

Neuromyelitis optica spectrum disorder presenting as Brown-Sequard syndrome

Abhinav Agrawal, Dennis Lourdusamy, Abhishek Agarwal,
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ABSTRACT

Introduction: Brown – Sequard syndrome (BSS) presents with features of ipsilateral motor function loss and contralateral loss of pain and temperature. It is most commonly caused from trauma/stab injuries and is also known to be caused by multiple sclerosis. Here, we are presenting a rare case of neuromyelitis optica spectrum disorder (NMOSD) presenting as Brown – Sequard syndrome. **Case Report:** A 70-year-old female presented the emergency room with a 2–3 day history of abrupt onset weakness in her left leg. Magnetic resonance imaging (MRI) scan of the spine showed T2 hyperintense signal seen within the upper thoracic cord centered at T2–T5 level with mild expansion of the cord. A diagnosis of neuromyelitis optica (NMO) spectrum disorder was made based on positive AQP4 antibody titres. **Conclusion:** This case underscores the clinical diversity of NMO spectrum disorder, and suggests that we should probably be checking NMO antibody status in more patients with unexplained myelopathy.

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Received: 19 December 2014

Accepted: 03 March 2015

Published: 23 March 2015

Keywords: AQP4 antibody, Brown – Sequard syndrome, Devic disease, Multiple sclerosis, Spinal cord, Traumatic injury, Neuromyelitis optica

How to cite this article

Agrawal A, Lourdusamy D, Agarwal A, Holland NR. Neuromyelitis optica spectrum disorder presenting as Brown-Sequard syndrome. *Edorium J Neurol* 2015;2:1–4.

Article ID: 100002N06AA2015

doi:10.5348/n06-2015-2-CR-1

INTRODUCTION

The Brown-Sequard syndrome (BSS) is usually the result of a partial traumatic injury (hemisection) of the spinal cord causing ipsilateral loss of motor sensation, vibration and proprioception and contralateral loss of pain and temperature sensation below the level of the lesion [1]. There have also been rare non-traumatic cases from spontaneous epidural hematomas or transverse myelitis. We report an elderly patient with acute BSS and normal cerebrospinal fluid who was ultimately diagnosed with neuromyelitis optica (NMO) spectrum disorder based on serologic testing.

CASE REPORT

A 70-year-old female presented the emergency room with a 2–3 day history of abrupt onset weakness in her left leg. She denied any trauma, recent infection, dizziness or changes in vision. Her past medical history was significant for hypothyroidism and depression.

On examination, she had 3/5 strength in the left leg and normal strength in the right leg. Temperature and pain sensitivity was markedly diminished in the right leg up to a truncal sensory level at the umbilicus. Proprioception was diminished in the left leg. The patellar and ankle reflexes were brisk on the left side. Magnetic resonance imaging (MRI) scan of the spine showed T2 hyperintense signal seen within the upper thoracic cord centered at T2-T5 level with mild expansion of the cord (Figures 1 and 2). MRI scan of the brain was normal. Spinal fluid analysis was normal – no pleocytosis and normal IgG index with no oligoclonal bands. She was initially suspected to have had a spinal cord infarct. However, computed tomography (CT) scan of the chest was negative for aortic dissection and trans-esophageal echocardiography was negative for embolic sources. Protein C, protein S, anti-cardiolipin antibodies and antithrombin was negative.

Ultimately, serum NMO (AQP4) antibody titers were found to be significantly positive with levels of 160 U/ml, suggesting a diagnosis of neuromyelitis optica (NMO) spectrum disorder. She was treated with high dose intravenous methyl prednisone, and her symptoms showed some improvement after a few days of therapy. She was ultimately transferred to an acute rehabilitation facility.

DISCUSSION

NMO, also referred to as Devic disease, is an autoimmune disorder characterized by features of

