Pakistan Journal of Neurological Sciences (PJNS)

Volume 9 | Issue 2 Article 7

4-2014

Spontaneous intracranial hypotension

Mehwish Sayed
Liaquat National Hospital, Karachi, Pakistan

Ahmed Asif
Liaquat National Hospital, Karachi, Pakistan

Bushra Reha
Liaquat National Hospital, Karachi, Pakistan.

Amanullah Khan
Liaquat National Hospital, Karachi, Pakistan.

SPONTANEOUS INTRACRANIAL HYPOTENSION

Mehwish Sayed[1], Ahmed Asif[2], Bushra Reha[3], Amanullah Khan[4]
[1] *Resident Department of Medicine, Liaquat National Hospital*
[2] *FCPS department of Neurology, Liaquat National Hospital*
[3] *FCPS Department of Radiology, Liaquat National Hospital*
[4] *Registrar Department of Radiology, Liaquat National Hospital*

Correspondence to: *Dr. MehwishSayed, Resident Medicine 4th year. Address: D-201 Saima Pride apartment, Main Rashid Minhas Road, Gulshan Block 10 A. Mob: 03343091503. Tel: 02135418974. Email:efficient_doctor@hotmail.com*
Date of Submission: *March 07, 2014,* ***Date of Revision:*** *May 15, 2014,* ***Date of Acceptance:*** *June 1, 2014*

ABSTRACT

Spontaneous intracranial hypotension (SIH) is uncommon and usually undiagnosed entity which present most commonly with postural headache. The diagnosis is difficult because of the nonspecific symptoms and lack of awareness about the disease .It is important to recognize SIH as if the disease goes undiagnosed it can cause a significant delay in initiation of the treatment and exposing patient to the risks associated with the treatment for the disorders mimicking intracranial hypotension . We describe a patient with SIH and outline the important clinical and radiological features of this syndrome. The aim of presenting this case report is to emphasize the need for being vigilant for this rare disorder which should be considered in the differential diagnosis of recurrent posture related headache.

Keywords: Postural Headache, Spontaneous Intracranial Hypotension

INTRODUCTION

Spontaneous intracranial hypotension or syndrome of spontaneous cerebrospinal fluid hypovolemia is a rare Syndrome. A condition being more commonly diagnosed in last two decades. Careful history taking and a high level of suspicion are essential to diagnosing this syndrome. Imaging studies, especially MRI are helpful in confirming the diagnosis and ruling out any other conditions. This condition is usually treatable with simple measures, though failure to recognize it, may subject the patient to unnecessary procedures, workup, and treatments. It is characterized by orthostatic headache (OH), low CSF pressure and DPME (diffuse pachymeningeal enhancement) on MRI in the absence of head trauma or lumbar puncture. It is defined as headache that occurs within 15 minutes of an upright position and is relieved within 30 minutes of recumbence.[1] The estimated annual incidence of SIH is 5/100,000. Peak incidence of SIH is at around the age 40 with male to female ratio of 1:2.[2, 3] To familiarize physicians with SIH, we present the following case.

CASE REPORT

A 46 year old previously healthy woman presented to a tertiary care hospital with one and a half month history of headache and vertigo which was associated with vomiting for the last two weeks. The pain began abruptly and was holocephalic, it used to be generally worse in the morning .She described that it partially relieved by analgesics but definitely settled after assuming recumbent position ,only to return after few minutes of assuming an upright position or movement of head. She felt her head spinning along with headache. Lately her symptoms became associated with nausea and vomiting 2 to 3 episodes a daythat was non projectile,occurred mostly in morning and relieved by lying straight. There was no history of skin rash, trauma, fever, and blurred or doubled vision, loss of consciousness or seizure activity. Prior to presenting there she had gone through her primary care physician and has remained admitted to another hospital where after routine workup no cause of her symptoms was found and had no significant improvement.Physical examination revealed an ill looking pale lady who was oriented. She was having fine tremors of her hands. Blood pressure was normal with no orthostatic hypotension. She was afebrile with normal respiratory rate and heart rate of around 100 beats per minute. General medical examination was within normal limits except for tachycardia and fine tremors. Neurological examination revealed normal cranial nerves without papiloedema. Her vision was intact and there was no nystagmus or nuchal rigidity. Strength, sensation and coordination were normal. Deep tendon reflexes were also normal, and planter responses were flexor bilaterally. Peripheral blood count was consistent with microcytic hypochromic anemia with the hemoglobin value of

