Case Series

A Case Series of Non-traumatic Brown-Sequard Syndrome with Rare and Diverse Etiologies

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This case series presents a rare collection of Non-traumatic Brown-Sequard Syndrome (BSS) cases from India, showcasing the diverse and unusual etiologies contributing to this condition. BSS is characterized by a unique pattern of sensory and motor deficits resulting from spinal cord hemisection. While typically associated with trauma, these cases highlight non-traumatic origins. The series includes cases with varied etiologies such as inflammatory disorders, spinal cord hematomata and infarction adding to the understanding of BSS's pathophysiology. The clinical presentations, diagnostic challenges and management strategies are discussed, emphasizing the need for a comprehensive approach to diagnosing and treating BSS. This series highlights the importance of considering rare and diverse etiologies in the evaluation of non-traumatic BSS.

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Brown-Sequard Syndrome (BSS) is a relatively uncommon neurological condition due to the hemi section of the spinal cord first described by Charles Eduard Brown Sequard in 1850. Clinical symptoms include motor weakness, loss of joint position, vibration sense on the same side of the lesion and loss of pain and temperature sense on the opposite side of the lesion. It is broadly classified as traumatic and non-traumatic based on aetiologies. The most common causes are spinal cord trauma and spinal disc herniations[1]. Non-traumatic causes can be divided into compressive and non-compressive which are further very rare. In this series of 4 nontraumatic cases with different aetiologies.

Case 1

A 22-year-old gym trainer presented with a 12-day history of progressive left leg weakness and numbness in the right leg in July, 2018. No back pain, radicular pain, or bladder issues were reported. Similar mild symptoms occurred in February and April, resolving within 7-10 days. The patient had no major trauma, smoking, or regular alcohol consumption and he was neither diabetic nor hypertensive.

Physical examination revealed normal mental functions, speech and cranial nerves. Motor examination showed Grade 3/5 power in the left lower limb with brisk reflexes, while the right side exhibited normal power and reflexes. Plantar reflexes were extensor on the left and flexor on the right.

Sensory examination showed absent vibration and proprioception on the left below the T10 dermatome, with a 75% loss of pin-prick and thermal sensations on the right below the T10 dermatome. Vibration and proprioception sensations were preserved.

A diagnosis of Brown-Sequard Syndrome was made, and an MRI of the cervical spine was obtained to rule out various differentials, such as disc herniation, epidural hematoma, cystic diseases, infection, low-grade glioma, neuromyelitis optica and Multiple Sclerosis (MS).

The MRI revealed multiple patchy hyperintensities in the cervical and dorsal spinal cord and the conus medullaris, with gadolinium enhancement at T10 and T11 (Fig 1). Brain MRI revealed multiple T2-FLAIR hyperintense lesions without diffusion restriction, meeting Revised McDonald's Criteria for relapsing-remitting MS as shown in Fig 2.

Further evaluation eliminated trauma and other differentials through MRI, abnormal Cerebrospinal Fluid (CSF) analysis with mild pleocytosis and positive tests for CSF oligoclonal bands and raised IgG index confirming the diagnosis of multiple sclerosis. The serum antibody tests for neu-romyelitis optica and MOG were negative. The patient received methylprednisolone and was advised Disease-modifying therapy (DMRD), resulting in significant motor strength improvement.

Editor's Comment :

- This case series highlights the rarity and diversity of non-traumatic BSS aetiologies, with a particular focus on the diagnostic challenges and atypical presentations in these cases.
- Diligent clinical examination with appropriate investigations is of paramount importance for early diagnosis and prompt appropriate treatment.
- These exceptional cases advance our knowledge of non-traumatic BSS.

