

Spontaneous Spinal CSF Leak: A Case Report and Review of Literature

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Abstract: *Spontaneous Intracranial Hypotension (SIH) is formed as a result of the leakage of cerebrospinal fluid (CSF) into the extradural space. The most common symptom caused by SIH is headache associated with changes in body position. Imaging, especially magnetic resonance imaging (MRI) of the head and spine with contrast, is the most important examination method in the diagnosis of SIH. In the case of no improvement after symptomatic treatment, surgery is very effective, especially in the case of finding the site of CSF leakage within the spinal canal. The patient described below had presented with features of intracranial hypotension and so had undergone MRI imaging of brain and spine which revealed a CSF leak at C2-C3 region into the extradural space. Hence the patient was posted for surgery in view of the severity of the symptoms and post CSF defect repair, the patient improved symptomatically.*

Keywords: Spinal CSF leak, Cerebrospinal fluid, intracranial hypotension, epidural blood patch

1. Introduction

Intracranial hypotension (IH) is a clinical syndrome which is characterized by absolute or relative hypovolemia of the cerebrospinal fluid (CSF) resulting in spectrum of neurological symptoms. An increasing number of publications on the subject in recent years suggests that IH is no longer a rare syndrome as described before. Although causes of IH are diverse, they may be classified as 1) spontaneous (primary), and 2) secondary. Secondary causes can be divided further into: 2-i) traumatic, mostly iatrogenic; 2-ii) degenerative spine disorders; 2-iii) truly hypovolemic, mostly from systemic dehydration; and 2-iv) over drainage of the CSF by neurosurgical procedures.

This syndrome is usually suspected in a patient with postural headache that occurs after head or spine trauma. However, such headaches can also occur spontaneously without an obvious etiology. Other symptoms of this disease include nausea with/without vomiting, blurring of vision or double vision, tinnitus, vertigo, and photophobia, hearing loss, facial weakness with or without pain, difficulty in speaking or swallowing, and strokelike symptoms. The combination of postural headache, low CSF pressure on diagnostic lumbar puncture, and meningeal enhancement on MRI without history of dural puncture should lead one to suspect this diagnosis.

Many patients with IH present without known predisposing factors or history of a major trauma, and this condition has been called "spontaneous". Most patients with spontaneous IH have a CSF leak in the spinal dural sac. A dural sleeve surrounding a spinal nerve root is the predilection site of a CSF leak, and meningeal diverticula, Tarlov's cysts, or "nude" nerve roots are occasionally found. Mechanical stress has an important role in its pathogenesis, by acting on the point of focal weakness in the dural sac. The location of spontaneous spinal CSF leak is more common at cervical and thoracic regions and rare in the lumbar region.

It is not uncommon for patients with spontaneous IH to recall having sustained a trivial trauma. Connective tissue disorders may be important risk factors for spontaneous IH.

Imaging studies often reveal multiple meningeal diverticula, indicating increased susceptibility of patients with connective tissue disorders to spontaneous IH.

It is rare for patients with IH to be seen by spine surgeons primarily: those patients almost invariably present with complaints of orthostatic headache, and neurologists. MRI of the brain with contrast is the imaging study of choice for patients suspected of spontaneous IH with features of intracranial hypotension followed by spine imaging to detect site of CSF leak. Low CSF pressure is the hallmark of IH, as suggested by its name. These patients are usually managed conservatively. Surgery is reserved for those not responding to conservative line of management.

Here, we report a case of intracranial hypotension with CSF leak in cervical region who responded well to surgery after no response to initial conservative line of management.

2. Case Report

A 43 year old male, presented with history of dull aching, headache since 2 months which worsened on bending forward, coughing and sneezing, interfering with daily activities and worsened since last 10 days. It was not associated with vomiting, cranial nerve deficit, trauma, focal neurological deficits. Patient did not have any comorbidities. Past history revealed past history of tuberculosis 22 years ago and taken anti tubercular drugs for 3 months and defaulted. There were no prior spinal or brain surgeries. There was no significant family history. On physical examination, vitals were stable. CNS examination revealed no cranial or focal neurological deficits.

