Case Report:
Sinking Brain: Unusual Cause of Orthostatic Headache.

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Abstract: We report a case presenting with an orthostatic headache. Brain magnetic resonance imaging (MRI) revealed typical pachymeningeal enhancement. CT myelography revealed leakage at the thoracic level. Patient was successfully treated by lumbar epidural blood patch (EBP).

Key Words: Sinking brain, Spontaneous intracranial hypotension

Introduction:
Sinking brain or sagging brain is seen in Spontaneous intracranial hypotension which has become a well-recognized clinical entity, but it remains an uncommonly, and probably underdiagnosed, cause of headache; its estimated prevalence is only one in 50,000 individuals. The clinical spectrum of spontaneous intracranial hypotension is quite variable and includes headache, neck stiffness, cranial nerve dysfunction, radicular arm pain, and symptoms of diencephalic or hindbrain herniation. Leakage of the spinal cerebrospinal fluid (CSF) is the most common cause of spontaneous intracranial hypotension. A combination of an underlying weakness of the spinal meninges and a more or less trivial traumatic event is often found to cause this event in these patients. Typical magnetic resonance imaging findings include diffuse pachymeningeal enhancement, subdural fluid collections, and downward displacement of the brain, sometimes mimicking a Chiari I malformation. Opening pressure is often, but not always, low, and examination of CSF may reveal pleocytosis, an elevated protein count, and xanthochromia. The use of myelography computerized tomography scanning is the most reliable method for the accurate localization of the CSF leak. If conservative treatment fails then treatment of choice is a lumbar epidural blood patch, regardless of the location of the CSF leak. If the epidural blood patch fails, the blood patch procedure can be repeated at the lumbar level, or a blood patch can be directed at the exact site of the leak. Surgical repair of the CSF leak is safe and generally successful.

Case Report:
A 35-year-old man presented to our institution with progressive headache for last 10 days. Headache was aggravated when he assumed an upright posture (while sitting or standing) and alleviated during lying down. Headache was not relieved by analgesics and he was unable to tolerate the pain when assuming the upright position. Patient was farmer by occupation and he was constructing his home for last 3 months and used carry heavy loads over his back. He denied any history of a traumatic accident or lumbar puncture examination.

Physical and neurological examinations performed were normal. The head CT only revealed mild high attenuation is is only one in 50,000 individuals. The clinical spectrum of spontaneous intracranial hypotension is quite variable and includes headache, neck stiffness, cranial nerve dysfunction, radicular arm pain, and symptoms of diencephalic or hindbrain herniation. Leakage of the spinal cerebrospinal fluid (CSF) is the most common cause of spontaneous intracranial hypotension. A combination of an underlying weakness of the spinal meninges and a more or less trivial traumatic event is often found to cause this event in these patients. Typical magnetic resonance imaging findings include diffuse pachymeningeal enhancement, subdural fluid collections, and downward displacement of the brain, sometimes mimicking a Chiari I malformation. Opening pressure is often, but not always, low, and examination of CSF may reveal pleocytosis, an elevated protein count, and xanthochromia. The use of myelography computerized tomography scanning is the most reliable method for the accurate localization of the CSF leak. If conservative treatment fails then treatment of choice is a lumbar epidural blood patch, regardless of the location of the CSF leak. If the epidural blood patch fails, the blood patch procedure can be repeated at the lumbar level, or a blood patch can be directed at the exact site of the leak. Surgical repair of the CSF leak is safe and generally successful.

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There was only slight improvement in his headache with conservative treatment, including bed rest, daily hydration with 2000 mL of normal saline, and administration of a nonsteroidal anti-inflammatory drug for 3 days. Further CT myelography showed that there was a dural tear and extradural leakage of contrast through defect at D1 to D4 level and cause was found to be intradural osteophyte at D1-D2 level. (Fig 5).