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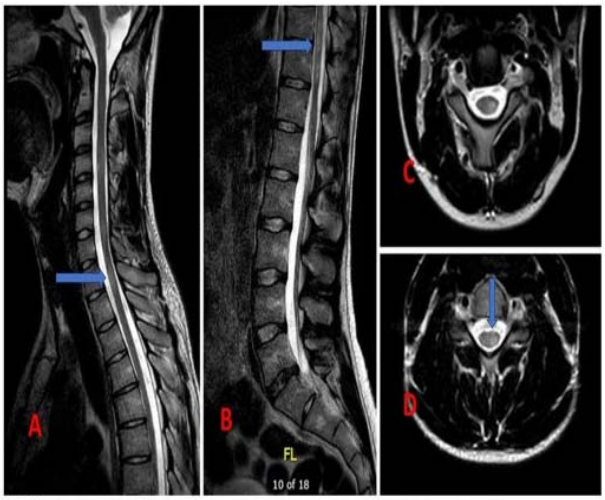

Fig 1 — Sagittal T2 MRI spine shows Multiple short segmental patchy focal areas of T2 hyperintensities are seen in the cervical and dorsal spinal cord as well as the conus medullaris (horizontal blue arrows in Images 1A and 1B). On Axial T2 imaging (Figs 1C and 1D) these lesions involves central cord and also of the peripheral sub pial location on left side of the spinal cord indicated by vertical blue arrow.

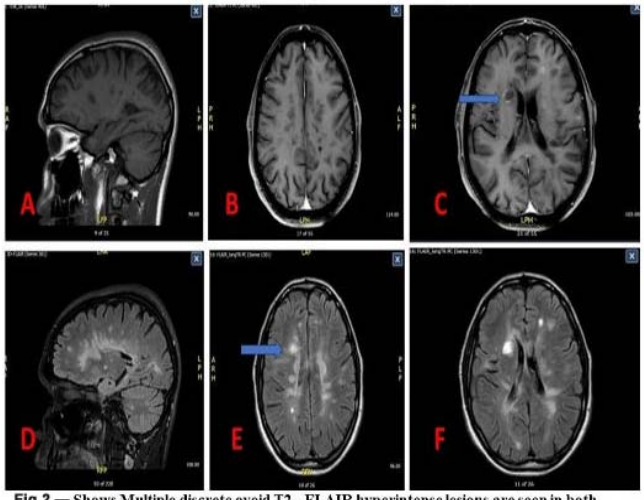

Fig 2 — Shows Multiple discrete ovoid T2 - FLAIR hyperintense lesions are seen in both supra as well as infratentorial brain parenchyma (Figs 2D, 2E &2F). These are seen in bilateral periventricular white matter involving the corpus callosum, bilateral fronto-parieto-temporal juxtacortical white matter more so in the both temporal lobes, bilateral internal capsules, cerebral peduncles, as well as deep white matter of cerebellum. Few of these lesions in the right frontal periventricular white matter indicated by blue horizontal arrow, left anterior frontal subcortical white matter show incomplete rim of enhancement (Fig 2C).

Case 2

In May 2019, a 64-year-old man with diabetes, hypertension and Myelodysplastic syndrome experienced sudden left lower limb weakness, which worsened over several days. He ultimately lost mobility in the left limb and experienced numbness in both lower limbs, with urinary retention. Subsequently, he developed right lower limb weakness.

Examinations revealed intact cognitive and cranial nerve functions. He exhibited heightened muscle tone, with left lower limb motor function at grade 1/5 and exaggerated deep tendon reflexes. Sensory examination showed reduced pain and temperature sensation on the right side below T4 and decreased joint position and vibration sensation on the left side. Follow-up exams indicated right lower limb motor function at grade 2/5 and left lower limb at 1/5, with extensor plantar reflexes and no additional sensory changes.

The diagnosis was Brown-Sequard Syndrome, a partial spinal cord injury causing one-sided motor and sensory deficits. Possible differentials included disc herniation, epidural hematoma, low-grade glioma, metastatic spread, infection, and bleeding diathesis with spinal cord hematoma.

Laboratory tests showed haemoglobin at 8.4 mg/dL, a platelet count of 48,000, WBC count of 4,400, INR of 1.1, prothrombin time of 11 seconds and APTT of 29.6/26 seconds.

