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Surgical management of spontaneous intracranial hypotension syndrome: a literature review

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ABSTRACT

Introduction. Spontaneous intracranial hypotension (SIH) is a highly disabling but often misdiagnosed disorder. The optimal management options for patients with SIH remain uncertain. The aim of this study was to review studies reporting the management of SIH with a special emphasis on the surgical treatment of SIH including clinical trials, case series and case reports related to the issue of various neurosurgical procedures performed for SIH treatment.

Objective. The clinical outcomes of patients diagnosed with SIH treated with either only surgery or with surgery as the primary method of treatment were analysed.

Material and methods. The PubMed, Scopus and Google Scholar databases were searched according to the established criteria.

Results. The literature search revealed seven clinical trials, five case series and eight case reports regarding surgical treatment of patients diagnosed with SIH. Manuscripts reporting at least five individuals treated surgically for SIH were considered as case series. In most published articles, surgery provided clinical benefit, resulting in a success rate of 82.6–100% for complete relief of SIH symptoms.

Conclusions. Our literature review has revealed that SIH can be diagnosed reliably by MRI and cisternography. The identification of the location of SIH is mandatory for its successful surgical treatment. The clinical outcome is related to the location of SIH in the spinal canal. Most often, cerebrospinal fluid leakage occurs in the thoracic region. Surgical treatment is very effective and the obtained treatment results are complete and permanent.

Key words: spontaneous intracranial hypotension, surgery for spontaneous intracranial hypotension, cerebrospinal fluid leak, orthostatic headache

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Introduction

The syndrome of spontaneous intracranial hypotension (SIH) is a condition that results from leakage of cerebrospinal fluid (CSF) into the extradural space [1–7]. Intracranial hypotension from a CSF leak can be classified as spontaneous, iatrogenic, or traumatic. Spontaneous cases result from dural tears, a meningeal diverticulum, or CSF-venous fistulas. The only known predisposing factor for SIH is hereditary disorders of connective tissue. Increased incidence of SIH has been noted in patients diagnosed with Marfan's Syndrome, Ehlers-Danlos, and adult polycystic kidney disease [8, 9].

The most common and typical symptom of SIH is orthostatic headache that is worse in the upright position [1–7]. It should be noted that the onset of SIH symptoms can occur at a wide range of times. Symptoms may occur within seconds of becoming upright, or not until hours later. [1, 10, 11] Generally, such orthostatic headache occurs or worsens within 15 minutes of obtaining an upright position. The SIH diagnostic criteria according to the International Classification of Headache Disorders (ICHD-3, 3rd Edition) are considered guidelines for a diagnosis of SIH, and these are set out in Table 1.

Conversely, orthostatic headache can improve within minutes of recumbency, or also not for hours [1, 10, 11].

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The positional aspect of the headache may be variable and change with time. These variable characteristics of headache have been attributed to physiological compensation over time. [13]. The headache may be diffuse or localised to the frontal, temporal, or parietal regions, but the most common locations are the occipital and suboccipital regions. The headache is the direct result of the downward displacement of the brain due to loss of CSF, impending on pain-sensitive structures such as dura mater. An alternative mechanism involves compensatory dilatation of the pain-sensitive intracranial venous structures.

Besides typical orthostatic headache, SIH can also be accompanied by a variety of other symptoms. These symptoms, depending on the CSF leak, are usually categorised into four types as shown in Table 2. Posterior neck pain with neck stiffness, dizziness, nausea or vomiting are the most common symptoms, being found in c.50% of patients. These symptoms point to meningeal irritation [14–16].

Table 1. Criteria for headache attributed to low cerebrospinal fluid pressure, International Classification of Headache Disorders, 3rd edition ICHD-3 [12]

A	Any headache fulfilling criterion C
B	1 Low cerebrospinal fluid (CSF) pressure (< 60 mm CSF)
	2 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

Tinnitus with impaired balance is another common symptom of SIH. This symptom can be explained by downward displacement of eighth cranial nerve complex. Visual blurring and visual field deficits are also attributed to downward displacement of the brain. Not only optic nerves and chiasm are affected by the downward brain shift, but also other sensitive cranial nerves located at the cranial base. Cranial nerve deficits due to SIH can involve diplopia (abducens nerve dysfunction, rarely trochlear or oculomotor nerves dysfunctions), facial numbness or facial pain (trigeminal nerve dysfunction), facial weakness or facial spasm (facial nerve dysfunction), and dysgeusia (chorda tympani or glossopharyngeal nerve dysfunction) [15–20].

