MR Imaging of Acute Cerebellar Involvement in Pediatric Anti-N-Methyl-D-Aspartate Receptor Encephalitis

Bruno P. Soares[1] Sonia F. Calloni[1,2] Shai Shrot[1] Andrea Poretti[1,3]

[1] Section of Pediatric Neuroradiology, Division of Pediatric Radiology, Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins University School of Medicine, Baltimore, Maryland, United States
[2] Department of Neuroradiology, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Università degli Studi di Milano, Milan, Italy
[3] Department of Neurogenetics, Kennedy Krieger Institute, Baltimore, Maryland, United States

Address for correspondence Bruno P. Soares, MD, Section of Pediatric Neuroradiology, Division of Pediatric Radiology, Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins University School of Medicine, 1800 Orleans St ayed Tower, Rm 4174 Baltimore, MD 21287 (e-mail: bruno.soares@jhmi.edu).

Neuropediatrics

Case Description

A 4-year-old boy presented with 10 days of fever, autonomical instability, confusion, sleeplessness, unstable gait, and choreoathetoid limb movements. Laboratory tests revealed leukocytosis and rhabdomyolysis. Brain magnetic resonance imaging (MRI) 3 days after admission was normal, but repeat MRI at 7 days revealed findings compatible with cerebellitis (►**Fig. 1**). Cerebrospinal fluid (CSF) studies showed lymphocytic pleocytosis and positive anti-N-methyl-D-aspartate (NMDA) receptor antibodies. Whole-body fluorine-18 fluorodeoxyglucose positron emission tomography showed diffuse bilateral cerebellar and occipital lobe hypometabolism (►**Fig. 2**), but no evidence of an occult malignancy. Electroencephalography showed diffuse high-voltage background slowing (2–5 Hz) with independent, bilateral high-voltage sharp waves predominantly in the frontal regions.

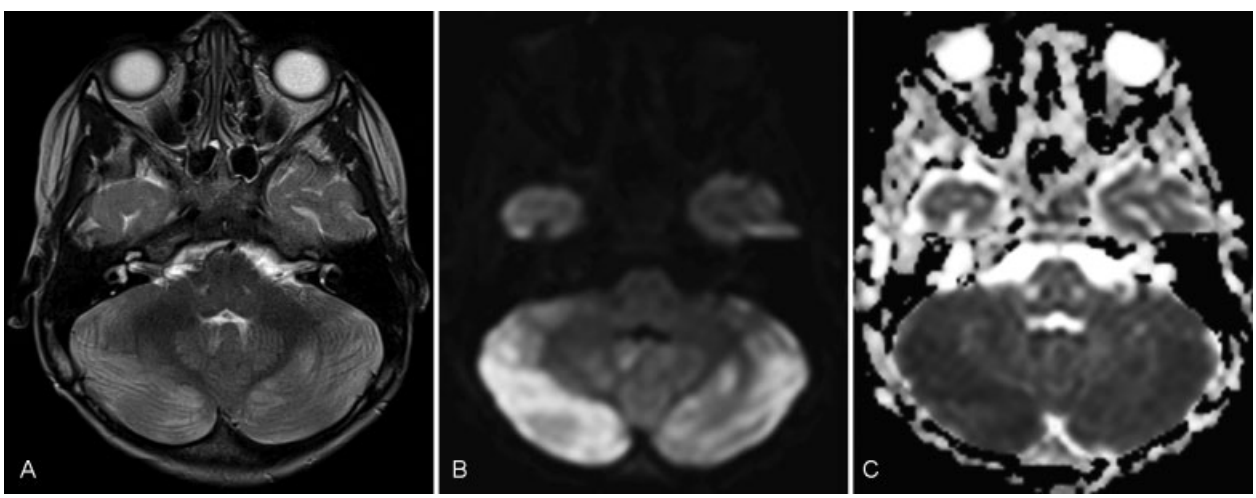

Fig. 1 Brain magnetic resonance imaging performed 7 days after hospital admission. (**A**) Axial T2-weighted image shows diffuse hyperintense signal in the cerebellar cortex and to a lesser extent in the dentate nuclei. (**B**) Axial diffusion-weighted image (DWI) shows increased signal in the cerebellar cortex bilaterally, more pronounced on the right. (**C**) Axial apparent diffusion coefficient map shows hypointense signal in the areas of DWI abnormality, confirming that they represent restricted diffusion.

received
November 23, 2016
accepted after revision
January 6, 2017

DOI http://dx.doi.org/10.1055/s-0037-1600111.
ISSN 0174-304X.

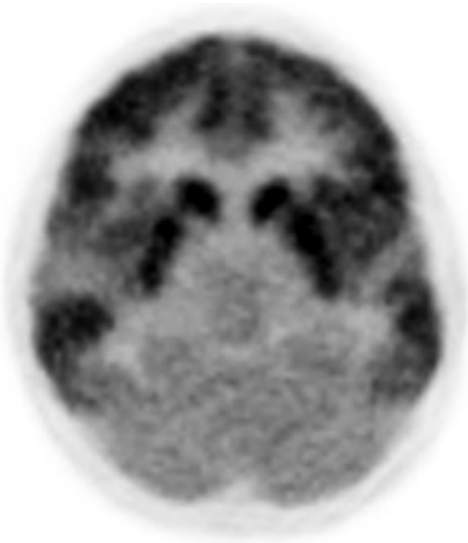

Fig. 2 Axial fluorine-18 fluorodeoxyglucose positron emission tomography image demonstrates symmetric bilateral cerebellar hypometabolism. The occipital lobes were similarly involved (not shown).

Intravenous immunoglobulin and high-dose methylprednisolone were started; however, repeat lumbar puncture showed persistent CSF pleocytosis. Rituximab was started approximately 4 weeks after admission and administered in 4 weekly doses. Three months after admission, the child had severe neurological sequelae including residual choreiform movements and dysphagia requiring a gastrostomy tube.

Discussion

Neuroimaging findings of acute cerebellitis include T2-hyperintense signal of the uni-/bilateral cerebellar cortex, white matter and/or dentate nuclei with/without global cerebellar swelling, contrast enhancement, and diffusion abnormalities.[1] Cerebellitis is commonly caused by infectious agents such as Epstein–Barr virus, varicella, rotavirus, mycoplasma, mumps, and human herpesvirus-6.[1] Cerebellar involvement is uncommon in anti-NMDA receptor encephalitis[2,3] and may be secondary to excitotoxicity to cerebellar granule cells.[4] Anti-NMDA receptor encephalitis should be considered as a differential diagnosis in children with neuroimaging findings suggestive of acute cerebellitis, particularly when the presentation includes extrapyramidal movement disorders.

References

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