9gm/dl consistent with iron deficiency anemia (MCV of 58 FL, M of 17.50pg and MCHC of 29.8 g/dl) and normal white cell and platelet count.Erythrocytes sedimentation rate was normal. Serum biochemistry revealed normal renal function and electrolytes but deranged thyroid function test that was consistent with hyperthyroidism (TSH of 0.01μiu/ml (NR: 0.27μiu/ml -4.2μiu/ml) FT4 was 4.0ug/dL(0.9ug/dL – 1.7ug/dL). Her liver function test showed billirubin of 0.34mg/dL, raised alanine aminotransferases (ALT) 68u/L (NR: up to 35) and alkaline phosphatase was 68u/l (<104). Viral serology was done for screening of hepatitis B and C which were negative and abdominal ultrasound showed hepatic fatty infiltration. Magnetic resonance imaging of brain with gadoliniumshowed minimal subdural effusion in bilateral frontoparietal region (Fig 1). There was circumferential thickening and enhancement of Dura involving cerebral and cerebellar hemispheres along with falx and tentorium cerebelli (Fig 2). There was slight descent of cerebellar tonsils through foramen magnum (Fig 3). Lumber puncture was not done as the diagnosis was evident by the brain imaging and clinical correlation. She was given anti thyroid drug i.e. Neomercazole along with metoprolol. Nonsteroidalanti inflammatorydrugs, antiemetic and labyrinthine sedatives were also given along with adequate intravenous hydration with normal saline and advised complete bed rest. Her condition improved over the course of 4 days and she was discharged home. Unfortunately she lost to follow up, so we were unable to assess the progress and repeat the brain imaging.

Figure 1: MRI axial T2 weighted image showing minimal bilateral subdural effusion in frontoparietal region

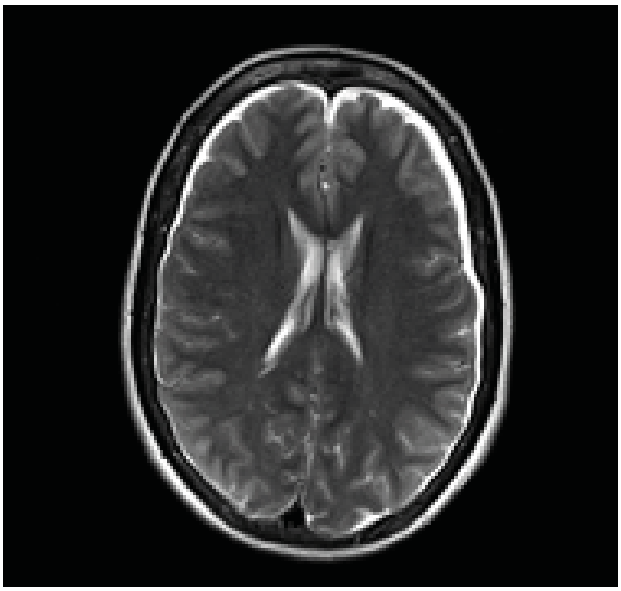


Figure 2: Axial and coronal T1 weighted post contrast images showing thickening and enhancement ofdura along the cerebral and cerebellar hemispheres, falxcerebri and along tentorium