SIH can cause so-called spinal manifestations such as local back pain at the level of the CSF leak, radiculopathy due to stretching of cervical nerve roots, or dilatation of the epidural venous plexus. Prominent CSF leak may lead to myelopathy with subsequent quadriplegia. The most dangerous life-threatening situations related to SIH are so-called severe intracranial manifestations caused by prominent displacement of the brain. These intracranial symptoms may include a decreased level of consciousness caused by diencephalon herniation causing stupor and coma. Other rare intracranial manifestations may produce cognitive dysfunction, dementia, and parkinsonism [21, 22]. These symptoms generally improve when conservative or surgical treatment of SIH remains effective. SIH may be responsible for cerebellar haemorrhage or ataxia. A summary of different clinical manifestations of SIH is set out in Table 2.

Table 2. Clinical symptoms are segregated into four types of clinical manifestation of spontaneous intracranial hypotension

Type of symptoms of SIH syndrome	Symptoms	Structures responsible for symptoms occurrence
Generalised symptoms	Orthostatic headache	Headache is a direct result of downward displacement of brain due to loss of CSF, impending on pain-sensitive structures like dura mater, or caused by compensatory dilatation of pain-sensitive intracranial venous structures
Generalised symptoms	Neck pain or neck stiffness, dizziness, nausea, vomiting	Meningeal irritation
Cranial nerve deficits	Visual blurring and visual field deficits	Downward displacement of brain, with pressure on optic nerves and optic chiasm
Cranial nerve deficits	Diplopia	Abducens nerve dysfunction, rarely trochlear or oculomotor nerves
Cranial nerve deficits	Facial numbness, facial pain	Trigeminal nerve dysfunction
Cranial nerve deficits	Facial weakness, facial spasm	Facial nerve dysfunction
Cranial nerve deficits	Dysgeusia	Chorda tympani or glossopharyngeal nerve dysfunction
Cranial nerve deficits	Phonophobia, muffled hearing, tinnitus	Vestibulo-cochlear nerve dysfunction
Spinal symptoms	Lower back pain	Meningeal irritation at level of CSF leak
	Radiculopathy	Stretching of cervical nerve roots or dilatation of epidural venous plexus
	Myelopathy	Prominent CSF leak may cause myelopathy with subsequent quadriplegia
Severe intracranial manifestations	Cognitive dysfunction	Mild dementia or subtle cognitive dysfunction caused by chronic subdural hygromas
	Dementia	
	Parkinsonism	Moderate to severe displacement of brain with diencephalic herniation
	Ataxia	Severe cerebellar herniation may cause cerebellar haemorrhage
	Stupor	with downward displacement of tonsils
	Coma	