CT guided autologous epidural blood patch at site of leakage was done in which 4ml of blood with 1ml of contrast was injected in epidural space between C7-T1. After one day of this procedure there was complete relief in his headache.

Discussion:
Spontaneous Intracranial Hypotension (SIH) was first described by Schaltenbrand in 1938 as a potential cause of postural headache, and 'spontaneous aliquorrhea' was the term coined by him. It is a rare condition presenting with cardinal postural headache and low cerebrospinal fluid (CSF) pressure. (1,2) The classical diagnostic triad of SIH includes postural headache, diffuse pachymeningeal gadolinium enhancement, and low CSF opening pressure, below 60 mmHg. (3)

This reduction in CSF pressure is thought to be caused due to a rupture of the arachnoid membrane resulting in the leakage of CSF into the subdural or epidural space. Spontaneous CSF leakage has been reported mostly as occurring at the cervicothoracic and thoracolumbar junction of the spine. (3) Our patient had the leak at upper thoracic level.

There are many causes of SIH, including meningeal diverticula, dural root sleeve tears, and Tarlov cysts. (4) The characteristic headache in SIH is similar to postlumbar puncture postural headache in that it is aggravated by sitting or standing and relieved by lying down. (4) Other associated symptoms, seen less commonly, include stiff neck, nausea and vomiting, diplopia, and cranial neuropathies, producing vertigo, tinnitus, photophobia, and changes in hearing. (4) Headache is mainly due to the traction on the pain-sensitive structures from the descent of brain. Compression or traction on cranial nerves or cervical roots is considered to be responsible for the neurological signs and symptoms. (3) Magnetic Resonance Imaging has revolutionized the understanding of SIH and has played a very important role in arriving at the proper diagnosis without having to resort to invasive procedures such as spinal puncture or intracranial
pressure monitoring. The five characteristic imaging features of SIH visible on MRI are:

1. Subdural fluid collection and presence of extrathecal CSF
2. Enhancement of the pachymeninges sparing of leptomeninges
3. Engorgement of venous structures
4. Pituitary hyperaemia and
5. Sagging of brain or downward displacement of brain.

In addition to these characteristic features, other finding may include: spinal meningeal diverticula, collapsed superior ophthalmic vein, reduction in the angle of vein of Galen and internal cerebral vein (the venous hinge sign). In our case, we found diffuse pachymeningeal enhancement, subdural venous collection, sagging of brain, and venous hinge sign. For detection of site of CSF leak Mokri et al have demonstrated in a study that the detection of CSF leaks with CT myelography was 67% as compared to 50% and 55% with spinal MR imaging and radionucleotide cisternography, respectively. CT myelography was done in our patients showed dural tear and CSF leaks.(6)

Treatment of SIH begins with conservative management, including bed rest, restoring the depleted CSF volume by intravenous fluid, intravenous or oral caffeine, theophylline. Patients who are refractory to the initial conservative management are candidates for EBP(epidural blood patching). A recent report suggests that early ‘blind’ epidural blood patching within one week of onset is effective; demonstrating complete cure in 77% of 30 patients (with or without typical MRI changes) after one (57%) or two (20%) blood patches. If treatment with EBP fails then direct surgical ligation of a leaking meningeal diverticulum, epidural packing using blood-soaked Gelfoam, muscle pledgets, or fibrin glue are primary techniques used to seal the site of CSF outlet.(3,4)

In the literature, only four cases of SIH secondary to osteophyte have been reported; only one case of spinal osteophyte originated SIH was treated successfully with EBP(8,9), while all other cases were managed by surgery after unsuccessful EBP treatment.(10,11) These cases are summarized in Table 1.

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<tr>
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<td>1</td>
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<td>EBP (subtotal improvement, patient refused further intervention)</td>
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Our patient had small osteophyte noticed at thoracic level during CT myelography which was the cause of dural tear and CSF leak, this patient was managed successfully with EBP. There was complete relief in his headache after one day of procedure.

References: