Case Report

Neuromyelitis Optica Spectrum Disorder Presenting Radiologically like Spinal Astrocytoma

Babawale Arabambi[1], Sikirat Yetunde Ashiru[1], Rashidat Amoke Olanigan[1], Shamsideen Abayomi Ogun[1,2]

[1]Department of Medicine, Neurology Unit, Lagos State University Teaching Hospital, [2]Department of Medicine, Lagos State University College of Medicine, Lagos, Nigeria

Abstract

Neuromyelitis optica spectrum disorder (NMOSD) is an infrequent antibody-mediated disorder of the central nervous system. The diagnosis is made in the setting of specific clinical syndromes and a positive aquaporin-4 antibody titer. Intramedullary spinal cord tumors may clinically and radiologically mimic NMOSD, necessitating careful differentiation, especially in individuals who meet the epidemiological profile of NMOSD. We report two cases of NMOSD in young female patients who had a neuroradiological diagnosis of spinal cord astrocytoma and were initially planned for spinal cord surgery. We aim to highlight the similarities between NMOSD and spinal cord tumors using these two clinical cases.

Keywords: Aquaporin-4 antibody, astrocytoma, neuromyelitis optica, spinal cord tumor

Introduction

Neuromyelitis optica spectrum disorder (NMOSD) is a typically antibody-mediated demyelinating disease affecting the central nervous system (CNS). Optic neuritis, intractable vomiting/hiccups, and long-segment transverse myelitis are some of its classical presentations.[1] The diagnosis of NMOSD can be made in the presence of at least one of the typical clinical characteristics with a positive aquaporin-4 antibody, after the exclusion of alternative diagnoses.[2]

Intramedullary spinal cord tumors are rare and may be challenging to distinguish radiological from spinal cord demyelinating disorders.[3] There have been reported cases of patients who had spinal cord surgery and biopsy, with histological findings of demyelination and serological confirmation of NMOSD.[4] It is crucial to consider the possibility of a demyelinating cord lesion in patients with a radiological impression of a spinal cord tumor.

We report two cases of NMOSD that was diagnosed neuroradiologically to be spinal cord tumors and were initially planned for spinal cord surgery.

Access this article online

Quick Response Code:

Website: www.njmonline.org

DOI: 10.4103/NJM.NJM_92_21

Case Report

Case one

A 24-year-old woman who had background rheumatoid arthritis presented with a 4-month history of low back pain and a 2-month history of progressive paraparesis with urinary retention and constipation. She had no history of visual impairment or recurrent vomiting and hiccups. She had reduced sensations up to her upper abdomen. Neurological examination revealed spastic paraparesis (power of 2/5 in the right lower limb and 1/5 in the left lower limb). She had loss of light touch, pain, and temperature up to T8 dermatome with loss of joint position sense in both lower limbs. Her visual acuity was intact, and she had no relative afferent pupillary deficits. A clinical assessment of thoracic myelopathy was made with noncompressive and compressive etiologies considered.

Address for correspondence: Dr. Babawale Arabambi, Department of Medicine, Neurology Unit, Lagos State University Teaching Hospital, Lagos, Nigeria.
E-mail: babawale.arabambi@gmail.com

How to cite this article: Arabambi B, Ashiru SY, Olanigan RA, Ogun SA. Neuromyelitis optica spectrum disorder presenting radiologically like spinal astrocytoma. Niger J Med 2021;30:713-6.

Submitted: 15-May-2021 **Revised:** 01-Oct-2021
Accepted: 02-Oct-2021 **Published:** 27-Dec-2021

Magnetic resonance imaging (MRI) of the spine [Figure 1] was reported as an expansile, long-segment diffuse intramedullary mass involving the thoracic spinal cord extending inferiorly (from T2-T10). The mass appears isointense on T1-weighted and hyperintense on T2-weighted images with peripheral contrast enhancement. These features were reported to be suggestive of a spinal astrocytoma. After discussions with the radiologists and neurosurgeons, she was planned for a spinal cord surgery with tumor biopsy. Cerebrospinal fluid (CSF) oligoclonal bands and serum Vitamin B12 were negative. Serum aquaporin-4 antibody titer was positive at 1:3200 (normal 1: <10, cell-based immune fluorescence assay), while antiextractable nuclear and antinuclear antibody titers were below cut-off levels.

