Cerebellitis in an Adult With Abnormal Magnetic Resonance Imaging Findings Prior to the Onset of Ataxia

Kirsten L. Gruis, MD; Paolo Moretti, MD; Stephen S. Gebarski, MD; Daniel D. Mikol, MD, PhD

Background: Brain magnetic resonance imaging (MRI) findings during acute cerebellar ataxia in cases of postinfectious cerebellitis are frequently normal. This has resulted in the use of other imaging modalities, such as single-photon emission computed tomography, to aid diagnosis.

Objective: To illustrate the chronologic occurrence of cerebellar ataxia, abnormal findings on MRI, and cerebral spinal fluid pleocytosis in an adult case of postinfectious cerebellitis.

Methods: Case report.

Results: A patient with a 6-week history of occipital headaches and only mild tandem gait difficulty had abnormal MRI findings that were consistent with cerebellar inflammation. As cerebellar ataxia progressed in parallel with cerebral spinal fluid pleocytosis, MRI findings indicative of cerebellar inflammation resolved, while single-photon emission computed tomography showed cerebellar hyperperfusion. Recovery of neurologic function was accompanied by clearing of the pleocytosis and residual MRI-detected cerebellar atrophy.

Conclusion: This case demonstrates that transient abnormalities can be detected by MRI before clinical manifestations of cerebellitis appear, while hyperperfusion detected by single-photon emission computed tomography is prolonged.

Arch Neurol. 2003;60:877-880

Cerebellitis or acute cerebellar ataxia is a neurologic complication that occasionally follows systemic viral or bacterial infections. Although presumed to be more common in children, adult cases of cerebellitis are well established, and the outcome of cerebellitis in young adults is considered quite favorable. While most cases are similar in describing the sudden onset of limb and/or gait ataxia, often with dysarthria and abnormalities of eye movements after an infectious prodrome, most cases have not shown cerebellar abnormalities on magnetic resonance imaging (MRI). We found only 2 reported cases of adults with cerebellitis and reversible MRI abnormalities. These cases described brain MRI abnormalities simultaneous with ataxia.

We describe an adult patient with postinfectious cerebellitis whose reversible MRI abnormalities preceded signs and symptoms of cerebellar ataxia. As cerebellar function improved, cerebrospinal fluid (CSF) and MRI evidence of cerebellar inflammation largely resolved, while single-photon emission computed tomography (SPECT) showed cerebellar hyperperfusion.

Methods

A 38-year-old right-handed woman had a 6-week history of episodic occipital headaches, difficulty focusing her gaze on objects that required sudden head and/or eye movements, and dizziness. She had a recurrent upper respiratory tract infection during the preceding 8 weeks, with clear rhinorrhea, nasal congestion, and sinus pain without objective evidence of fever. There was no recent history of travel or ingestion of toxins. A skin melanoma had been removed 8 years prior with no recurrence.

On physical examination, her blood pressure was 115/70 mm Hg, and pulse rate was 100/min. Signs of meningeal irritation were absent. Results of a complete neurologic examination were normal except for a slight difficulty with tandem gait (Table). An MRI performed on the day prior to this examination (Figure 1A and B) demonstrated diffuse...
Cerebellitis Clinical Course and Paraclinical Studies

<table>
<thead>
<tr>
<th>Week</th>
<th>1 (Initial Clinical Findings)</th>
<th>2 (Hospital Admission)</th>
<th>4</th>
<th>12</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Neurologic examination</strong></td>
<td>Normal except for tandem gait</td>
<td>Cranial nerves: dysmetria of saccadic eye movements, multidirectional rotatory nystagmus and dysarthria</td>
<td>Abnormal, but improved with resolution of saccadic dysmetria, nystagmus, and limb ataxia; continued dysarthria, dysdiadochokinesia, and gait ataxia</td>
<td>Normal, including speech, eye movements, rapid alternating movements, and casual and tandem gait</td>
</tr>
<tr>
<td>Mental status:</td>
<td>full orientation, intact language</td>
<td>Motor, sensory and deep tendon reflex examination results unchanged</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cranial nerves:</td>
<td>unremarkable without nystagmus, and normal speech</td>
<td>Coordination: mild limb ataxia on bilateral finger-nose-finger and heel-knee-shin testing and prominent dysdiadochokinesia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Motor, sensory, deep tendon reflexes, and coordination functioning intact, including finger-nose-finger, heel-knee-shin testing, and rapid alternating movements of limbs</td>
<td>Gait: significant truncal and gait ataxia; patient unable to walk unassisted</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gait: narrow-based but mild difficulty with tandem gait</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>CSF analysis</strong></td>
<td>WBC, 62/mm (97% lymphocytes), elevated protein level 58 mg/dL (580 g/L) (reference range, 5-45 mg/dL [150-450 g/L])</td>
<td>WBC, 10/mm (95% lymphocytes); normal glucose level; protein, 41 mg/dL (410 g/L)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Imaging studies</strong></td>
<td>MRI: abnormal findings, with cerebellar swelling, contrast enhancement, and ectopia</td>
<td>MRI: normal findings except for cerebellar ectopia; resolution of cerebellar swelling and contrast enhancement</td>
<td>SPECT: abnormal findings with increased cerebellar perfusion in comparison with cerebral cortex</td>
<td>MRI: normal findings except for mild cerebellar gyral atrophy; resolution of swelling, contrast enhancement, and ectopia</td>
</tr>
<tr>
<td><strong>Abbreviations:</strong> CSF, cerebrospinal fluid; MRI, magnetic resonance imaging; SPECT, single-photon emission computed tomography; WBC, white blood cell count.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Cerebellar edema, pathologic enhancement after intravenous gadolinium contrast, and mild mass effect causing low-grade cerebellar tonsillar ectopia.

