

Spontaneous intracranial hypotension: diagnostic and therapeutic implications in neurosurgical practice

*Franzini Angelo, Messina Giuseppe, Mea
Eliana, Chiapparini Luisa & Bussone
Gennaro*

Neurological Sciences
Official Journal of the Italian
Neurological Society

ISSN 1590-1874

Neurol Sci
DOI 10.1007/s10072-011-0699-9



Your article is protected by copyright and all rights are held exclusively by Springer-Verlag. This e-offprint is for personal use only and shall not be self-archived in electronic repositories. If you wish to self-archive your work, please use the accepted author's version for posting to your own website or your institution's repository. You may further deposit the accepted author's version on a funder's repository at a funder's request, provided it is not made publicly available until 12 months after publication.

Spontaneous intracranial hypotension: diagnostic and therapeutic implications in neurosurgical practice

Franzini Angelo · Messina Giuseppe · Mea Eliana ·
Chiapparini Luisa · Bussone Gennaro

© Springer-Verlag 2011

Abstract Spontaneous intracranial hypotension syndrome (SIH) is a complex pathological entity due to reduction of cerebrospinal fluid (CSF) volume and results into a variegated symptomatology, ranging from orthostatic headache to more severe conditions, such as stupor and coma; several attempts have been done in the past to understand the exact pathophysiological mechanisms which underlie its phenomenology, as well as different therapeutic approaches to relieve symptoms and reverse the reduction of overall CSF volume, both focally (in the case of a clear localized CSF leak) or by reversing the pressure gradient between the venous system and the subdural and epidural compartments (in the case of idiopathic SIH). The Authors' experience is here reported, along with some pathophysiological and neuroradiological implications and considerations.

Keywords Intracranial hypotension · Spinal blood patch · Cerebrospinal fluid leak

Introduction

Spontaneous intracranial hypotension (SIH) is a clinical syndrome secondary to a reduction of the cerebrospinal

fluid (CSF) volume, mainly characterized by orthostatic headache associated with nausea, vomiting, diplopia, dizziness and tinnitus [1]. Brain Magnetic Resonance Imaging (MRI) with contrast medium is the study of choice for the diagnosis and during the follow-up of the syndrome.

SIH is often misinterpreted and imaging features may be confused with other neurologic conditions, the herniation of the cerebellar tonsils may be mistaken, for example, with a Chiari I malformation (CMI).

In this case, wrong diagnosis and loss of recognition of SIH may lead to a wrong and even dangerous surgical treatment with occipital decompression, worsening of the cerebellar descent and exacerbating of the intracranial hypotension and of the neurological symptoms.

The first treatment of SIH is bed rest and hydration. In a certain percentage (about 50% of hospitalized patients in our experience), the patients need a "blood-patch", that is the epidural injection of the patient's blood with or without fibrin glue mixed to a contrast medium. The goals of the epidural injection are to repair the CSF leak and/or to balance the negative epidural pressure and the pressure of the venous drainage in the epidural space [2, 3]. In SIH the rupture of the dura usually occurs at weak points that are present along the spinal root sleeves, where meningeal diverticula are demonstrated often in the presence of connective tissue disorders [4].

Our experience in 74 consecutive patients affected by SIH who underwent EBP and with long-term follow-up is reported and discussed.

Patients and methods

Between 2000 and 2010, 74 patients (mean age 42, range 22–60 years, 44 females) of the Neurological Headache

F. Angelo (✉) · M. Giuseppe
Department of Neurosurgery, Fondazione IRCCS Istituto
Nazionale Neurologico "Carlo Besta", Milan, Italy
e-mail: bsvjf@tin.it

M. Eliana · B. Gennaro
Department of Neurology, Fondazione IRCCS Istituto Nazionale
Neurologico "Carlo Besta", Milan, Italy

C. Luisa
Department of Neuroradiology, Fondazione IRCCS Istituto
Nazionale Neurologico "Carlo Besta", Milan, Italy

Unit of our Institute have been admitted to our Neurosurgical Unit to perform epidural blood patch. All these patients previously had no complete successful medical treatment for at least two months.

Clinical picture

In all patients the heralding and prominent symptom of the disease was headache and/or neck pain appearing or worsening within 15 min after sitting or standing. In addition to these symptoms, some patients complained nausea (19 patients), tinnitus (29 patients), hypoacusia (45 patients). Mild endocrine dysfunction related to ADH deficiency was present in two cases. The median time elapsed between symptom's onset and diagnosis was 4 months (from 15 days to 45 months).

Radiological findings

Characteristic brain MRI features of SIH are: diffuse thickening and linear enhancement of the pachymeninges along the convexities, the cerebral falx, the tentorium and the clivus (Fig. 1a), subdural fluid collections or hematomas (Fig. 1b, c), sagging of the brain with caudal diencephalic displacement, descent and enlargement of the pons, flattening of the peripontine cisterns and, in some case, downward tonsillar displacement (Fig. 2a, b, c). Additional findings may include pituitary enlargement and engorgement of the dural venous sinuses. Almost one of these findings was observed in all the patients of this series.

In most of the patients spinal MRI findings included: epidural fluid collections within the spinal canal and engorgement of the venous plexuses in the cervical

epidural anterior space. The search for CSF leak has been performed by MRI myelograms in 39 patients and by isotopic myelography in 14 cases. The CSF leak has been demonstrated in four patients within the cervical (2 patients), thoracic (1 patients) and lumbar (1 patients) spinal canal. No evidence of CSF leaks occurred in the remnant 35 patients.

