The delineating CNS demyelinating disease

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ABSTRACT

Background: Brain and spine magnetic resonance imaging (MRI) in Neuromyelitis Optica Spectrum Disorder (NMOSD) is well described in the diagnostic criteria of Wingherchuk 2015 criteria. However, despite this, the pattern of lesion remains incomprehensive.

Case Presentation: We report the case of a 65-year-old female with acute onset of drowsiness, dysphagia, aphasia, and respiratory distress. Brain and spine MRI showed extensive white matter lesions in the subcortical area and cystic lytic patchy lesions in spine. Extensive blood work up was conclusive of anti Aquaporin 4 channel IgG positive suggestive of NMOSD.

Conclusion: The bilateral extensive white matter lesions are rare in presentation in NMO disorders which may mimic adult leukodystrophy. This feature can be considered as a radiological marker for future references.

Keywords : NMOSD, Extensive white matter lesion (EWML), CNS demyelination, Adult Leukodystrophy, Magnetic Resonant Imaging (MRI)

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Background

CNS demyelinating diseases are a heterogeneous group of disorders that may or may not be associated with antibodies. Antibodies are formed either against neuronal (autoimmune encephalitis, NMO MOG, glial fibrillary acid proteins) or intracellular (paraneoplastic disorders) antigens. The spectrum of CNS demyelinating disease without antibody includes Multiple Sclerosis (MS) (discovered in 1968 by Charcot), Bechet's disease, CNS vasculitis, Susac Syndrome. Neuromyelitis Optica (1894 by Devic) was for long considered a form of MS until antibodies against astrocytic water channels aquaporin, AQP4, were discovered. Thereafter the diagnostic consensus criteria for adults, by Wingherchuk 2015, was proposed which has core characteristics as optic neuritis, transverse mellitus, area postrema or extensive form with brainstem lesions called as Neuromyelitis Optica spectrum disorder (NMOSD). Core clinical features: (1) optic neuritis; (2) acute myelitis; (3) area postrema syndrome (hiccups, nausea, and/or vomiting); (4) acute brainstem syndrome; (5) narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic syndrome; (6) Symptomatic Cerebral Syndrome [1,2].

Magnetic resonance imaging (MRI) requirements for AQP4antibody-negative NMOSD:
(1) optic neuritis with either normal/nonspecific MRIorT2-hyperintense or T1-weighted gadolinium-enhancing lesions extending over >1/2 optic nerve length or involving optic chiasm; (2) acute myelitis with intramedullary MRI lesion extending over three or more vertebral segments (longitudinal extensive transverse myelitis: LETM); (3) area postrema syndrome: dorsal medulla/area postrema lesions in MRI; (4) acute brainstem syndrome: periependymal brainstem lesions in MRI [2].
The bilateral extensive white matter lesions (EWMLs) are rare presentation in NMO disorders which may mimic adult leukodystrophy. This feature can be considered as a radiological marker for future references.

Case Presentation

This is a case of a 65-year-old female, Makonde ethnicity, presented with inability to walk, inability to swallow solid or liquid, not responding to verbal stimulus progressing for 3 weeks. She was mechanically ventilated in view of poor glasgow coma scale and respiratory acidosis. Laboratory work showed sepsis (raised total leucocytic count and C reactive protein). A provisional diagnosis of

