Chronic cerebellar hemorrhage in spontaneous intracranial hypotension: association with ventral spinal cerebrospinal fluid leaks

Clinical article

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Object. Spontaneous intracranial hypotension is an important cause of new-onset daily persistent headache. Cerebellar hemorrhage has been identified as a possible feature of spontaneous intracranial hypotension. The authors reviewed the MR imaging studies from a group of patients with spontaneous intracranial hypotension to assess the presence of cerebellar hemorrhage.

Methods. Medical records and radiological images were reviewed in 262 cases involving patients with spontaneous intracranial hypotension who had undergone MR imaging of the brain as well as spinal imaging.

Results. Chronic cerebellar hemorrhages were found in 7 (2.7%) of the 262 patients with spontaneous intracranial hypotension. These hemorrhages were found in 7 (19.4%) of the 36 patients with a ventral spinal CSF leak and in none of the 226 patients who did not have such a CSF leak (p < 0.0001). The degree of hemosiderin deposits was variable, ranging from mild involvement of the cerebellar folia to widespread superficial siderosis. Only the 1 patient with superficial siderosis had symptoms due to the hemorrhages. The time period between the onset of symptoms due to spontaneous intracranial hypotension and MR imaging examination was significantly longer in those patients with cerebellar hemorrhage than in those with a ventral spinal CSF leak and no evidence for cerebellar hemorrhage (mean 19.6 years vs 2.3 months, p < 0.0001).

Conclusions. Chronic cerebellar hemorrhage should be included among the manifestations of spontaneous intracranial hypotension. The severity is variable, but the hemorrhage generally is asymptomatic. The underlying spinal CSF leak is ventral and mostly of long duration. (DOI: 10.3171/2011.5.SPINE10890)

Key Words • cerebellar hemorrhage • cerebrospinal fluid leak • headache • intracranial hypotension • superficial siderosis
For statistical analysis, ANOVA and the Fisher exact test were used to compare groups and associations between the groups and patient characteristics, respectively.

Results

During the 9-year study period, a total of 273 patients with spontaneous intracranial hypotension were evaluated. Two patients did not undergo brain MR imaging and 9 patients did not undergo any spinal imaging, leaving a total of 262 evaluable cases.

Chronic cerebellar hemorrhages were found in 7 (2.7%) of the 262 patients with spontaneous intracranial hypotension (Figs. 1–7). These hemorrhages were found in 7 (19.4%) of the 36 patients with a ventral spinal CSF leak and in none of the 226 patients who did not have such a CSF leak (p < 0.0001).

The mean age of the 5 men and 2 women was 39.6 years (range 15–63 years) at the time of onset of symptoms due to spontaneous intracranial hypotension and 59.1 years (range 41–78 years) at the time of their first MR imaging study demonstrating the cerebellar hemorrhages (Table 1). In 2 patients, the symptoms of spontaneous intracranial hypotension had resolved (over 2- and 4-year periods, respectively) decades before the diagnosis of cerebellar hemorrhage, and none of the typical imaging features of spontaneous intracranial hypotension were present on MR images. The remaining 5 patients remained symptomatic, and in these patients, MR imaging showed the typical features of spontaneous intracranial hypotension in addition to the cerebellar hemorrhages.

The MR imaging appearance of the cerebellar hemorrhages was characterized by bilateral and symmetrical linear foci of hemosiderin staining involving the superior or middle regions of the cerebellar hemispheres, usually also involving the superior vermis. All hemorrhages were chronic, without gliosis or edema in the parenchyma. The degree of hemosiderin deposits was variable, ranging from mild to moderate involvement of the cerebellum in 6 patients; there was typical widespread superficial siderosis involving the whole neuraxis, but most pronounced in the cerebellum, in 1 patient. Only this latter patient, who presented with the classical clinical manifestations of progressive ataxia and sensorineural hearing loss, was symptomatic from the chronic hemorrhages. Two other patients also underwent audiometry for “aural fullness” but results were normal. Repeat MR imaging scans were performed in all 7 patients between 6 months and 4 years after the initial study (mean 2.2 years), and no change in the cerebellar hemorrhages was noted.

The time period between the onset of symptoms due to spontaneous intracranial hypotension and MR imaging examination was longer in those patients with cerebellar hemorrhage (mean 19.6 years, range 1 month–43 years) than in those with a ventral spinal CSF leak and no evidence for cerebellar hemorrhage on MR imaging (mean 2.3 months, range 2 days–30 months; p < 0.0001; Table 2). Xanthochromia was found in 2 of the 7 patients with cerebellar hemorrhage, but in none of the 29 with a
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ventral spinal CSF leak and no evidence for cerebellar hemorrhage on MR imaging ($p = 0.04$). Lumbar puncture and CSF examination were performed for the evaluation of superficial siderosis in 1 patient and at the time of CT myelography in the remaining 6 patients. No other statistically significant differences were found between the 2 groups of patients with ventral spinal CSF leaks.

