What Every Radiologist Should Know About Intracranial Hypotension: A Misdiagnosis and Mismanagement

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Purpose:
1. Illustrate multimodal imaging findings of intracranial hypotension syndrome.
2. Illustrate imaging findings of cranial and spinal CSF leak as they relate to intracranial hypotension.
3. Discuss the correlation of MRI features with clinical manifestations.
4. Discuss MRI findings and pitfalls to avoid inappropriate or delayed management.

Introduction:
References to intracranial hypotension syndrome, both spontaneous and secondary, date back more than six decades.1 Despite significant patient morbidity, intracranial hypotension syndrome continues to remain underdiagnosed both clinically and by imaging. The clinical and neuroradiologic signs and symptoms of intracranial hypotension are often confused with those of other neurologic diseases.2 Common symptoms include positional headache, neck pain/stiffness, hearing abnormalities, and nausea/vomiting.3 Misdiagnosis of intracranial hypotension syndrome has been demonstrated in as often as 94% of cases in some studies wherein ‘migraine’ and ‘meningitis’ were the most frequent misdiagnoses.4 The role of imaging in intracranial hypotension syndrome management is vital for accurate diagnosis to ensure appropriate early treatment, though limited studies exist about the use of neuroimaging in the initial evaluation.1 In this exhibit we demonstrate the different multimodal imaging findings of intracranial hypotension syndrome presenting at our institution.

Table 1.

<table>
<thead>
<tr>
<th>Pachymeningal Enhancement</th>
<th>Post-operative Changes, Meningiomas, Metastatic Disease, Granulomatous Disease, Secondary CNS Lymphoma</th>
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<tbody>
<tr>
<td>Pituitary Hypo(</td>
<td>)enhancement</td>
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<tr>
<td>Cerebellar Tonsil Displacement</td>
<td>Chiari I Malformation</td>
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Discussion:
Intracranial hypotension is most commonly due to a CSF leak along the neuroaxis, and can be primary (spontaneous) or secondary (often iatrogenic). Common causes of spontaneous leakage are weakness in the dura or arachnoid cysts, often due to a connective tissue disorder. Due to clinical and neuroimaging mimickers of intracranial hypotension, misdiagnosis and inappropriate or delayed management is frequent. While enhancement of the pachymeninges, brainstem slumping, subdural hygroma or hematoma, pituitary enlargement, slit-like ventricles, and dural venous sinus engorgement are common hallmarks of intracranial hypotension, these findings in isolation can often be mistaken for other diagnoses. Common radiographic findings that mimic intracranial hypotension syndrome are listed in Table 1. We suggest an MRI protocol as part of the initial diagnosis and evaluation of patients with intracranial hypotension. Correlation of collective MRI features with clinical manifestations is critical for prompt diagnosis. Further confirmation of suspected intracranial hypotension can be performed with myelography.

Treatment for intracranial hypotension syndrome consists of epidural blood patching for persistently symptomatic patients with surgical ligation or repair only considered for cases not responsive to at least two attempts.5

Common MR pitfalls include misdiagnosis of intracranial hypotension syndrome as Chiari malformation and subsequent treatment with decompressive surgery. While all hallmark features are not present in every case of intracranial hypotension, high clinical suspicion and correlation of collective MRI features with known common clinical features is necessary for accurate and prompt diagnosis.

Our comprehensive examination of neuroradiography in patients across Tulane Medical Center will contribute to the scarce literature on neuroradiography in intracranial hypotension syndrome to optimize delivery of specialized tertiary care.