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LEADING TOPIC

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Headache associated with intracranial hypotension: diagnostic challenges and difficulties in everyday neurological practice

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ABSTRACT

Low pressure of cerebrospinal fluid (CSF) is a rare cause of headache, except when the patient undergoes a lumbar puncture. Headache associated with a low CSF pressure i.e. intracranial hypotension causes diagnostic difficulties. Headaches related to spontaneous intracranial hypotension (SIH) pose a significant diagnostic challenge in everyday neurological practice. Patients with headaches due to SIH are usually diagnosed only after a long delay. Diagnostic problems may result in unnecessary invasive diagnostic procedures, or even neurosurgical operations. Diagnosing headaches attributed to SIH requires the consideration of several clinical scenarios, and the disease's features causing primary or secondary disturbances. In this review, we discuss the differential diagnosis of SIH-related headaches with reference to accumulated knowledge, including meta-analyses, guidelines, casuistry, and the applicable criteria of the International Classification of Headache Disorders. In addition, we discuss head and spine magnetic resonance imaging abnormalities, which may indicate intracranial hypotension.

Keywords: intracranial hypotension, spontaneous intracranial hypotension, orthostatic headache

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Introduction

The definition of intracranial hypotension (ICH) is a cerebrospinal fluid (CSF) pressure of less than 60 mm H_2O causing various clinical symptoms, including headache. However, in over 50% of cases, pressures are above 60 cm H_2O [1]. This applies to lumbar puncture opening pressure in the lateral decubitus position. It is possible that rather than low CSF pressure, it is in fact low CSF volume that is the primary cause of the symptoms of ICH.

Therefore, the term 'CSF hypovolemia' has also been used to describe this syndrome [2].

The symptoms of ICH are orthostatic headache, usually accompanied by nausea or vomiting, neck pain or neck stiffness, tinnitus, hearing disturbances, and visual symptoms including photophobia [3].

The most common reason for ICH is a CSF leak, either traumatic or iatrogenic. Otherwise, ICH can be a spontaneous condition. Spontaneous intracranial hypotension (SIH) is challenging to diagnose because of the variability of clinical symptoms and feasibly subtle imaging findings. SIH can be easily overlooked, and thus patients may await a correct diagnosis for many years.

Diagnostic criteria and clinical picture of headache resulting from low cerebrospinal fluid pressure

According to the 3rd edition of the International Classification of Headache Disorders, a headache resulting from ICH is classified as a secondary headache attributed to a non-vascular intracranial disorder caused by low CSF pressure [4]. Intracranial hypotension headaches are categorised

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Table 1. Diagnostic criteria of headache resulting from intracranial hypotension according to the International Classification of Headache Disorders [4]

Headache attributed to low CSF pressure:
A. Headache fulfilling criterion C
B. Either or both of:
a) Low CSF pressure (< 60 mm CSF)
b) Evidence of CSF leakage on imaging
C. Headache has developed in temporal relation to low CSF pressure or CSF leakage or led to its discovery
D. Not better accounted for by another ICHD-3 diagnosis.
Postdural puncture headache:
A. Headache fulfilling criteria for headache attributed to low CSF pressure, and criterion C below
B. Dural puncture has been performed
C. Headache has developed within five days of dural puncture
D. Not better accounted for by another ICHD-3 diagnosis.
CSF fistula headache:
A. Headache fulfilling criteria for headache attributed to low CSF pressure, and criterion C below
B. A procedure has been performed, or trauma has occurred, known sometimes to cause persistent CSF leakage (CSF fistula)
C. Headache has developed in temporal relation to procedure or trauma
D. Not better accounted for by another ICHD-3 diagnosis.
Headache attributed to spontaneous intracranial hypotension:
A. Headache fulfilling criteria for headache attributed to low CSF pressure, and criterion C below
B. Absence of a procedure or trauma known to be able to cause CSF leakage
C. Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage or has led to its discovery
D. Not better accounted for by another ICHD-3 diagnosis.

CSF — cerebrospinal fluid; ICHD-3 — International Classification of Headache Disorders 3rd edition

as either a postdural puncture headache (PDPH), a CSF fistula headache, or a SIH-related headache. Table 1 sets out the diagnostic criteria for headaches related to ICH [4].

The hallmark of a headache attributed to ICH is frequently orthostatic, usually but not always related to changing position from supine to standing. It has been postulated that a decrease in CSF volume and the downward displacement of the brain cause traction on pain-sensitive structures [4]. The other current theory indicates that venous dilatation causes the symptoms (which we discuss later).