Conventional cerebral angiography was performed in 3 patients and MR or CT angiography in 4. No source for the cerebellar hemorrhages was detected.

All 5 patients with persistent headaches due to the ventral spinal CSF leaks underwent between 2 and 7 epidural blood patches with complete and sustained resolution of symptoms in 2 patients. The 3 remaining patients underwent surgical repair of the ventral spinal CSF leak with complete and sustained resolution of symptoms in 2 patients and partial resolution in 1 patient.

The 1 patient with superficial siderosis also was found to have a middle fossa arachnoid cyst (Fig. 1), and he underwent a craniotomy. No abnormal fragile vasculature was encountered, and his symptoms continued. The patient declined treatment for his ventral spinal CSF leak.

Clinical follow-up was complete through September 2010 and ranged from 3 to 49 years (mean 24.6 years) from the onset of symptoms, from 3 to 9 years from the initial MR imaging examination (mean 4.6 years), and from 6 months to 5 years (mean 2.9 years) from the last treatment for spontaneous intracranial hypotension.

**Discussion**

In this study, we found evidence for chronic cerebellar

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Fig. 2. **A** and **B**: Axial and sagittal T2-weighted MR images showing symmetrical signal hypointensities in the superior cerebellum (arrows) consistent with hemosiderin. **C** and **D**: Sagittal and axial T2-weighted spinal MR images showing a ventral CSF collection (arrows).

Fig. 3. **A**: Sagittal T1-weighted MR image showing brain sagging with inferior displacement of cerebellar tonsils (arrow). Note the downward displacement of the optic chasm and distortion of brainstem. **B** and **C**: Axial T2-weighted and coronal gradient echo MR images showing symmetrical linear and punctate signal changes of hemosiderin along the vermis and cerebellar hemispheres. **D**–**F**: Sagittal and axial T2-weighted MR images (**D** and **E**, respectively) and CT myelogram (**F**) showing the ventral epidural CSF collection (arrows).
Fig. 4. A: Axial T1-weighted MR image showing diffuse pachymeningeal enhancement. B: Axial T2-weighted MR image revealing subtle hypointense signal in the upper cerebellar folia. C and D: Coronal gradient echo MR images demonstrating accentuation of the abnormal dark signal (arrows) related to chronic hemosiderin. E and F: Sagittal and axial T2-weighted MR images showing a ventral epidural CSF collection.

Fig. 5. A: Sagittal T1-weighted image showing brain sagging with flattening of the pons (arrow). B: Axial T2-weighted MR image revealing linear dark signal in the upper vermis (arrows). C–F: Coronal gradient echo, axial susceptibility-weighted, and sagittal gradient echo MR images demonstrating accentuation of the signal loss due to hemosiderin in the cerebellar vermis and upper folia. G and H: Sagittal and axial T2-weighted MR images showing the ventral epidural CSF collection (arrows).
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hemorrhage in about 2.5% of patients with spontaneous intracranial hypotension. There was a strong correlation with the type of underlying spinal CSF leak. Cerebellar hemorrhages were seen in about one-fifth of patients with ventral spinal CSF leaks but in none of the patients with other types of CSF leaks. Among the patients with ventral spinal CSF leaks, duration of symptoms was strongly associated with the development of cerebellar hemorrhage.

A variable degree of hemosiderin deposits was observed, ranging from mild cerebellar involvement to classic superficial siderosis involving the entire neuraxis. However, in most patients only the cerebellum was involved and the hemorrhages were asymptomatic. The one patient with superficial siderosis syndrome presented with the classical clinical manifestations of sensorineural hearing loss and ataxia. Superficial siderosis is defined as a degenerative disorder characterized by widespread deposition of hemosiderin in the leptomeninges and sub-pial layers of the brain and spinal cord.\textsuperscript{5,12,13} Previously reported patients with a diagnosis of superficial siderosis syndrome and spontaneous intracranial hypotension all had ventral spinal CSF leaks.\textsuperscript{10,14,16,17} These typical ventral spinal CSF collections also have been described in other patients with superficial siderosis syndrome who did not carry a diagnosis of spontaneous intracranial hypotension.\textsuperscript{4,11,15,19,20,27} However, it appears that in some of these patients in whom superficial siderosis syndrome was diagnosed, only the cerebellum was involved.\textsuperscript{14,16}