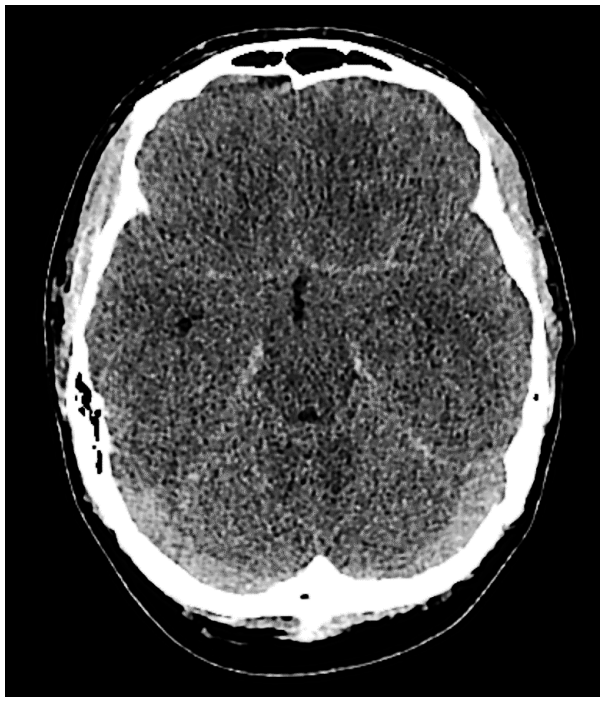

Figure 1. Computed tomography (CT) image showing increased attenuation in basal cisterns and diffuse pachymeningeal enhancement. Increased attenuation, especially in sylvian fissures, can mimic subarachnoid haemorrhage

Imaging findings in spontaneous intracranial hypotension syndrome

Computed tomography (CT) is often the first diagnostic tool performed in emergency units in patients with severe, disabling orthostatic headache. Cranial CT may show subdural fluid collection, effacement of cerebral cisterns, and downward displacement of cerebellar tonsils.

Moreover, CT may show increased attenuation in the sylvian fissures, basal cisterns, mimicking a subarachnoid haemorrhage (Fig. 1) [23]. A further diagnostic tool is magnetic resonance imaging (MRI), which is diagnostic in 80% of cases of SIH. The MRI findings are characteristic and are summarised in the mnemonic 'SEEPS', standing for: subdural fluid collections, enhancement of the pachymeninges, engorgement of venous structures, pituitary engorgement, and sagging of the brain. All these MRI findings together allow a proper and prompt diagnosis. [10, 24–29]. The MRI findings description is beyond the scope of this review article, and more information on MRI findings in SIH can be found in other review articles [30–33].

The recently described 'MRI sign' found in patients diagnosed with SIH is oedema of the corticospinal tracts in the midbrain [34]. Causes of such oedema are only speculative at present and are secondary to long-standing compression or to injury of axons within the midbrain caused by stretching along their axons [34].

Another described MRI finding is superficial siderosis detected on gradient-recalled echo or susceptibility-weighted MR imaging. Hemosiderin depositions are located on the pial surface of the brain or spinal cord as the result of chronic recurrent bleedings in the subarachnoid space. These bleedings are caused by brain sagging and stretching of superior cerebellar bridging veins or intraspinal friable vessels. The superficial brain or spinal siderosis is usually found in patients with long-standing SIH [34].

Other diagnostic tools that can help locate subtle CSF leaks include computed tomography (CT) myelogram or spinal MRI. Adjusting CT timing in brisk leaks with prompt imaging and in slow leaks with delayed imaging, can increase CSF leak detection and localisation [35]. Small ventral leaks or leaks associated with dural diverticula along root sleeves may be better localised using a dynamic CT myelogram [36]. Spinal MRI may be of value in detecting meningeal enhancement, extradural fluid collections, dilated venous structures, or meningeal diverticula. MRI myelograms, especially T2 weighted sequences or intrathecal gadolinium applications, have comparable sensitivity to a CT myelogram in localising CSF leaks [37, 38]. Yet even the aforementioned CT or MRI myelography will fail to localise the site of a CSF leak in approximately half of patients with a fistulous connection between CSF and paraspinal veins [39]. Such CT myelogram-occult CSF leaks require a more sophisticated detection method such as digital subtraction myelography (DSM). This detecting method is especially helpful in detecting CSF leaks secondary to CSF-venous fistula. Recent findings have shown a hyperattenuated paraspinal vein in close proximity to a CSF leak [24]. This so-called 'hyperdense paraspinal vein sign' represents the rapid passage of myelographic contrast into the venous system through the fistula. This finding correlates on DSM images as an opacified paraspinal vein. The recognition of this sign may be helpful in localising CSF leak (fistula) in an otherwise myelogram-occult CSF leak [39, 40].

In summary, DSM can be a very helpful diagnostic tool in high volume CSF (fistula) spinal leaks, which benefit from real-time imaging. DSM offers superior leak detection compared to a CT myelogram with an inherent time delay in ventrally located CSF venous fistulas. Even in small, brisk CSF leaks, by the time CT is performed, the contrast can spread over many levels and the exact location of the dural tear remains unknown [41, 42].

