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Case Report

A Case Report of Low Cerebrospinal Fluid Pressure Headache Due to Cerebrospinal Fluid Leak

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Abstract

We, here, present a case of 24-year-old female, who was admitted to our hospital with history of headache off and on since last 1 month that was aggravated by rising from bed in the morning and on standing posture. With this scenario, she was started nonsteroidal anti-inflammatory drugs and antimigraine regimen. Since there was no relief, she was investigated and found to have low cerebrospinal fluid pressure headache. Who improved on treatment with epidural blood patch.

Key words: Cerebrospinal fluid, Computerized tomography, Intracranial hypotension, Nonsteroidal anti-inflammatory drugs, Spontaneous intracranial hypotension

INTRODUCTION

The most common cause of intracranial hypotension (ICH), or low cerebrospinal fluid (CSF) pressure in the brain, is CSF leak. Low CSF pressure headache is caused by an internal spinal fluid leak and may range from obvious and disabling to subtle and nagging.

The syndrome of ICH is a single pathophysiological entity of diverse origin. Usually, it is characterized by an orthostatic headache, that is, one that occurs or worsens with upright posture, although patients with chronic headaches or even no headache have been described.[1]

The nature and location of the headache vary greatly from patient to patient, but consistently the pain is exacerbated by laughing, coughing, jugular venous compression, and Valsalva maneuver, and is resistant to treatment with analgesic agents.[2,3]

In addition to headache, patients may experience nausea, vomiting, anorexia, neck pain, dizziness, horizontal diplopia, changes in hearing, galactorrhea, facial numbness or weakness, or radicular symptoms involving the upper limb, all of which are orthostatic in nature.[4] The ICH generally is considered to be a benign condition and most cases resolve with conservative management.

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CASE REPORT

A 24-year-old female patient admitted with c/o recurrent type headache since 1 month, nausea, vomiting, and neck pain intermittently since last 1 month.

The patient was alright before 1 month to start with then later started with continuous type of headache, located to occipital, frontal and temporal region, gradually progressive in nature, relieved on lying down and after taking tea/coffee, and aggravated by sitting and standing position.

Later, she also started c/o intermittent type nausea, vomiting, and neck pain. For all these complaints she use to prescribed nonsteroidal anti-inflammatory drugs (NSAIDS) and migraine medications by local practitioners and over counter medication.

Initially, she use to get relief with same but had many repeated episodes and now admitted for same because headache becomes unbearable to her.

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On examination, BP/PR/TEMP found to be normal, B/L pupils equally reacting to light. There was no postural drop in BP. There was no neurodeficit and neck rigidity. Rest systemic examination was found to be normal.

On laboratory examination, she was having HB=11.9, TLC=5200, PLATELETS = 2.39 lakh, Sr. ctreatinine = 0.81, PT/INR = 12.5/1.03, HIV/HBS Ag = Negative.

Magnetic resonance imaging (MRI) cervicodorsal spine shows mild cerebellar tonsillar herniation through foramen magnum, tonsil appears pear shaped.

CSF intensity signal in anterior epidural region extending from upper border of D1 vertebra to midlevel of D3 vertebral body s/o possibility of CSF leak in anterior epidural space.

For confirmation computerized tomography (CT) myelogram done, which was s/o extravasation of contrast in above mentioned area so our diagnosis was confirmed to have low CSF pressure headache secondary to CSF leak.

Initially, patient was treated with NSAIDS, head low position, coffee, and bed rest. Later on planned for epidural blood patch (EBP), after giving EBP, patient was significantly improved symptomatically.

No second setting required. Patient came to follow-up asymptomatic after 15 days.

DISCUSSION

In 1939,[5] Georg Schaltenbrand, a German neurologist, using the term "aliquorrhea" described spontaneous occurrence of a syndrome of orthostatic headache and a few other symptoms associated with low CSF opening pressure (OP).[6] This later came to be known as spontaneous ICH (SIH).[7,8] Modern neuroimaging has revolutionized our understanding of this entity. The original theory of Schaltenbrand that the disorder was due to decreased CSF production has never been substantiated. It is now recognized that almost all cases of SIH result from spontaneous CSF leaks. The overwhelming majority of these spontaneous leaks occur at the spinal level and only rarely from the skull base.[9] Although the triad of orthostatic headaches, low CSF pressures, and diffuse pachymeningeal enhancement is the classic hallmark of this disorder, the variability is indeed substantial.[10] The core factor in pathogenesis, and the independent variable is loss of CSF volume; while CSF pressures, clinical manifestations, and MRI abnormalities are variables dependent on the loss of CSF volume. The term "SIH" no longer appears broad enough to embrace all these variations. Therefore, terms such as "CSF hypovolemia" or "CSF volume depletion" as well as "spontaneous CSF leaks" have appeared in the literature and have been used interchangeably.[8,11,12]

Etiology

The etiologies of CSF volume depletion are listed in Table 1.

