Neuromyelitis Optica Involving the Area Postrema in a Nigerian Female: A Case Report

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Introduction: Neuromyelitis optica spectrum disorders (NMOSD) are a group of inflammatory diseases associated with demyelination of the central nervous system involving the optic nerves, brain and spinal cord.1 The clinical features, findings on neuroimaging, immunology, and histopathology are distinct.2 Literature on NMOSD in Africans is scarce.

Aim: This is to report a case of neuro-myelitis optica involving the area postrema in a Nigerian female.

Case Presentation: A 35-year-old Nigerian female presented to the University of Abuja Teaching Hospital with an 11-month history of sudden painless vision loss in the right eye associated with hiccup, nausea and non-projectile vomiting. She also complained of headaches. There was no history of seeing haloes around light nor bumping into objects by the side. Eleven months before presentation, she was diagnosed with retrobulbar optic neuritis and received oral steroids for a couple of weeks, at a different hospital, but noticed a marginal improvement in vision. However, she noted that her hiccups, nausea and vomiting persisted. Her presenting visual acuity was 6/60 in the right eye which did not improve with pinhole; and 6/6 in the left eye. Her colour vision was 2/10 in the right eye, and 6/10 in the left eye. She had a grade 2 relative afferent pupillary defect (RAPD) in the right eye. Dilated fundal examination revealed a moderately pale disc on the right, with indistinct margins temporally. In both eyes, she had a vertical cup to disc ratio of 0.5 and the ISNT rule was maintained. Intraocular pressure measured at 1.15 pm was 10mmHg in both eyes. She had an optical coherence tomography (OCT) that showed ganglion cell loss and retinal nerve fibre layer loss in the right eye (Figure 1). All other neurological examination was essentially normal. Magnetic Resonance Imaging of the brain and orbits with and without contrast showed hyperintense lesions in both sides of the dorsal medulla more on the right near area postrema (Figure 2). It also showed some hyperintensities in both optic nerves worse

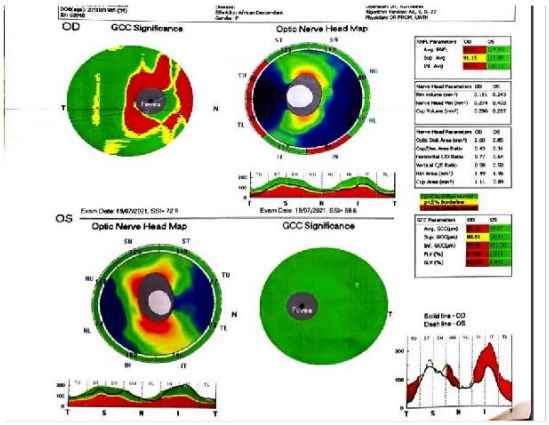

Figure 1: Optical coherence tomogram of the patient showing diffuse ganglion cell and retinal nerve fibre loss in the right eye typical in neuromyelitis optica spectrum of diseases

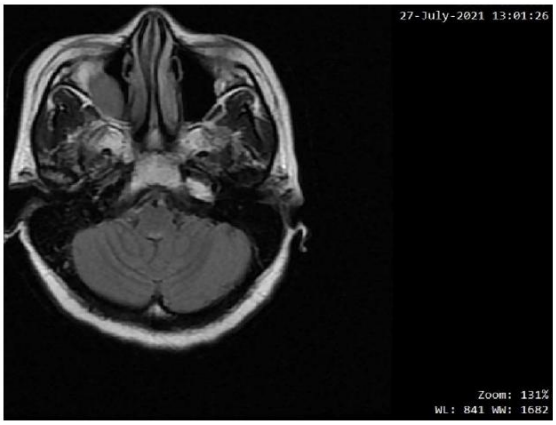

Figure 2: Axial T2 Flair image of patient showing a hyperintense lesion in the dorsal medulla