Management of spontaneous intracranial hypotension syndrome by epidural blood patching (EBP)

SIH syndrome is very often misdiagnosed, which has a profound influence on diagnostic and therapeutic management of patients with SIH. It has been observed that many cases of SIH resolve spontaneously without any specific treatment [10]. Several therapy options exist to treat patients with SIH.

Table 3. Types of CSF leak causing SIH, their characteristics on imaging studies, extradural CSF collections as well as different surgical techniques used to close these four types of CSF leak causing SIH syndrome

Leak type in spontaneous intracranial hypotension	Characterisation of leak type	Extradural CSF collection	Surgical management
Type 1a CSF leak	Ventral CSF leaks, usually related to an injury from adjacent bony abnormality	Yes	Identification of dural tear under microscopic inspection, direct repair of dural tear with sutures or placing a small muscle near dural tear with fibrin glue
Type 1b CSF leak	Posterolateral CSF leaks	Yes	Usually placing a small muscle near dural tear with fibrin glue
Type 2a CSF leak	Significant single or multiple meningeal diverticula	Yes, in 20% of cases or sometimes dilated dural sac found	Surgical treatment in cases of multiple meningeal diverticula is directed at largest diverticula Diverticula can be safely closed with small titanium aneurysm clips Direct suturing or muscle graft to directly repair CSF leak
Type 2b CSF leak	Complex meningeal diverticula or dural ectasia	Yes, in 20% of cases or sometimes dilated dural sac found	In cases of dural ectasia, coating dura with artificial dura graft
Type 3 CSF leak	Direct CSF-venous fistula	Extradural collection of CSF not present on imaging	Single venous channel may be successfully treated by applying small aneurysm clip A network of dilated veins should be treated by electrocautery to close small network veins
Type 4 CSF leak	Leaks without any dedicated spinal imaging modality and remaining otherwise intermediate as to above mentioned type	Extradural CSF collection in c.50% of cases	Lumbar dural reduction surgery performed through laminectomy (a strip of dura is resected and dural defect is closed). This manoeuvre increases intracranial CSF volume and pressure Implementation of epidural catheter

It should be noted that to date no randomised clinical trial (RCT) has been done to assess the treatment outcomes in patients with SIH [10].

Once a patient is diagnosed with MRI findings consistent with SIH, first line treatment includes conservative management such as bed rest, intravenous hydration, the use of an abdominal binder, or generous caffeine intake. An unknown percentage of SIH resolve spontaneously, so first-line conservative treatment is recommended especially for uncomplicated presentations. Patients with prominent disabling clinical signs of SIH cannot be expected to gain substantial and durable clinical benefit from conservative treatment for SIH [10].

In these cases, epidural blood patching (EBP) should be considered. EBP is performed by injecting the patient's own blood into the epidural space. It is not necessary to inject the blood at the level of the CSF leak, and the injection is typically administered in the lumbar region. The mechanism of action of an EBP injection includes compression of the thecal sac to immediately increase lumbar and intracranial pressure. Moreover, the injected blood clot over the CSF leak promotes an inflammation process that facilitates healing. Initial injection volume is from 10 to 20 mL of the patient's own blood. One third of patients undergoing this procedure achieve relief of symptoms. If the initial EBP is ineffective, a larger volume of up to 100 ml is recommended. The larger-volume EBP is usually done at two levels (one at the thoracolumbar junction and the second at the lower lumbar level). The volume of administered blood is limited by the development of local back pain or radiculopathy. Two-level large-volume blood administration forms a more stable dural tamponade, thereby sealing the CSF leak. After administration of EBP, the patient is left either in a supine, a prone or a lateral position for 30–60 minutes to allow the blood to move over many segments in the epidural space which may facilitate the sealing of the CSF leak. When several EPBs are required, a repeat procedure can be done, leaving at least a five day interval. If repeat EPBs fail to provide relief, and if the location of the CSF leak is known, targeted percutaneous placement of fibrin sealant is recommended. This is effective in one third of patients [10].