The source of cerebellar bleeding in our patient population has not been established. Previously we have

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{fig6.png}
\caption{A: Axial T1-weighted MR image showing diffuse pachymeningeal enhancement. B: Axial gradient echo MR image showing linear dark signal along the superior cerebellar folia. C and D: Digital subtraction myelogram (C) showing the leakage site (arrow) and resultant ventral fluid collection, that is confirmed on the axial CT myelogram (D, arrow).}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{fig7.png}
\caption{A: Axial T1-weighted MR image shows diffuse pachymeningeal enhancement. B and C: Coronal gradient echo MR images demonstrate punctate and linear foci of dark signal (arrows) indicating chronic hemosiderin staining. D and E: Sagittal T2-weighted MR and axial CT myelogram images show a ventral epidural CSF collection (arrows).}
\end{figure}
TABLE 1: Summary of demographic and clinical characteristics of 7 patients with spontaneous intracranial hypotension and cerebellar hemorrhage

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs) at Sx Onset, Sex</th>
<th>Presenting Sx</th>
<th>Age (yrs) at MRI</th>
<th>MRI Findings</th>
<th>Extent of Ventral CSF Leak</th>
<th>Opening Pressure (cm H₂O)</th>
<th>Xanthochromia</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>35, M</td>
<td>orthostatic headache</td>
<td>78</td>
<td>superficial siderosis</td>
<td>C6–T7</td>
<td>0</td>
<td>no</td>
</tr>
<tr>
<td>2</td>
<td>22, M</td>
<td>orthostatic headache</td>
<td>59</td>
<td>cerebellar hemorrhage</td>
<td>C2–T12</td>
<td>3</td>
<td>no</td>
</tr>
<tr>
<td>3*</td>
<td>54, F</td>
<td>quadriplegia, headache</td>
<td>54</td>
<td>meningeal enhancement, sagging, cerebellar hemorrhage</td>
<td>C6–T10</td>
<td>2</td>
<td>no</td>
</tr>
<tr>
<td>4</td>
<td>63, M</td>
<td>orthostatic headache</td>
<td>76</td>
<td>subdural hematoma, meningeal enhancement, cerebellar hemorrhage</td>
<td>T1–7</td>
<td>3</td>
<td>yes</td>
</tr>
<tr>
<td>5</td>
<td>37, F</td>
<td>orthostatic headache</td>
<td>48</td>
<td>sagging, meningeal enhancement, pituitary hyperemia, cerebellar hemorrhage</td>
<td>C6–T11</td>
<td>2.5</td>
<td>yes</td>
</tr>
<tr>
<td>6</td>
<td>51, M</td>
<td>orthostatic headache</td>
<td>58</td>
<td>subdural hematoma, sagging, meningeal enhancement, cerebellar hemorrhage</td>
<td>C3–L5</td>
<td>6</td>
<td>no</td>
</tr>
<tr>
<td>7</td>
<td>15, M</td>
<td>orthostatic headache</td>
<td>41</td>
<td>sagging, meningeal enhancement, pituitary hyperemia, cerebellar hemorrhage</td>
<td>C6–T7</td>
<td>14</td>
<td>no</td>
</tr>
</tbody>
</table>

* This case was reported previously. Quadruplegia was likely the result of severe hindbrain herniation associated with compression of the upper cervical spinal cord.

TABLE 2: Demographic and clinical characteristics of 36 patients with spontaneous intracranial hypotension and ventral spinal CSF leaks*

