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Spontaneous intracranial hypotension and a comparison of available treatment options. A case report

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Abstract : We report the case of a patient presenting with postural headache and suffering from spontaneous intracranial hypotension. A general review of the management options, mainly focusing on imaging techniques and treatment, is discussed in this paper. Another cause of postural headache is post-dural puncture headache. Both pathologies are referred to the anesthesiologist for management, the key treatment being an epidural blood patch. Hence, anesthesiologists need to be aware of the similarities and differences between management options and efficacy of the blood patch in each case. This is why we attempt a comparison between the two in this paper.

Key words : case report ; spontaneous intracranial hypotension ; blind and targeted epidural blood patch ; post-dural puncture headache.

Introduction

Spontaneous intracranial hypotension (SIH), was first described by Schaltenbrand in 1938 (1), who named it 'aliquorrhea' and described the syndrome as presenting with a headache typical to that observed after lumbar puncture. This pathology has been increasingly recognized over the last few decades owing mainly to increasing availability of imaging techniques. Similarities exist with the more 'classical' post-dural puncture headache but mechanisms involved are different and management options and treatment might differ.

Case Report

A 55 year-old female patient was admitted to our hospital for intense headaches. She presented with nuccalgia and holocranial headaches worsening over the last 48 hours. She also complained of nausea but without any other symptoms. Her past medical history was insignificant except for a history of gastric ulcers. On physical examination, neck stiffness was noted with no other neurological signs being present. The headaches were also noted to worsen in the standing position suggesting orthostatic headache. Her vital signs were normal.

A blood test showed normal RBC and WBC count and no sign of inflammation.

An initial CT-Scan of the head and the spine proved to be normal. She was then hospitalized in the neurology department for a further medical investigation. Analgesics were initiated (paracetamol and ibuprofene) but the symptoms persisted, and the headaches only subsided in the supine position. A brain MRI showed diffuse pachymeningeal enhancement and a diagnosis of Spontaneous Intracranial Hypotension was made. The patient was then referred to the anesthesiology department for an epidural blood patch.

In the absence of any contra-indication for the procedure and in aseptic conditions, she underwent a blood patch in the L3/L4 lumbar region using the loss of resistance technique, with the injection of 15ml of autologous blood using a 17G Tuohy needle. Injection was stopped when the patient experienced back pain. The patient remained in the supine position thereafter for 2hrs and was then progressively switched to the sitting then to the standing position. Her symptoms gradually disappeared in the hours following the blood patch and she was discharged home 36 hours later. Informed consent was obtained from the patient for presentation of this case.

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Discussion

SIH is a rare condition, with an estimated incidence of 5 out of 100 000. It has a higher incidence in females, affecting 2 females for every male and has a peak incidence between 40 to 50 years of age (2). Although more common in adults, some cases of SIH in children have also been reported (3).

Etiology

Cerebrospinal fluid (CSF) leakage has been suggested as the cause for SIH. This leakage can be present anywhere along the spine but mainly in the thoracic and lumbar region although leaks at the base of the skull have also been reported (4). However, mechanisms underlying the CSF leakage can be multiple and remain undetermined in some cases. Mokri et al. reported associations with the presence of connective tissue abnormalities and marfanoid symptoms (retinal detachment, hyper-flexible joints, and carotid artery dissection) in 9 out of 58 patients presenting with SIH (5). Another potential cause can be minor trauma resulting in rupture of preexisting spinal epidural or perineural cysts. This minor trauma can be the result of a fall, coughing/sneezing or even any vigorous sports activity (5). A case of CSF leakage after horse riding has recently been reported by Karsidag et al. (6). Presence of degenerative bone disease and bone spurs may provoke dural tear and cause SIH (7). The presence of a CSF venous fistula draining the CSF into epidural veins has also been described (8). It is to be noted, however, that in most cases the initial cause remains unidentified, which was also the case in our patient.

Symptoms

Postural headache is the typical symptom encountered in these patients. Its presentation can vary in terms of onset (sudden versus gradual) and intensity (moderate versus intense pain) (2). In some cases and if untreated these headaches may also develop into chronic headaches and lose their postural component (9). Traction on the brain and nervous structures can also lead to a wide range of other symptoms. Concomitant neck pain or stiffness, nausea and vomiting, and hearing anomalies are most frequently observed, although photophobia, diplopia, Parkinsonism and cognitive abnormalities have also been reported (10).

Imaging

Once SIH is suspected, imaging studies are necessary firstly to confirm the diagnosis and secondly to exclude other possible causes of headache. Magnetic resonance imaging (MRI) of the brain and spine has been recommended as the best initial imaging option. Available evidence showed a sensitivity of 83% for brain MRI and 94% for spinal MRI (11). Several features have been identified as being strongly suggestive for SIH, namely (5) :

- Diffuse pachymeningeal enhancement: the most common finding seen in SIH being present in 80% of cases. It is the result of hyperemia from the dilatation of subdural vessels.
- Subdural fluid collections: Present in 50% of cases. Fluid collections can be either hygromas or hematomas. In some cases, a mass effect with the consequent symptoms may be present. One study including 40 patients with SIH showed an association between the presence of subdural hematomas (SDH) and worsening headaches and neurological deficits (12). Another study showed that delay in diagnosis and treatment of SIH may lead to the formation of SDH (13).
- Pituitary gland hyperemia: An enlargement of the gland can be seen. It can be mistaken for adenoma or hyperplasia of the gland.
- Sagittal sag: Ventricular collapse and descent of cerebellar tonsils below the foramen magnum are signs suggestive of sagging.
- Engorged cerebral venous sinuses.
- Epidural fluid collections and dural sac collapse can most commonly be found on spinal MRI.