A diagnosis of NMOSD was made based on the positive antibody titer and long-segment myelitis. She received pulsed methylprednisolone for five days and was started on prednisolone and azathioprine. At follow-up three months after presentation, she had made significant clinical improvement with resolution of sphincteric dysfunction and improvement of lower limb power to 4/5. There was the return of all sensory functions.

Case two

A 39-year-old woman with a 2-month history of worsening quadriparesis, weakness started in the right lower limb before involving the other limbs. She had a history of recurrent vomiting up to five months before the onset of limb paresis. There was sphincteric dysfunction with urinary urgency with urge incontinence and constipation. She had no visual impairment or features that suggested an autoimmune disease. There was significant weight loss due to persistent vomiting and inadequate nutrition.

On examination, she had a spastic quadriparesis with a power of 0/5 in the right limbs and 1/5 in the left limbs. There was an impaired sensation to light touch, pain, and temperature up to the C4 dermatome. She also had loss of joint position sense in all limbs.

The neurologist read preliminary MRI films [Figure 2] as a longitudinally extensive cervical transverse myelitis. A possible diagnosis of NMOSD was considered, and the patient was commenced on pulse methylprednisolone over five days. However, the radiological report of the MRI revealed an expansile intramedullary lesion within the cervical cord extending from the C1-C7 vertebra. The lesions appear hypointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images with no signal dropout on short tau inversion recovery images. There was a narrowing of the surrounding CSF column. The conclusion was that the lesion was most likely a spinal astrocytoma.

The neurosurgeons reviewed the patient and counseled her for a surgical biopsy for histologic diagnosis, but she declined surgical intervention.

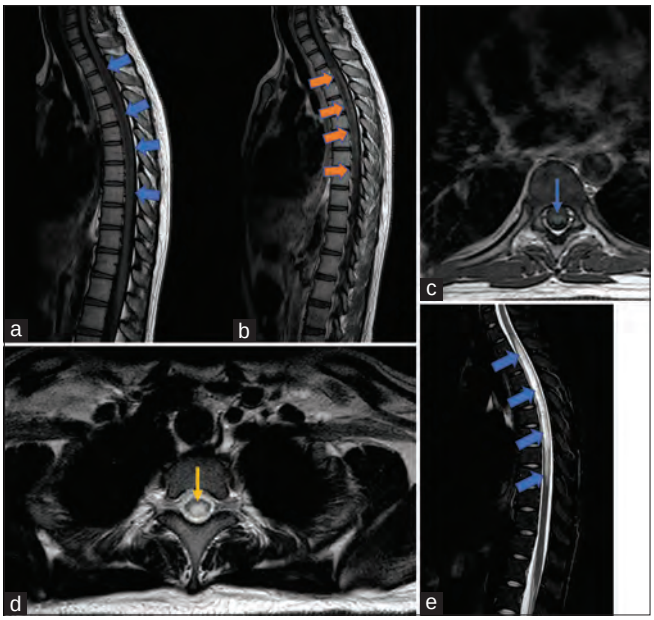

Figure 1: An expansile, long-segment diffuse intramedullary mass involving the thoracic spinal cord extending inferiorly (from T2-T10). The mass appears isointense on T1 weighted (a) and hyperintense on T2-weighted images (d and e) with peripheral contrast enhancement (b and c). Features reported to be suggestive of a spinal astrocytoma