Symptoms can occur immediately after standing up, and resolve quickly after resuming a supine position, or may show delayed response to postural change, worsening after minutes or hours of being upright, and improving after minutes or hours of being horizontal [4].

In most cases, the onset of headache is gradual, reaching maximal intensity in a period ranging from several minutes to several hours. The severity of headache varies from mild and non-bothersome to very severe, resembling a sub-arachnoid haemorrhage [3]. It may be extremely painful and debilitating [5].

The orthostatic type of headache and posture-related component of this symptom may become less evident over time, leading to frequently delayed diagnosis and misdiagnosis [3].

Post-dural puncture headache

PDPH is the best-known type of headache caused by ICH. Previously, it was referred to as 'post-lumbar puncture headache' or 'post-puncture syndrome'. It is estimated that PDPH may affect up to 40% of patients who undergo a diagnostic or therapeutic lumbar puncture (LP) [6, 7]. The time correlation between the LP and the occurrence of orthostatic headache usually helps to establish the diagnosis, although there are interesting case reports of PDPH with extended latencies [8, 9]. Risk factors for PDPH include female gender, age 30-50, a previous history of this type of headache, and an orientation of the needle bevel perpendicular to the long axis of the spinal column during the LP [4]. The use of an atraumatic needle enables fewer complications after lumbar puncture [10].

Typically, PDPH occurs early, i.e. during the first attempts to tilt after LP, but it can also manifest after a delay. The current diagnostic criteria assume five days from dural puncture [3]. Emphasising this issue is especially important in the era of constant reductions in the length of hospitalisations after diagnostic or therapeutic LP.

The essential elements to effectively prevent PDPH include using atraumatic puncture needles and injecting the needle at an oblique angle to the spinal axis [7]. According to the study by Cognat et al. [6], other prophylactic methods such as hydration and bed rest are ineffective and might even contribute to a more severe course of PDPH. Optimal PDPH management has been precisely detailed in the recent multidisciplinary guidelines [11].

Cerebrospinal fluid fistula headache

Headaches caused by CSF leakage can cause enormous diagnostic difficulties. When discussing this type of headache, it is worth considering two clinical situations: a) a headache in a patient after a medical procedure; and b) a headache as a part of the complex symptoms in a patient after injury or trauma.These are the fistulas caused by rupture of meninges, as distinct from CSF venous fistula.

Headache caused by CSF leakage after a medical procedure is usually described in patients operated on due to spinal column diseases (i.e. disc herniation, spinal canal stenosis caused by herniated disc, and/or degenerative changes) [12]. This type of headache is often not recognised early enough for several reasons, e.g. the short duration of the patient's stay in the surgical ward, the treatment used in the postoperative period including parenteral hydration and analgesics, and even the marginalising of patient complaints and omitting this diagnosis in order to limit the number of adverse events in a clinical centre. All these factors may contribute to an underestimation of the incidence of CSF fistula headache. If there is no spontaneous healing during the convalescence

period, a patient with such a headache is usually referred to a neurologist. The longer the time between the procedure and the onset of symptoms, the more difficult it is to connect these facts and to establish the correct diagnosis and treatment.

Diagnosing headache due to CSF fistula is particularly difficult when a headache is a component of the post-traumatic symptoms due to brain/spinal injury. Symptoms of the disease can present up to several months after such an injury. This headache may not have a typical orthostatic nature. In addition, due to treatment that includes a bed regimen and analgesic treatment, symptoms may not appear at first. The symptoms of low CSF pressure may occur only during rehabilitation while standing, and then may be interpreted as symptoms of autonomic dysfunction and dysautonomia resulting from the prolonged supine position of a bedridden patient [13]. LP should be performed and the CSF pressure should be measured to verify the diagnosis [4]. This classification does not meet with the current recommendations. Most headache specialists rather deny the use of LP to diagnose ICH. In the most specialised clinical centres, there is a possibility of assessing the opening CSF pressure during neuroimaging (CT myelogram or DSM imaging) that allows the localisation of a CSF leak. In this clinically severe situation, a LP may be perceived as bizarre in a patient who has experienced injury or multiple injuries. Therefore, understanding the possible abnormalities in non-invasive imaging tests that can be performed in this group of patients will be extremely helpful in raising a suspicion of low CSF pressure. Findings in imaging studies that indicate ICH will be discussed in the next part of this article.

