

Brown-Séquard Syndrome as a First Presentation of Multiple Sclerosis

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Abstract

A female patient aged 48 years presented with sub-acute onset of weakness in right upper and lower limb over the past one month and numbness over left side of body below neck level. Multiple sclerosis (MS) presenting as Brown-Séquard syndrome is very rare. We present a case of hemicord myelitis which presented as Brown-Séquard syndrome as a first manifestation, which was later diagnosed as MS during subsequent relapses.

Keywords: Brown-Séquard syndrome, hemicord myelitis, multiple sclerosis, paraparesis, idiopathic myelitis

Introduction

Brown-Séquard syndrome is a rare neurological syndrome first described by Charles Eduard Brown Séquard in 1850 as ipsilateral upper motor neuron motor weakness below the level of lesion and lower motor neuron type at the level of lesion with loss of ipsilateral proprioception with contralateral loss of pain and temperature sensations below the level of lesion. This clinical picture resembles that of hemisection of cord. Trauma to cervical cord (1) and cervical disc herniations (2) are the usual cause of this syndrome, but this syndrome is also rarely reported in spinal cord tumours, infarctions, hematoma, tuberculosis, herpes zoster, syringomyelia, decompression sickness (3–5), and multiple sclerosis (MS). We hereby report a case of Brown-Séquard syndrome as a first presentation of MS.

Case Report

A female patient aged 48 years presented with sub-acute onset of weakness in the right upper and lower limb over the past one month and numbness over the left side of body below the neck level. She is a diabetic and is on oral hypoglycemic drugs, for the last one year. Past history revealed that she had been earlier evaluated with Magnetic Resonance Imaging (MRI) of the brain and electroencephalography for one episode of generalised tonic-clonic seizures, a year back after which she was started on sodium valproate. Both her MRI and electroencephalography studies were normal at that time. Her general physical examination was normal. Mental functions, speech and cranial nerve examination were normal. Motor system examination revealed Medical Research Council (MRC) Grade 2/5 power on the right upper and lower limb with normal power on the left side. Deep tendon reflexes were brisk on the right side of body and were normal on the left side. Right side plantar reflex showed extensor response and

left side was flexor. Sensory system examination revealed 75% loss of pin-prick and thermal sensations below the left C2 dermatome and was normal on the right side. Proprioception was impaired on the right side of body.

In view of the above findings a diagnosis of Brown-Séquad syndrome was made and MRI of cervical spine was obtained which showed hyperintensity in the short segments of the right hemicord from C2–C3 to C6–C7 levels in sagittal section (Figure 1) and T2 axial section (Figure 2), which confirmed the diagnosis of Brown-Séquad syndrome. The imaging differential diagnosis were tuberculosis, low grade neoplasm and MS. MS seemed less likely because MRI of brain was normal at this instance (Figure 3). Cerebrospinal fluid analysis revealed two lymphocytes, sugar 63 mg/dl, protein–67 mg/dl and adenosine deaminase–0.37 U/L. Therefore a diagnosis of idiopathic myelitis was made and was started on injection methylprednisolone 1 g for 5 days and the patient improved substantially and was discharged on 7th day. Patient was asymptomatic at the follow up visit after a month and continued to do so.



Figure 1. T1W sequence of MRI of the cervical spine sagittal section during the first attack with arrow showing short segment hyperintensity in the right hemicord at C2–C3 and C6–C7 levels 87 mm × 127 mm (150 × 150 DPI)

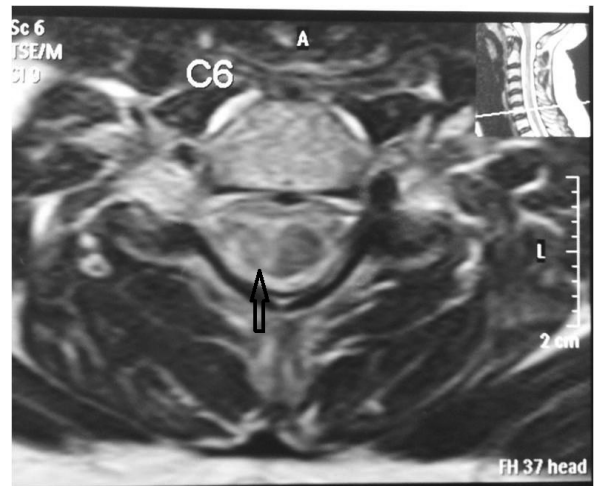


Figure 2. MRI of the cervical spine T2W axial section at C6 level during the first attack (arrow showing hyperintensity in the right hemicord) 133 mm × 110 mm (150 × 150 DPI)