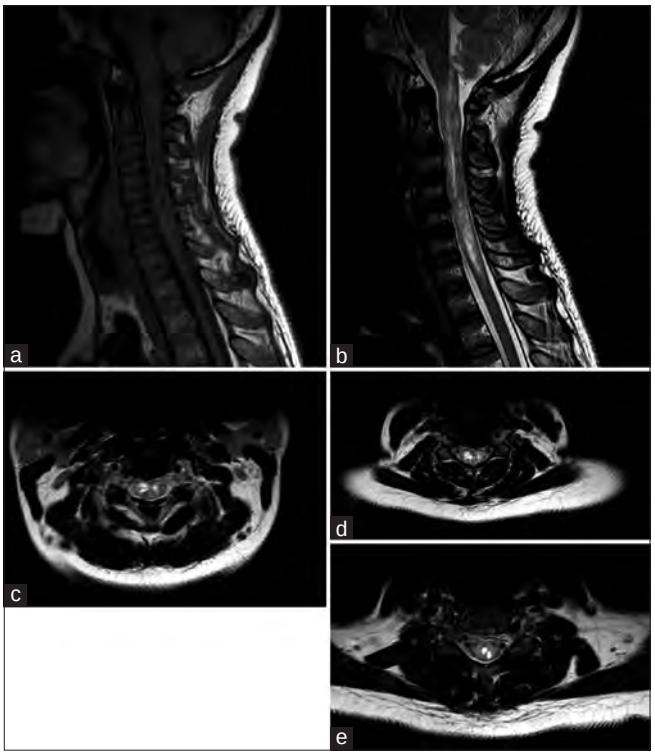

Figure 2: An expansile intramedullary cord lesion within the cervical cord extending from C1 to C7 vertebra. The lesions appear hypointense on T1-weighted image (a), heterogeneously hyperintense on T2-weighted images (b-e) with no signal drop out on STIR images (not shown). There is narrowing of the surrounding cerebrospinal fluid space. Findings were reported as likely spinal astrocytoma

Serum antibody to aquaporin-4 titer done was positive with a titer of 1:132 (normal 1: <10, cell-based immune fluorescence assay). A diagnosis of NMOSD was confirmed based on the positive antibody titer. She was later discharged on oral prednisolone and azathioprine. Her power gradually improved to 3/5 in the right limbs and 4/5 in the left limbs by the 3rd-month postpresentation.

Discussion

The cases presented were similar – both had long-segment myelitis and were diagnosed with spinal cord tumors. Confirmation of a spinal cord tumor requires biopsy and histology. There have been reported cases of NMOSD, which radiologically mimicked cord tumors and, in some cases, were biopsied, with the demyelination seen on histology.[4] The diagnosis of NMOSD requires a high index of suspicion and recognition of the core clinical syndromes, including acute myelitis, optic neuritis, area postrema syndrome, acute diencephalic clinical syndrome, acute brainstem syndrome, and symptomatic cerebral syndrome. The former three are the more common findings.[5] Serum aquaporin-4 titer (also called NMO-IgG) is highly useful in the diagnosis of NMOSD, with the median specificity ranging between 97% and 99% depending on the type of assay done. The cell-based assay has the highest diagnostic accuracy compared to the other types.[6] The diagnostic criteria for NMOSD include a positive NMO-IgG with the presence of one or more core clinical syndromes.[5]

The antibodies involved in NMOSD binds to aquaporin-4 (AQP4) receptors in the CNS. The IgG-1 isotype (NMO-IgG) is the predominantly expressed type. AQP4 receptors are found widely in the water channels of the brain, spinal cord, and optic nerves near the blood–brain barrier. The binding of these antibodies to the receptors leads to cytokine-mediated damage to the blood–brain barrier and complement activation with eventual demyelination from loss of the supporting function of the astrocytes to its surrounding cells.[7]

NMO-IgG-positive NMOSD is more common in the female sex, with a female-to-male ratio as high as 9:1. The disorder is also common in people of African ancestry, especially compared to multiple sclerosis. The mean age at onset is approximately 40 years, but younger-onset age has been reported.[8,9] The presented cases were both females, with the second case closer to the reported mean age at onset. NMOSD is associated with several other autoimmune diseases – it is considered to coexist with rather than complicate other autoimmune conditions.[10] Our first case had a background rheumatoid arthritis which further increased the possibility of a coexisting NMOSD.

The two cases had different titer values of NMO-IgG at 1:3200 versus 1:132, respectively. Although NMO-IgG positivity is associated with a higher rate of relapse, the antibody titer is not a useful predictor of the extent of the lesion, clinical severity, relapse rate, or prognosis.[11,12]

Radiologically, MRI spine appearance of NMOSD and intramedullary spinal cord tumors may be similar, with multisegmented central T2 hyperintensity. However, tumors are more likely to be heterogeneous due to the presence of hemorrhagic areas. The presence of cord expansion may also suggest a tumor, although there have been such reports in NMOSD.[4] The two cases received a radiological diagnosis of spinal cord astrocytoma because of central T2 hyperintensity, peripheral contrast enhancement, and cord expansion.[13]