Spinal MRI revealed intraparenchymal haemorrhage in the cervicodorsal cord from T2 to T4 with surrounding long-segment intraparenchymal T2 hyperintensity from C6 to T6 suggestive of spinal cordoedema was observed as shown in Figs 3 & 4. Patient managed conservatively as it was intra parenchymal oedema with correction bleeding diathesis with platelet infusion therapy. Patient improved slowly with residual left leg weakness however he was able to walk with support.

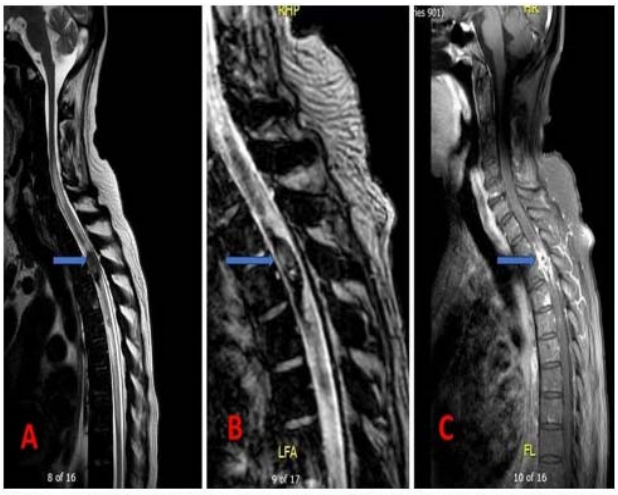

Fig 3 — Sagittal T2 spinal MRI revealed long-segment intraparenchymal T2 hyperintensity seen extending from C6 to T4 vertebral levels suggestive of oedema (Fig 3A). Focal intraparenchymal haemorrhage is seen as hypo intensity on GRE images in spinal cord along T3 &T4 segments (Fig 3B).

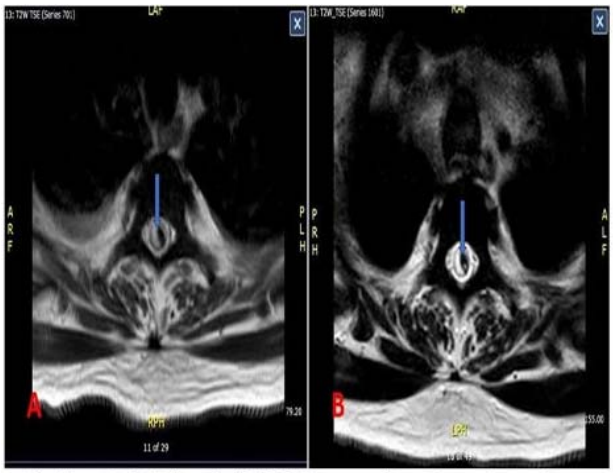

Fig 4 — Axial T2 spinal MRI images show hypo intensity suggestive of intraparenchymal haemorrhage on left side with surrounding hyper intensity suggestive of cord oedema indicated by blue vertical arrows (Figs 4A & 4B).

Case 3

The 80-year-old male patient presented with a two-day history of progressive right leg weakness and difficulty walking. He also experienced numbness in his left leg accompanied by pain. Over the past day, he developed urinary retention, necessitating Foley catheterization and he couldn't stand or walk without support. Additionally, he complained of paraesthesia in his right forearm and had suffered from lower back pain radiating to the right leg for the past 4 to 5 years, with progressive intensity. He has a known history of ischemic heart disease and underwent Coronary Artery Bypass Grafting (CABG) in 1993. He has been taking Clopidogrel and aspirin and has a past history of hypertension but no diabetes.

