International Journal of Advances in Medicine
Aryadevi CP et al. Int J Adv Med. 2022 Dec;9(12):1224-1227
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: https://dx.doi.org/10.18203/2349-3933.ijam20223026

Case Report

Neuromyelitis optica presenting as neurogenic bladder

C. P. Aryadevi[1], Geethika Pradeep[1], P. V. Bhargavan[1], Shiji P. V.[2]*

[1]Department of Medicine, Baby Memorial Hospital, Kozhikode, Kerala, India
[2]Department of Medicine, Govt. Medical College, Kozhikode, Kerala, India

Received: 24 August 2022
Accepted: 31 October 2022

***Correspondence:**
Dr. Shiji P. V.,
E-mail: shiji_pv@yahoo.com

ABSTRACT

Neuromyelitis optica (NMO) is an immune mediated disease of central nervous system primarily affecting optic nerves, spinal cord and brain stem. This case report describes a 24-year-old male with no comorbidities presented with high grade fever followed by proximal lower limb weakness and bilateral hydrouretero nephrosis without any lower urinary tract obstruction. He had clinical features of conus- cauda lesion with MRI spine showing features of longitudinally extensive transverse myelitis (LETM) and brain MRI showing involvement of splenium of corpus callosum. He improved with steroid therapy with in a period of 2 weeks and oral steroid was tapered of and stopped within a period of two months. There was no relapse of symptoms so far.

Keywords: Neuromyelitis optica, Neurogenic bladder, Lower limb weakness

INTRODUCTION

In 1870, Albutt was the first to report a case of neuromyelitisoptica, but Devic was first to describe the disorder in detail.[1] Neuromyelitis optica was initially thought to be a part of multiple sclerosis, but now it is described as a separate entity in the spectrum of inflammatory central nervous system demyelinating disorders which has disease specific antibodies, aquaporin -4 antibody which was not detected in this patient.[2] The facility for checking anti MOG antibody was not available in our institution or state and hence could not be done. He improved with appropriate treatment and back to normal after a period of 2 months.

CASE REPORT

A 28-year-old male, an airport employee presented with high grade intermittent fever, head ache and myalgia of 10 days duration. On admission he was febrile (Temperature-38.33 degree celsius), pulse rate-100/min, regular with normal volume. Blood pressure-120/80 mmHg. There was no rash, jaundice or lymphadenopathy. Systemic examination was unremarkable. His total WBC count was 10,900 with 70% neutrophil. ESR-16 mm/hr. liver function test, renal function test and peripheral smear were with in normal limits. Blood Widal and Weil felix tests were also negative. Urine routine examination was normal. Blood cultures were sterile. Suspecting a bacterial infection, he was empirically started on parenteral antibiotics. The fever spikes continued despite treatment. After 2 days of admission, he developed acute urinary retention. He was catheterised and drained 500 ml of clear urine. Urine routine examination from catheterised sample was normal. USG abdomen showed grossly distended urinary bladder with bilateral hydroureteronephrosis. Urology evaluation was done to rule out any obstructive lesion. CT urogram (Figure 1) showed the same findings without any evidence of obstruction like bladder stone, enlarged prostate or prostatic abscess. He was on continuous bladder drainage. Next day, he developed paraesthesia of both lower limbs extending from both feet up to the knee. He had mild unsteadiness on walking. Neurological examination revealed normal higher mental function and there was no cranial nerve dysfunction. Optic fundi were normal. Motor system examination showed no wasting and tone was normal. Upper limbs were normal.

