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Intracranial hypotension masking a complex lymphatic anomaly: a case of skull base CSF-lymphatic leak

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Introduction
Spontaneous intracranial hypotension (SIH) is a debilitating condition typically resulting from cerebrospinal fluid (CSF) leaks. While most cases are idiopathic or related to spinal discogenic microspurs, rare systemic conditions can predispose individuals to these leaks. Gorham-Stout Syndrome (GSS) is an exceptionally rare lymphatic anomaly characterized by progressive osteolysis where bone is replaced by proliferating lymphatic tissue. We present a unique case of GSS manifesting as SIH via a CSF-lymphatic fistula, highlighting the clinical challenges in diagnosing rare etiologies for refractory headaches.

Case presentation
A 20-year-old woman with medical history for meningitis approximately 16 months prior presented with a history of sudden-onset, exertional cranial pain triggered by physical activities such as boxing and gym workouts. The headache was associated with tinnitus, aural fullness, and episodes of intense pain accompanied by neck stiffness and nausea. Brain MRI confirmed intracranial hypotension, and lumbar puncture revealed a low opening pressure (<1 cmH2O). Spine and cranial imaging identified multifocal T1/T2 hyperintense bone lesions from the skull base to the sacrum. CT myelography pinpointed bone resorption at the petrous apex, creating a CSF-lymphatic fistula in the left middle cranial fossa, allowing a direct connection between the subarachnoid space and the abnormal lymphatic structures. Following a GSS diagnosis, the patient underwent endoscopic surgical repair, resulting in significant clinical resolution of her symptoms.

Discussion
While SIH is typically spinal in origin, this case identifies an unconventional cranial etiology: a CSF-lymphatic fistula. In GSS, the replacement of bone with lymphatic tissue compromises the structural integrity of the skull base. Literature suggests that involvement of the petrous apex is particularly rare, creating a direct shunt between the subarachnoid space and pathological lymphatic channels. Diagnostically, this presentation is challenging because traditional spinal imaging may not show the "extradural fluid collections" typical of SIH. Instead, the diagnosis relies on identifying multifocal lytic bone lesions and using CT myelography to pinpoint the cranial defect. The patient's history of meningitis underscores the risk of these fistulas acting as a bidirectional conduit for CSF loss and retrograde infection. Unlike spinal leaks, these cranial-lymphatic connections are refractory to epidural blood patches and necessitate targeted surgical closure.

Conclusion
This case highlights GSS as a rare but significant cause of SIH through the formation of a CSF-lymphatic fistula. For patients presenting with SIH and multifocal bone lesions, a thorough diagnostic evaluation is crucial. Recognizing this rare lymphatic anomaly as an underlying cause of CSF leaks is vital for ensuring effective surgical intervention and long-term management.

Keywords:
Gorham-stout syndrome
Intracranial hypotension
Skull base
CSF-lymphatic

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