It may be complicated to differentiate the symptoms of a demyelinating cord lesion such as NMOSD from those of a spinal cord tumor using clinical and radiological features. Cases that epidemiologically fit the profile of NMOSD should warrant appropriate serological investigations, eye assessments, including fundoscopy and visual evoked potentials to rule out the involvement of the optic nerve. CSF analysis may show pleocytosis with oligoclonal bands typically absent.[3,14] Confirmed NMOSD cases with positive NMO-IgG are treated acutely with corticosteroids, intravenous immunoglobulin, or plasmapheresis. Options for the prevention of a relapse include azathioprine, rituximab, Mycophenolate mofetil, methotrexate, and tocilizumab. Duration of treatment varies, but some authors have suggested treatment for at least 5 years. The response to treatment varies, with only about 40% of cases achieving total recovery.[15,16] The presented cases were treated with prednisolone and azathioprine with satisfactory and sustained response.

Conclusion

NMOSD diagnosis should be ruled out in patients who are suspected to have an intramedullary spinal cord tumor to avoid a potentially harmful surgical intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Huda S, Whittam D, Bhojak M, Chamberlain J, Noonan C, Jacob A. Neuromyelitis optica spectrum disorders. Clin Med (Lond) 2019;19:169-76.
2. Weinshenker BG, Wingerchuk DM. Neuromyelitis spectrum disorders. Mayo Clin Proc 2017;92:663-79.
3. Woo PY, Chiu JH, Leung KM, Chan KY. Seropositive neuromyelitis optica imitating an intramedullary cervical spinal cord tumor: Case report and brief review of the literature. Asian Spine J 2014;8:684-8.
4. Brinar M, Rados M, Habek M, Poser CM. Enlargement of the

spinal cord: Inflammation or neoplasm? Clin Neurol Neurosurg 2006;108:284-9.
5. Flanagan EP. neuromyelitis optica spectrum disorder and other non-multiple sclerosis central nervous system inflammatory diseases. Continuum (Minneap Minn) 2019;25:815-44.
6. Ruiz-Gaviria R, Baracaldo I, Castañeda C, Ruiz-Patiño A, Acosta-Hernandez A, Rosselli D. Specificity and sensitivity of aquaporin 4 antibody detection tests in patients with neuromyelitis optica: A meta-analysis. Mult Scler Relat Disord 2015;4:345-9.
7. Tait MJ, Saadoun S, Bell BA, Papadopoulos MC. Water movements in the brain: Role of aquaporins. Trends Neurosci 2008;31:37-43.
8. Hor JY, Asgari N, Nakashima I, Broadley SA, Leite MI, Kissani N, *et al*. Epidemiology of neuromyelitis optica spectrum disorder and its prevalence and incidence Worldwide. Front Neurol 2020;11:501.
9. Kissani N, Cherkaoui Rhazouani O, Berrhoute L, Chraa M, Louhab N. An update on epidemiology, diagnosis & management of NMO-SD in the tertiary neurology department of Marrakech (Morocco). Rev Neurol (Paris) 2021;177:80-4.
10. Pittock SJ, Lennon VA, de Seze J, Vermersch P, Homburger HA, Wingerchuk DM, *et al*. Neuromyelitis optica and non organ-specific autoimmunity. Arch Neurol 2008;65:78-83.
11. Jitprapaikulsan J, Fryer JP, Majed M, Smith CY, Jenkins SM, Cabre P, *et al*. Clinical utility of AQP4-IgG titers and measures of complement-mediated cell killing in NMOSD. Neurol Neuroimmunol Neuroinflamm 2020;7:e727.
12. Kessler RA, Mealy MA, Jimenez-Arango JA, Quan C, Paul F, López R, *et al*. Anti-aquaporin-4 titer is not predictive of disease course in neuromyelitis optica spectrum disorder: A multicenter cohort study. Mult Scler Relat Disord 2017;17:198-201.
13. Ogunlade J, Wiginton JG 4th, Elia C, Odell T, Rao SC. Primary spinal astrocytomas: A literature review. Cureus 2019;11:e5247.
14. Bergamaschi R. Importance of cerebrospinal fluid examination in differential diagnosis of Devic's neuromyelitis optica by multiple sclerosis. Neurol Sci 2003;24:95-6.
15. Kleiter I, Gold R. Present and future therapies in neuromyelitis optica spectrum disorders. Neurotherapeutics 2016;13:70-83.
16. Wingerchuk DM, Weinshenker BG. Neuromyelitis optica. Curr Treat Options Neurol 2008;10:55-66.