Management of spontaneous intracranial hypotension syndrome by surgery with presentation of clinical outcomes

However, in patients in whom the application of a blood patch or fibrin sealant have failed to bring SIH symptoms relief, surgical treatment is indicated [10]. Generally, surgical treatment is safe and efficacious in patients with a structural abnormality or when a focal CSF leak is precisely identified [43–45]. To perform surgical repair of a CSF, the location of the CSF level and the direction of CSF leak (i.e. ventral or dorsal to the dural sac) play the major role in choosing the surgical approach (i.e. anterior or posterior spinal approach) [1, 46–50]. Posterior CSF leaks and leaks along nerve roots are usually approached by hemilaminectomy or laminectomy [1, 6, 27, 48, 50–54].

In general, ventral CSF leaks are more challenging surgically than dorsal leaks, and the anterior or posterior approach depends greatly on the spinal CSF leak level [24, 55]. Ventral CSF

Table 4. Case reports of SIH treated by surgery

First author and year of publication	Age and gender	Clinical symptoms	Type of CSF leak	Type of diagnostic examination	Location of cerebrospinal fluid leak	Surgical approach	Follow-up in months or years	Final outcome
Davenport RJ (1995) [26]	19 F	Orthostatic headache, nausea, vomiting	Multiple diverticula Type 2b	Radioisotope cisternography	T1-T3	Epidural catheter and a saline infusion.	6 weeks and 3 months	Headache score 3–4/10
Vishteh AG (1998) [46]	32 M	Orthostatic headache	Single meningeal diverticula Type 2a CSF leak	MRI	C5-C6	Discectomy C5/C6 with osteophysectomy, dura mater closure. Dura suturing	ND*	Asymptomatic
Inamasu J (2004) [49]	41 M	Orthostatic headache	Single meningeal diverticula Type 1b	MRI and CT myelography	C1-C2	C1 laminectomy and partially C2	1 month	Asymptomatic
Witiw CD (2012) [57]	46 F	Orthostatic headache, nausea, vomiting	Single meningeal diverticula Type 1b	MRI	C4-C5	Discectomy C4/C5 with osteophysectomy, dura substituted membrane and fibrin glue	2 months	Asymptomatic
Fehnel KP (2015) [50]	34 M	Postural headaches, shoulder discomfort	Multiple meningeal diverticula Type 2b CSF leak	MRI	T9-T10	T9-T10 laminectomy	1 month	Asymptomatic
Turel MK (2018) [52]	50 F	Progressive lower-back pain, postural headaches	Multiple diverticula Type 4 CSF leak	MRI and CT myelography	T10-T11	T10-T11 laminectomy	6 weeks	Asymptomatic
Shahab S (2020) [53]	46 F	Orthostatic headache, vomiting	Type 4 CSF leak	MRI	T11-T12	Laminectomy T11-T12	2 months	8 years laer, patient is still having mild on and off headaches
Sobczyk P (2022) [48]	28 M	Orthostatic headache	Multiple meningeal diverticula Type 1b	MRI and CT	C1-C2	Laminectomy and fibrin glue	6 months	Asymptomatic

ND* — no data

leaks located at the cervical spine are approached by anterior corpectomy or discectomy [26, 56, 57]. Ventral thoracic leaks require a transdural or transpedicular approach, and ventral lumbar leaks a posterior approach between the nerve roots.

The recently defined four-grade classification system for cases of SIH may help select the most appropriate surgical management of a CSF leak. This classification system in the surgical management of SIH leaks was devised by Schievenk et al. [10]. The classification with different types of CSF leaks with preferred surgical technique utilised is presented in Table 3.

The individual case reports of surgical treatment for SIH are presented in Table 4. All patients reported in these case reports remained asymptomatic after surgery for intractable SIH symptoms [26, 46, 48–50, 52, 53, 57]. Case series and clinical studies reporting the outcomes after surgery for SIH are presented in Table 5. Most patients after neurosurgical treatment remained asymptomatic, and the successful rate for operation of SIH varied between 82.6–100% [1, 27, 43, 47, 51, 54, 56, 58–60]. Only one case series presented a successful rate of 50% for SIH symptom relief [51].

