Advanced neuroimaging findings of pseudotumoral hemicerebellitis in an elderly male requiring surgical decompression

Case report

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Acute cerebellitis is an inflammatory process that usually affects bilateral cerebellar hemispheres in the pediatric population. Pseudotumoral hemicerebellitis is an extremely rare presentation in which unilateral cerebellar involvement mimics a tumor that can exert significant mass effect on the surrounding structures, which may require surgical intervention. Magnetic resonance imaging characteristics that suggest cerebellitis include cerebellar swelling, T2 hyperintensity, and pial enhancement. Advanced neuroimaging, including MR perfusion and MR spectroscopy, may be helpful in excluding other diagnoses. The authors present the case of pseudotumoral hemicerebellitis in the oldest documented patient, a 73-year-old man who required surgical decompression, and they provide a brief discussion of advanced neuroimaging findings.

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Key words • cerebellitis • advanced neuroimaging • MR spectroscopy • MR perfusion • surgical decompression • infection

Abbreviations used in this paper: CBF = cerebral blood flow; Cho = choline; Cr = creatine; CRP = C-reactive protein; ESR = erythrocyte sedimentation rate; LDD = Lhermitte-Duclos disease; MRS = magnetic resonance spectroscopy; NAA = N-acetylaspartate; ppm = parts per million.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Pseudotumoral hemicerebellitis requiring decompression

Initial physical examination revealed partial left cranial nerve IV, VII, and XII palsies, nystagmus, and left dysmetria. Complete blood count testing showed a leukocyte count of 13.6 tho/μl, neutrophil differential of 96%, an elevated erythrocyte sedimentation rate (ESR) of 52 mm/hour, and C-reactive protein (CRP) of 72.5 mg/L.

A head CT scan obtained at an outside hospital showed an ill-defined hypodense and expansive lesion predominantly involving the left cerebellar hemisphere, with significant mass effect obstructing the fourth ventricle and causing acute hydrocephalus with transependymal flow of CSF.

Contrast-enhanced 3-T MRI of the brain, performed at our hospital, revealed an expansive, infiltrative T2-weighted and FLAIR-hyperintense mass involving the entire left cerebellar hemisphere and extending to the vermis and into the medial aspect of the right cerebellar hemisphere (Fig. 1). The mass was predominantly non-enhancing, showing only mild adjacent leptomeningeal enhancement (Fig. 2). The abnormal signal also extended into the left brachium pontis, pons, and left midbrain; there was mass effect and effacement of the fourth ventricle, causing acute obstructive hydrocephalus (Fig. 3). There was also crowding at the foramen magnum and tonsillar herniation.

The affected area demonstrated predominantly facilitated diffusion, with apparent diffusion coefficient values of $0.874 \times 10^{-3}$ mm$^2$/sec compared with the normal brain parenchyma value of $0.683 \times 10^{-3}$ mm$^2$/sec (Fig. 4). Dynamic susceptibility perfusion MRI demonstrated decreased relative cerebral blood volume compared with the contralateral cerebellar hemisphere.

Multivoxel 2D$^1$H magnetic resonance spectroscopy (MRS) was performed by using a spin echo (point-resolved spectroscopy) sequence with water suppression by means of selective excitation. Sequence parameters included the following: TR 1700 msec; TE 135 msec; NEX 3; FOV 16 x 16 cm$^2$; section thickness 20 mm; bandwidth 1200 Hz; and matrix size 16 x 16. Volume of interest was selected to include the area of signal abnormality in the left cerebellar hemisphere as well as areas of normally appearing contralateral cerebellum. Magnetic resonance spectroscopy demonstrated a choline/creatine (Cho/Cr) ratio of 2.01 (compared with 0.8 in the contralateral normal parenchyma), and an N-acetylaspartate (NAA)/Cr ratio of 1.42 (compared with 1.21 in the contralateral normal parenchyma). Also observed were a succinate peak at 2.4 parts per million (ppm) and lipid peaks at 0.9–1.3 ppm (Fig. 5).

The patient required an emergency ventriculostomy on admission; thereafter a bilateral suboccipital craniectomy was performed for decompression and biopsy sampling of the left cerebellar infiltrative lesion. Histopathological analysis of the cerebellar cortex revealed edema and marked acute inflammation of the cerebellar white matter, with only focal acute inflammation of the granul...

Fig. 1. Axial T2-weighted MR image showing expansive T2 prolongation within the left cerebellar hemisphere extending to the medial aspect of the right cerebellar hemisphere, with complete effacement of the fourth ventricle.

