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Short-segment myelitis presenting as Brown-Séquard-plus syndrome as the initial attack of multiple sclerosis

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Abstract

Brown-Séquard syndrome is an uncommon condition accounting incomplete spinal cord injury, manifesting as ipsilateral weakness and proprioception loss, associated with contralateral pain and temperature sensation loss. Brown-Séquard-plus syndrome (BSPS) has not quite a well-stablished definition, but usually is defined as a Brown-Séquard syndrome associated with clinical findings compatible with another spinal cord tract. We present a pearls & oysters article naming the most relevant findings and aspects to evaluate in this pathology, throughout a clinical case where a patient presents a Brown-Séquard-plus syndrome as the initial attack in multiple sclerosis.

Key words: Brown-Séquard-plus syndrome, short-segment myelitis, multiple sclerosis.

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Introduction

Brown-Séquard syndrome is an uncommon condition accounting incomplete spinal cord injury, manifesting as ipsilateral weakness and proprioception loss, associated with contralateral pain and temperature sensation loss. Brown-Séquard-plus syndrome (BSPS) has not quite a well-stablished definition. Issaivanan described BSPS as a classic Brown-Séquard syndrome associated with additional neurologic findings, which may involve autonomic symptoms (bowel or bladder disfunction) or bilateral proprioceptive loss [2] Other authors suggest the term Brown-Séquard-plus syndrome for patients that present similar but not the full characteristics of a Brown-Séquard syndrome, as a case reported with classic BSS with Horner Syndrome and bilateral extensor response [3]. We report a case of a short-segment myelitis presenting as Brown-Séquard-plus syndrome as the initial attack of multiple sclerosis.

Case report

A 55-year old woman without any relevant past medical history (nor trauma) initiated with progressive right hemihypesthesia. She noticed a slight decrease in temperature sensation in her right side of the body when she was taking a shower in the morning, until she was unable to distinguish between hot and cold water in the afternoon. A week later, she noticed progressive weakness in her left arm and leg combined with sensory ataxia, until she could not lift objects nor walk. 5 days after her weakness started, she was incapable to voluntarily control her urinary sphincter. She was evaluated in the emergency room where we found left side hemiparesis with a 3/5 Medical Research Council score (Both upper and lower extremity), ipsilateral hiperreflexia and left extensor response. Regarding the sensitive examination, she had absent pain and temperature sensation in her right side of the body, a sensory level below C2, complete bilateral loss of tactile, vibratory

and position sensation and absence of voluntary urinary function. A clinical Brown-Sequard-plus syndrome was made.

Cervical magnetic resonance imaging (MRI) revealed an intramedullary lesion affecting the C2 spinal cord, comprising the left half of the spinal cord and bilateral posterior columns, with punctate gadolinium enhancement (Figure). Methylprednisolone pulses were given for 5 days (1 gram/day) with considerable improvement in both sensitive and motor functions. The patient was able to walk, regained temperature sensation, and she also was able to control urinary and anal sphincters again. Further workup revealed present oligoclonal bands in cerebrospinal fluid and brain MRI showed several periventricular lesions with demyelinating characteristics. According to the McDonald revised criteria of 2017, the patient proved to have a clinical attack with objective clinical evidence, dissemination in space implicating a different CNS site and dissemination in time demonstrated by CSF-specific oligoclonal bands, concluding the diagnosis of multiple sclerosis [1].