Clinical Features and Related Mechanisms

Headaches

Headache is the most common clinical manifestation. This is often orthostatic (present when upright and relieved in recumbency). The latency of headache onset or resolution from change in posture classically should be only a few minutes, but in reality, the variability is substantial, and with chronicity, this latency may become even further prolonged. The headache may be throbbing, but more commonly it is not, and is described as a pressure sensation of variable intensity, sometimes quite intense. It is typically, although not invariably, bilateral.[13] It may be bifrontal, occipital, bifrontal-occipital, or holocephalic. Occasionally, it may start as a focal or unilateral headache and evolve into a holocephalic headache if the patient continues to be up and about. The headaches are often aggravated by Valsalva-type maneuvers and occasionally are even triggered by such maneuvers. At this point, it should be emphasized that not all orthostatic headaches are due to ICH or CSF leaks (this will be discussed later in this communication), and not all headaches in CSF leaks are orthostatic.

Sinking of the brain and the resultant traction on pain-sensitive suspending structures of the brain is thought to be the main cause of the orthostatic headaches in CSF

Table 1: The etiologies of CSF volume depletion are listed

True hypovolemic state (reduced total body water)
Traumatic CSF leaks
Definite trauma (MVAs, sports injuries, etc.)
Thecal holes and rents from LPs and epidural catheterizations
Spinal and cranial surgeries including skull base and some sinus surgeries
Proximal brachial plexus avulsion injuries, nerve root avulsions
CSF shunt overdrainage
Spontaneous CSF leaks
Undetermined cause
Preexisting weakness of the dural sac, surgical anatomical observations
Meningeal diverticula
Disorders of connective tissue matrix
Marfan syndrome, Marfanoid features
Joint hypermobility
Retinal detachment at young age
Abnormalities of elastin and fibrillin in cultured dermal fibroblasts
Trivial trauma in the setting of preexisting dural weakness
Spondylotic spurs, herniated discs

leaks. Dilatation of the cerebral veins and venous sinuses may also be a participatory mechanism and in some situations perhaps even the dominant mechanism.

Some patients with stubborn orthostatic headaches, in recumbency, may report an earlier and a more effective relief in certain positions or postures, such as Trendelenburg position,[14] or by lying prone with the head dropped somewhat at the edge of the bed. It has been demonstrated that CSF OP is significantly higher in prone than in lateral decubitus position.[15]

Clinical Features Other Than Headaches

- Spinal pain (neck, interscapular, and less commonly lower back), sometimes orthostatic
- Nausea with or without emesis (often orthostatic)
- Diplopia
- Cochleovestibular manifestations (tinnitus, change in hearing, and dizziness)
- Photophobia, visual blurring
- Upper limb numbness, paresthesias
- Gait unsteadiness[16]
- Facial numb feeling
- Change in level of consciousness (i.e., encephalopathy,[17] lethargy, stupor,[18] coma[19])
- Personality change, memory decline, apathy, frontotemporal dementia-like picture[20,21]
- Meniere-like syndrome[22]
- Upper limb radiculopathy[23]
- Trouble with bowel or bladder control.[24]

Diagnosis

LP and CSF analysis

In search of inflammatory, infectious or neoplastic disease, CSF OP is low in the large majority; but in a significant minority, perhaps in about one fourth of patients, it is within normal limits. The OP is uncommonly atmospheric and rarely is even negative (Table 2).

Radioisotope cisternography

Indium-111 is the radioisotope of choice. It is introduced intrathecally (IT) via an LP and its dynamics are followed by sequential scanning at various intervals of up to 24 or even 48 h. Normally, after 24 h, though often earlier, ample radioactivity can be detected over the cerebral convexities while no activity outside the dural sac is noted, unless there has been inadvertent injection of part of the radioisotope extradurally or if some of the IT-injected radioisotope has extravasated through the dural puncture site or CSF leaks.

MRI abnormalities of head and spine and their related mechanisms

Head and spine MRI abnormalities of CSF leaks and CSF hypovolemia are listed.

Table 2: CSF analysis

Color	Clear/xanthochromic
Protein concentration	Normal/high
Leukocyte count	Normal/lymphocytic pleocytosis
Erythrocyte count	Normal/high
	Glucose concentration, cytology, and bacteriology should all be normal

Head MRI abnormalities in CSF leaks

- The most common and most reliable head MRI abnormality in spontaneous CSF leaks is cephalad opening of aqueduct of sylvius as seen in midline sagittal views
- Diffuse pachymeningeal enhancement: Uninterrupted, nonnodular, can be thick or thin, no leptomeningeal abnormality
- Descent ("sagging" or "sinking") of the brain
  1. Descent of cerebellar tonsils at or below the foramen magnum (may mimic type I Chiari)[25]
  2. Descent of the brainstem and mesencephalon, occasionally without descent of cerebellar tonsils to or below foramen magnum
  3. Increase in anteroposterior diameter brainstem resulting from distortion of the brainstem
  4. Descent of iter‡ below the incisural line 10[13]
  5. Obliteration of prepontine or perichiasmatic cisterns
  6. Crowding of the posterior fossa
  7. Flattening of the optic chiasm
  8. Flattening of the anterior pons
- Subdural fluid collections, typically hygromas, infrequently hematomas
- Enlargement of the pituitary (may mimic pituitary tumor or hyperplasia)[26]
- Engorged cerebral venous sinuses
- Decrease in size of the ventricles ("ventricular collapse").