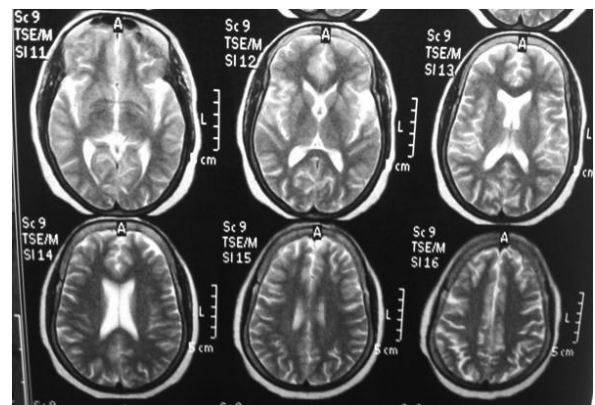


Figure 3. First MRI of the brain axial T2W Section during the first episode showing no significant abnormality 151 mm × 104 mm (100 × 100 DPI)

After a year, she again presented with sudden onset of paraplegia. She had MRC Grade 1/5 power in both lower limbs with normal power in the upper limbs. She had reduced sensory level from T5 dermatome downward. MRI spine sagittal section revealed multifocal short segment hyperintense plaques at C1–C2, C5–C6 and T1–T3 levels (Figure 4). MRI of the brain sagittal section at this time showed multifocal hyperintense lesions in the white matter of right corona radiata, centrum semiovale, frontoparietal lobes and bilateral post central gyrus and specifically perpendicular to the ventricles (Dawson’s fingers) which were

highly suggestive to represent demyelinating plaques typical of MS (Figure 5). Therefore, keeping in mind the gradual disease progression and appearance of new hyperintense multifocal lesions in MS typical areas, the diagnosis of relapsing and remitting type of MS was made, which met Revised McDonald's Criteria (10) with two clinical attacks and objective clinical evidence of two lesions, which was also supported by MRI which showed T2 hyperintense lesions in more than one anatomical regions namely periventricular and spinal cord. The patient was treated with injection methylprednisolone 1g for 5 days and was discharged. Patient improved significantly in terms of lower limb strength. Subsequently she has had another two relapses of paraplegia. She is currently on Injection Glatiramer acetate therapy.



Figure 4. MRI of the spine T2W sagittal section 9 months after the first attack with arrows showing short segment multifocal hyperintense plaques at C1–C2, C5–C6 and T1–T3 levels
76 mm × 150 mm (67 × 67 DPI)

Discussion

Brown-Séquard syndrome occurs due to involvement of ipsilateral corticospinal tract and dorsal column which accounts for ipsilateral weakness and loss of proprioception and contralateral spino-thalamic tract which causes contralateral loss of temperature and pin-prick sensations (6). Pure Brown-Séquard syndromes exhibiting all the features are very rare, instead patients present with only a part of syndrome, which then presents as a diagnostic difficulty. The hemisection syndrome may also occur with additional symptoms and signs. The primary etiologies for Brown-Séquard syndrome are associated most commonly with a traumatic injury to the spinal cord. Extramedullary spinal neoplasm, cervical disc herniation, cystic diseases, spinal epidural hematoma, MS and myelitis are reported as uncommon causes.

Our patient presented with subacute onset of symptoms suggestive of Brown-Séquard syndrome or cord hemisection which was confirmed by MR imaging of spine. In our course of evaluation detailed history provided no significant history of trauma in the past and other differentials like cervical disc herniation, epidural hematoma and cystic diseases were ruled out on MRI. Other relatively uncommon differentials namely infection, low grade glioma, neuromyelitis optica and MS were thought of. Cerebrospinal fluid (CSF) analysis ruled out infection as the cause and neuromyelitis optica (NMO) antibody test was not performed. However, the diagnosis of NMO was less likely based on short segment of spinal cord involvement, no optic nerves involvement from clinical history, examination as well as confirmed by VEP, and the subsequent brain MRI is more suggestive of MS rather than NMO. Then the differential diagnosis came down to MS and glioma. For evaluation of MS, MRI of the brain was done, which turned out to be normal. Since patient did not want to undergo spinal cord biopsy to rule out glioma, injection methylprednisolone was started empirically. Patient improved dramatically and was discharged and closely followed up. When the patient presented again with paraparesis, a repeat MRI of the spine showed multifocal hyperintense plaques at new levels (Figure 3) and also on the contralateral spine. MRI of the brain also showed multifocal hyperintense plaques in typical locations which confirmed the possibility of MS (Figure 5 and Figure 6). The

relapsing-remitting nature of the disease and the lesions which were dissociated in time and space as depicted by the initial and successive MR images strongly led to the diagnosis of demyelinating disease of MS which in our case presented initially as a cord hemisection or Brown-Séquad like syndrome.

