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CASE REPORT

Transverse myelitis in a teenager

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Abstract

The objective of this study is to report the case of an adolescent patient with an acute myelopathy evolving to paraplegia in a few hours, with alteration in neuroimaging compatible with complete transverse myelitis, referred to our service with diagnosis of optic neuromyelitis.

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INTRODUCTION

Transverse myelitis (TM) is a term used to describe a spinal cord inflammation, with varied etiology. It may be inflammatory, vascular, paraneoplastic, radiation-related, and idiopathic¹. The diagnosis is of exclusion and neuroimaging is initially performed to rule out other causes, such as compression disorders². Once ruled out, cases may be subdivided into acute idiopathic transverse myelitis or acute transverse myelitis associated with some secondary disorder, such as multiple sclerosis.

The prevalence of TM is estimated to be between 1.34 and 4.6 per million, and the incidence is up to 3.00 million patients per year (0.003%). The most common symptoms are back pain - usually in the area of the injury-corresponding dermatome, lower limb paraparesis, sensory disorders and autonomic dysfunction³.

When transverse myelitis involves three or more vertebral segments, it is called extensive longitudinal transverse myelitis (MTLE)⁴. This condition is commonly associated with optic neuromyelitis (ONM), an autoimmune disease of the central nervous system, characterized by the involvement of the optic nerves and medulla spinal. However, extensive spinal cord injury may be present in other diseases⁵.

CASE REPORT

Patient CCB, 16 years old, male, brown, single, came to the Gaffrée and Guinle University Hospital (HUGG) accompanied by his mother. The young man reported that six months ago, in July of 2015, during a rehearsal for the end of year holiday celebration, he presented with intense sudden pain in the lumbar region that ascended to the thoracic region, evolving to paraplegia in around 2 hours. The patient received immediate medical attention and remained in the ICU for 23 days with a bladder catheter due to urinary incontinence. He was submitted to skull and spine neuroimaging, cerebrospinal fluid testing, general laboratory tests, and underwent methylprednisolone pulse therapy without improvement. Subsequently, he was transferred to the SARAH medical care network, where he was submitted to a visual evoked potential (PEV) test, when Devic's disease was suggested as a possible diagnosis. After hospital discharge, he was referred to the neuroimmunology clinic.

During the consultation at the HUGG, the young man said he had had no episode of upper airway infection, exanthema, or vaccination prior to his symptoms, denied smoking, alcoholism, and comorbidities. At the time of the neurological examination, he had been restricted to the wheelchair, with a bladder catheter, and he was clear-minded and oriented in time and space/ his pupils were isochoric light-reactants; he had normal cranial nerves test and his motor, sensory and coordination functions were preserved in the upper limbs. He was hypotonic in his lower limbs with global areflexia. His tactile sensitivity had been spared, but he had painful and thermal anesthesia at the xiphoid appendix (T6) level. He

also had distal symmetrical hypopallesthesia, and preserved segmentary position.

Complementary tests: His magnetic resonance imaging (MRI) of the skull (07/22/2015) was normal.

His thoracic spine MRI showed degenerative discopathy in D6-D7; posteromedial focal disc protrusion in D5-D6, touching the ventral face of the dural sac; left posterolateral focal disc protrusion in D8-D9. Schmorl nodules on vertebral plateaus affixed to D6-D7, D7-D8, D9-D10, D10-D11, D11-D12; normal spinal canal amplitude. Intrinsic signal change of the anterior portion of the thoracic spine at the level of D6-D7 to D10-D11. (Annex - Figure 1 and Figure 2).



Figure 1. spine MRI - sagittal cross-section Intra-substance signal change from the thoracic spine at the levels of D6-D7 to D10-D11.

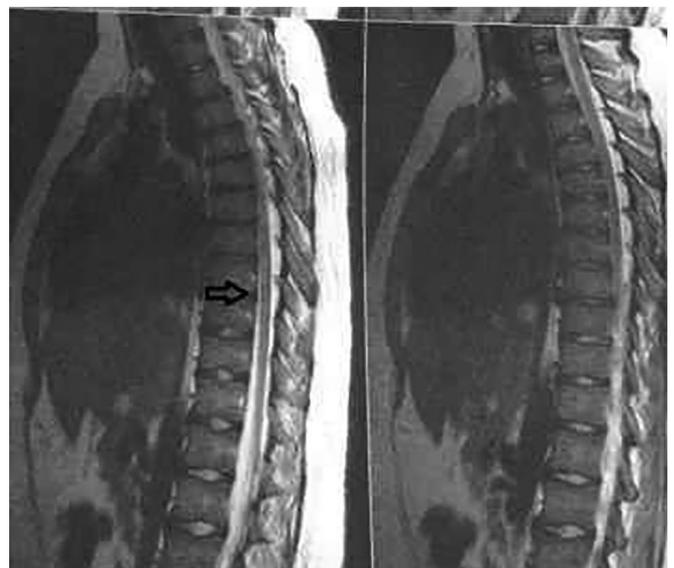


Figure 2. Spine MRI Intra-substance signal change from the anterior portion of thoracic spine at the levels of D6-D7 to D10-D11.