Spine MRI abnormalities in spontaneous CSF leaks

- May or may not be the actual site of leak, even when the diverticulum is large, although larger diverticula may be more prone to be the site of the leak
  1. Extra-arachnoid fluid collections (often extending along several spinal levels)[27-29]
  2. Extradural extravasation of fluid (extending to paraspinal soft tissues)
     a. May identify the level of the leak (i.e., cervical, thoracic or lumbar), not uncommon
     b. May identify the actual site of the leak, uncommon[30]
  3. Meningeal diverticula, a single or multiple, various sizes, any level of spine
  4. Spinal dural enhancement[31]
- Engorgement of spinal epidural venous plexus.

CT myelography (CTM)

CTM thus far is the most accurate study for demonstrating the exact site of the spinal CSF leakage.[1] Similar to radioisotope cisternography, it also provides an opportunity to measure the CSF OP at the time of dural puncture. In addition to its accuracy in revealing the site of the leak, it can show meningeal diverticula, dilated nerve root sleeves, extra-arachnoid fluid collections, and extradural egress of contrast into the paraspinal tissues.

Treatment

For spontaneous spinal CSF leak, a variety of treatment modalities have been tried as follows:

1. Conservative measures
    a. Bed rest (those with substantial orthostatic headaches remain reclined much of the time anyway)
    b. Coffee
    c. Hydration (actually overhydration since most patients are not dehydrated)
    d. Time
2. Medications
    a. Analgesics
    b. Caffeine
    c. Theophylline
    d. Corticosteroids
3. Abdominal binder
4. Epidural injections of:
    a. Homologous blood ("EBP")
        i. Targeted
        ii. Distant at lumbar level or bilevel, "blind EBP"
    b. Fibrin glue
    c. Fibrin glue and blood
5. Surgical repair or the leak
6. Other measures in special situations
    a. Intrathecal fluid injection (volume replacement)
    b. Epidural saline infusion
    c. Intravenous saline infusions
    d. Epidural infusion of dextran.

EBP is now recognized as the treatment of choice in those patients who have not responded to the initial trial of conservative management.[32] EBP works via two separate mechanisms: (1) The immediate effect related to volume replacement by compression of the dural sac (decreasing the volume of the container); (2) sealing of the dural defect, which may be delayed from the first one. Therefore, it is not uncommon to note an initial quick response in connection with the first mechanism, recurrence of symptoms within merely a day or two, and then, a gradual and often variable improvement after several days. Variability is, however, substantial. The efficacy of each EBP is about 30%.[1] A previous EBP failure should not be taken as a signal that a subsequent EBP will fail. Indeed, many patients may require more than one EBP and some have required several. At times, a cumulative effect from multiple EBPs may be noted. Similarly, a previous success will not guarantee success of a future EBP. The site of the leak in spontaneous CSF leaks is mostly at levels above the lumbar spine where most of the epidural block patches are placed. Therefore, the odds are that many of these will be nontargeted and distant from the site of the leak. (3) The dural defect in spontaneous CSF leaks, as opposed to post-lumbar puncture leak, often is not a simple hole or rent instead it is frequently a preexisting zone of attenuated dura with or without associated diverticula where an unsupported arachnoid may finally give way and ooze CSF from one or more sites. Surgical anatomical observations[33] have clearly identified such defects in many patients who have ended up with surgery.

Surgery in well-thought-of cases is effective and can be tried when less invasive measures (such as EBP) fail.

Furthermore, some patients may have CSF leaks from more than one site and at different levels. It is strongly emphasized that thorough pre-operative neurodiagnostic studies should be conducted to identify the actual site of the leak before surgery is undertaken.

The fundamental purpose of the surgery in the treatment of CSF leaks is to stop the leak.

Complications of Spontaneous CSF Leaks

1. Subdural hematoma
2. Rebound intracranial hypertension
3. Cerebral venous sinus thrombosis
4. Bibrachial amyotrophy
5. Superficial siderosis
6. Natural history and outcome
7. Recurrence of CSF leaks.

CONCLUSION

A 24-year-old case of spontaneous CSF leak treated with EBP significantly improved symptomatically and clinically.

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