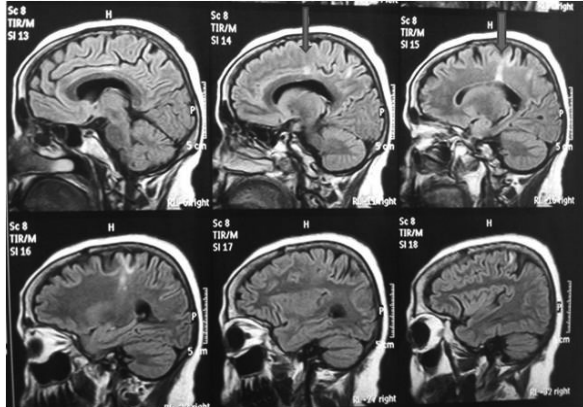


Figure 5. MRI of the brain T2W sagittal section during the second attack after 9 months with arrow showing multifocal hyperintense lesions in the white matter of right corona radiata, centrum semiovale, frontoparietal lobes and bilateral post central gyrus and specifically perpendicular to the ventricles(Dawson’s fingers)
169 mm × 89 mm (96 × 119 DPI)

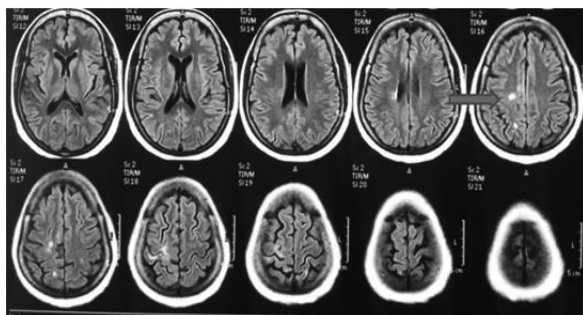


Figure 6. MRI of the Brain axial FLAIR section after 9 months during the second attack:arrow showing hyperintense demyelinating plaques in the periventricular and juxtacortical areas
169 mm × 84 mm (82 × 81 DPI)

The co-association of MS and diabetes mellitus (DM) type 2 has been reported in literature which may be because of abnormalities in fat, calcium, and vitamin D metabolism. Also, there is evidence of disruption of myelin due to changes in glucose levels. Diabetes may also occur during the course of MS because of steroid therapy. According to various epidemiologic studies women with type 1 diabetes are at a highly increased risk of MS. Further epidemiologic studies are needed to confirm these findings. MS is associated with DRB1*0405-DQA1*0501-DQB1*0301 and DRB1*0301-DQA1*0501-DQB1*0201 in Sardinia and not DRB1*15-DQA1*0102-DQB1*0602, as it is elsewhere in the world. The DRB1*0301-DQA1*0501-DQB1*0201 haplotype is also associated with type 1 diabetes and other autoimmune diseases, such as celiac disease and autoimmune thyroid disease, which are also highly prevalent in sardinia. Genetic studies are also required to evaluate the DRB1*0301, DRB1*0405, and DRB1*15 extended haplotypes for common alleles at other loci that may contribute to the familial clustering of MS and type 1 diabetes. The haplotypes which show common association with diabetes and MS are relatively rare except in Sardinia owing to their unique distribution which still needs to be investigated and studied (11). In our patient, diabetes was present prior to diagnosis of MS which was definitely a challenge in management and the linking of both disorders is mere a speculation which warrants further research. Keeping in view the current clinical knowledge and taking into consideration the relative paucity of literature linking both of these diseases, the prognosis of MS in this patient appears to be meek and the patient is more likely to be disabled in the future as it is co-existing with dia.

Till date MS stands out as one of the rarest sole presenting features of cord hemisection or Brown-Séquad like syndrome with very few cases reported in the literature (7, 8). However the exact percentage for this association could not be found out reflecting its relative rarity.

In our case, Brown-Séquad syndrome was the first and sole presenting feature of MS which manifested fully with successive attacks. We report this case due to its relative rarity and depicting how a progressive demyelinating disease like MS can masquerade initially as highly atypical presentations.

Author's Contributions

Conception and design: TKR, RS

Analysis and interpretation of the data: TKR, RS, SS, RS

Drafting of the article: CB, SS, RS

Critical revision of the article for important intellectual content: TKR

Administrative, technical, or logistic support: CB

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