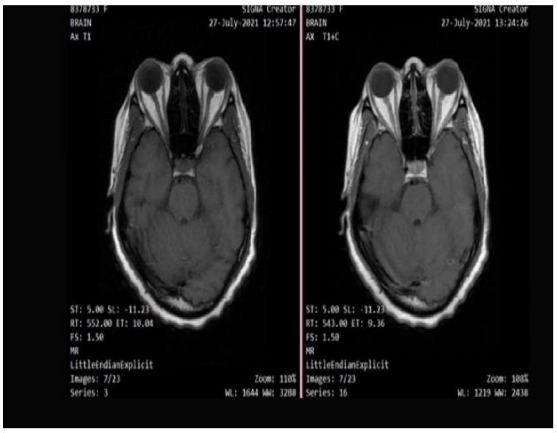

Figure 3: Axial T1 and T1+C image of patient showing a hyperintense lesion in the right optic nerve

on the right especially on T1 with contrast (Figure 3).

Discussion: Neuromyelitis optica spectrum disorders typically involve autoimmune destruction of aquaporin 4 (AQP4) channels in the central nervous system.[1] Brain lesions are typical in areas like the area postrema which has high AQP4 expression.[4,5] NMO prevalence is about 0.5-10 per 100,000 people and it is more common in females with a median age of onset between 32.6-45.7 years.[6,7] Our patient is 35 years old.

Hallmark symptoms of NMOSD include visual loss, limb weakness, sensory loss and bladder dysfunction with a remitting-relapsing course. Occasionally some patients present with nausea, vomiting, and hiccups. Symptoms, like the ones described above, are characteristic, while none of them is disease-specific, so clinical judgement is always necessary. A study from Mayo clinic revealed that 14% of their patients who were diagnosed with NMOSD had nausea and vomiting as their initial presentation. Most of the patients developed other neurological symptoms as the disease progressed.

The International Panel for NMO Diagnosis revised the criteria in 2015 (Table 1) and came up with the following guidelines for diagnosis as outlined in NMOSD diagnostic criteria for adults.[6] Diagnosis is primarily based on the presence of core clinical characteristics: AQP-4 antibody status, and MRI features. However, in patients whose AQP-4 antibody status is not available, diagnosis of NMOSD can also be made using clinical features and peculiar MRI features as shown below in Table 1. Our patient could not have AQP-4 antibody testing done because the test is not available in Abuja at the time of this report but she did meet the criteria for diagnosis based on the presence of 2 core clinical characteristics: optic neuritis and episodes of unexplained hiccups in addition to optic nerve enhancement and area postrema syndrome with associated dorsal medulla lesions as shown on MRI.

Only 33% of patients with NMO present with brain stem syndromes.[3,4] Dandu *et al* reported a case of NMOSD presenting with isolated area postrema syndrome with intractable nausea and vomiting.[3] Figure 4 shows axial T2 Flair of the patient-reported by Dandu et al with the red arrow pointing to the hyperintensity in the dorsal medulla as similar to the location of the hyperintensity in our case as shown in Figure 2.

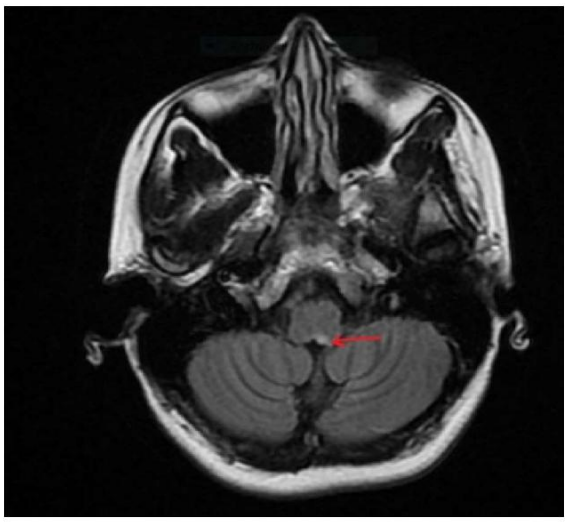

Figure 4: MRI brain FLAIR sequence showing demyelinating lesion in left medulla-area postrema (arrow pointing)