In most studies, most of the affected individuals who underwent surgery were women, and the female-to-male ratio was almost 2:1 [1, 43, 51, 54, 58–60]. The patients operated for SIH were in their fourth or fifth decades of life [1, 6, 11, 54, 59, 60]. In almost all cases, MRI was used to diagnose SIH, which remains the basic diagnostic tool [1, 5, 54, 61, 62]. Some patients were diagnosed with CT myelography [42, 47, 49, 56, 58, 59]. A typical MRI case of SIH in a patient with spontaneous CSF leak is presented in Figures 2 and 3.

Interestingly, among the reviewed cases, the most common site of CSF leak was thoracic spine, followed by cervical spine [1, 27, 43, 47–54, 56, 58–60, 63, 64].

Table 5. Clinical studies and case series reporting outcomes of surgery for SIH. Case series include at least five individuals. Studies reporting more than five individuals are considered as clinical studies. Limit on number of individuals was set arbitrarily due to relatively small number of patients treated by surgery worldwide

First author and year of publication	Number of individuals in study	Mean age of patients [yrs]	Mean follow-up in weeks	Percentage of operated patients [%]	Percentage of successful operation (asymptomatic) [%]	Percentage of unsuccessful operations (symptomatic) [%]
Schievink WI (1998) [43]	10	42.3	–	100	100	0
Eros EJ (2002) [56]	3	43	64	66.6	100	0
Farhat HI (2011) [47]	4	37	91	100	100	0
Chai CM (2014) [58]	2	54	5	100	100	0
Idrissi AL (2015) [60]	24	46	24	8	100	0
Schievink WI (2016) [1]	568	45.7	–	50.2	–	–
Beck J (2016) [67]	15	45.7	12	93.3	86.7	6.65
Beck J (2018) [27]	47	44.3	18.5	100	96	4
Wang TY (2020) [5]	20	51.3	64	83.3	90	10
Majeed K (2021) [59]	3	47.6	12	100	100	0
Kamenova M (2021) [51]	5	50.2	14.75	100	50	40
Häni L (2022) [54]	86	46.7	12	80.2	82.6	17.4

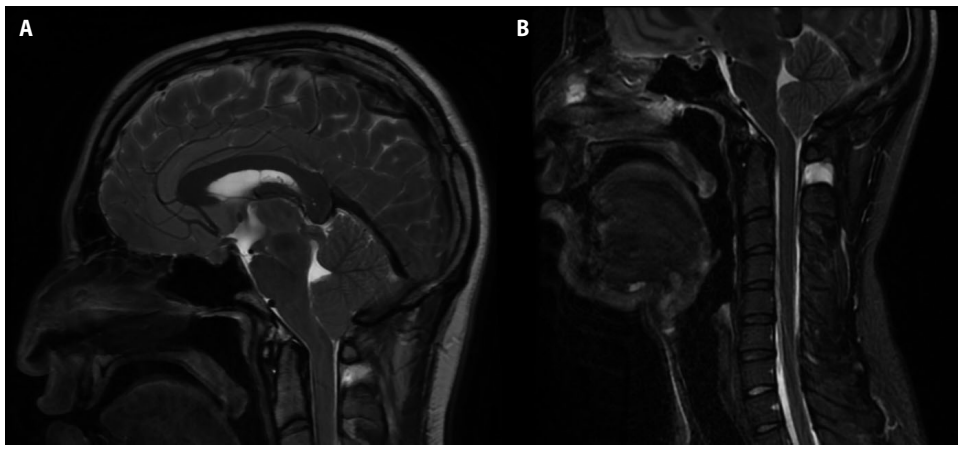

Figure 2. A. Sagittal MRI image T2-weighted sequences, showing changes in spontaneous intracranial hypotension. Lack of cerebrospinal fluid in basal cisterns and interpeduncular fossa are noticeable. **B.** Sagittal MRI image T2-weighted sequences, sagittal wedging of cerebellar tonsils into foramen magnum with confirmed CSF leak at level C1/C2 located dorsally to dural sac