Fig. 2. Postcontrast axial T1-weighted MR image demonstrating mild pial enhancement in the inferior aspect of the mass (arrows).
lar cell layer and sparing of the molecular layer and leptomeninges (Fig. 6). Numerous vessels of the white matter displayed fibrinoid necrosis and associated microhemorrhages. The surrounding inflammatory infiltrate was composed of predominantly neutrophils and macrophages with scattered lymphocytes (Fig. 7). There was no evidence of lymphoma or any other malignancy. Gram, Grocott, and acid-fast bacillus stains were negative for bacterial, fungal, and acid-fast organisms. Immunohistochemical stains for cytomegalovirus, herpes simplex viruses 1 and 2, varicella-zoster virus, John Cunningham virus, and toxoplasma were negative. Cerebrospinal fluid analysis only revealed slightly elevated glucose at 82 mg/dl, low protein at 8 mg/dl, and 1 white blood cell/μl. The CSF was negative for routine and anaerobic cultures. Cytology was negative for malignant cells. Microbiological culture of the cerebellar tissue was not performed. Although a causative organism could not be identified, the pathological findings were most consistent with an infectious etiology. The final pathological diagnosis was acute cerebritis involving the cerebellum.

Empirical treatment with intravenous ampicillin, cefepime, and vancomycin was started and the patient’s leukocytosis, ESR, and CRP trended down. Follow-up MRI 3 weeks after the initial study demonstrated significantly improved signal abnormality in the left cerebellar hemisphere, left middle cerebellar peduncle, and left pons with resolution of signal abnormality in the left midbrain (Fig. 8). The mass effect on the fourth ventricle and cerebral aqueduct had also resolved. The patient’s physical examination findings resolved, and the patient improved clinically and was discharged to a rehabilitation facility.

Discussion

Cerebellitis, most common in the pediatric population, is extremely rare in the adult or elderly population. To the best of our knowledge, the oldest previously reported case was of a 66-year-old Korean man with post–influenza vaccination cerebellitis. Gruis et al. reported a case of postinfectious cerebellitis in a 38-year-old woman. Pseudotumoral hemicerebellitis represents a rare manifestation of acute cerebellitis in which a unilateral cerebellar expansive, infiltrative lesion mimics a tumor exerting mass effect on the surrounding structures. An extensive literature search revealed only 7 patients with this condition who required surgical decompression (Table 1).

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Fig. 3. Axial FLAIR images. The abnormal signal also extends into the left brachium pontis (A), pons (B), and left midbrain (C).

Fig. 4. Diffusion imaging. Left: Diffusion-weighted imaging displays increased signal surrounding the lesion. Right: The corresponding apparent diffusion coefficient map shows increased signal in the same areas consistent with facilitated diffusion.

Fig. 5. Results of an intermediate MRS image (TE 135 msec) showing increased Cho/Cr and decreased NAA/Cr ratios with the presence of a succinate (Suc) peak (2.4 ppm) and lipid (Lip) peaks (0.9–1.3 ppm).
The modality of choice to evaluate cerebellitis is MRI. Classic imaging features include expansive T2 prolongation within the cerebellar white matter with or without enhancement. Pial enhancement along the cerebellar folium is often observed. Interval improvement of imaging findings and the absence of a well-defined mass are characteristic. The majority of cases show facilitated diffusion, as was seen in our patient. However, Jaggi et al. reported a case of bacterial cerebellitis in which restricted diffusion was present. Reports on perfusion imaging in cases of cerebellitis are sparse. Nagamitsu et al. described decreased relative cerebral blood flow (CBF) in the affected area of 5 patients with postinfectious cerebellitis shown using $^{123}$I-N-isopropyl-4-iodoamphetamine SPECT. This finding may help differentiate this condition from other more aggressive neoplasms, which are known to have increased perfusion.

There have been several reports involving the use of MRS in cerebellitis that provide nonspecific findings—decreased NAA/Cr and elevated Cho/Cr ratios—with interval increase in NAA/Cr and Cho/Cr ratios—on follow-up MRS. In our patient, the NAA/Cr ratio was also decreased and the Cho/Cr ratio was increased compared with the reference ratios of 2 and 1.2, respectively. In the case presented by Jaggi et al., MRS demonstrated acetate, lactate, and succinate peaks in their patient with anaerobic bacterial cerebellitis and they reported that the presence of acetate, along with lactate, is specific of anaerobic infection. Succinate, acetate, and lactate are known end products arising from microorganisms and, except for lactate, are usually reported in human brain tissue in conjunction with an infection. Magnetic resonance spectroscopy was very helpful in our case as the presence of a succinate peak was a strong imaging indicator of an infectious etiology.