Table 1: The 2015 Revised international panel for NMOSD diagnosis

Diagnostic criteria for NMOSD with AQP4-IgG
1. At least one core clinical characteristic
2. Positive AQP4-IgG
3. Exclusion of other alternative diagnoses

Diagnostic criteria for NMOSD without AQP4-IgG or NMOSD with unknown AQP4-IgG status
1. At least two core clinical characteristics occurring as a result of one or more clinical attacks and meeting all the below requirements:
   a) At least one core clinical characteristic must be optic neuritis, acute myelities with LETM, or area postrema syndrome
   b) Dissemination in space (two or more different core clinical characteristics)
   c) Fulfilment of additional MRI requirements, as applicable
2. Negative tests for AQP4-IgG using best available detection method or testing unavailable
3. Exclusion of alternative diagnoses

Core Clinical Characteristics
1. Optic neutistis
2. Acute myelitis
3. Area postreme syndrome: episode of otherwise unexplained hiccups or nausea and vomiting
4. Acute brain stem syndrome
5. Symptomatic narcole[sy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions

Additional MRI requirements for NMOSD without AQP4-IgG and NMOSD with unknown AQP4-IgG
1. Acute optic neuritis requires brain MRI showing (a) normal findings or only nonspecific white matter lesions, OR (b) optic nerve MRI with T2-hyperintense lesion or T1-weighed gadolinium-enhancing lesion extending over >1/2 optic nerve length or involving optic chiasm.
2. Acute myelitis requires associated intramedullary MRI lesion extending over three contiguous segments (LETM) OR greater than or equal to three contiguous segments of focal spinal cord atrophy in patients with history compatible with acute myelitis.
3. Area postrema syndrome requires associated dorsal medulla/area postrema lesions.
4. Acute brain stem syndrome requires associated periependymal brainstem lesions.

Conclusion: Neuromyelitis optica spectrum disorders with involvement of the area postrema is an atypical presentation and is rare in Africans. Prompt referral to the Neurologist is vital to prevent severe vision loss and death.

References

1. Mealy MA, Wingerchuk DM, Greenberg BM, Levy M. Epidemiology of neuromyelitis optica in the United States: a multicenter analysis. Arch Neurol. 2012; 69: 1176–1180.
2. Pandit L, Asgari N, Apiwattanakul M, Palace J, Paul F, Kleiter I *et al.* Demographic and clinical features of neuromyelitis optica: a review. Mult Scler. 2015;21:845–853.
3. Dandu V, Siddamreddy S, Meegada S, Muppidi V, Challa T. Isolated Area Postrema Syndrome Presenting as Intractable Nausea and Vomiting. Cureus. 2020; 12(2):e7058.
4. Kremer L, Mealy M, Jacob A, Nakashima I, Cabri P, Bigi S *et al.* Brainstem manifestations in neuromyelitis optica: a multicenter study of 258 patients. Mult Scler. 2014;20:843–847.
5. Pittock SJ, Weinshenker BG, Lucchinetti CF, Wingerchuk DM, Corboy JR, Lennon VA. Neuromyelitis optica brain lesions localized at sites of high aquaporin 4 expressions. Arch Neurol. 2006;63:964–968.
6. Wingerchuk D, Banwell B, Bennett J, Cabre P, Caroll W, Chitnis T *et al.* International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology. 2015;85:177–189.
7. Kunchok A, Malpas C, Nytrova P, Havrdova E, Alroughani R, Terzi M *et. al.* Clinical and therapeutic predictors of disease outcomes in AQP4-IgG+ neuromyelitis optica spectrum disorder. Mult Scler Relat Disord. 2020;38:1018-1068.