He was initially managed conservatively with analgesics and IV fluids, however had no symptomatic relief. Hence Magnetic resonance imaging of brain was done which revealed intracranial hypotension features in the form of sagging brainstem with decreased mamillopontine distance, downward dropping of splenium of corpus callosum and cerebellar tonsillar herniation (fig 1). MR myelogram of whole spine was done in view of suspected CSF leak. It revealed 6 x 5 mm dural defect involving posterolateral

spinal dura matter at C2-C3 level with CSF leak from subarachnoid space into the adjacent right posterior para spinal region (Fig 2). His hemoglobin, renal function, liver function tests were within normal limits.

Patient was admitted for surgery for repairing CSF leak in view of severe symptoms. Patient was induced and positioned in sitting position. Incision was taken frominion to C5. Following laminectomy from C2 to C4, there were adhesions seen in right epidurals space in anterolateral area with CSF leaking through. On releasing adhesion, CSF leak was found at C2-C3 level, which was concordant to imaging (Fig 3). CSF leak was confirmed on Valsalva maneuver. Generous fat graft was used to plug the CSF defect. There was no CSF leak on repeat Valsalva maneuver post repair. Hemostasis was achieved. The wound was closed back in layers. Post operatively, patient was extubated immediately and had no post op complications. 2 months post operatively, headache had significantly decreased and did not require analgesics.

3. Discussion

Presentation

IH is usually suspected in a patient with postural headache that occurs after head or spine trauma. Rarely, it may be spontaneous as well, as in our patient who denied history of trauma and did not have connective tissue disorder.

In a study done in Minnesota, there was a clear female preponderance for the syndrome: a male to female ratio was between 1:1.75-4¹. The mean age at the time of presentation was 38-42 years in both genders which is similar to our case (1).

Imaging studies

Our patient underwent MRI brain in view of suspected IH followed by spinal imaging. In literature, use of imaging studies of the spine has become increasingly common in patients with spontaneous IH, as it became known that a dural tear or defect in the spinal dural sac is the most common pathology which will aid in therapeutic decision. Computed tomography (CT) myelography, spinal MRI, and radionuclide cisternography are the imaging studies used in for spinal imaging. CT myelography is considered to be the most reliable imaging modality for localizing the actual site or spinal level of a CSF leak. A "positive" CT myelogram does not always translate into the actual site of a CSF leak, however. Because of a lack of the "gold standard" imaging study, the sensitivity or specificity of each imaging study cannot be evaluated.

In others, magnetic resonance myelography has been able to identify the site of a CSF leak. Radionuclide cisternography seems to have a relatively high "sensitivity" with its panoramic view of the entire spine, and may be useful in certain subset of patients. Because of its lower spatial resolution capacity compared with CT myelography or spinal MRI, however, it is rarely helpful in identifying the actual site of a CSF leak.

CSF studies

80-90% of patients with spontaneous IH have an opening CSF pressure of less than 60 mm of water (normal, 65-195 mm of water)³⁻⁵. The CSF studies may even be contraindicated in massive subdural fluid collection in the cranium, because of the risk of an acute tonsillar herniation by LP. The great majority of patients with spontaneous IH have a moderately elevated level of CSF protein and pleocytosis with lymphocyte dominancy, a finding that may be confused with viral meningitis.

Treatment

Initially conservative management, including bed rest and administration of intravenous fluids is given which partially restores the depleted CSF volume of patients. Pharmacological therapy, including intravenous or oral caffeine, theophylline, and steroids, is also a part of the conservative management.

Patients who are refractory to the initial conservative management usually receive Epidural blood patch (EBP). The initial tamponade effect of a blood clot over a dural tear or defect and subsequent scar formation is the suggested mechanism of action of EBP. The success rate of EBP was 70% to 90% in most studies^{2,6}. Factors that may affect the success rate include the timing of the treatment, the severity of patient's symptoms, and the amount of the autologous blood injected.