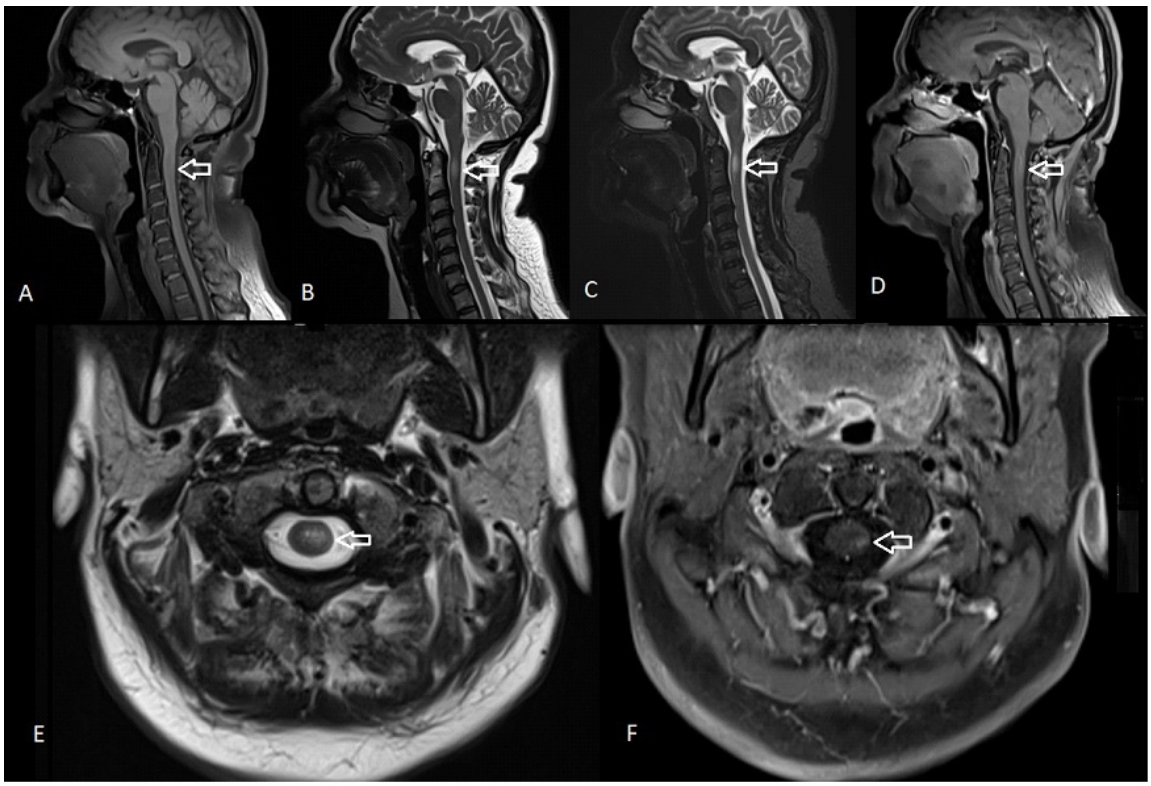

Figure. Cerebral and spine MRI. A. Sagittal T1-wigther cervical spine image reveals short segment hypointensity at the level of C2. B and C. Sagittal T2-weighted and STIR images showing short segment hyperintensity. D. Post-gadolinium linear enhancement on T1-weighted image. E. Axial T2 showing left lateral intramedullary hyperintensity and (F) punctacte gadolinium enhacement on T1-weigthed image.

Discussion

Short-segment myelitis (<3 vertebral segments) as the initial attack in multiple sclerosis is uncommon [4]. There are few case reports of Brown-Sequard syndrome as the initial manifestation of multiple sclerosis [5, 6, 7]. All cases were retrospectively analyzed, because multiple sclerosis diagnosis needs to be constructed with multiple clinical and imaging findings. Another thing to consider regarding these cases, is that the diagnostic criterion used depend the year of the case report. Although rare, the principal etiologies related to myelitis presenting as Brown-Sequard syndrome are infection with cytomegalovirus, herpes-zoster, diphtheria and tetanus vaccination, tuberculosis and syphilis [8, 9].

Koehler, et al. in 1986 divided clinical presentation of hemimedular syndrome as Brown-Séquard Syndrome and Brown-Séquard-Plus syndrome, and found 3 cases of association with multiple sclerosis, but with no specification on whether they considered them in the group of Brown-Séquard syndrome or Brown-Séquard-plus syndrome. The case series also considered incomplete forms of Brown-Séquard syndromes as Brown-Séquard-plus [10].

One interesting point in this case is the clinical progression. The patient initiated with diminished pain and temperature sensation, which may imply that the pattern of inflammation started in the spinothalamic tract. Later, the patient presented weakness, sensory ataxia, and proprioceptive loss, implicating that inflammation probably compromised the corticospinal tract, and both posterior columns. This is interesting, because when the patient consulted with us in the emergency department, her clinical manifestations were in that moment compatible with a Brown-Séquard-plus syndrome. The clinical evolution demonstrates the demyelinating progression in time and its relationship with the clinical picture. This addresses two important questions yet to be answered: which is the more frequent clinical progression in short segment myelitis in cases with multiple sclerosis diagnosis? and does the clinical picture suggests the pattern and degree of inflammation?.

Based on the different descriptions of the cases and series reviewed, we intend to propose a definition that appropriately addresses full characteristics of this syndrome. Our proposal for the clinical definition of a Brown-Séquard-plus syndrome is the picture of a classical Brown-Séquard Syndrome (ipsilateral weakness and proprioceptive loss, and contralateral pain and temperature sensation loss) plus symptoms and/or signs of other spinal cord pathway affection. This may include the sympathetic pathway (ipsilateral Horner syndrome), the contralateral posterior columns (bilateral proprioceptive loss), contralateral corticospinal pathway (bilateral weakness) and autonomic pathways implicating bowel and bladder function (urinary retention and bowel disfunction).

This definition intends to name appropriately certain medullary syndromes that don't fulfill criteria neither for complete medullary syndrome nor for a classical Brown-Séquard syndrome.

Conclusion

Clinical diagnosis of Brown-Séquard-plus syndrome includes a classical Brown-Séquard syndrome (ipsilateral weakness and proprioception loss, associated with contralateral pain and temperature sensation loss) plus another clinical sign or symptom that implies another spinal cord tract affection. Sclerosis multiple should always be considered as part of the etiological workup in patients with subacute-onset Brown-Séquard-plus syndrome without trauma history.

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Artículo sin conflicto de interés