patient described here received a cycle of intravenous corticosteroids with an adequate response, so it was decided to give him a second cycle to obtain the maximum recovery during the acute phase.

The presentation of this patient is interesting because it is a rare, pediatric case of Brown-Séquard syndrome as a clinical presentation of idiopathic transverse myelitis. Although traumatic injuries are the most frequent causes of this syndrome, inflammatory causes should be considered in the differential diagnosis. ■

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