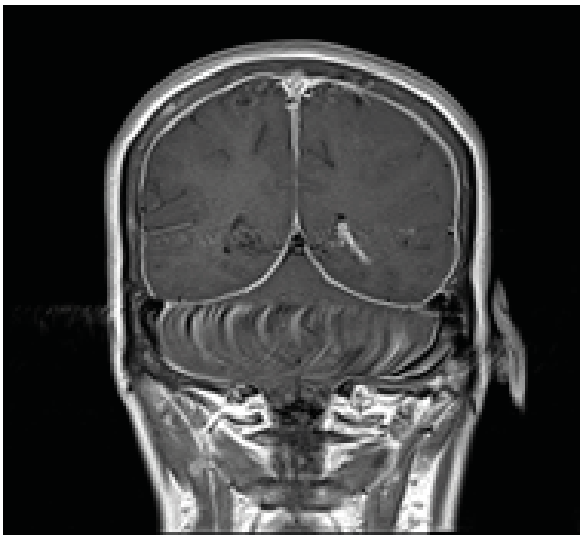
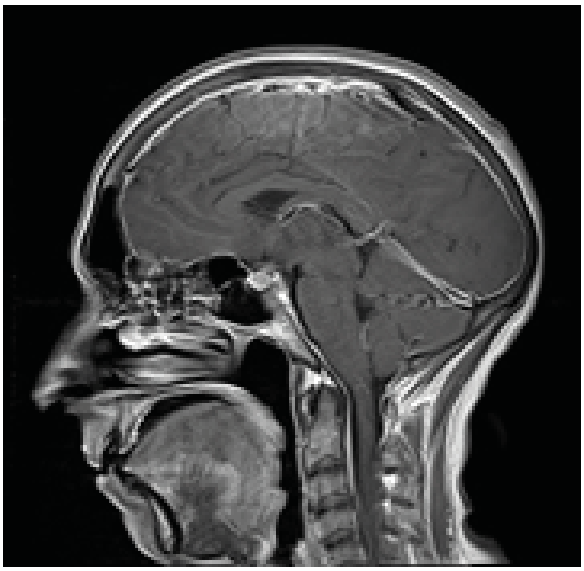


Figure 3: Saggital T 1 weighted post contrast image showing mild cerebellar tonsil descend, low position of corpus callosum with compression of 3rd ventricle, crowding of posterior fossa structure, flattening of pons and prepontine cistern and bulging of pituitary gland.



DISCUSSION

The syndrome of spontaneous intracranial hypotension (SIH) was first proposed in 1983 by schaltenbrand. [4] It shares many similarities with the post lumber puncture headache to which many physicians are familiar with. [5] Women are effected more commonly then men. The pathogenesis is usually considered to be an occult CSF