Headache attributed to spontaneous intracranial hypotension

The real diagnostic challenge among headaches attributed to ICH is the headache during SIH. The incidence of SIH is estimated at 3.7-5/100,000/year in the general population [14, 15]. Middle-aged people (30-50) are most often affected, although the disease can occur even in the first or the ninth decades of life. Women are affected more often than men [3, 14]. The literature highlights two critical issues of SIH. The first is a long delay from the onset of symptoms to the diagnosis, which may last several years. A second important aspect is the variety of the clinical picture of SIH-related headaches. The differentiation concerns the nature and location of the headache, changes such as pain over time, and a large diversity of symptoms accompanying the headache. All of these factors may contribute to confusion about this type of headache, with others more common in the population i.e. primary and secondary headaches.

The most common cause of SIH is a spontaneous CSF leakage through a tear or a defect in the dura of the spinal cord, leakage through a meningeal diverticulum, or a CSF-vein fistula [16]. The differentiator between a headache attributable to SIH and a CSF fistula headache is the absence (in the case of SIH) of a procedure or trauma known to be able to cause CSF leakage. Various conditions can lead to spontaneous leakage of CSF, e.g. congenital absence or focal weakness of dura around the nerve root sheaths, congenital connective tissue disorders causing structural abnormalities, osteophyte protrusions, or spinal disc herniation.

The symptoms of SIH are thought to be the result of pulling the meninges (dura mater), stretching of the brain, compression, and stretching of the structures of the posterior fossa (especially the dura mater, cranial nerves, and the opening of vestibular canaliculus) [17]. The SIH headache phenotype is discussed later.

Differential diagnosis of intracranial hypotension

Clinical symptoms that may occur during SIH are set out in Table 2 [3, 16, 17]. To emphasise the broad clinical picture and the diagnostic pathway of SIH, we have decided to present below two clinical situations that neurologists will meet in everyday clinical practice.

Diagnosis of headache due to spontaneous intracranial hypotension in a neurological emergency room: sudden-onset headache

In this section, we analyse a scenario when a patient with SIH is referred for urgent neurological evaluation with a sudden-onset headache. Misdiagnoses described in the literature are related to two main clinical scenarios. In the first, the headache is accompanied by symptoms indicating meningeal syndrome (which is defined as a group of symptoms caused by damage of meningitis for any possible reason). The second clinical picture is mainly marked by symptoms suggesting cerebrovascular diseases (e.g. transient ischaemic attack, TIA).

Scenario 1: Headache due to spontaneous intracranial hypotension with meningeal-like symptoms

Headache attributed to SIH can present with a sudden onset headache. Patients can indicate the exact time of onset. This type of headache's severity can sometimes be comparable to that of a primary thunderclap headache. Many symptoms can accompany headaches attributed to SIH, some of which are consistent with the symptoms of meningeal syndrome. These symptoms include, but are not limited to, nausea and vomiting, visual and hearing disturbances (including sensitivity to light and sound), disturbances of consciousness, neck pain, and stiffness. In such cases, SIH-related headaches should be differentiated from those in subarachnoid haemorrhage and those attributable to meningitis [18].

The exclusion of a subarachnoid haemorrhage is obligatory in the differential diagnosis of sudden onset headache, which has occurred for the first time in the patient's life. Most often, the first diagnostic test performed in the A&E is a head computed tomography (CT) scan, which is widely available

Table 2. Symptoms other than headache of spontaneous intracranial hypotension [3, 16, 17]

CNS structures	Symptoms
CN II (and/or visual pathway)	Photophobia, transient visual obscurations, visual blurring, visual field defects
CN III, IV, VI	Diplopia: abducens nerve, oculomotor nerve, or trochlear nerve palsy
CN V	Facial pain or facial numbness
CN VII	Facial weakness/palsy, facial/hemifacial spasms, dysgeusia
CN VIII	Tinnitus, auditory disturbance ('underwater feeling', muffled heating, aural fullness), phonophobia, vertigo, dizziness; hypoacusis, unilateral hearing loss, acute hearing loss, hyperacusis
Posterior fossa	Nausea, vomiting
General brain dysfunction	Cognitive deficits/symptoms (cognitive impairment, behavioural changes, slow thinking)
	Unconsciousness, coma
	Seizures, status epilepticus
Basal ganglia and/or cerebellar dysfunction	Parkinsonism, tremor, chorea, ataxia
Hypothalamus, hypophysis	Hyperprolactinemia, galactorrhea
Brainstem	Bulbar palsy
Brain vessels	Posterior reversible encephalopathy syndrome (PRES)
	Reversible cerebral vaso-constriction syndrome (RCVS)
	Dural sinus and/or vein thrombosis
Meninges	Meningismus, posterior neck pain, neck stiffness
	Complex symptoms of leptomeningeal hemosiderosis (sensorineural hearing loss, ataxia, dementia)
Spinal cord	Cervical myelopathy, tetraplegia, progressive paraparesis due to secondary spinal cord herniation
Nerve roots	Radicular arm pain, arm numbness, interscapular pain, back pain