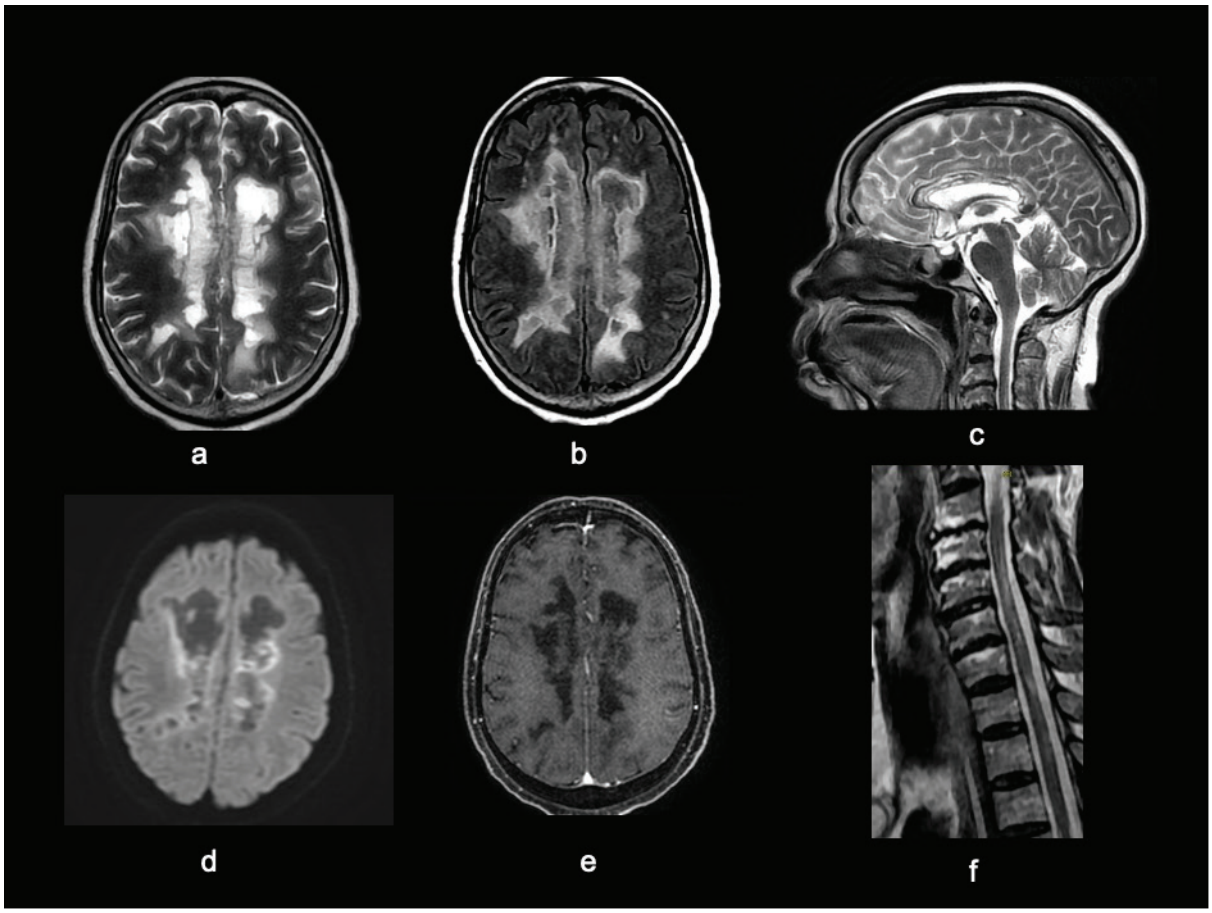

Figure 1. *Contrast-enhanced MRI of the brain reveals confluent areas of T2 (a) and FLAIR (b) hyperintense signal intensity in bilateral periventricular white matter. There is partial central signal suppression on FLAIR. Sagittal T2Wt image (c) shows diffuse involvement of the corpus callosum. Axial diffusion-weighted image (d) reveals mild peripheral diffusion restriction on both sides. No significant contrast enhancement is seen on axial T1Wt post-contrast image (e). T2Wt sagittal image of the cervical spine reveals longitudinally extensive T2 hyperintense signal intensity involving the cervical cord. A mildly reduced bulk of the cervical cord is also noted. Imaging findings suggest an underlying chronic demyelinating etiology.*

septic encephalopathy was kept and was started on intravenous antibiotics and supportive treatment. Brain and spine MRI showed extensive white matter lesions in the subcortical area and cystic lytic patchy lesions in the spine Figure 1. Extensive blood work up (anti nuclear antibody, anti neutrophil cytoplasmic antibody, hepatitis B surface antigen, human immunodeficiency virus, venereal disease research laboratory, autoimmune encephalitis panel) was conclusive of anti Aquaporin 4 channel IgG positive suggestive of NMOSD. She was started on intravenous methylprednisolone over 5 days. Thereafter the patient was weaned off from the ventilator. She continues to be in a stuporous state and was discharged for domiciliary care. She was prescribed oral steroids and azathioprine due to financial constraints.

Discussion

This case describes a very rare radiological presentation of NMOSD which can be easily misdiagnosed as a case of adult leukodystrophy. The involvement of white matter with extensive lesions bilaterally with acute presentation has not been much reported. Brain MRI changes can be deceptive in concluding the diagnosis of a treatable neurological disease. Brain MRI revealed extensive confluent periventricular white matter lesions along with diffuse callosal involvement. It also showed a peripheral pattern of diffusion restriction without significant contrast enhancement. MRI of the cervical spine revealed longitudinally extensive signal alteration involving the cervical cord. It also showed mildly reduced bulk. Considering the above-mentioned imaging findings and clinical presentation possibility of chronic demyelination was considered. Differentials included Marchiafava Bignami Syndrome (MBS) in view of extensive callosal involvement. MBS presents in the age group of 40-60 years with a history of chronic alcohol intake. They have motor or cognitive disturbances, apraxia, hemialexia, dementia, and/or seizures. However, in our patient, there was no history of significant alcohol intake or signs of malnutrition. A remote possibility of adult-onset leukodystrophy was also kept in view of near symmetrical and extensive involvement of periventricular white matter as well as indolent clinical history. There was no obvious thalamic, brainstem, or deep gray matter involvement was noticeable. The absence of optic

neuritis, which is mentioned as the primary presentation in other case reports with similar EWML, makes our case more unique and difficult to diagnose. Rituximab could not be administered to the patient due to financial issues.[3,4,5]

What is new?

The bilateral EWML are rare presentation in NMO disorders. The absence of optic neuritis, which is mentioned as the primary presentation in other case reports with similar EWML, makes the authors' case more unique and difficult to diagnose. Rituximab could not be administered to the patient due to financial issues.

Conflicts of interest

The authors declare that they have no competing interests.

Consent for publication

The patient gave written informed consent for information and images to be published.

Funding

None.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	Patient (gender, age)	65 year old female
2	Final diagnosis	NMO spectrum disorder
3	Symptoms	progressively worsening sensorium.
4	Medications	Pulsed steroid Therapy (iv methyprednisolone) for 5 days
5	Clinical procedure	Serology, MRI
6	Specialty	Neurology