Power of extensors and adductors of hip joints were grade 4/5 bilaterally. Gait could not be assessed due to unsteadiness on standing. No abnormal movement was detected. Corneal, conjunctival, abdominal and cremasteric reflexes were normal. Bulbocavernosus reflex and anal reflex were absent. Anal tone was reduced. Plantar response was extensor on both sides with absent knee jerk and brisk ankle jerk bilaterally. Touch, pain, temperature, vibration and joint position sense were diminished on both lower limbs below the knee. No peripheral nerve thickening. He had no signs of meningeal irritation. The possible etiologies considered are 1) demyelinating disorder, 2) intramedullary cord compression and 3) Arachanoiditis. CSF analysis showed lumphocytic pleocytosis. MRI spine with brain screening showed multifocal discontinuous segments of abnormal central cord signal intensities in cervical and dorsal spinal cord. Hyper intense lesion in posterior part of body and splenium of corpus callosum. Features were suggestive of longitudinally extensive myelitis (more than three vertebral segments) with hyper intense signal intensities at C3- C4, D2-D4, D5-D7, D7, D9 - D11 levels in the dorsal column predominantly involving central part of cord suggestive of NeuromyelitisOptica (Figure 2). Further work up to exclude a connective tissue disorder or vasculitis showed normal ANA profile. P- ANCA and C ANCA were negative. Antibody against viral capsid antigen (IgG) for EBV was positive. Anti-aquaporin 4 antibody was absent. Visual evoked potentials were normal. MOG antibody could not be done.

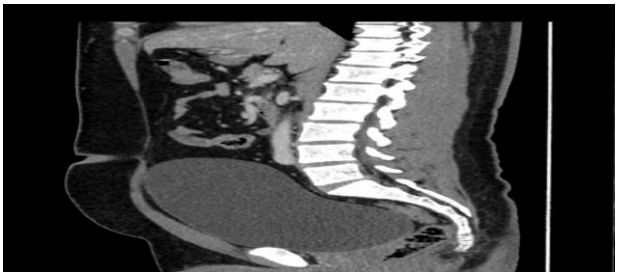

Figure 1: CT urogram of grossly distended urinary bladder.

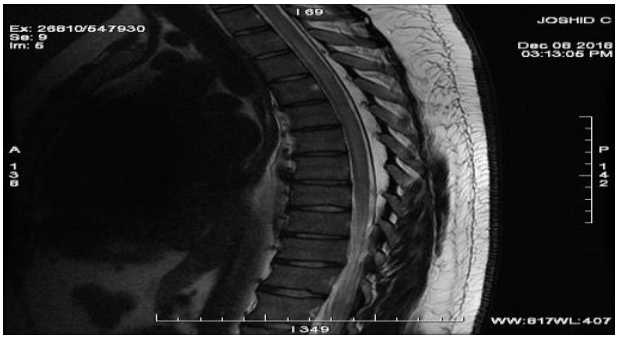

Figure 2: MRI whole spine of multifocal discontinuous segments of abnormal central cord intensities with each segment involving more than 3 vertebral segments suggestive of longitudinally extensive transverse.

He was started on inj. methylprednisolone 1 gm daily for 3 days followed by oral steroids which was gradually tapered and stopped within 4 weeks. After 24 hours of starting steroids, his fever and paresthesia subsided and he was on continuous bladder drainage on discharge through foley's catheter. He was followed up after 2 weeks in OPD, when he was able to walk without support and clinical improvement was dramatic. A follow up after 2 months, he was absolutely normal.

DISCUSSION

The diagnosis of neuromyelitis optica spectrum disorder require the presence of the absolute criteria. Revised criteria published in 2015 depends on the presence of cor clinical characteristics, Aquaporin 4 antibody status and characteristic MRI findings.[3,4]

Clinical characteristics-Optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, aymptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD with typical.

Diencephalic MRI lesion-Symptomatic cerebral syndrome with NMOSD with typical brain lesion.