On examination, there was increased tone in the right lower limb with pyramidal-type weakness (3/5), hyperreflexia, and an extensor plantar response. Sensory examination revealed contralateral pain and temperature loss below T12-L1 (distal to the knee), while proprioception and vibration were preserved. Sensation was decreased in the lumbar and sacral dermatomes. These clinical findings suggested a partial Brown-Séquard Syndrome on the right at the T12 level.

In light of sub-acute right lower limb weakness and Brown-Sequard-like sensory disturbances, infectious and inflammatory causes were initially considered. However, laboratory studies, including hematologic, biochemical, and immunologic investigations, returned normal results. Cerebrospinal fluid analysis from a lumbar puncture was unremarkable. Although the MRI of the head didn't show any obvious abnormalities, diffusion-weighted imaging of the spinal cord revealed restricted diffusion (Fig 5) in the thoracic spinal cord, suggesting of infarct.

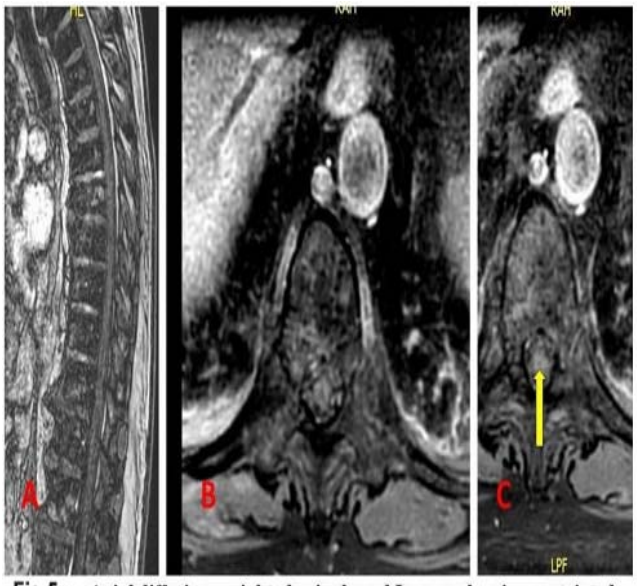

Fig 5 — Axial diffusion weighted spinal cord Images showing restricted diffusion in spinal cord corresponding to vertebral segments T8 & T10 indicated by yellow vertical arrow (Figs 5A & 5B)

Case 4

A 46-year-old female with a history of hypothyroidism presented to the emergency room with a 3 day history of sudden-onset motor weakness in her left side with right leg paraesthesia's. She denied any recent trauma, infection, dizziness, or changes in vision. On examination, she exhibited 3/5 strength in the left upper limb and lower limb while maintaining normal strength in the right side. Temperature and pain sensory deficits were noted in the right leg,extending to a truncal sensory level at the chest. Additionally, proprioception was diminished in the left leg. Patellar and ankle reflexes were brisk on the left side. Her vision was normal, fundus examination was normal with No RAPD.

Magnetic Resonance Imaging (MRI) of the spine revealed a T2 hyperintense signal within the upper cervicothoracic cord, centred at the C3-T1 level, with mild cord expansion as shown in Fig 6.

Notably, MRI of the brain displayed no abnormalities. Spinal fluid analysis yielded normal results, with no pleocytosis and a normal IgG index, and no presence of oligoclonal bands. She was also evaluated for vascular aetiologies. Chest Computed Tomography (CT) scan was negative for aortic dissection and echocardiography were normal. Her ANA ESR and CRP level was normal. Her serum NMO (AQP4) antibody titres were found to be significantly elevated, supporting the diagnosis of Neuromyelitis optica. High-dose intravenous methylprednisolone was initiated, leading to some improvement in symptoms after several days of therapy. Subsequently, the patient was started on immuno-suppressant and neuro-rehabilitation. She recovered well on follow up.