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Total Group</th>
<th>Pts w/ Cerebellar Hemorrhage</th>
<th>Pts w/o Cerebellar Hemorrhage</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>no. of pts</td>
<td>36</td>
<td>7</td>
<td>29</td>
<td></td>
</tr>
<tr>
<td>male sex</td>
<td>36%</td>
<td>71%</td>
<td>28%</td>
<td>0.07</td>
</tr>
<tr>
<td>age at onset of Sx (yrs)</td>
<td>41.4</td>
<td>39.6</td>
<td>41.8</td>
<td>0.62</td>
</tr>
<tr>
<td>mean</td>
<td>42.5</td>
<td>37.0</td>
<td>43.0</td>
<td></td>
</tr>
<tr>
<td>median</td>
<td>12.5</td>
<td>24.0</td>
<td>10.0</td>
<td></td>
</tr>
<tr>
<td>IQR</td>
<td>15–63</td>
<td>15–63</td>
<td>19–55</td>
<td></td>
</tr>
<tr>
<td>range</td>
<td>15–63</td>
<td>15–63</td>
<td>19–55</td>
<td></td>
</tr>
<tr>
<td>age at first MRI (yrs)</td>
<td>45.3</td>
<td>59.1</td>
<td>41.9</td>
<td>0.0001</td>
</tr>
<tr>
<td>mean</td>
<td>43</td>
<td>58</td>
<td>43</td>
<td></td>
</tr>
<tr>
<td>median</td>
<td>12.5</td>
<td>16.5</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>IQR</td>
<td>19–78</td>
<td>41–78</td>
<td>19–55</td>
<td></td>
</tr>
<tr>
<td>range</td>
<td>19–78</td>
<td>41–78</td>
<td>19–55</td>
<td></td>
</tr>
<tr>
<td>time from Sx to MRI (mos)</td>
<td>47.6</td>
<td>235</td>
<td>2.3</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>mean</td>
<td>0.5</td>
<td>155.9</td>
<td>0.3</td>
<td></td>
</tr>
<tr>
<td>median</td>
<td>4.4</td>
<td>270</td>
<td>0.7</td>
<td></td>
</tr>
<tr>
<td>IQR</td>
<td>0.1–516</td>
<td>0.9–516</td>
<td>0.1–30</td>
<td></td>
</tr>
<tr>
<td>range</td>
<td>13.4 ± 4.9</td>
<td>13.0 ± 5.6</td>
<td>13.5 ± 4.8</td>
<td>0.82</td>
</tr>
</tbody>
</table>

* IQR = interquartile range; Pts = patients.
† Extent of CSF leak.
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hypothesized that 2 mechanisms could play a role: 1) occlusion of superior cerebellar bridging veins due to brain sagging resulting in hemorrhagic infarctions and 2) actual rupture of these bridging veins. A venous source of hemorrhage is consistent with the distribution of the hemosiderin deposits bilaterally along the cerebellar folia, which is similar to that seen following iatrogenic spinal CSF loss. This pattern of hemosiderin distribution and the lack of cerebellar gliosis argue against venous hemorrhagic infarcts as the underlying pathology. Others have suggested that friable vessels at the site of the ventral dural tear are the likely source of chronic bleeding in patients with superficial siderosis and ventral spinal CSF leaks.1,16

Although not common in our study, only patients with ventral spinal CSF leaks who had cerebellar hemorrhages were found to have xanthochromia on CSF examination. Xanthochromia has been described previously in patients with spontaneous intracranial hypotension26 and also is a frequently reported finding in patients with superficial siderosis.8,10,14,15

The present study demonstrates that long duration of symptoms due to spontaneous intracranial hypotension is strongly associated with the occurrence of cerebellar hemorrhage, possibly explaining why only ventral spinal CSF leaks were found in these patients. Compared with other types of spontaneous spinal CSF leaks, ventral CSF leaks are less likely to resolve spontaneously, are less responsive to treatments such as epidural blood patching, and are more likely to recur (W.I. Schievink et al., unpublished data). Also, the volume of CSF loss generally is more extensive in patients with spontaneous intracranial hypotension from ventral CSF leaks compared with the other types of CSF leaks (W.I. Schievink et al., unpublished data). In reviewing the literature, it is apparent that the interval between the onset of symptoms from spinal CSF leaks (that is, positional headache) and the development of symptoms from superficial siderosis also is typically long, measured in years or decades.10,17,18 Thus, the typical patient with chronic cerebellar hemorrhage and a ventral spinal CSF collection presents with a longstanding or remote history of positional headaches. Because the positional headaches can be so characteristic and debilitating, it is not unusual for patients to recount such headaches even if they occurred in the distant past. This study suggests that cerebral angiography is not indicated for the diagnostic evaluation of patients with a spontaneous ventral spinal CSF leak who exhibit these characteristic cerebellar hemosiderin deposits. Conversely, spinal imaging should be considered in patients who are found to have these cerebellar hemosiderin deposits, and patients should be asked about a history, possibly remote, of positional headaches. Chronic cerebellar hemorrhage should be included among the manifestations of spontaneous intracranial hypotension. Other types of intracranial hemorrhage associated with spontaneous intracranial hypotension include, in order of decreasing frequency, subdural hematomas,15 hemorrhagic venous infarctions,22 and Duret hemorrhages.2

Conclusions

Variable amounts of cerebellar hemosiderin deposits should be included among the manifestations of spontaneous intracranial hypotension. The underlying spinal CSF leak is ventral and generally of long duration.

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: Schievink. Acquisition of data: Schievink, Maya. Analysis and interpretation of data: all authors. Drafting the article: Schievink. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Schievink. Statistical analysis: Nuño.

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Please include this information when citing this paper: published online July 8, 2011; DOI: 10.3171/2011.5.SPINE10890.

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