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Clinical Observations

Cerebellar Mutism in Acute Disseminating Encephalomyelitis

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ABSTRACT

BACKGROUND: Cerebellar mutism in children occurs after posterior fossa tumor resection and can have lasting effects on cognition, language, and behavior. Cerebellar mutism in acute disseminated encephalomyelitis is rare. **PATIENT:** A 7-year-old boy with a 3-day history of fever, vomiting, and diarrhea presented with altered mental status and expressive aphasia. Magnetic resonance imaging showed new diffusion restriction in the bilateral dentate nuclei and right cerebellum. With treatment, he began to verbalize again but had long-term cognitive and language difficulties. **CONCLUSION:** Acute disseminated encephalomyelitis is commonly a benign process, but its effect on the cerebellum can be long-lasting.

Keywords: cerebellar mutism, acute disseminating encephalomyelitis, aphasia, dentate nucleus

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Introduction

Cerebellar mutism is a well-described phenomenon of aphasia and anarthria seen most commonly after posterior fossa tumor resection for which the exact pathophysiologic mechanism is unknown.[1] Despite its time-limited and transient nature, cerebellar mutism can have lasting effects on cognition, language, and behavior.[2] We describe cerebellar mutism in a nonsurgical case of a boy with acute disseminating encephalomyelitis (ADEM).

Case Report

A 7-year-old boy presented with altered mental status after a 3-day history of fever, vomiting, and diarrhea. On the day of admission, he had acute onset of expressive aphasia at home and was unable to verbally respond to questions or spontaneously talk to his parents. He flailed his extremities randomly but had no rhythmic or jerking movements. There was no history of trauma or ingestion. Recent medications included acetaminophen and a probiotic. He had received his first measles, mumps, and rubella vaccine 8 days prior without initial reaction.

On examination, he was afebrile with normal vital signs, sleepy but arousable, and unable to communicate verbally. Receptive language appeared intact as he would follow one-step commands, but with a significant delay. He had otherwise normal general and neurological examinations. Diagnostic evaluation revealed normal serum chemistries except bicarbonate of 18 mEq/L. Other normal studies included: complete blood count, blood and urine toxicology, stool cultures, and brain computed tomography. Cerebrospinal fluid (CSF) demonstrated white blood count $3 \times 10^3/\mu L$, 9 red blood cell, protein 9 g/dL, and glucose 51 mg/dL with negative gram stain.

The child was hospitalized and received intravenous antibiotics and acyclovir. Brain magnetic resonance imaging/venogram (MRI) showed focal restricted diffusion centrally within the splenium of corpus callosum with T2/fluid-attenuated inversion recovery (FLAIR) hyperintensity without evidence of sinus venous thrombosis (Fig 1). Electroencephalography showed nonspecific slowing in right temporal lobe. CSF cultures/studies were normal/negative except for CSF cytomegalovirus polymerase chain reaction, which detected 1000 copies DNA/mL; repeat testing 1 week later was negative. The infectious disease department was consulted and reported this initial detection of cytomegalovirus was not the primary cause of his symptoms and antiviral therapy was not recommended.

Two days later, his condition worsened, with intermittent episodes of unresponsiveness and lethargy, dysmetria, and continued expressive aphasia. Repeat brain MRI at this time demonstrated new diffusion restriction in the bilateral dentate nuclei and right cerebellum without change in prior restriction of splenium with T2/FLAIR hyperintensity (Fig 2). Repeat CSF examination revealed mild pleocytosis but was

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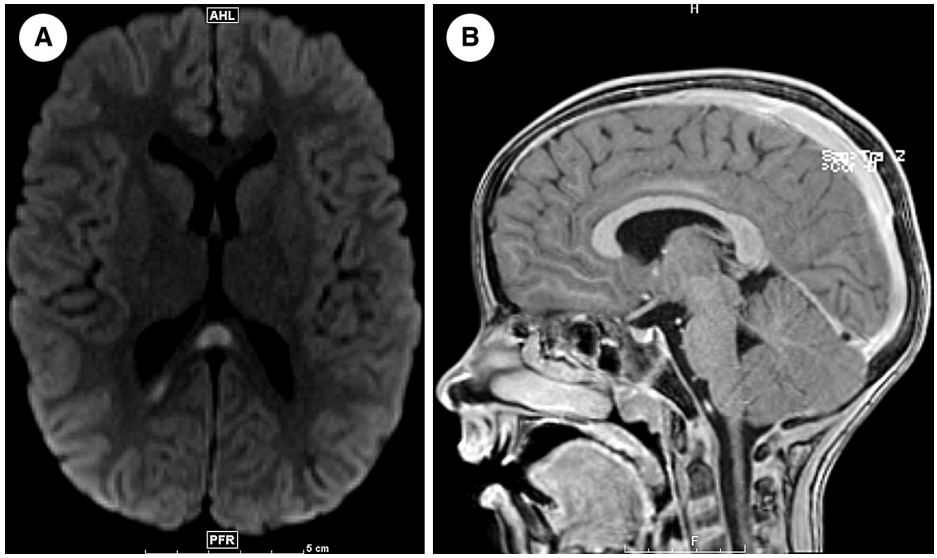

FIGURE 1.
Magnetic resonance imaging of the brain (day of admission). (A) Focal area of restricted diffusion centrally within the splenium of corpus callosum with T2/fluid-attenuated inversion recovery hyperintensity. (B) No occlusive venous sinus thrombosis identified on postcontrast venogram.

otherwise unremarkable (white blood cell 12 $\times$ $10^3/\mu L$, 1 red blood cell, protein 27 g/dL, glucose 75 mg/dL).