CSF analysis (19/06/2015) showed 4 red blood cells, 31 leukocytes (0 neutrophils, 0 eosinophils, 87 lymphocytes, 13 monocytes, and 0 macrophages), 91 mg/dl of protein and 55 mg/dl of glucose. Bacterioscopy and serology were negative, and there were no oligoclonal bands. The final report was compatible with nonspecific inflammatory process. Other tests performed for diagnostic investigation were negative serology for antibody to aquaporin 4 in the cell-based assay (CBA), another normal visual VEP. He also had normal cerebral arteriography. As for treatment, he was referred for motor rehabilitation, and is followed up by the neuroimmunology department. One year and six months after the event, he showed a slight improvement with right lower limb plegia and lower left limb paresis (grades III/V), painful and thermal hypoesthesia up to T6, normal tactile sensitivity, without urogenital control.

DISCUSSION

The case reported is about a young, previously healthy patient, who presented a complete transverse myelitis preceded by pain of acute installation, evolving to plegia within 2 hours. Immediate investigation by neuroimaging examination showed a signal alteration in the anterior portion of the thoracic spine at D6-D7, level with extension to D10-D11 (in annex - figure 1 and 2), and the cerebrospinal fluid examination had inflammatory characteristics, leading to suspected autoimmune etiology. Pleocytosis greater than 10 cells, with absence of oligoclonal bands and normal cranial magnetic resonance imaging reduced the possibility of a multiple sclerosis diagnosis. However, it initially suggests myelitis associated with the spectrum of optic neuromyelitis (NMO), due to the medullary involvement extension - more than three segments affected. However, the serological investigation for anti-aquaporin 4 antibody by cell-based assay (CBA), with specificity around 90%, was negative. Although the negativity of this test does not totally exclude the possibility of an NMO complex syndrome, the onset characteristics and lack of changes to the optic nerves decreases the likelihood of such this condition.

After ruling out NMO, we suspected of an idiopathic transverse myelitis, but among the exclusion criteria, the patient had clinical distribution pointing to an occlusion of the anterior spinal artery.

The neuro MRI showed a longitudinally extensive spinal cord lesion, for which the main diagnostic considerations are inflammatory myelitis secondary to infection, autoimmune disease and spinal cord ischemia. Medullary vascular accident is rare, approximately 0.3-1% of all strokes, and it is classified into upper VA (neck) and lower VA (thoracolumbar). It is a rare cause of acute myelopathy, accounting for only 1% of all ischemia and 5% to 8% of acute myelopathy⁶.

The most common clinical condition is the anterior spinal artery syndrome, which is clinically characterized by an acute onset of symmetrical motor weakness and bilateral spinothalamic sensory deficit below the level of the lesion in conjunction with autonomic sphincter dysfunction, but with preservation of vibration and proprioceptive sensitivity⁷.

Nowadays, atherosclerotic disease and thoracoabdominal surgery are among the most frequent causes of spinal infarction, other causes are patent foramen ovale, bacterial endocarditis, vasculitis, infection, hematological disorders, decompression sickness, iatrogenic. Among the causes, fibrocartilaginous embolism is due to an increase in the intradiscal pressure of the nucleus pulposus, leading to medullary infarction by retrograde embolization to the central artery, and some theories consider disc herniation and Schmorl nodules as risk factors, but it is yet to be proven⁸. Some criteria for fibrocartilaginous embolism are sudden onset of symptoms, with or without a trigger (such as small trauma, physical exertion), MRI changes compatible with ischemic myelopathy and/or evidence of disc herniation or disc degeneration, and few vascular rich factors. In the case reported, the patient had no history of trauma and had no cardiovascular risk factors, but the MRI presented degenerative discopathy, disc protrusions and Schmorl nodules^{7,8}.

CONCLUSION

We have shown a case of longitudinally extensive myelitis, of probable ischemic etiology, initially diagnosed as NMO. The purpose of our study was to compare the clinical and radiological characteristics of these disorders, and to emphasize that early identification of the underlying etiology is vital to initiate appropriate therapy.

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