These findings are in line with the Monro-Kellie hypothesis which states that the total volume of the brain, CSF and intracranial blood are constant and that thus a decrease in one of the components leads to an increase in another (14) : loss of CSF resulting in veno-dilation, hyperemia and eventually hygromas/hematomas. In some of the cases, however, MRI results may be normal (15).

In cases of doubt or to target treatment, spinal imaging may be necessary. Spinal MRI can identify an area of CSF collection. A more invasive imaging technique called myelography consists of injecting contrast material intrathecally and identifying CSF leaks using MRI or CT. This technique may be used in cases in which treatment is ineffective and targeted treatment is needed or in which treatment options like surgery are being considered. Expertise of the radiological team is of great importance for this technique. Williams et al. have shown that

in 50% of cases, the CSF leak site remains undetected (16). In one study involving 17 patients, myelography with MRI showed CSF collections in 24% of patients in which the initial brain MRI was negative. Another diagnostic tool is radionuclide cisternography (RNC) where a tracer is injected intrathecally and CSF leak sites identified. In one study on 30 patients, RNC showed direct signs of CSF leak in 80% of them (17).

Treatment

Several treatment options in SIH are available with epidural blood patch (EBP) being the cornerstone. Conservative measures like hydration and bed rest are the first line treatment. Medications like caffeine, theophylline and corticosteroids have also been used but their effectiveness is still under question. In rare cases, surgery might be needed to fix the CSF leak site. These are all available treatment options, but in this report, we will mainly focus on EBP.

EBP needs to be considered in the absence of early resolution of symptoms. A certain volume of autologous blood is thereby injected in the epidural space. Up to 30 ml of autologous blood can be injected intrathecally at the lumbar level (18). The injectate will firstly fill in the missing volume by exerting a mass effect, limiting traction on the nervous structures, and may thus have an immediate relief on symptoms. Secondly, a clot may form and seal the site of CSF leakage.

Controversy still exists about the EBP procedure, some advocating a targeted EBP technique for the treatment of SIH rather than a blind EBP technique. A retrospective study including 56 patients with SIH demonstrated 87,1% effectiveness with targeted EBP in 31 patients compared to 52% effectiveness with a blind EBP technique at first attempt in the remaining 25 patients (19). In the blind EBP group, 6 patients had a thoracic EBP and 19 had a lumbar EBP. The EBP procedure was done with the patient in the lateral decubitus position and maintaining a 30-degree Trendelenburg position for 20 minutes post-procedure in the blind EBP group, whereas in the targeted group, the patients remained supine after the EBP. The volume of autologous blood used for treatment also varied with lower volumes used for the targeted group (up to 15 ml compared to 20 ml for the blind group).

Thomas et al. reported a full recovery in a series of 4 patients who had targeted EBP following CSF leak detection using radionuclide cisternography (20). Notably, in this series, the leak was localised in the lumbar region in 2 out of 4 cases. It is worth noting that targeted EBP can also lead to some serious side-effects like compression of the spinal cord and nerve roots, chemical meningitis, intrathecal blood injection and neck stiffness (19).

Another study followed 30 patients and showed an efficacy of 77% for the blind EBP technique at the lumbar level (21). Of the 77%, 57 % of the patients needed one EBP while 20% showed full recovery only after a second EBP. The patients remained supine two hours after the EBP procedure. The authors describe their low success rate by presuming that some CSF leaks might have been present in the thoracic region so that EBP in the lumbar region might have not been effective, even though high volumes of autologous blood of up to 40 ml were used in this study.

Ferrante et al. reported promising results with blind lumbar EBP for treating SIH (22). In this observational study, 42 patients were included who remained symptomatic after conservative measures. Spinal MRI or myelography was used to detect the CSF leakage site, and spinal neuroimaging was used to evaluate blood patch spread after the procedure. The EBP procedure was done in the lumbar region between the L2-L5 lumbar spaces. There were two particularities linked with the EBP procedure in this report. Firstly, the patients were premedicated with acetazolamide (250 mg 18h and 6h before the procedure). Secondly, the patients were maintained in a 30-degree Trendelenburg position one hour before, during and 24h after the procedure. The authors hypothesized that acetazolamide decreased CSF production and intracranial pressure. This effect associated with the Trendelenburg position might limit CSF leakage and favor the approach of the borders at the leakage site and hence maximize the efficacy of the EBP. The mean volume of autologous blood injected was quite high at 28 ml. The CSF leakage site was identified in 17 patients with 6 being at the cervical level, 7 at the cervicothoracic junction, 1 at the dorsal level and the remaining 3 at the lumbar level. The leakage site remained unidentified in 25 patients. Interestingly, complete relief was obtained in 90 % of patients after a single lumbar EBP. The remaining 10% required up to 3 EBP. Another interesting observation is that spinal neuroimaging post-EBP showed a lumbar to cervical spread in 63% of patients.

Conclusion

No guidelines or consensus exist for the treatment of SIH. The paucity of controlled studies

and the necessity for invasive imaging techniques makes the question between blind versus targeted EBP technique very debatable. Blind EBP have comparable efficacy compared to targeted-EBP in light of the available literature and should thus be the first choice of treatment. A trend also exists towards high autologous blood volumes for EBP. Acetazolamide pre-medication associated with the Trendelenburg position might have a positive impact on treatment efficacy. If, however, targeted EBP is being considered, one has to bear in mind its potential side effects and expertise of the radiological team might also be crucial. As anesthesiologists, we are most often confronted with post-dural puncture headache, but we need to be aware of the differences in treatment that exist with SIH.

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