

Spontaneous intracranial hypotension and its complications

Hipotensão intracraniana espontânea e complicações

Marília Maria Vasconcelos Girão¹, Rachid Marwan Pinheiro Sousa¹, Mayani Costa Ribeiro², Tânia Aparecida Marchiori de Oliveira Cardoso², Marcondes Cavalcante França Júnior², Fabiano Reis¹

ABSTRACT

Spontaneous intracranial hypotension (SIH) is a syndrome that was unknown until the advent of magnetic resonance imaging (MRI). It is a cause of orthostatic headache, which remains underdiagnosed and, rarely, can result in several complications including dural venous sinus thrombosis, subdural hematoma and subarachnoid hemorrhage. Some of these complications are potentially life-threatening and should be recognized promptly, mainly by imaging studies. We reviewed the MRI of nine patients with SIH and describe the complications observed in three of these patients. Two of them had subdural hematoma and one had a dural venous sinus thrombosis detected by computed tomography and MRI. We concluded that MRI findings are of great importance in the diagnosis of SIH and its complications, which often influence the clinical-surgical treatment of the patient.

Keywords: intracranial hypotension; subdural hematoma; sinus thrombosis, intracranial; magnetic resonance.

RESUMO

Hipotensão Intracraniana Espontânea (HIE) é uma síndrome desconhecida até o advento das imagens de Ressonância Magnética (RM). É uma causa de cefaleia ortostática que permanece subdiagnosticada e raramente resulta em complicações, como trombose de seios venosos durais, hematoma subdural e hemorragia subaracnoidea. Algumas dessas complicações são potencialmente ameaçadoras à vida e devem ser prontamente reconhecidas pelos estudos de imagem. Nós revisamos as RM de 9 pacientes com HIE e descrevemos as complicações observadas em 3 casos. Dois deles tiveram hematoma subdural e um teve trombose de seio venoso dural detectados por tomografia computadorizada e RM. Concluímos que achados de RM são de grande importância no diagnóstico de HIE e suas complicações, frequentemente influenciando o tratamento clínico-cirúrgico do paciente.

Palavras-chave: hipotensão intracraniana; hematoma subdural; trombose dos seios intracranianos; ressonância magnética.

Headache is a common complaint with a wide range of differential diagnoses. Spontaneous intracranial hypotension (SIH) is an uncommon cause of novel onset persistent headaches, typically one that occurs or worsens upon standing and is relieved by lying down, and is often an underdiagnosed condition^{1,2,3}.

Intracranial hypotension has an estimated incidence of 5/100,000 and typically results from a cerebrospinal fluid (CSF) leak, which leads to a decline in CSF volume. The leak can be explained by structural weakness of the dura mater, associated with minimal or no history of trauma. It is known that decrease in CSF volume, rather than CSF pressure decrease, is the core pathogenic factor. The disease is slightly more common in women and can occur in all age groups, with a typical age of incidence around 40 years^{1,2,4,5,6,7,8}.

Delay or failure to diagnose SIH may lead to life-threatening complications such as dural venous sinus thrombosis, subdural hematoma and subarachnoid hemorrhage³.

Imaging exams, particularly magnetic resonance imaging (MRI), play an important role in detecting such abnormalities. Other complications described include bibrachial amyotrophy, superficial siderosis, syringomyelia, cranial nerve palsies and rebound intracranial hypertension^{1,4,5}.

We report a series of patients with SIH, some of whom had complications detected by MRI of the brain.

METHODS

We reviewed the medical records and MRI studies of nine patients with SIH and describe the complications observed

¹Universidade Estadual de Campinas, Faculdade de Medicina, Departamento de Radiologia, Campinas SP, Brasil;

²Universidade Estadual de Campinas, Faculdade de Medicina, Departamento de Neurologia, Campinas SP, Brasil.

Correspondence: Fabiano Reis; Rua Tessália Vieira de Camargo, 126; 13083-887 Campinas SP, Brasil; E-mail: fabianoreis2@gmail.com

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in three of these patients, evaluated by neurologists at the University of Campinas, São Paulo, Brazil, in 2016. The patients had given their informed consent. All patients met previously-reported diagnostic criteria for spontaneous spinal CSF leaks and intracranial hypotension. All of them underwent at least one brain MRI. All radiological images were reviewed by at least one board-certified neuroradiologist. We also reviewed the literature on possible complications of SIH.