Surgical treatment:

Our patient underwent surgery in view of debilitating headache. Surgical treatment is usually reserved for patients who fail multiple attempts of EBP. The rate of patients with spontaneous IH who required open surgery ranged from 0%-72%^{6,7}. The great variability may reflect the difference in treatment strategy of each institution as well as in patients' demographics and referral pattern. Patients in whom preoperative imaging studies reveal the presence of underlying dural/arachnoidal lesions, particularly meningeal diverticula, are treated by ligation of the diverticula. In patients in whom the spinal level of a CSF leak is determined but no underlying lesion is visualized by the imaging studies, exploratory surgery, consisting of a laminectomy (with or without foraminotomy) and inspection of the spinal dural sleeves, is performed. If the site of a CSF leak could not be identified intraoperatively, the patients underwent subsequent packing of the epidural space with muscle fragments, fibrin glues, and Gelfoam. The patient in the current report had adhesions in the right epidural space where the CSF leak was noted. He had not undergone EBP or any other procedures and hence the cause for the adhesions at the CSF leak site which looks like a sequelae of previous inflammatory condition has not been determined.

Our patient had good symptomatic response post-surgery. In literature, despite the high success rate of surgical treatment, recurrence of a CSF leak are not uncommon. In a recent study, 5 of 18 (28%) patients with spontaneous IH treated by surgery developed a recurrent CSF leak⁸. Treatment of patients in whom the imaging studies fail to identify the spinal level of a CSF leak or those with multiple CSF leaks is most challenging. Repeated attempts of EBP rather than an exploratory surgery may be indicated for such patients.

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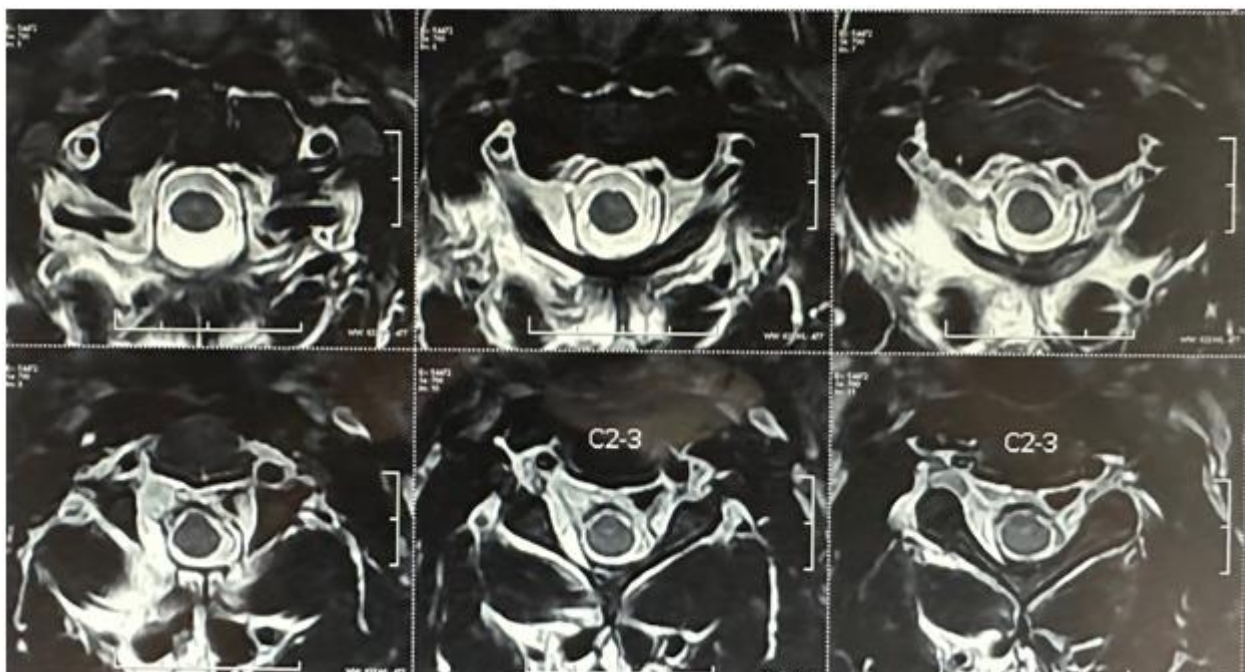


Figure 1: MRI cervical spine showing CSF leak at C2-C3 level into extrusion into right posterior paraspinous region.