leak through small defects in the meninges with a resultant decrease in CSF volume and pressure[6]. Although exact cause of CSF leaks often remains unknown. Two factors are frequently suspected one is Mechanical factors including minor trauma or an inciting event like sudden sneezing or sports activity, and the other is an underlying connective tissue disorder thatmay result in dural weakness and play a role in the development of spontaneous low cerebrospinal fluid pressure. [7] Theseinclude occurrence of meningeal diverticulae and CSF leaks in patients with Marfan'ssyndrome. [8] Our patient found to have hyperthyroidism. Although on literature search no direct association between these two entities have been established. Hyperthyroidism is mostly autoimmune in etiology; work up of connective tissue diseases and levels of anti-TPO antibodies should also have been done. As connective tissue disorders aremostly autoimmune in etiology which can be an underlying causative factor of SIH The cardinal clinical manifestation in SIH is orthostatic headache i.e. a headache in upright position that is relieved by recumbency. It is typically bilateral and may or may not be throbbing in nature, there is however considerable variability. Other associated features include pain or stiff feeling in neck, Nausea some time emesis, diplopia typically horizontal with or without unilateral or bilateral 6th cranial nerve palsy, visual blurring, photophobia and visual field defect, [9] along with hyperacusis, echoing, tinnitus, vertigo, dizziness, unsteadiness of gait, hiccups, facial numbness, facial weakness, and dysgeusia.[1, 2] Clinical picture can sometimes mimic dementia, and sagging of the brain can cause lesions in the brainstem leading to stupor and coma.[10] Other rare manifestations include Parkinsonism, ataxia, cerebellar hemorrhage, and cerebral venous thrombosis.[1] Descent of brain occurs as a consequence of CSF leak and CSF volume depletion. This leads to traction or distortion of various anchoring structures of the brain that are pain sensitive,[11] and therefore, leads to orthostatic headache. Secondary vasodilatation of the cerebral vessels to compensate for the low CSF pressure may contribute to the vascular component of the headache by increasing brain volume. Similarly traction and distortion of some of the cranial nerves may also be responsible for the cranial nerve palsies, or even dizziness and altered hearing. Visual blurring and visual field defects have been attributed to compression, traction, or vascular congestion of intracranial portions of the optic nerves. Diagnosis is suggested by the sign and symptoms and is confirmed by the Magnetic resonance imaging (MRI). Lumber puncture usually reveals low opening pressure of less than 60mm of H2O, or the measurement of pressure may not be possible.[12] Common CSF abnormalities include moderate lymphocytic pleocytosis (up to 50cells/mm3).Presence of red blood cells and raised protein (up to 100mg/dL).[6] CSF hypovolemia rather than CSF hypotension has also been proposed as underlying cause of headache syndrome as patients with normal CSF pressure have been described who have clinical and radiographic features that are otherwise typical of orthostatic headache. [5] Confirmation of the diagnosis requires evidence of low CSF pressure by MRI which shows following features; diffuse meningeal enhancement is most universal MRI finding while Subdural hematomas or hygromas presumably from rupture of the bridging veins as the CSF volume decreases is seen in 50% of cases.[2, 3, 13] Saggingof the brain with effacement of prepontine cistern, cerebellar tonsillar herniation and descent of the brainstem mimicking a Chiari 1 malformation is very characteristic feature of SIH.[2] Beside that there may be engorgement of cerebral venous sinuses, pituitary enlargement, flattening of optic chiasma and increased anteroposterior diameter of brainstem . MRI may be normal in 20% of patients.[2] The acronym SEEPS (for subdural fluid collections, Enhancement of the pachymeninges, Engorgement of the venous structures, Pituitary enlargement, and Sagging of the brain) recalls the major features of spontaneous intracranial hypotension on brain MRI.[2] Diagnosis should be considered in patientswho present with orthostaticheadache, with or without associated symptoms, perhaps in the setting of minor trauma, and in the absence of a history of dural puncture or other cause of CSF fistula, confirmation of the diagnosis requires evidence of low CSF pressure by MRI or LP, and/or evidence of a CSF leak on neuroimaging.[14] Diagnostic criteria of SIH as recommended by international headache society are shown in Table 1.[1]

Our patient presented with continuous headache which settles after assuming an upright position, associated with vertigo nausea and vomiting. Her MRI also revealed classical findings diagnostic of SIH. Lumber puncture was not done as diagnosis was confirmed by the brain imaging. Majority of patients respond to conservative and supportive therapies, which include strict bed rest and avoidance of the upright position; strategies aimed at restoring cerebrospinal fluid (CSF) volume include oral or intravenous fluids, highoral caffeine intake and high salt intake. There is also some evidence of intravenous caffeine use.[15] Glucocorticoids have been reported to be of some benefit, but remains unproven, while analgesics are often recommended as first line treatment, generally, they provide little relief, beside that narcotics have also some role in pain management.[2] When conservative treatment fails, epidural blood patch (EBP)is the treatment of choice. [2] EBPtherapy involves

the infusion of 10-20 cc of autologous blood into the epidural space. EBP provides complete, long lasting relief in most patients.[16] Patching with fibrin glue and surgical repair of dural rent or meningeal diverticula may be required when EBP fails.[2] Our patient was managed with the conservative treatment i.e. bed rest intravenous hydration with normal saline and analgesics along with the treatment of hyperthyroidism with antithyroid drugs and beta blocker; metoprolol. She responded well to the medical and conservative management in a course of 3-4 days. Low CSF pressure headaches generally resolve spontaneously within 2 weeks.[6] Occasionally it may last for months, and rarely year. Furthermore, some patients have persistent symptoms despite documented resolution of CSF leakage with therapy.