Patient 1

A 49-year-old male truck driver, with a history of diabetes mellitus type 2 and previous tension-type headaches, reported a two-week history of headache that started when he was carrying out his usual activities. It was a severe holocranial headache (10/10 intensity), maximum at the onset, which improved with lying down and worsened with orthostasis/sitting up, associated with nausea, photo/phonophobia and rotational vertigo. He also had a history of a minimal head contusion on an aluminum plate a week before the onset of the symptoms, and took acetylsalicylic acid in the first two days of headache, without any improvement.

Computed tomography (CT) of the brain without contrast at admission showed chronic right and left subacute subdural hematomas, decrease in size of the ventricles and enlargement of the pituitary. He was discharged with symptomatic treatment.

The patient came back three days later without having had pain relief. He was evaluated by neurologists and had an MRI, which showed typical findings of SIH: subdural fluid collections (hygromas), diffuse pachymeningeal enhancement, decrease in the size of the ventricles, enlargement of the pituitary, and cerebral venous sinus engorgement. There were also bilateral subdural hematomas (thickness of 1.0 cm on the left and 0.5 cm on the right), creating a discrete midline shift (Figure 1).

The diagnosis of a spontaneous CSF leak was confirmed by radionuclide cisternography (lumbosacral spontaneous dural fistula between L4-L5 on the right and bilateral L5-S1). He was treated by clinical measures (lying down, analgesics, hydration, pure coffee twice a day) and an epidural blood patch. A subdural hematoma drainage procedure was performed as the symptoms did not improve. The patient was discharged with improvement of the headache and without neurological deficits.

Patient 2

A 35-year-old unemployed man with hypertension, presented at the emergency department with a history of three months of bilateral frontotemporal progressive headache, 7/10 intensity, which worsened with orthostasis/sitting up and was associated with nausea, vomiting and otalgia, with slight improvement with the use of NSAIDs.

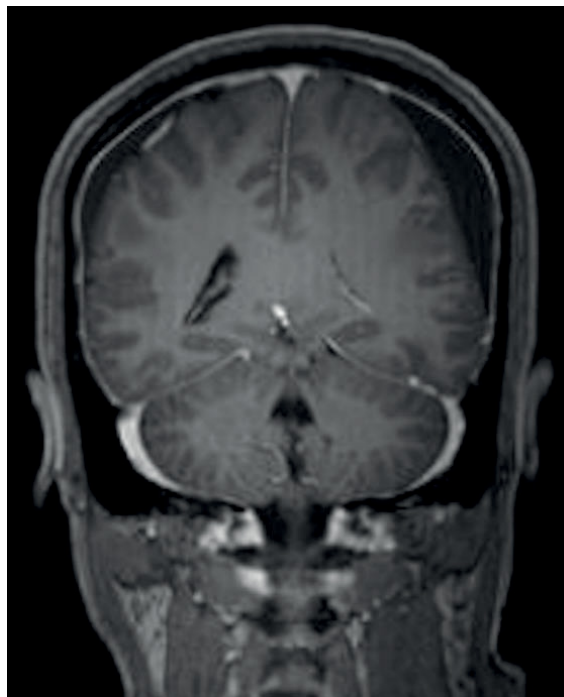


Figure 1. Coronal contrast-enhanced MR image with diffuse pachymeningeal enhancement and bilateral subdural hematoma, showing a decrease in size of the ventricles and discrete dislocation of centroencephalic structures.

He denied any history of trauma and reported that he had stopped working because he could not sit at work due to worsening of the headache. With the hypothesis of SIH, he underwent a CT scan of the brain, which showed a decrease in size of the ventricles and cisterns and a left frontoparietal laminar subdural hematoma. A few days later, he underwent an MRI to complement the investigation, which showed a reduction in the size of the ventricles, diffuse pachymeningeal thickening and enhancement, volumetric increase of the pituitary, engorgement of the venous sinuses, bilateral subdural effusions and bilateral frontotemporal laminar (thickness less than 0.5 cm) subdural hematomas (Figure 2).

The diagnosis of spontaneous CSF leak was confirmed by neuroaxis MRI (enlargement of the posterior epidural space with thin liquid in the epidural fat, and apparent foci of discontinuity in the dura mater at the lower thoracic levels, notably on the left), and radionuclide cisternography (altered CSF dynamics, without progression of the radiotracer from the base cisterns to the cerebral convexity even after 24 hours, and thoracolumbar spontaneous dural fistulas between T8-T12 bilaterally, notably T8-T9 on the left). He was treated with the same clinical measures as Patient 1, with improvement, and needed an epidural blood patch. There was no need for hematoma drainage and the patient was discharged with improvement of symptoms.