Figure 2: MRI myelography whole spine (plain) showing C2-C3 level CSF leak.

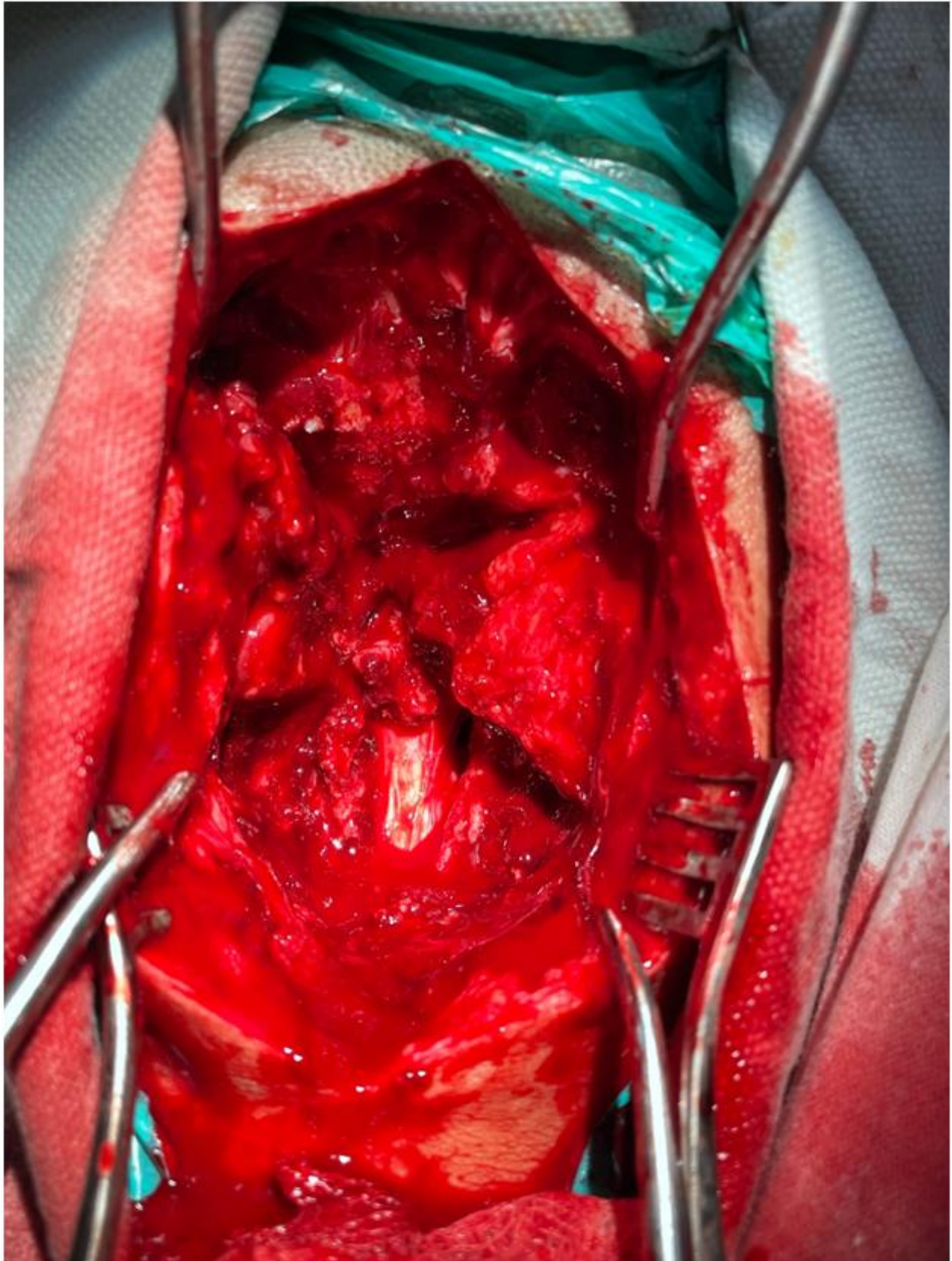


Figure 3: Intraoperative image showing dural defect at right anterolateral region.