CN II — optic nerve; CN III — oculomotor nerve; CN IV — trochlear nerve; CN VI — abducens nerve; CN V — trigeminal nerve; CN VII — facial nerve; CN VIII — vestibulocochlear nerve

and sensitive in detecting the presence of intracranial haemorrhage. However, a head CT scan may not detect signs of subarachnoid haemorrhage, or sometimes these symptoms can be missed due to their small extent. According to a systematic review and meta-analysis by Carpenter et al., the sensitivity of head CT is 94% in the first few hours. In such cases, the following diagnostic tests should be performed: LP and CSF assessment. The LP may provoke complications (including headaches), but they are irrelevant when we consider the possibility of overlooking a life-threatening condition, which is what SAH is. Some difficulties may arise in differentiating between headaches in the course of SIH and headaches in the course of subarachnoid haemorrhage. Firstly, a head CT scan, especially one without contrast, may not show any changes suggestive of SIH.

The proper neuroimaging examination to diagnose SIH is the head magnetic resonance imaging (MRI) with contrast [19]. Secondly, the performed LP and CSF examination may be non-diagnostic of SIH or can give a misleading picture.

It should be noted that the diagnosis of all headaches attributed to ICH includes a criterion of CSF pressure of less than 60 cm H_2O. Fulfillment of this criterion confirms the diagnosis of ICH, but the converse is not true, i.e. CSF pressure equal to or greater than 60 cm H_2O does not exclude an ICH headache. According to the previously cited meta-analysis by D'Anton et al., CSF pressure in SIH patients was normal in 32% of cases, but in 3% of cases was elevated [3]. The assessment of CSF can also be misleading. In patients with SIH, the technical aspects of LP may be difficult, which promotes CSF bleeding [18]. Cases of xanthochromic CSF during SIH have also been reported. The consequences of misdiagnosis may include further invasive diagnostic procedures for vascular pathology exclusion [17, 18].

In an emergency setting, headache caused by SIH should also be differentiated from headache attributed to intracranial infection. Differentiating between these two types of headache should not be a challenge. Therefore, one should consider the explanations behind the diagnostic errors described in the literature. Given the aetiology of meningitis, the most likely misdiagnosis is viral/aseptic meningitis. Meningitis can proceed without a strong expression of inflammatory symptoms and can be milder than neuroinfections of bacterial and fungal aetiology [20, 21]. In addition, attention should be drawn to the unreliable nature of elevated body temperature (fever or low-grade fever) as a symptom differentiating between these two diseases. A significant increase in body temperature is not a permanent symptom of neuroinfection. Furthermore, a patient with severe headaches will most likely take or be given painkillers and anti-inflammatory medications before going to Emergency Room. Hussein et al. [22] observed an increase in body temperature in SIH, which could be explained by inadequate activation of the thermoregulatory centre in the diencephalon as a consequence of the stresses resulting from stretching the brain along its long axis, compression by swollen veins, or activation of the thermoregulatory centre by cytokines as a result of damage to the blood-brain barrier.

Finally, a CSF analysis result can settle the argument regarding diagnosing and treating neuroinfections alone. CSF in patients with SIH may show lymphocytic pleocytosis, which may exceed 200 cells/mm^3. Elevated protein concentrations and decreased glucose concentrations have also been described in CSF taken from patients with SIH [17, 23].

Scenario 2: Headache due to spontaneous intracranial hypotension with transient ischaemic attack-like symptoms

Due to the disease's varied symptomatology and possible sudden onset, SIH-related headaches may also be misdiagnosed as headaches during TIA or even stroke [24]. Symptoms of SIH that may direct the diagnosis towards cerebrovascular diseases include dizziness, balance disorders, numbness or paresthesia of the face or limbs, vision and hearing disorders, taste disorders, and abducens nerve palsy [25, 26]. From a practical point of view, the most helpful tool in verifying the diagnosis will be a head MRI (with contrast). This examination also makes it possible to decide on the diagnosis i.e. TIA or ischaemic stroke [27].