Patient 3

A 35-year-old female patient, unemployed (she was a pharmacist), with a history of bariatric surgery, use of oral contraceptives and ex-smoker, presented at the emergency department with a two-week history of severe (8/10) holo-cranial headache (in the occipital region), with a progressive pattern and a clear correlation with the orthostatic position, which improved with lying down. It was associated with nausea, vomiting, photophobia, phonophobia, hypoacusis and was barely responsive to common analgesics.

An MRI was performed and showed signs of intracranial hypotension (engorgement of the pituitary, sagging of the brain

with herniation of the cerebellar tonsil, homogeneous and diffuse pachymeningeal enhancement, posterior fossa with reduced size, diffuse venous sinus engorgement, subdural effusions) and right transverse sinus thrombosis (Figure 3A). A CT angiography also confirmed the right transverse sinus thrombosis (Figure 3B).

The diagnosis of a CSF leak was made by cisternography, which showed a spontaneous dural leak at the cervical and thoracic levels, more evident between C7 and T1.

The patient's oral contraceptive was discontinued and she was discharged with anticoagulation and significant improvement of headache after clinical treatment (lying down, hyper-hydration and caffeine).

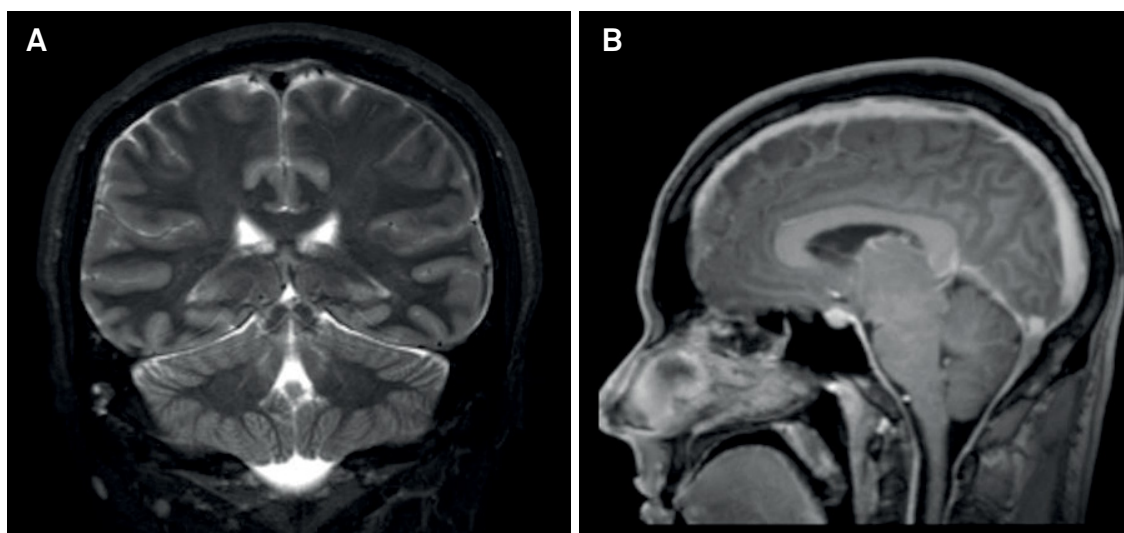


Figure 2. Coronal T2-weighted MR image (A) showing hypointense left frontotemporal laminar subdural hematoma; (B) Contrast-enhanced sagittal T1-weighted MR image showing diffuse pachymeningeal thickening and enhancement, enlargement of the pituitary gland and engorgement of the venous sinuses.