CONCLUSION

SIH is a rare syndrome that should be considered in the differential diagnosis of daily persistent headache. A good history would help in suspecting the diagnosis which can be confirmed by low CSF pressure and typical findings on contrast enhanced Magnetic resonance imaging (MRI). Failure to recognize can subject the patient to unnecessary procedures, treatment and prolong morbidity. Once diagnosed, majority of patients respond well to conservative treatment. In refractory cases EBP provides long-lasting relief in most patients.

Table 1: Diagnostic criteria for headache attributed to spontaneous (idiopathic) low CSF pressure

Diffuse and/or dull headache that worsens within 15 min. after sitting or standing, with at least one of the following (and fulfilling criterion D):
Neck stiffness
Tinnitus
Hypacusia
photophobia
Nausea
Atleast one of the following:
Evidence of low CSF pressure on MRI
Evidence of CSF leakage on conventional Myelography, CT Myelography, or Cisternography
CSF opening pressure < 60mmH_2o in sitting position
No history of dural puncture or other cause of CSF fistula
Headache resolves within 72 hours after epidural blood patching

REFERENCES

1. Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders. Cephalalgia. (2nd ed) 2004;24(Suppl 1):9–160
2. Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension.J Am Med Assoc. 2006;295:2286–96.
3. Ramesha KN, Chandrashekaran K, Thomas SV. Cerebrospinal fluid hypovolemia syndrome with benign course. Ann Indian Acad Neurol. 2010; 13:293–6.
4. Khurana RK: Intracranial hypotension. Semin Neurol. 1996;16:5-9
5. Mokri B. Spontaneous cerebrospinal fluid leaks: from intracranial hypotension to cerebrospinal fluid hypovolemia. Mayo ClinProc 1999;74:1113-23
6. Rando T, Fishman R. Spontaneous intracranial hypotension: report or two cases and review of the literature. Neurol. 1992;42:481-48
7. Fishman R, Dillon D. Dural enhancement and cerebral displacement secondary to intracranial hypotension. Neurol. 1993;43:609-11
8. Davenport RJ, Chataway SJ, Warlow CP: Spontane ous intracranial hypotension from a CSF leak in a patient with Marfan's syndrome. J NeurolNeurosurg Psychiatry 1995, 59;516–19.
9. Horton JC, Fishman RA: Neurovisual findings in the syndrome of spontaneous intracranial hypotension from dural cerebrospinal fluid leak. Ophthalmol. 1994;101:244–51
10. Agarwal P, Menon S, Shah R, Singhal BS. Sponta neous intracranial hypotension: Two cases includ ing one treated with epidural blood patch. Ann Indian Acad Neurol. 2009;12:179–82.

11. Fay T. Mechanism of headache. Arch Neurol Psychiatry. 1937;37:471–74.
12. Rabin BM, Roychowdhury S, Meyer JR, Cohen BA, LaPat KD, Russell EJ. Spontaneous intracranial hypotension: Spinal MR findings. Am J Neuroradiol. 1998;19:1034–9.
13. Lin WC, Lirng JF, Fuh JL, Wang SJ, Chang FC, Ho CF, et al. MR findings of spontaneous intracranial hypotension. ActaRadiol. 2002;43:249–55.
14. Chung SJ, Kim JS, Lee MC. Syndrome of cerebral spinal fluid hypovolemia: Clinical and imaging features and outcome.Neurol. 2000;55:1321–7.
15. Chio A, Laurito CE, Cunningham FE. Pharmacologi cal management of postdural puncture headache. Ann Pharmacother. 1996 30:831-39
16. Safa-Tisseront V, Thormann F, Malass- ine P, Henry M, Riou B, Coriat P, et al. Effectiveness of epidural blood patch in the management of post-duralpuncture headache. Anesthesiol. 2001; 95:334-39.

Conflict of Interest: Author declares no conflict of interest.

Funding Disclosure: Nil

Author's Contribution:

Mehwish Sayed1: Concept and design, data collection, data analysis, manuscript writing, manuscript review

Ahmed Asif: Concept and design,data collection, data analysis, manuscript writing, manuscript review

Bushra Reha: Data analysis, manuscript writing, manuscript review

Amanullah Khan: Data analysis, manuscript writing, manuscript review