Previously, this diagnosis was established based on the time criterion, i.e. if the symptoms of vascular brain dysfunction persisted for more than 24 hours, then stroke was diagnosed, and if it subsided within 24 hours, then TIA was diagnosed. In the presence of ischaemic focus in the brain, an ischaemic stroke is diagnosed, even if the symptoms of its damage have subsided within the first 24 hours of the disease. Contrast-enhanced head MRI can reveal abnormalities indicative of SIH and establish the diagnosis at this stage.

At this point, we must underline that brain venous thrombosis (i.e. cerebral venous stroke or sinus thrombosis) occurs more often in patients with SIH. The diagnosis of venous infarction itself is a diagnostic challenge. This is the least common type of stroke, usually with multifactorial aetiology and an often confusing clinical picture. It may manifest as a new daily persistent headache [28, 29].

Diagnosis of headache attributed to spontaneous intracranial hypotension in a neurology outpatient clinic: chronic headache

Due to the previously described diagnostic difficulties associated with SIH, the patient is rarely correctly diagnosed at the onset of the disease, even when the disease has an abrupt onset and is associated with orthostatic headache. Orthostatic headache occurs in most (c.80%) of patients, but in the remaining c.20% of cases it can take a different form, including pain independent of body position. In addition, even if the headache was initially orthostatic, it may lose this feature over time, persist, or even intensify, after assuming the supine position [16].

In the literature on headache due to SIH, the most common false diagnoses are tension-type headache, migraine, cervical headache, cervical radiculopathy, headache attributed to Chiari malformation type I, headache attributed to somatisation disorder, and cough headache. Suspicion of SIH requires differentiation from postural orthostatic tachycardia syndrome (POTS) and orthostatic hypotension.

Misdiagnosis of headaches attributed to SIH as either tension-type headaches or migraines can be explained by the high prevalence of these primary headaches in the general population. According to the Global Burden of Disease study published in 2022, it appears that the most common primary headaches in the population are tension-type headaches (26% of the population) and migraines (14% of the population) [30]. Therefore, these are the most frequently diagnosed headaches in everyday neurological practice [31].

The most important issue is to carefully collect the medical history from the beginning of the disease, even if this extends over several years. Headache due to SIH is usually chronic from the onset, whereas tension-type headaches and migraines evolve from episodic to chronic forms during the course of the disease. Another feature that may lead to misdiagnosis is the possible location of the headache attributed to SIH, which usually involves the occipital area, often with coat hanger distribution pain or the entire head. However, it can also be located in the temporal area or cover the top of the head. Limiting neuroimaging to head CT or MRI without contrast may favour diagnosing a primary headache, e.g. tension-type headache [3, 32]. In cases of concomitant visual disturbances, photosensitivity, nausea, or vomiting, migraine may be misdiagnosed. Patients with SIH-related headaches often experience reduced activity during the day because the symptoms tend to worsen in the second half of the day. This feature can be wrongly interpreted as typical migraine headaches that worsen with physical exertion [3, 32].

In particular, headaches attributed to SIH may be misdiagnosed as vestibular migraine. This mistake is facilitated by the frequent occurrence of balance disorders or symptoms of dizziness, which affects about half of patients with SIH [33].

Headache during SIH should be considered in the differential diagnosis of cervicogenic headache, neck pain, cervical radiculopathy, and cervical syndrome. Due to their common characteristics, these diseases will be discussed together.

The cervical part of the spine is the second location of discopathic and degenerative changes in the general population [34, 35]. This diagnosis is usually considered first in a patient with neck pain radiating to the back of the head. The typical characteristics of the headache, accompanied by stiffness in the neck and limitation of movement, may, therefore, be confusingly similar to the chronic pain associated with SIH. In addition, during SIH, numbness and paresthesia of the limbs may occur, which suggests the diagnosis of radicular syndrome. However, patients who experience dizziness and balance disorders may be misdiagnosed with cervical syndrome [3, 16, 17]. Incorrect results of imaging tests of the cervical spine, showing discopathy and/or degenerative changes, may contribute to persistent misdiagnosis of the symptoms of spinal disease.

At this point, it should be noted that an MRI of the cervical spine may reveal the abnormalities discussed below, indicating SIH.