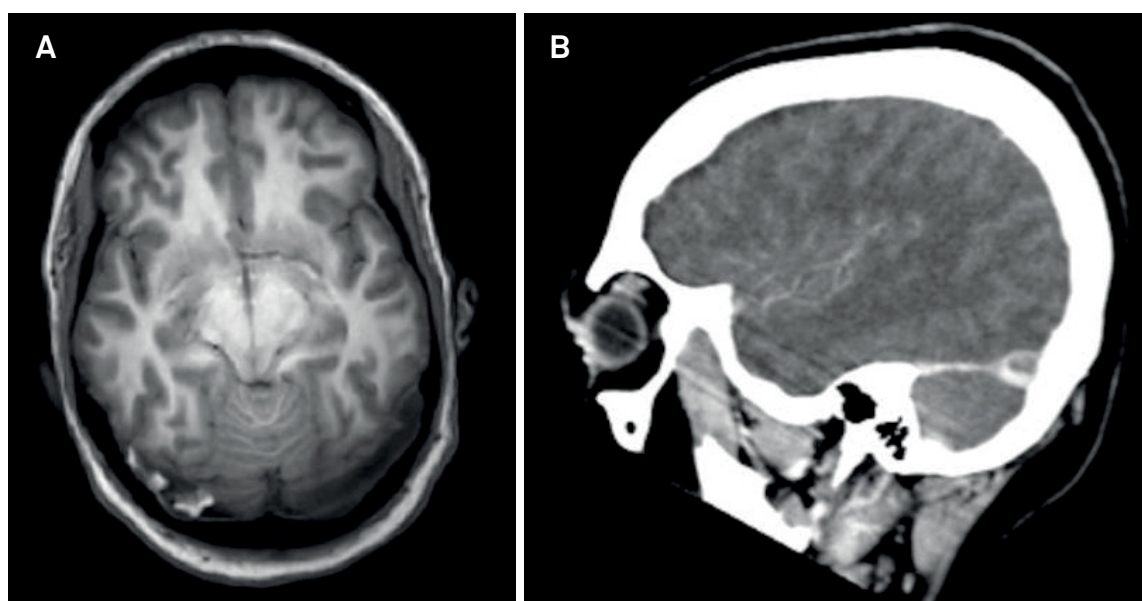


Figure 3. Axial T1-weighted MRI (A) demonstrating hyperintensity in the right transverse sinus; (B) sagittal CT angiography confirming the right transverse sinus thrombosis.

DISCUSSION

Headache is a common complaint with a wide range of differential diagnoses. Spontaneous intracranial hypotension is an uncommon cause, which remains underdiagnosed and should be considered in cases of orthostatic headache¹⁻⁵.

In this context, it is important to perform imaging of the brain, and that the radiologist suspect the diagnosis. Tomographic examination can be used for screening, especially in the emergency department, and ventricular collapse, descent of the cerebellar tonsils¹² and even pachymeningeal enhancement can be observed if an iodinated contrast is used. Hemorrhagic complications are easily detected in a CT and venous sinus thrombosis can also be seen in this method. However, in general, the accuracy of MRI is much higher than that of CT, and a complementary MRI should be performed in patients with a high clinical suspicion, even when a CT scan does not reveal major alterations. For example, small ventricles may not draw much attention in a young patient's CT, and pachymeningeal enhancement areas are not as well defined as they are in the MRI. When a CT raises suspicion of an SIH, it is also important to perform an MRI with gadolinium, which is better able to depict typical findings and complications. Some classic findings of SIHs seen in MRIs are diffuse pachymeningeal enhancement, decrease in the size of the ventricles, subdural fluid collections, enlargement of the pituitary, flattening of the pons against the clivus, cerebral venous sinus engorgement and sagging of the brain^{1,2,4,5,6,9,10,11}.

However, there are some pitfalls to avoid, such as the downward displacement of the cerebellar tonsils into the spinal canal, which may mimic a Chiari malformation (pseudo-Chiari malformation), subdural fluid collections mimicking a primary subdural hematoma, pachymeningeal enhancement that may be mistaken for an infectious or neoplastic disease, and pituitary hyperemia mimicking a pituitary tumor, such as an adenoma^{8,11,12}.

Complications associated with SIH are rare, but potentially serious, and early radiological diagnosis favors proper management. Complications described include subdural hematoma, cerebral venous and venous sinus thrombosis, bibrachial amyotrophy, superficial siderosis and syringomyelia.^{3,4,5} We have reviewed the literature on these conditions.

Subdural fluid collections in SIH range from simple thin hygromas to massive subdural hematomas. Subdural fluid collections are common in SIH, occurring in about 50% of patients. Mostly, they represent hygromas and are bilateral, thin, usually found over the cerebral convexities, and do not cause significant mass effect^{1,2,5,13,14}. However, subdural hematomas with varying degrees of mass effect (seen in Patient 1), sometimes shifting the midline, are not uncommon and have been described in about 20% of patients with SIH, this being the most common complication of this entity. Subdural hematomas are typically bilateral but asymmetrical and the maximal thickness usually ranges from 1 cm to 3 cm^{4,13}.