We considered several other differential diagnoses in addition to hemicerebellitis in our case, as there were no previous reports of infectious cerebellitis in this age group. The leading differential diagnosis was Lhermitte-Duclos disease (LDD) because it has similar MRI features with unilateral T2 hyperintense cerebellar involvement. Our case lacked the characteristic findings of preserved cortical striations in a “tiger-striped” or “corduroy” pattern. The T2 appearance was not typical and the presence of pial enhancement helped us to exclude LDD. Additionally, in LDD relative CBF is increased on perfusion imaging, and Cho/Cr, as well as myo-inositol, is decreased on MRS, whereas in our case there was decreased perfusion and
TABLE 1: Cerebellitis in 7 cases requiring surgical decompression

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs)</th>
<th>Imaging Findings</th>
<th>Clinical Findings</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asenbauer et al., 1997</td>
<td>4</td>
<td>cerebellar swelling, tonsillar herniation, hydrocephalus</td>
<td>persistent signs/symptoms of intracranial hypertension</td>
<td>decompression</td>
</tr>
<tr>
<td>Omeis et al., 2002</td>
<td>16</td>
<td>rt cerebellar swelling, mass effect w/ 4th ventricular displacement</td>
<td>ataxia, somnolence, diplopia, rt trigeminal neuralgic pain, &amp; rt-sided dysmetria</td>
<td>decompression</td>
</tr>
<tr>
<td>Jabbour et al., 2003</td>
<td>13</td>
<td>rt cerebellar swelling, mass effect w/ 4th ventricular displacement</td>
<td>persistent signs/symptoms of intracranial hypertension after 5 days of steroids</td>
<td>decompression</td>
</tr>
<tr>
<td></td>
<td>6</td>
<td>cerebellar swelling, disappearance of basal cisterns, deformed 4th ventricle, triventricular hydrocephalus</td>
<td>persistent signs/symptoms of intracranial hypertension after ventricular drain placement</td>
<td>decompression &amp; resection of cerebellar tonsils</td>
</tr>
<tr>
<td>de Ribaupierre et al., 2005</td>
<td>14</td>
<td>fullness of posterior fossa, slight transtentorial &amp; foramen magnum herniation, hydrocephalus</td>
<td>persistent headaches, photophobia, nausea, voice changes, &amp; bradycardia after ventricular drain placement</td>
<td>decompression &amp; resection of cerebellar tonsils</td>
</tr>
<tr>
<td>Wagel et al., 2010</td>
<td>4</td>
<td>diffuse cerebellar swelling w/ significant mass effect &amp; tonsillar herniation</td>
<td>persistent signs/symptoms of intracranial hypertension w/ acute clinical deterioration on Day 4</td>
<td>decompression w/ removal of C-1 vertebral arch</td>
</tr>
<tr>
<td>Morais et al., 2013</td>
<td>15</td>
<td>lt cerebellar swelling, deviated 4th ventricle &amp; tonsillar herniation w/ mild hydrocephalus</td>
<td>persistent signs/symptoms of intracranial hypertension after 1 day of antibiotics, antivirals, &amp; steroids</td>
<td>decompression</td>
</tr>
</tbody>
</table>

Conclusions

Hemicerebellitis is an extremely rare entity in the elderly population. The noninvasive diagnosis of pseudotumoral hemicerebellitis may be difficult without a biopsy. The diagnosis should be suspected in cases involving an atypical clinical presentation, with characteristic imaging features of cerebellar swelling, T2 prolongation, and leptomeningeal enhancement. Advanced neuroimaging features such as decreased CBF on perfusion MRI and the presence of a succinate peak on MRS may aid in the exclusion malignancy. Most cases of cerebellitis are self-limited and may respond to medical therapy alone, with more invasive surgical procedures reserved for fulminating cases that result in rapid clinical deterioration requiring surgical decompression.

Elevated Cho/Cr level. In view of our patient’s history, other diagnostic considerations included neoplasms such as lymphoma and glial tumors. The lack of enhancement and characteristic findings on diffusion and perfusion MRI argued against these. Acute disseminated encephalomyelitis is also considered in the differential diagnosis; however, there is usually subcortical white matter, deep gray matter, and spinal cord involvement, which was not seen in our case. Karmon et al. described a case of paraneoplastic cerebellar degeneration initially presenting as acute infectious cerebellitis that eventually turned out to be Hodgkin’s disease. However, the case lacked the MRI features characteristic of cerebellitis, which were present in our case. Our patient was ultimately diagnosed as having acute infectious pseudotumoral hemicerebellitis because of the consistent neuroimaging and pathological findings as well as clinical and radiological improvement that the patient exhibited once he started receiving antibiotics.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Mohan, Gupta, Maralani. Acquisition of data: all authors. Analysis and interpretation of data: Mohan, Chawla, Gopal. Drafting the article: Mohan, Gupta, Maralani, Gopal. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Statistical analysis: Chawla. Administrative/technical/material support: Mohan. Study supervision: Mohan.

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