The differentiation of headache attributed to SIH from headache due to Chiari type I malformation deserves separate discussion. In this case, a false diagnosis may result in an unnecessary suboccipital craniotomy. Both conditions share common symptoms: headache, dizziness, balance disorders, sensory disturbances, and cranial neuropathies [32]. The differentiating feature is the duration of the pain. In the case of Chiari type I malformation, the pain episodes are brief and, as emphasised in the diagnostic criteria, last up to five minutes, whereas headache during SIH lasts for hours or is a chronic pain. In both cases, the headache can be aggravated by coughing, Valsalva manoeuvre, or anything that increases intrabdominal pressure. Pain typically localises in the occipital location in both conditions.

One of the radiological signs described in SIH is the displacement of the cerebellar tonsils into the foramen magnum. It is strongly recommended to seek SIH features in neuroimaging to avoid misdiagnosing Chiari type I malformation [36]. CSF leakage should be excluded in all patients with headaches and features of this malformation in head MRI.

A possible reason for SIH-related headaches is heritable connective tissue disorders, e.g. Marfan syndrome and Ehlers-Danlos syndrome. Abnormalities of collagen, fibrillin, or elastin can cause dural weakness, which eventually plays a role in causing spontaneous low CSF pressure due to cryptic CSF leaks [37–39]. Dural diverticulum and perineural cysts are generally common and theoretically might be prone to rupture during a sudden sneeze, sports activity, fall, or benign trauma. Nevertheless, which dural cysts are more prone to rupture in healthy patients and people with connective tissue diseases is unclear. The presence of dural cysts and heritable connective tissue disorders does not necessarily increase the likelihood of a diagnosis of SIH if no active leakage is identified on imaging [40]. In rare cases, a CSF venous fistula can develop between the subarachnoid space and the adjacent spinal epidural veins. There is direct drainage of CSF from subarachnoid space into spinal epidural veins [41].

SIH-related headaches often worsen during coughing; therefore, a primary cough headache should also be included in the differential diagnosis [3, 42]. This type of headache has a different characteristic in that it starts shortly after coughing, and usually peaks within seconds. Unlike SIH-related headache, it should subside within two hours [4]. A feature that may lead to misdiagnosis is the location of the primary cough headache, which, as in SIH, usually affects the occipital area.

Due to the broad spectrum of accompanying symptoms of headache attributed to SIH, there is a risk of false diagnosis of headache attributed to somatisation disorders. Patients with headaches attributed to somatisation disorders (as well as in SIH) can experience nausea, vomiting, coordination and balance disturbances, diplopia, seizures, and sometimes even disturbances of consciousness. Headache can be constant or intermittent with fluctuations of other symptoms. To establish the correct diagnosis, it is vital to have a reliable approach and verification in additional tests of the patient's complaints. It is worth remembering that somatisation disorder is always a diagnosis of exclusion of somatic diseases, and should be established as the last possible option based on the psychiatric consultation [41].

In the SIH diagnostic and therapeutic consensus published in 2023, POTS and orthostatic hypotension were included in the differential diagnosis of this disease [19]. In both conditions, an orthostatic headache may occur. Accompanying symptoms may include dizziness, balance, vision, and consciousness disorders. Early initial verification of the diagnosis of both conditions is possible. However, caution should be taken when performing diagnostic tests due to the possibility of symptoms intensification during the orthostatic test, including loss of consciousness [43].

Abnormalities in head and spine magnetic resonance imaging associated with intracranial hypotension

The most important and widely available tests that can bring the clinician closer to a correct diagnosis are head MRI with contrast and spine MRI. The 2023 guidelines for the diagnosis of SIH define the protocols according to which head and spine MRI should be performed [19]. However, there are no unequivocal criteria for assessing abnormalities in MR neuroimaging in patients with SIH.

The results of studies on this issue differ in the frequency of individual signs in the studied groups of patients. Therefore, at least one MRI finding that may indicate SIH should increase diagnostic consideration and encourage the search for other findings, but cannot of itself result in a diagnosis of SIH.

Abnormalities in head magnetic resonance imaging in patients with spontaneous intracranial hypotension

Contrast-enhanced head MRI is the most important examination for every patient with symptoms (the so-called brain sagging and signs of venous engorgement) that may indicate SIH because of CSF leakage. The second of these pathomechanisms can be explained by referring to the Monro-Kellie doctrine. According to this, the sum of the volumes of the three components filling the skull (the brain tissue, the CSF, and the blood) is a constant value. As a result of the reduction in CSF volume, blood volume increases.