The decrease of CSF volume can lead to compensatory enlargement of the subdural/subarachnoid resulting in subdural hygromas; on the other hand, subdural hematomas are probably caused by bleeding from enlarged veins in the subdural zone or tearing of bridging veins^{13,14}.

It is important to point out that a careful anamnesis should include risk factors for hemorrhagic events, such as acetylsalicylic acid (seen in Patient 1) or anticoagulant use, or history of hemophilia, which, in addition to SIH, increase the risk of these complications.

The heterogeneous appearance and attenuation of the subdural hematoma in the imaging indicates that recurrent bleeding within the subdural space is common in untreated SIH. Studies on this entity have shown no significant differences in symptomatology, age, sex or location of the CSF leak among patients with or without subdural hematomas as a complication of SIH¹³.

Resolution of subdural hygromas (and also the typical MRI imaging features of SIH) can be demonstrated within days of successful treatment of the CSF leak. For subdural hematomas with mass effect, however, resolution occurs more slowly and may take up to a few months¹³.

The main treatment of the collection of subdural fluid is the management of the underlying CSF leak, almost always without the need for neurosurgical intervention. Besides, a craniotomy may increase sagging of the brain. If subdural hematomas are evacuated but the CSF leak is not treated, there is a high risk of persistent or recurrent subdural hematomas^{4,13}.

Cerebral venous thrombosis is found in about 2% of the population with SIH¹⁵.

The Virchow's triad comprises three categories of factors that contribute to venous thrombosis: venous stasis, hypercoagulability (hyperviscosity) and vessel damage^{16,17}. Spontaneous intracranial hypotension is a risk factor for cerebral venous thrombosis according to the following mechanisms that make up the triad: venous engorgement due to the loss of CSF (Monro-Kellie doctrine)¹⁸ results in the slowing of venous blood flow (venous stasis); the loss of CSF reduces absorption of CSF into the cerebral venous sinuses leading to an increase of blood viscosity (hypercoagulability); and sinking of the brain due to the loss of buoyancy may reflect in damage of cerebral veins and sinuses (vessel damage)^{15,19}.

As previously mentioned, a thorough anamnesis is very important, and should include other risk factors for thrombotic events, such as a previous history of thrombosis, use of contraceptives with estrogen (seen in Patient 3), pregnancy, puerperium, and thrombogenic disorders.

As concern imaging findings, a CT scan can show a spontaneous hyperdensity, or a T1-weighted sequence in MRI may reveal a hyperintensity in the sinus or vein location, which represents the thrombus. A CT or MRI angiograph may show the 'empty delta sign' (classically described for sagittal sinus thrombosis) at the confluence of the sinus, or a filling failure in the thrombosed sinus²⁰.

Central venous thrombosis is a serious complication, potentially life threatening, with a patient fatality rate of up to 5% and it may also complicate a cerebral venous infarction, which may occur in up to 40% of the patients, according to some studies. Successful treatment combines anticoagulation and the treatment of the spinal CSF leak^{15,19}.

Bibrachial amyotrophy is characterized by weakness and atrophy of a few contiguous cervical myotomes (segmental amyotrophy) and it is associated with an extra-arachnoid encapsulated fluid collection that causes chronic pressure on the ventral aspect of the cord. These extra-arachnoid fluid collections may be associated with a CSF leak. This may be mistaken for a motor neuron disease, and sensory symptoms are minimal or absent^{4,21}.

Superficial siderosis, in the context of SIH, is a result of hemosiderin deposition in the subpial layers of the brain

and spinal cord and may be caused by repeated hemorrhage into the subarachnoid space. This may happen due to bleeding from friable vessels of a dural defect or due to exudation from engorged vessels (frequently seen in CSF hypovolemia). Impaired hearing and ataxia may be seen as neurologic manifestations, especially when the superficial siderosis occurs in the posterior fossa and cranial nerves, and cerebellar involvement occurs^{4,21,22}.

Syringomyelia is a rare complication associated with significant downward displacement of the cerebellar tonsils. It may vary from very small to quite extensive⁴.

It is important that the radiologist identifies not only the classic imaging findings related to SIH, but also the possible associated complications, which often influence the clinical or surgical treatment of the patient.

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