Case Report

Syndrome of spontaneous cerebrospinal fluid hypovolemia: Report of six cases

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Syndrome of spontaneous cerebrospinal fluid hypovolemia (SCH) is a rare cause of new onset headache. We report six cases of SCH presenting with new onset headache. All six cases were females. Acute onset orthostatic headache and neck pain were the chief characteristics of SCH in our cases. The MRI brain showed pachymeningeal gadolinium enhancement in all patients. Spinal extradural CSF collection was demonstrable on MRI in three cases. All cases improved with conservative therapy. High index of clinical suspicion and contrast enhanced MRI brain is the key to accurate diagnosis in the majority of cases.

Key words: Cerebrospinal fluid hypovolemia, orthostatic headache, spontaneous intracranial hypotension

Summary of six cases

Six cases of SCH were seen over a two-year period at our center. The clinical and laboratory features of all six cases are summarized in Table 1. All were females and had typical OH at the onset of the illness. In Case 1 and 4, headache character subsequently changed to persistent global headache. Other common symptoms included neck pain (5/6 cases), vertigo (3/6), nausea and vomiting (3/6); all symptoms worsened with upright posture.

Spontaneous cerebrospinal fluid hypovolemia (SCH) is increasingly recognized as an important yet often misdiagnosed cause of new onset headaches.[1] Classically, patients with SCH present with acute onset “orthostatic headache” (OH).[1,2] Though secondary OH following lumbar puncture is well known, “spontaneous” cerebrospinal fluid (CSF) hypovolemia is less often recognized.[1,2]

We report six cases of SCH seen at our center over a two-year period. To our knowledge this is the first reported series of SCH from India.

Case 1 and 3 had history of abnormal neck movements temporally related to the onset of headache.

Gadolinium enhanced MRI brain was performed in all cases prior to performing lumbar puncture. All cases had diffuse pachymeningeal enhancement [Figures 1a, 1b]. The FLAIR images showed hyperintense rim of the thickened pachymeninges in three cases [Figure 1c]. The MRI spine was performed in four cases; three cases had cervical extradural CSF. None had subdural hygromas, suboccipital CSF accumulations or any features of brain herniation. The CSF analysis was performed in three cases. Opening CSF pressure was low in all three cases. The CSF analysis results were available for two cases; both had normal sugar and mild elevation in protein. Minimal pleocytosis was noted in one case.

All patients were treated conservatively with bed rest, analgesics, oral theophylline and plenty of oral and intravenous fluids. All patients except Case 1 and 4 improved significantly within a week of the treatment. Case 1 and 4 had persistent mild symptoms lasting six weeks (Case 1) and three months (Case 4). Detailed description of Case 2 is given below.

A 35-year-old woman presented with acute onset severe bi-temporal headache of one-week duration. She had a five-year history of infrequent vascular headache. She was initially diagnosed to have an acute episode of migraine and was treated with analgesics. On further enquiry, she reported typical features of orthostatic headache. Headache was associated with nausea and vomiting which was worse on upright posture. She had a history of neck pain for which she was doing unsupervised neck exercises. On examination, she had a marfanoid habitus with long slender fingers and hyper-mobile joints. Neurological examination was unremarkable. The MRI with contrast revealed mild patchy pachymeningeal enhancement; extradural CSF collection in the cervical spine was noted [Figure 2]. She improved with conservative measures. She developed orthostatic vertigo during her hospital stay and was...
Figure 2: Cervical spine MRI shows extradural CSF accumulation. Extradural CSF appears hyperintense compared to the intradural CSF.

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Figure 1: A. Gadolinium enhanced T1-weighted axial images show diffuse thick pachymeningeal enhancement. B. Gadolinium enhanced T1-weighted coronal images show thick pachymeningeal enhancement. C. FLAIR images show hyperintense rim corresponding to the engorged pachymeninges.

In this report, we describe six cases of new onset headache due to spontaneous cerebrospinal fluid hypovolemia seen over a two-year period at our center. Paucity of reports of SCH from India may suggest that this disorder may be under-recognized. Syndrome of SCH can be readily recognized in its classical presentation as “orthostatic headache”. Orthostatic headache is defined by the International Headache Society, as a headache that occurs in less than 15 min after assuming upright posture and disappears or improves in less than 30 min after resuming the recumbent position.[3] Chung et al. considered the presence of at least two of the following three criteria to diagnose SCH - OH, CSF pressure less than 60 mm of CSF and diffuse pachymeningeal enhancement. The CSF pressure was low in all three cases tested.

Though OH is seen in the majority, other types of headache like non-orthostatic chronic headaches, acute thunderclap headache, cervical or inter-scapular pain can also occur.[2] The typical orthostatic pattern of headache often disappears with time in patients with long-standing symptoms as seen in Case 1 and Case 4. Neck pain and vertigo were the other common symptoms seen in our cases. Rarely, cranial nerve palsies, radicular pain, low backaches, parkinsonism, ataxia, bulbar palsy and galactorrhea have been described.[2]

The CSF analysis is not mandatory in a classical case with typical presentation and positive MRI findings. Though low CSF pressure is a characteristic feature of SCH, Chung et al. reported normal CSF pressure in 18% cases in their series of 30 patients.[4] Similar findings were observed by other authors and this led them to postulate CSF “hypovolemia” rather than “hypotension” as the chief cause of symptoms.[4-6] Cerebrospinal fluid volume depletion results in descent of brain which leads to traction and distortion of pain-sensitive anchoring structures of the brain. This descent will be accentuated in the upright position resulting in orthostatic symptoms. Traction of cranial nerves, brainstem and diencephalon may be responsible for some of the manifestations. Apart from low CSF opening pressure, mild pleocytosis and moderately elevated protein can occur; CSF sugar is always normal.[2,5,6] In our series, lumbar CSF pressure was measured in only three cases, as we believed that additional dural puncture might aggravate the symptoms of SCH. Lumbar puncture should be avoided in patients with tonsillar or brainstem herniation.