Given the unremarkable neurologic examination findings in the patient despite 6 weeks of headaches and the aforementioned symptoms, we followed her closely as an outpatient with symptomatic treatment of her headaches using anti-inflammatory medications. A lumbar puncture was arranged to investigate possible infectious, inflammatory, or neoplastic causes.

Two weeks later, the patient had worsening occipital headache, nausea and emesis, difficulty walking, and slurred speech, necessitating admission to the inpatient service. On physical examination, her oral temperature was 37°C; pulse rate, 96/min; and supine blood pressure, 117/85 mm Hg. She had significant ataxia of speech, extracocular eye movements, limbs, and gait (Table). The remainder of her examination showed that her condition had not changed during the 2 weeks.

The peripheral white blood cell count was $4.9 \times 10^3/\mu L$, with normal electrolyte and liver function measurements. A VDRL test was nonreactive, and the sedimentation rate was 2 mm/h. Tests for serum Borrelia antibodies, extractable nuclear antibodies, human immunodeficiency virus, and antineuronal antibodies were all negative. The first lumbar puncture on the second hospital day demonstrated pleocytosis and elevated protein level (Table 1). A second MRI was obtained on hospital day 1 and demonstrated resolving edema, no pathologic enhancement after intravenous gadolinium contrast, and less mass effect (Figure 1C and D).

Subsequent CSF studies found 2 oligoclonal bands that were not present in the serum, normal granulorm, negative bacterial, fungal, and viral cultures, negative herpes simplex virus polymerase chain reaction, normal myelin basic protein levels, and a normal cytologic review. Influenza virus type A and B antibodies were not detected in the CSF, and serum influenza A and B IgM titers were negative. Results of computed tomographic studies of the chest, abdomen, and pelvis were unremarkable. A cisternogram with radioactive tracer and nasal pledgets was performed given the patient’s recurrent upper respiratory tract symptoms, with no evidence of a CSF sinus leak.

The patient was hydrated and treated with antiemetics and intramuscular nonsteroidal anti-inflammatory medications for symptomatic relief of her nausea and headache. After 1 week, her condition was unchanged. She remained unable to ambulate unassisted. She was treated with 1000 mg of methylprednisolone intravenously for each of 5 consecutive days. By hospital day 13, she had improved but continued to have cerebellar ataxia at 4 weeks despite improvement of the abnormalities seen on MRI (Table). Hence, we performed SPECT (Figure 2), since abnormal cerebellar uptake of radioactive tracer has been demonstrated in postinfectious cerebellitis when MRI results were normal.8,12 A repeated CSF analysis showed resolving pleocytosis and a normal protein level (Table).

Twelve weeks after the onset of her initial symptoms, the patient’s illness had fully resolved (Table). A follow-up brain MRI (Figure 1E and F) demonstrated resolution of signs of cerebellar inflammation, but mild cerebellar atrophy was now apparent when compared with the previous study.

We described a patient whose clinical course and CSF analysis results fit previous reports of postinfectious cerebellitis.1,2,4,8,10-13 Abnormal findings on MRI in cerebell-
litis have rarely been described. There are only 2 reports of serial MRI improvement in young adults with reversible cerebellitis.\textsuperscript{10,11} This has led to alternative imaging techniques, such as SPECT, to facilitate diagnosis and determine prognosis in these patients.\textsuperscript{8,12} Single-photon emission computed tomography demonstrates cerebellar hyperperfusion in patients with acute cerebellar ataxia at a time when the MRI result is normal. Cerebellar hyperperfusion may reflect the inflammatory component of cerebellitis.\textsuperscript{12} At present, there is no evidence of efficacy for immunosuppressive treatment in postinfectious cerebellitis. Single-photon emission computed tomographic findings were reported to normalize after immunosuppression with intravenous immunoglobulin or steroids in 2 respective case reports,\textsuperscript{8,12} although this may also have occurred spontaneously. We found no reports of untreated cerebellitis with abnormal SPECT findings that normalized spontaneously.

Because of occipital headaches, our patient underwent her first MRI (Figure 1A and B) before developing cerebellar ataxia. This MRI demonstrated cerebellar abnormalities, including cerebellar edema, pathologic contrast enhancement, and mass effect. By the time the physical examination showed cerebellar dysfunction, a second MRI (Figure 1C and D) demonstrated reduced cerebellar edema and no pathologic contrast enhancement. Most of the previous case reports of cerebellitis and normal brain MRI results have not described the temporal relationship between neurologic dysfunction and the imaging study, and they have not mentioned the use of intravenous contrast.\textsuperscript{1,3,4,8,12} In a study of 11 adults with acute-onset cerebellitis, all with full recovery, patients underwent MRI 1 year after the onset, and there was no mention of an imaging study being performed in the acute phase.\textsuperscript{2} All of the patients reported to have complete resolution of cerebellar ataxia were reported to have normal brain MRI findings. It is unlikely that these patients received an MRI prior to developing ataxia, as our case did. Therefore, comparisons between early and convalescent brain MRIs may be needed to detect subtle cerebellar atrophy (Figure 1E and F). Our case demonstrates that abnormalities indicative of cerebellar inflammation on brain MRI are reversible and may already be resolving by the time the patient’s ataxia is significant enough to warrant investigation, thus explaining why so many case reports of cerebellitis have shown normal MRI findings. These findings underscore the utility of SPECT imaging in the

\begin{figure}
\centering
\includegraphics[width=\textwidth]{figure1}
\caption{T1-weighted sagittal sections after intravenous administration of gadolinium (A, C, and E) and T2-weighted axial sections through the cerebellum (B, D, and F). Findings at the initial examination (A and B) demonstrate contrast enhancement of foliar surfaces (vertical arrows, A), the folia are tightly packed, and there is cerebellar tonsillar ectopia (horizontal arrow, A) consistent with cerebellar swelling. At week 2 (C and D), contrast enhancement is no longer seen (vertical arrows, C), the folia are more loosely packed, there is less cerebellar ectopia (horizontal arrow, C), and overall T2-weighted signal (D) is decreased compared with the first image (B), consistent with resolving cerebellar swelling. At week 12 (E and F), there is no cerebellar ectopia (horizontal arrow, E) and there is more separation of the folia (C) compared with the first image (E), consistent with mild cerebellar atrophy.}
\end{figure}

\begin{figure}
\centering
\includegraphics[width=\textwidth]{figure2}
\caption{Axial single-photon emission tomographic sections through the cerebellum (A) and cerebral hemispheres (B). High photon flux is seen in the more superficial aspects of the cerebellum (arrowheads, A), consistent with increased perfusion. Normal cerebral photon flux is present.}
\end{figure}
diagnostic workup of symptomatic patients, when MRI may be less sensitive. Finally, this case suggests that while neurologic function may return to normal, cerebellar atrophy may occur.

Accepted for publication August 9, 2002.

Author contributions: Study concept and design (Dr Gruis); acquisition of data (Drs Gruis and Moretti); analysis and interpretation of data (Drs Gebarski and Mikol); drafting of the manuscript (Drs Gruis and Mikol); critical revision of the manuscript for important intellectual content (Drs Moretti, Gebarski, and Mikol); administrative, technical, and material support (Drs Gruis, Moretti, Gebarski, and Mikol); study supervision (Dr Mikol).

We would like to thank Sid Gilman, MD, FRCP, William J. Herdman Professor of Neurology, chairman, Department of Neurology, University of Michigan Medical School, Ann Arbor, for his thoughtful remarks and review of this case report.

Corresponding author and reprints: Kirsten L. Gruis, MD, Department of Neurology, Taubman Center 1324/0322, 1500 E Medical Center Dr, Ann Arbor, MI 48109-0322 (e-mail: kgruis@umich.edu).

REFERENCES