NMOSD with aquaporin 4 IgG antibody requires the following: At least one core clinical characteristics, a positive test for AQP4-IgG using cell-based assay, exclusion of alternate diagnosis, diagnostic criteria of NMOSD with negative or unknown AQP4-IgG antibody status requires, at least two core clinical characteristics occurring as a result of one or more clinical attacks and meeting all of the following requirements. A) At least one core clinical characteristic must be optic neuritis, acute myelitis with longitudinally extensive transverse myelitis or area postrema syndrome, b) Dissemination in space (2 or more different core clinical characteristics), c) fulfillment of additional MRI requirements as applicable-Negative tests after AQP4-IgG (NMO-IgG) using best available detection method and exclusion of alternative diagnosis

Our patient has one core criteria, i.e., LTEM which is characteristic of NMOSD than spinal form of multiple sclerosis and brain MRI showing hyperintense lesion in the posterior part of the body and splenium of corpus callosum. Ideally testing for AQP4 antibody status should be performed during attacks and before immunotherapy. Since conversion to seronegative status may occur with immunotherapy.[1] In our case it was done on the day of illness before starting an IV Methylprednisolone. A retesting for AQP4 should not have been done as there was strong clinical and MRI features suggestive of NMOSD.

The presence of anti MOG autoantibodies may define a distinct clinical syndrome which can be differentiated from AQP4 antibody associated NMOSD by the following features:[5] More likely to involve the optic nerve than the spinal cord, more likely to present with simultaneous

bilateral optic neuritis, more likely to be monophasic or to have fewer relapses, less likely to be associated with other autoimmune disorders, proportionally more brainstem and cerebellar lesions and fewer supratentoprial lesions, spinal cord lesions mainly occur in the lower portion of the spinal cord, the spectrum of disease may be wider and include acute disseminated encephalomyelitis (ADEM), particularly in children and the male-to-female ratio is close to 1:1, unlike the predominance of women in NMOSD with AQP4-IgG antibodies.

In our patient visual evoked potentials was absent which may help to rule out optic / retro bulbar neuritis. However optic coherence tonography studies in NMOSD report micro-optic macular oedema of the inner nuclear layer which is commoner in NMOSD. As a diagnostic tool this test is not well established.[2,11]

NMOSD must be distinguished from autoimmune or inflammatory disease including SLE, Sjogren's syndrome, neuro Behcet disease, sarcoidosis, multiple sclerosis, para infectious disorders, as longitudinally extensive spinal cord lesions are were well described in the above-mentioned conditions.[6]

The clinical course, prognosis, underlying pathology as well as responsiveness to disease modifying therapies help to differentiate NMOSD from other demyelinating disorders like multiple sclerosis, autoimmune and inflammatory disorders.[7]

Aquaporin negative NMO is a rare variant of neuromyelitis optica spectrum disorder. Our patient presented with high grade fever, myalgia and headachee of 10 days duration. Two days after admission he developed acute urinary retention due to non-obstructive hydro-uretero-nephrosis due to spinal cord or lumbosacral radicular involvement. Subsequently he developed features of conus cauda lesion without any higher function or cranial nerve dysfunction. His unsteadiness on standing was due to proximal muscle weakness and posterior coloumn dysfunction. There was no involvement of cerebellum, optic nerve or retrobulbar neuritis as his visual evoked potentials were normal. MRI spine with brain screening showed longitudinally extensive hyperintense signal intensity involving central part of the cord suggestive of demyelination due to neuromyelitis optica.

Brain MRI screening showed involvement of posterior part of corpus callosum. This callosal lesion in NMO is often edematous and heterogenous creating a marbled pattern.[8] Sometimes it may present as thickening of splenium in a unique arch bridge pattern. It can also extend to the cerebral hemisphere forming an extensive white matter lesion[9]This is not seen in our case. Sometimes these lesions may produce dysfunction of cognition which is not present in our patient. Callosal lesions in MS are discrete and perpendicular to the ventricles and involve inferior aspect of corpus callosum which helps to distinguish multiple sclerosis from NMOSD. Using the revised NMO criteria, incidence of brain MRI abnormalities is almost 85%. Brain abnormality was predominantly located within the periventricular region of third ventricle, fourth ventricle, supratentorial and infratentorial white matter, midbrain and cerebellum.[5] The lesions are predominantly seen in anti-AQP4 antibody seropositive than seronegative patients. Mass effect is usually absent and is usually vasogenic edema associated with acute inflammation.[11] Our patient had no hemiparesis, encephalopathy or visual field defects. Features suggestive of longitudinally extensive myelitis involving the central part of cord predominantly cervical and upper thoracic than lower thoracic and lumbar cord is characteristic of NM. Lumbosacral cord revealed no anatomical abnormality on MRI suggesting lumbosacral radiculopathy. In Multiple Sclerosis spinal cord lesions are short and multiple and it will remain peripheral, asymmetrical and often posterior.