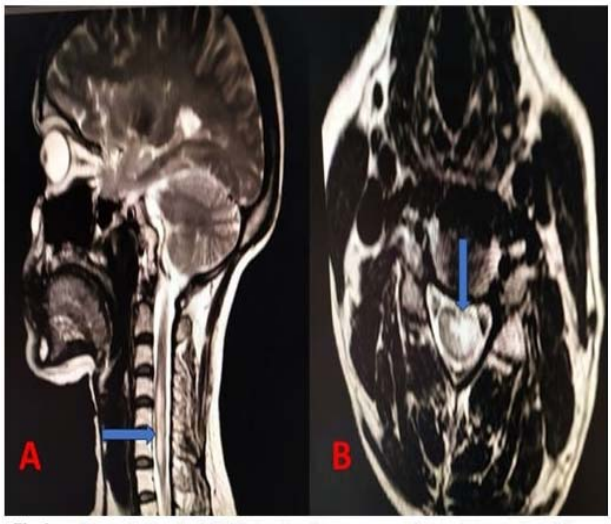

Fig 6 — T2 sagittal spinal MRI showing long segment hyper intense signal in cervical cord indicated by horizontal blue arrow (Fig 6A). The axial T2 Images shows hyper intensity involving central spinal cord extending to left half indicated by blue vertical arrow (Fig 6B).

Discussion

Brown-Sequard Syndrome (BSS) resulting from non-traumatic etiologies is an unusual clinical entity. Cases of non-traumatic BSS without compressive factors are even rarer. In this discussion, we explore the diverse etiologies and clinical manifestations of non-traumatic BSS, shedding light on the diagnostic challenges posed by this condition.

Traditionally, BSS is predominantly associated with traumatic spinal cord injuries, most commonly resulting from penetrating and non-penetrating trauma. Non-penetrating traumatic causes include vertebral fractures and atypical disc herniations. On the other hand, non-traumatic BSS can be categorized into compressive and non-compressive etiologies. Compressive causes encompass conditions such as syringomyelia, hematomyelia, spinal epidural hematoma and meningioma, while non-compressive causes include inflammatory myelopathies, decompression sickness, and vasculitis induced by heroin abuse[2].

Inflammatory Myelopathies : Non-traumatic BSS due to inflammatory myelopathies is a notable subset. While idiopathic transverse myelitis, Multiple Sclerosis (MS), and Neuromyelitis Optica Spectrum Disorder (NMOSD) are common aetiologies, they typically manifest as complete transections of the spinal cord. However, in some cases, patients exhibit atypical clinical manifestations with unilateral involvement mimicking BSS. MS is a chronic neurological disorder primarily affecting young females, diagnosed using MacDonald's criteria. Instances of MS presenting as BSS are exceptionally rare but have been documented in various case reports[3]. NMOSD, which usually presents with features like optic neuritis and longitudinally extensive transverse myelitis, is increasingly recognized for its atypical and rare presentations, including BSS[4].

Spinal Cord Infarction : Non-compressive partial BSS due to spinal cord infarct is another intriguing non-traumatic aetiology. Spinal cord infarction is relatively uncommon, accounting for a small fraction of vascular neurological pathologies. The specific hemi-cord localization seen in these cases raises questions about atypical spinal cord infarcts. MRI findings in spinal cord infarction can vary, with spinal cord swelling and T2 abnormalities developing over time and gadolinium enhancement appearing at a later stage. Timely diagnosis and repeated MRI can enhance the sensitivity and specificity in patients suspected of acute spinal cord ischemia. Recently there are case reports of spinal cord infarctions presenting as BSS[5].

Hematomyelia : Non-traumatic hematomyelia is an extremely rare condition that can present as a Brown-Séquard type of syndrome. Its diagnosis relies on precise MRI imaging. This condition may be associated with various factors such as blood dyscrasias, bleeding disorders and other underlying conditions[6].

Conclusion

In Conclusion, this case series highlights the rarity and diversity of non-traumatic BSS aetiologies, with a particular focus on the diagnostic challenges and atypical presentations in these cases. Diligent clinical examination with appropriate investigations is of paramount importance for early diagnosis and prompt appropriate treatment. These exceptional cases advance our knowledge of non-traumatic BSS.

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