

Importance of diagnosing spontaneous intracranial hypotension with uncommon clinical presentation

Spontaneous intracranial hypotension (SIH) results from loss of cerebrospinal fluid (CSF), most commonly from a CSF spinal leak, without history of recent surgery, spinal procedure or trauma. Patients typically present with orthostatic headache, i.e. headache upon standing up that improves when lying down. This condition is considered to be underdiagnosed and frequently confused with others such as craniocervical Chiari malformations. Awareness of SIH has heightened in recent years. Management includes first medical treatment (rest, hydration, caffeine) with indication of epidural blood spinal injection (blood patch) when symptoms persist. Surgical repair of the spinal dural defect is reserved for cases that fail to respond to conservative measures.

Normal



Brain sagging

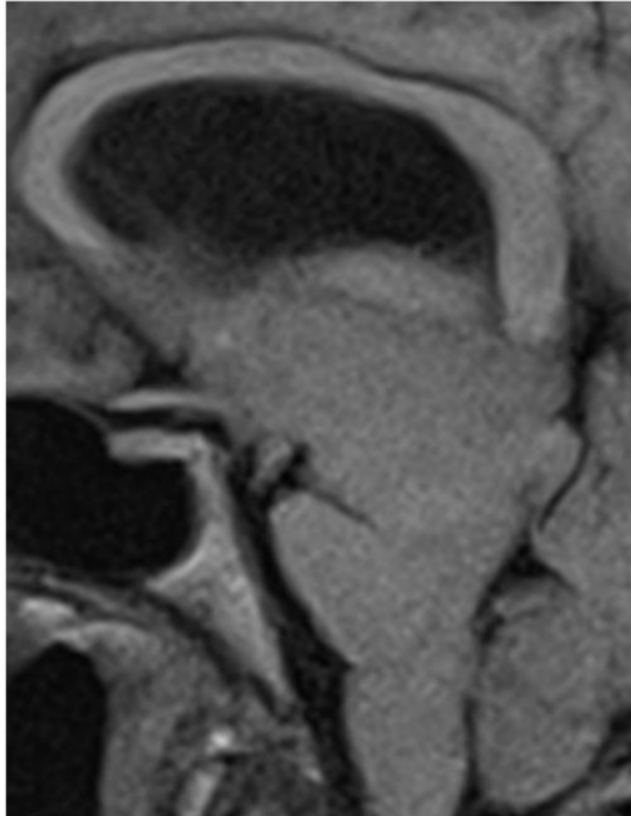


Fig. 1. Normal brainstem anatomy on sagittal MRI (left) compared to severe brainstem sagging from SIH (right)

Atypical clinical presentations of SIH have been sparsely reported in the literature and include:

obtundation, stupor, deafness, memory deficit, dementia with behavioral changes, abnormal involuntary movements and incoordination. On brain MRI (see Figure below) classical features of SIH are: downward brain sagging with effacement of perimesencephalic cisterns and pituitary and dural engorgement, which are sometimes associated with subdural hematoma.

We retrospectively studied 33 patients with classical and 8 patients with atypical SIH. Atypical SIH patients were older than classical SIH and had a trend for higher representation of men over women, while women are more commonly affected than men in classical SIH. Atypical SIH patients had longer clinical history and lower odds of becoming symptom free compared to classical SIH. The degree of brain sagging as seen on brain MRI was more severe in atypical versus classical SIH patients without significant differences in other radiographic signs of intracranial hypotension such as dural enhancement, fluid collections or cerebellar tonsillar herniation.

The longer clinical duration and less favorable outcome of atypical SIH may relate to a weaker dura from underlying connective tissue anomaly and / or insufficient physiologic adaptation to CSF volume deficit leading to brain stretching. The potential for reversibility of symptoms and signs even in chronic cases with atypical presentation after tamponade of the CSF leak underscores the importance of diagnosing SIH and distinguishing it from other conditions.

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