The symptoms most reported on head MRI include diffuse dural enhancement, the presence of epidural fluid collections, dilatation of the intracranial venous structures, and 'sagging' of the brain and pituitary engorgement [3, 16, 44, 45].

We suggest adopting the rule of assessing the MRI results from the top to the bottom of the head. It is important to distinguish between different types of sections and MRI sequences. During such an assessment of the head MRI, attention should be paid to:

- The presence of subdural fluid collection, hygromas, or subdural haematomas [3, 16]. SIH can also lead to epidural haematomas (up to 25% in patients aged under 60), including recurrent ones [46].
- Diffuse contrast enhancement of the dura mater. This is the most frequently reported abnormality in head MRI during SIH [3, 16, 44, 45]. Particular attention should be paid to the current strong trend of limiting contrast-enhanced examinations (to reduce the incidence of contrast-induced nephropathy, gadolinium deposition, and toxic effects on the brain) [47, 48]. This tendency is sometimes unnecessary and can affect the sensitivity of the radiological examination.
- Enlargement of the dural venous sinuses [3, 16, 44, 45]. The symptoms are particularly visible when assessing the shape of the sagittal sinus in the coronal sections along its course on the skull vault. Typically, the lumen of the sinus should have a triangular cross-section, while in SIH the edges of the sinus created by the dura are bulging towards the cranial cavity.
- Presence of superficial siderosis, resulting from bleeding of overflowing and drawn tiny veins [45]. Superficial siderosis (SS) of the CNS is caused by repeated slow haemorrhage into the subarachnoid space with resultant hemosiderin deposition in the subpial layers of the brain and spinal cord [49]. The most common aetiology of SS is dural pathology: cerebral amyloid angiopathy, CNS tumours, arteriovenous malformations, head or spinal trauma, and craniospinal surgery [50]. Infratentorial SS has been recognised in 3.6% of patients with SIH due to CSF leaks, and the reasons were ventral CSF leak (10.4%), dural ectasia (3.9%), CSF-venous fistula (2.6%), and simple meningeal diverticula (0.9%) [51].
- Signs of brain sagging are most easily noticed in sagittal sections [3, 16, 44, 45].

Decreased dimensions of the CSF subarachnoid cisterns resulting from the vacuum effect from spinal CSF leakage are most easily noticed on sagittal sections. The effacement of the prepontine cistern has also been described in SIH. The sagittal sections also may show an opened pontomesencephalic angle. Furthermore, MRI reveals midbrain descent, kinking of the midbrain and pons toward the clivus, and the descent of the cerebellar tonsils to the foramen magnum.

The signs listed above do not include all the abnormalities in SIH patients described in the literature. Table 3 sets out the symptoms and parameters assessed by head MRI in patients with SIH. Some of them require measurement. Nevertheless, a neurological awareness of the radiological evaluation of a head MRI is beneficial in this troubleshooting diagnostic process. On referral for a head MRI in a patient with suspected SIH, a neurologist should ask the radiologist to take or extend the measurements to specific ones.

As mentioned above, there is no generally accepted scale for assessing changes in head MRI in patients with SIH. The current diagnostic criteria also do not specify radiological abnormalities during the diagnosis. However, the so-called Bern score is worthy of attention in diagnosing SIH [47]. This index requires an assessment of six findings in a head MRI. Three of them are referred to as 'major criteria'. These are the engorgement of venous sinuses, pachymeningeal enhancement, and effacement of the suprasellar cistern of 4 mm or less. The occurrence of each of the abovementioned abnormalities results in a score of 2 points. Signs assigned to 'minor criteria' are the presence of subdural fluid collection, effacement of theprepontine cistern by 5.0 mm or less, and a mamillopontine distance of 6.5 mm or less. One point is scored for meeting each minor criterion. The points make up a total, wherein the maximum score is 9 points. In this scale, the risk of SIH is defined as low if the total is up to 2 points, medium if it is 3-4, and high if it is 5 or more [52].

When discussing the importance of head MRI in diagnosing SIH, it should be noted that this examination may not show any abnormalities in up to 20% of patients with SIH [3]. Therefore, a head MRI that does not show any changes typical for SIH does not exclude such a diagnosis.