The diagnosis of postinfectious ADEM and cerebellar mutism was made, and he received a 5-day course of high-dose intravenous methylprednisolone followed by slow tapering doses. On day 4 of treatment, he began to verbalize monosyllabic words with continued improvement over the next several days. At onset of speech production, he had dysarthria impacting speech intelligibility and prosody related to motor planning deficits. Both dysmetria and ataxia markedly improved over the next 7 to 10 days.

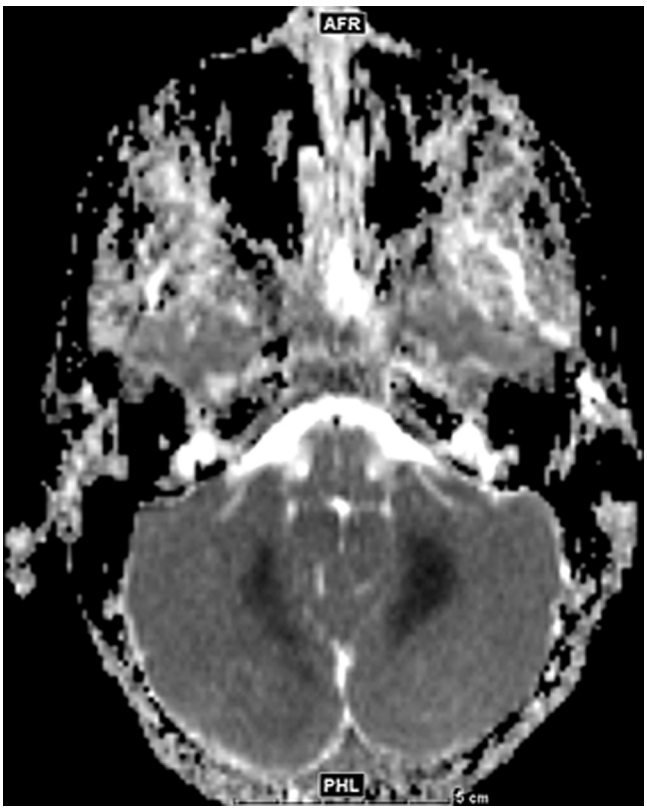

FIGURE 2.
Magnetic resonance imaging of the brain (day 2 of admission). New diffusion restriction in the bilateral dentate nuclei and right cerebellum with corresponding decreased apparent diffusion coefficient attenuation in the left is greater than in the right. There was subtle T2 hyperintensity in regions of diffusion restriction (not shown).

Repeat MRI 1 week later demonstrated resolution of previous lesions in the splenium and cerebellum but incidentally found a large sinus venous thrombosis of the superior sagittal sinus and right sigmoid confluence (Fig 3). He became more withdrawn and less vocal the next day. Follow-up MRI revealed mild propagation of the previously described thrombus; he began intravenous heparin and had rapid clinical improvement.

Throughout his admission, multiple infectious (respiratory viral panel, stool cultures, mycoplasma, anti-streptolysin O, meningoencephalitis panel, CSF herpes simplex virus, CSF Enterovirus, Epstein-Barr virus, CSF cytology), autoimmune (antineutrophil cytoplasmic antibody, neuromyelitis optica immunoglobulin G, CSF angiotensin-converting enzyme, antithyroglobulin, double-stranded DNA, antinuclear antibody) and pro-thrombotic studies (protein C, protein S, plasminogen, lupus anticoagulant, antithrombin III, lipoprotein A, factor V Leiden, activated protein C resistance, anti-phospholipid antibody, homocysteine, and factor VIII activity) were obtained. His only remarkable finding was a heterozygous prothrombin G20210 A mutation.

The patient was ultimately discharged to an inpatient rehabilitation facility to complete a 6-week tapering course of prednisone and to continue anticoagulation with enoxaparin.

Six weeks later, his parents reported marked improvement of motor symptoms; he was walking and running without assistance, and had mild dysmetria on examination. They noted continued word-finding troubles, slowed verbal output, dysprosody with lack of intonation, and persistent cognitive impairment with difficulty sustaining attention. He continues to undergo speech therapy with a neurology follow-up biannually.

Discussion

ADEM is a demyelinating disease of the central nervous system with multifocal neurological symptoms and encephalopathy, typically presenting after an intercurrent illness in children.[3,4] Prognosis for ADEM is usually good with both clinical and radiographic resolution after treatment with intravenous corticosteroids.[4] Cerebellar