Generally, surgical treatment of SIH is regarded as effective and safe [49]. It is also worth noting that none of the presented patients experienced a deterioration in their health. The sealing material used by most authors has included a fat patch, a muscle piece, fibrin glue or cyanoacrylate-based preparations. [43, 47, 57, 65]. The applied surgical treatment turned out to be a very effective method, as evidenced by the fact that more than two thirds of the patients presented with no further symptoms or complaints. As mentioned above, patients after surgery for SIH rarely complain of severe orthostatic headache or other pre-existing disabling symptoms. However, a change of pattern of headaches could indicate a rebound transient intracranial hypotension, and dural sinus thrombosis should also be considered [15, 66].

The recurrence of clinical SIH symptoms after successful surgical treatment may indicate a recurrent CSF leak [45]. It is estimated that c.10% of operated patients have recurrent CSF leaks. Outcomes studies have shown that patients with typical MRI findings for SIH and a localised CSF leak have very favourable results, in contrast to patients with normal MRI findings and multilevel spinal CSF leaks, who have worse outcomes [10]. The surgical treatment of SIH is rarely performed, as shown by this literature review, but it is very effective [48, 53, 54, 56]. However, due to the small amount of research conducted into this subject, the choice of treatment should be always individualised [48, 53].

Conclusions

SIH syndrome is very often misdiagnosed and can constitute a diagnostic challenge, although in recent times SIH syndrome has become better recognised due to the broad

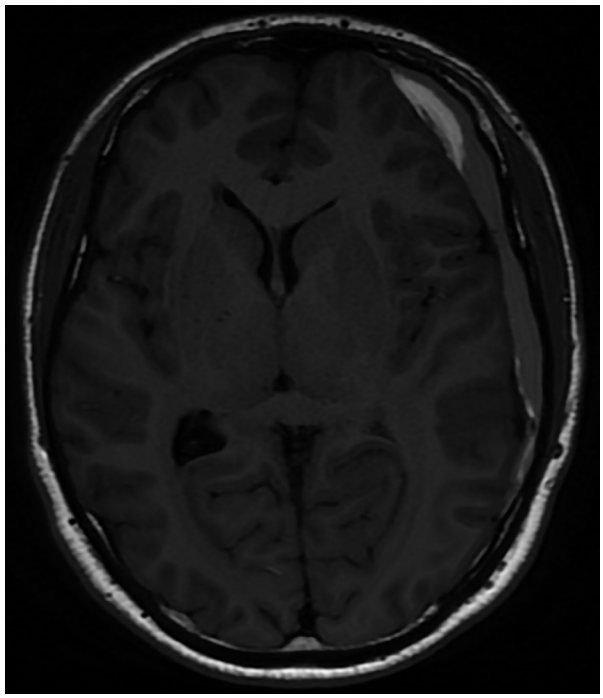

Figure 3. Axial MRI image T1-weighted sequence, showing example of subdural collection (SDC) due to SIH secondary to massive CSF leak in the cervical region

application of MR imaging. Pathological MRI findings are easily ascertained and allow proper and prompt diagnosis combined with clinical manifestations of SIH.

It should be noted that SIH syndrome is not always associated with only orthostatic headache, but can also represent a mixture of different symptoms grouped as general symptoms, cranial nerve deficits, and spinal and intracranial manifestations of SIH. Some patients require more demanding diagnostic modalities such as a CT myelogram or even a digital subtraction myelogram before a final diagnosis of SIH syndrome. An unknown number of SIH cases resolve spontaneously, and the first-line treatment for SIH, if needed, is EBP.

If the EBP fails to control the CSF leak, surgery should be reserved for symptomatic patients with a known CSF leak location. Our review has shown that different surgical approaches and methods of CSF leak sealing are safe and highly effective, and therefore surgery for SIH should not be regarded as only a 'last resort' treatment modality. There is also no doubt that the issue of the surgical treatment of SIH requires further research.

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