Pachymeningeal gadolinium enhancement, subdural fluid collections, prominence of the cerebral venous sinuses and pituitary hyperemia may be seen on an MRI.[2,4,6,7] Obliteration of subarachnoid cisterns, crowding of the posterior fossa and tonsillar herniation treated with labyrinthine sedatives. She was discharged after a week. At one-year follow-up, she remained symptom-free.

Discussion

In this report, we describe six cases of new onset headache due to spontaneous cerebrospinal fluid hypovolemia seen over a two-year period at our center. Paucity of reports of SCH from India may suggest that this disorder may be under-recognized. Syndrome of SCH can be readily recognized in its classical presentation as “orthostatic headache”. Orthostatic headache is defined by the International Headache Society, as a headache that occurs in less than 15 min after assuming upright posture and disappears or improves in less than 30 min after resuming the recumbent position.[3] Chung et al. considered the presence of at least two of the following three criteria to diagnose SCH - OH, CSF pressure less than 60 mm of CSF and diffuse pachymeningeal enhancement on MRI.[4] All cases in our series had OH and diffuse pachymeningeal enhancement. The CSF pressure was low in all three cases tested.

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has been described. The descent of cerebellar tonsils may mimic Type 1 Chiari malformation. Spinal pachymeningeal enhancement, engorgement of spinal epidural venous plexus and spinal extradural CSF collection may be noticed. Three patients in our series had spinal extradural collection in the cervical region. Pachymeningeal enhancement may be absent in some patients. Lack of pachymeningeal enhancement may imply inability to compensate for CSF volume loss and hence indicate less favorable prognosis.

Bed rest with good hydration orally or intravenously is the usual initial treatment for SCH. Addition of caffeine and steroids may be helpful. Dural defects heal spontaneously over days to weeks with conservative treatment. All our patients improved with these measures. Case 1 and 4 probably warranted a more aggressive approach in the initial phase as they took nearly three months to recover. In refractory cases, other treatment options include epidural infusion of saline or dextran and epidural blood patch or fibrin glue injection. Epidural blood patch (EBP), which involves injection of 20 ml of autologous blood into the lumbar epidural space, is a widely accepted treatment. All our patients improved with these measures.

In conclusion, we report six cases of new onset headache due to spontaneous cerebrospinal fluid hypovolemia. Orthostatic nature of headache, associated neck pain and vertigo should alert to the possibility of SCH. Gadolinium enhanced MRI brain and CSF pressure estimation often confirms the diagnosis.

References


Table 1: Clinical features, CSF findings, MRI features and outcome of six cases of SCH

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age/ gender</th>
<th>Symptoms</th>
<th>Potential predisposing/ precipitating factors</th>
<th>CSF pressure (mm of CSF)</th>
<th>CSF Protein/Sugar</th>
<th>CSF cells</th>
<th>MRI brain</th>
<th>MRI spine</th>
<th>Time to recovery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>35/F</td>
<td>OHA, Later PHA, NP, Vo, Ve, T</td>
<td>Startling arousal from a nightmare</td>
<td>ND</td>
<td>-</td>
<td>-</td>
<td>DPME</td>
<td>ED CSF</td>
<td>6 weeks</td>
</tr>
<tr>
<td>2</td>
<td>35/F</td>
<td>OHA, NP, Vo, Ve</td>
<td>Manframoid habitus, unsupervised neck exercises</td>
<td>ND</td>
<td>-</td>
<td>-</td>
<td>DPME</td>
<td>ED CSF</td>
<td>1 week</td>
</tr>
<tr>
<td>3</td>
<td>60/F</td>
<td>OHA, NP</td>
<td>Radiation for carcinoma rectum</td>
<td>45</td>
<td>56/100</td>
<td>7 cells; 90% lymphocytes</td>
<td>DPME</td>
<td>ND</td>
<td>2 weeks; asymptomatic at 6 months follow-up</td>
</tr>
<tr>
<td>4</td>
<td>35/F</td>
<td>OHA, Later PHA, Ve</td>
<td>None</td>
<td>Very low</td>
<td>NA</td>
<td>NA</td>
<td>DPME</td>
<td>ND</td>
<td>1 week</td>
</tr>
<tr>
<td>5</td>
<td>31/F</td>
<td>OHA, NP, Vo</td>
<td>Hypermobility of joints</td>
<td>ND</td>
<td>-</td>
<td>-</td>
<td>DPME</td>
<td>ED CSF</td>
<td>1 week</td>
</tr>
<tr>
<td>6</td>
<td>29/F</td>
<td>OHA, NP</td>
<td>None</td>
<td>60</td>
<td>51/56</td>
<td>2 cells; all lymphocytes</td>
<td>DPME</td>
<td>Normal</td>
<td>1 week</td>
</tr>
</tbody>
</table>

OHA—Orthostatic headache; PHA—Persistent headache; T—Tinnitus; Vo—Vomiting; NP—Neck pain; Ve—Vertigo; DPME—Diffuse pachymeningeal enhancement; ND—Not done; ED CSF—Extradural CSF collection; NA—Not available.