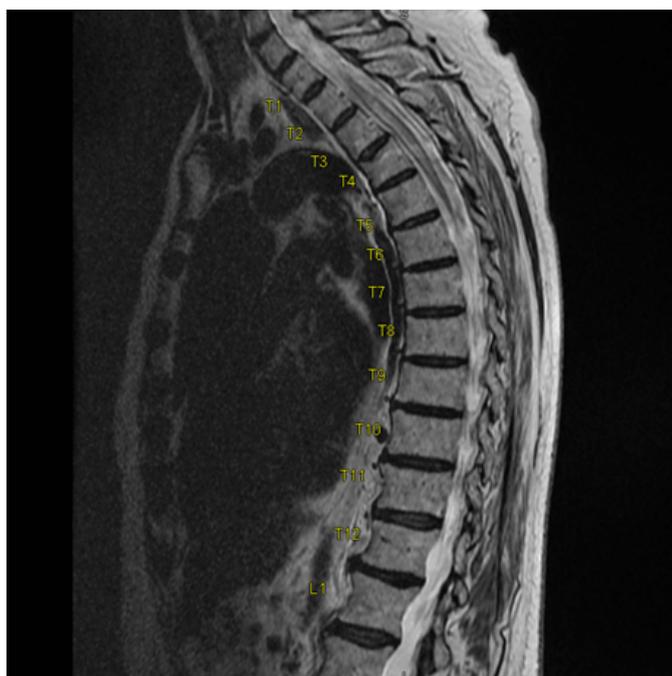


Figure 1: Sagittal T2 weighted MRI showing a diffuse high signal intensity lesion spanning from T2 through T5 with slight expansion of the cord.

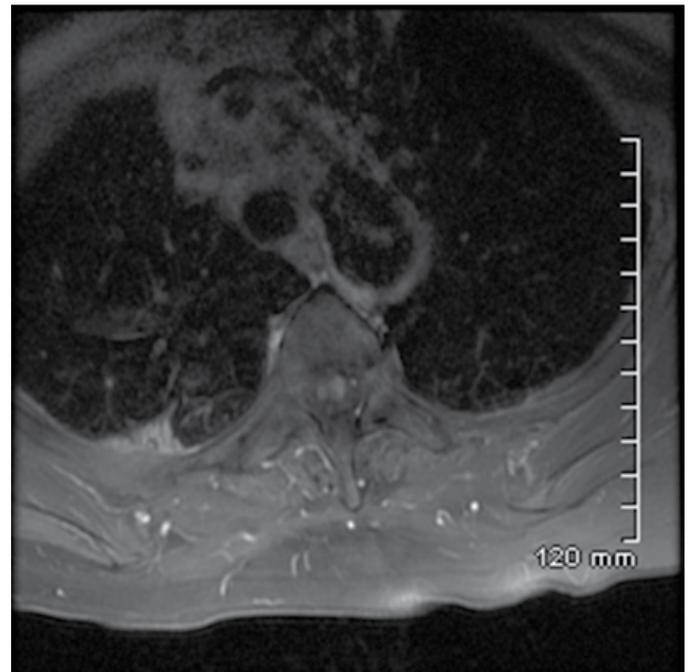


Figure 2: Axial T1 fat saturated post-gadolinium images showing patchy nodular enhancement predominantly within the left dorsal aspect of the cord.

bilateral or rapidly sequential optic neuritis and transverse myelitis [2]. The pathogenesis of NMO is primarily mediated by the humoral immune system. NMO-IgG (anti-AQP4) plays a direct role in the pathogenesis of NMO [3]. Aquaporin-4 (AQP4), the target antigen of NMO-IgG, is a water channel protein highly concentrated in spinal cord gray matter, periaqueductal and periventricular regions, and astrocytic foot processes at the blood-brain barrier [4]. Testing for AQP4 autoantibodies in patients suspected of having NMO is now routinely recommended [5]. Recently, some patients with less classical presentations of isolated myelitis or recurrent/bilateral optic neuritis have been found to have positive NMO/anti-aquaporin-4 (AQP4) antibodies, and these cases are referred to as NMO-spectrum disorder (NMOSD) [6–8]. Myelitis in NMO/NMOSD is typically a bilateral symmetric longitudinally extensive lesion of the spinal cord [7].

Our patient had an atypical clinical presentation with unilateral involvement of spinal cord, presenting as BSS. We are not aware of any previously reported cases of NMO/NMOSD presenting as BSS. Furthermore, the median age of onset for NMO is 32–41 years. Our patient's onset of symptoms occurred far later at the age of 70. Although cases have been described previously in both older and pediatric population, presentation at this age is usually uncommon [9]. Finally, most NMO patients present with CSF pleocytosis (82% in the relapsing group) [10], and our patient had normal CSF findings.

CONCLUSION

This case underscores the clinical diversity of NMO spectrum disorder, and suggests that we should probably be checking neuromyelitis optica (NMO) antibody status in more patients with unexplained myelopathy.

Author Contributions

Abhinav Agrawal – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Dennis Lourdasamy – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Abhishek Agarwal – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Neil R. Holland – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Agrawal A, Lourdasamy D, Agarwal A, Holland NR. Neuromyelitis Optica: spectrum disorder presenting as Brown-Sequard syndrome. Edorium J Neurol 2015;2:1-4.



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