An Adult Case of Hemifacial Spasm as the Predominant Manifestation of Spontaneous Intracranial Hypotension

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**Introduction**
- Spontaneous intracranial hypotension (SIH) is increasingly recognized cause of secondary headaches.\(^1\)
- Cerebral nerve palsy are associated with SIH, with CN VIII as the most commonly reported and less frequently III, IV, and VI.\(^1\)
- Hemifacial spasm refers to unilateral involuntary contractions of the facial muscles.\(^2\)
- It is caused by facial nerve demyelination from aberrant vascular contact at the nerve root exit zone, akin to classic trigeminal neuralgia, although there can be other etiologies.\(^3\)
- This report is the first described case in the literature of hemifacial spasm secondary to SIH

**Case Report**
- In 2017, a 63 yo F developed new onset headaches which would start abruptly in the upper neck and radiate "shocks" to the occiput. They were uniquely triggered by bending over and Valsalva maneuvers and were associated with ear fullness.
- The patient was diagnosed with tension headaches and cervical neuralgia. Diet, exercise, and weight loss improved her initial headaches.
- In 2019, she developed right periorbital twitching, right lower face weakness, and a "pulling" sensation in her right lower face which would occur multiple times per hour throughout the day and worsened towards the end of the day.
- The patient would experience improvement upon waking up from sleep in the morning. She also developed dizziness, hearing difficulty, and subjective memory impairment.
- A diagnosis of Bell's Palsy and steroid treatment was administered without improvement.
- The patient presented to Dr. Fermo for a second opinion. On exam, there was mild weakness of the right lower face that overwhelmed with activation. There were also continuous contractions in the right periorbital and perioral regions at rest, and more so with activation. Her exam was otherwise unremarkable, including a normal eye-exam.

**Radiographic Findings**
- MRI of the brain with and without contrast demonstrated diffuse, smooth pachymeningeal enhancement, engagement of the venous sinuses, pituitary engorgement, cerebellar tonsillar ectopia, and signs of brain sagging. (Table 1), highly suggestive of intracranial hypotension.\(^4\)
- There were no aberrant vessels near the anterior fontanelle.

**Table 1. Key radiographic signs of SIH and their change in response to EBP**

<table>
<thead>
<tr>
<th><strong>Radiographic Signs of Intracranial Hypotension</strong></th>
<th><strong>Brain MRI 7/17/19 (Before treatment)</strong></th>
<th><strong>Brain MRI 9/5/19 (After first blind EBP)</strong></th>
<th><strong>Brain MRI 2/22/20 (After second blind EBP and one targeted EBP)</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Engorged Venous Sinus</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Pachymeningeal Enhancement</td>
<td>Yes</td>
<td>Yes (improved)</td>
<td>Yes</td>
</tr>
<tr>
<td>Subarachnoid Collections</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Supraoptic Cistern (abnormal &lt; 4 mm)</td>
<td>&lt; 4 mm</td>
<td>&lt; 4 mm</td>
<td>&lt; 4 mm</td>
</tr>
<tr>
<td>Preoptic Cistern (abnormal &lt; 5 mm)</td>
<td>1.6 mm</td>
<td>2.3 mm</td>
<td>3 mm</td>
</tr>
<tr>
<td>Mamillo-Interthalamic Distance (abnormal &lt; 6 mm)</td>
<td>3.4 mm</td>
<td>4.6 mm</td>
<td>5.2 mm</td>
</tr>
<tr>
<td>Diameter of Basal Infratentorial Tract</td>
<td>8.1 mm</td>
<td>7.7 mm</td>
<td>6.6 mm</td>
</tr>
</tbody>
</table>

**Figure 1. T1 sagittal and coronal MRI brain with contrast before treatment (A, B) after first blind EBP (C, D) and after targeted EBP (E, F).**

**Discussion and Conclusion**
- SIH causes traction, compression, displacement or congestion of critical brain structures, leading to symptoms.\(^4\)
- Improvement of spasms "attacks" and radiographic improvement of brain sag and venous sinus congestion after targeted EBP suggests that downward traction on CN VII and sub-radiographic, venous congestion close to the nerve root were the most likely initial mechanisms of the patient's hemifacial spasm.\(^4\)
- This is further supported by the original history of end-of-day symptoms and improvement of symptoms with downward traction on CN VII, suggesting CSF leakage with prolongation to the end-of-day headache.\(^4\)
- SIH is a condition that can present in a highly variable manner. When investigating the etiology of typical headaches, SIH and its physiologic effects on other structures within the CRN should be considered. Imaging allows for precise characterizations of SIH, and EBP is an effective means of treatment of SIH.