Our patient is anti AQP4negative. Owing to lack of facilities and financial constraints we were unable to investigate for antibodies against myelo oligo glycoproteins (MOG). Patients with anti MOG positive antibodies and anti AQP4 negative serotype were suggested to have fewer attacks, more caudal myelitis and recover better than patients with positive anti AQP4 antibodies, than those who are seronegative for both antibodies.[10] Patients presenting with NMOSD phenotype with anti MOG antibodies may have a distinct underlying disease mechanism with a better prognosis than those with anti AQP4 antibodies This requires further study.

CONCLUSION

We have presented a case history of 28-year-old male who had longitudinally extensive transverse myelitis from cervical to thoracic cord and involvement of posterior part of body and splenium of corpus callosum. He is AQP4 negative and anti MOG antibody could not be tested. There was no optic nerve involvement. He improved with steroid therapy and is back to normal.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Jarius S, Wildemann B. The history of neuromyelitis optica. J Neuroinflammation. 2013;10:8.
2. Glisson C. Neuromyelitisoptica spectrum disorders. Waltham, MA: UpToDate. 2020.
3. Kim W, Park MS, Lee SH. Characteristic brain magnetic resonance imaging abnormalities in central nervous system aquaporin-4 autoimmunity. MultScler 2010;16:1229-36.
4. Wingerchuk DM, Lennon VA, Pittock SJ, Lucchinetti CF, Weinshenker BG. Revised diagnostic criteria for neuromyelitisoptica. Neurology. 2006;66:1485-9.

5. Matsushita T, Isobe N, Matsuoka T. Extensive vasogenicedema of anti-aquaporin-4 antibody-related brain lesions. Mult Scler. 2009;15:1113-7.
6. Kitley JL, Leite MI, George JS, Palace JA. The differential diagnosis of longitudinally extensive transverse myelitis. Multiple Sclerosis J. 2012;18(3):271-85.
7. Di Pauli F, Berger T. Myelin oligodendrocyte glycoprotein antibody-associated disorders: toward a new spectrum of inflammatory demyelinating CNS disorders? Frontiers Immunol. 2018;9:2753.
8. Nakamura M, Misu T, Fujihara K. Occurrence of acute large and edematouscallosal lesions in neuromyelitisoptica. Mult Scler. 2009;15:695-700.
9. Kim W, Park MS, Lee SH. Characteristic brain magnetic resonance imaging abnormalities in central nervous system aquaporin-4 autoimmunity. Mult Scler 2010;16:1229-36.
10. Sortirchos ES, Saidha S. Byraiah G. *In vivo* identification of morphologic retinal abnormalities in neuromyelitis optica. Neurology. 2013;80:1406.
11. Asgari N, Skejoe HP, Lillevang ST, Steenstrup T, Stenager E, Kyvik KO. Modifications of longitudinally extensive transverse myelitis and brainstem lesions in the course of neuromyelitisoptica (NMO): a populationbased, descriptive study. BMC Neurol. 2013;13:33.

Cite this article as: Aryadevi CP, Pradeep G, Bhargavan PV, Shiji PV. Neuromyelitis optica presenting as neurogenic bladder. Int J Adv Med 2022;9:1224-7.