Abnormalities in magnetic resonance imaging of spinal canal in patients with spontaneous intracranial hypotension

MRI of the spinal canal with heavily weighted, fat-saturated sequences should be performed in each case of suspected SIH [19]. The most common abnormalities reported in patients with SIH are epidural fluid collections that usually extend over five segments of the spine. These reservoirs are called 'spinal longitudinal extradural collections/fluid (contrast)' (SLEC) and are more likely to be detected on axial images. The chance of SLEC occurring depends on the location and cause of the CSF leak. SLEC occurs when the dura mater ruptures on the dural sac's ventral surface or the spinal nerves' root sheath proximal to the intervertebral foramen leaks. SLEC is unlikely to occur in patients with CSF leakage of the spinal nerve root sheath located distally to the intervertebral foramen or through the CSF-vein fistula [53].

The C1-C2 symptom is the other important feature of SIH. In the sagittal sections in a T2-weighted sequence with attenuation of the fat signal, a reservoir with the CSF signal may be observed between the spinous processes of the C1 and C2 vertebrae. This symptom is called 'false localising C1--C2 symptom'. This image may rarely correspond to the location of the CSF leak [54], but usually does not.

According to the Monro-Kellie doctrine, the filling and engorgement of the venous structures increase to maintain pressure within the spinal canal in a CSF leak. This mechanism is responsible for the dilation of the epidural veins and the thickening of the epidural ventral venous plexus. Dural vasodilatation and engorgement may result in dural contrast enhancement. Such reinforcement is usually of a uniform and circular nature [47, 48, 53-55]. The pathology responsible for CSF leakage is most often located in the thoracic region (c.50% of dural tears in the upper

Table 3. Head magnetic resonance imaging findings in intracranial hypotension/ spontaneous intracranial hypotension [4, 12, 13]

No measurement required:
– Subdural fluid collection, hygromas, haematomas, subdural haematomas
– Diffuse gadolinium pachymeningeal enhancement, hyperintense pachymeninges (FLAIR)
– Venous sinus enlargement
– Superficial siderosis
– Brain sagging: midbrain descent, kinking of midbrain and pons toward clivus, reduced distance from optic chiasm to pituitary gland, effacement of perichiasmatic and prepontine cisterns, descent of cerebral aqueduct, effacement of subarachnoid space, tonsillar descent
– Distended inferior intercavernous sinus
– Pituitary gland enlargement, posterior lobe pituitary haematomas
Measurement required:
Size measurement in mm:
– Effacement of suprasellar cistern of 4.0 mm or less
– Effacement of prepontine cistern of 5.0 mm or less
– Mamillopontine distance of 6.5 mm or less
– Tonsillar descent
– Midbrain descent*
– Pituitary height*
– Difference of pituitary height to age-adjusted and sex-adjusted reference*
– Tonsillar descent
Size measurement in mm^2:
– Area cavum veli interpositi*
– Angle measurements:
– Venous-hinge, pontomesencephalic
Other assessments:
– Superior surface of pituitary (concave, flat, convex)

*parameters rarely assessed, normal values are not yet established

thoracic spine) [53], then in the cervicothoracic junction, next in the cervical region, and finally in the lumbar region. To visualise the exact location and determine the nature of the lesion, it is necessary to perform more advanced tests, which according to the current guidelines include CT myelography, digital subtraction myelography, and ultrafast CT myelography [19]. Usually, performing advanced diagnostic procedures requires directing the patient to the specialistic centres.

Discussion of these subsequent tests ordered by neurosurgeons, neuroradiologists, or neurologists is beyond the scope of this article. Interested physicians are referred to the extensive literature available describing all the abovementioned imaging methods.

Summary

Headaches associated with ICH, especially in the case of SIH, are a frequent diagnostic challenge for neurologists.

A wide range of symptoms and similarities to other neurological diseases often lead patients to struggle with this problem for many years without improving after inadequate treatment. This article has presented several tips and possible clinical scenarios indicating the need of considering SIH as the cause of headaches in patients diagnosed in the neurological emergency department and outpatient neurological clinics.

To sum up, the diagnosis in such cases should be verified based on the medical history, in particular, the onset of the disease, the occurrence of any headache depending on the body position (especially if the headache is accompanied by varied neurological symptoms that also worsen when upright and improve when horizontal), an unsatisfactory effect of, or even complete resistance to, the applied treatment, and verification of the assessment of head and spine via a head MRI with contrast and a full spine MRI, as well as searching for easily overlooked or not typically described symptoms in these examinations that may suggest SIH.

A separate part of this paper has described abnormalities in the MRI of the head and spine that indicate SIH. These tests are widely available to neurologists and should be performed and interpreted appropriately. We hope that such a presentation of this problem will allow neurologists to improve their knowledge and more easily establish the proper diagnosis.

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