In these series the search for the CSF leaks was mainly performed between 2000 and 2005, while in the last 5 years diagnostic work-up has gradually been modified and spinal studies have been fairly abandoned because of the adoption of a standardized blood patch procedure always performed within the lumbar region.

Epidural blood patch

The blood patch procedure has always been performed in local anaesthesia in prone position and assisted by fluoroscopy. A 16-gauge french Thuoy needle has been used for the localization of the epidural space trough the detection of the negative epidural pressure witnessed by the rapid aspiration of saline through a connected syringe in a closed system. The mixture injected within the lumbar epidural space (L1–L4) through the needle consisted of 6–8 cc of patient's venous blood, 1–3 cc of hydrosoluble contrast medium and 2–5 cc of fibrin glue. Fluoroscopy assisted and confirmed the correct epidural injection of the mixture (Fig. 3).

Results

After the first blood patch procedures, 65 (87%) patients obtained complete symptoms regression within 3 months.

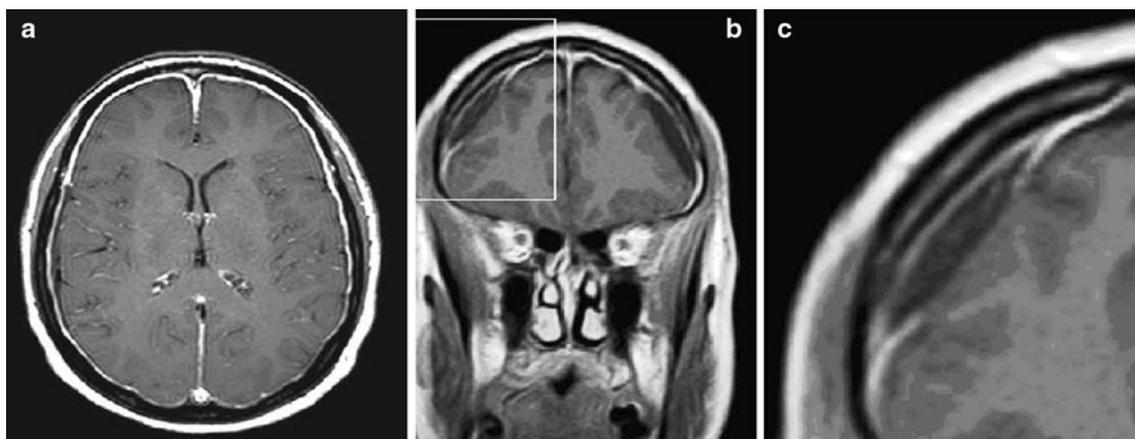


Fig. 1 a–c Axial T1-weighted image (*left*) after intravenous contrast medium demonstrate intense, linear and diffuse pachymeningeal enhancement along the cerebral convexities and the falx cerebri. Coronal T1-weighted image (*middle*) after contrast medium

demonstrated frontal bilateral subdural fluid collections and pachymeningeal enhancement. The magnified image of boxed area (*right*) shows that subdural hygroma is, actually, intradural

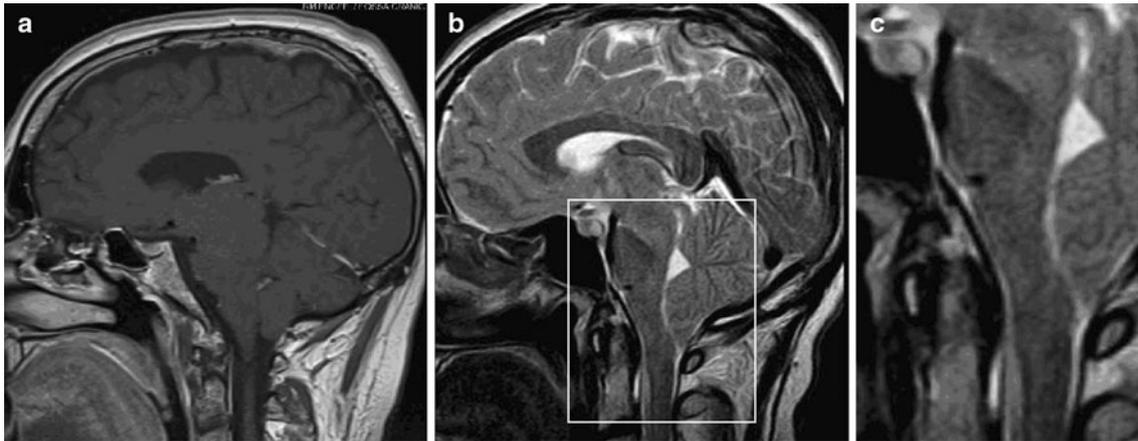


Fig. 2 a–c Sagittal T1-weighted image with contrast medium (*left*) and Sagittal T2-weighted image (*middle*) show severe brain sagging with deep brain swelling causing fairly complete obliteration of the perisellar cisterns and downward displacement and closure of the

interpeduncular cistern. The magnified image of boxed area (*right*), underlines tonsillar herniation and flattening of the pons against the clivus with obliteration of the pre-pontine cisterns



Fig. 3 Sagittal reconstructed lumbar CT performed immediately after blood patch demonstrate the compound in the posterior epidural space at the level of L1–L2, L2 and L2–L3. Note the compression and the anterior dislocation of the dural sac. A small amount of air is also visible in the epidural spaces

In nine patients (13%) first EPB was not effective and a second treatment has been performed: in three of these complete recovery was obtained, in six, recovery was not observed and they still complain the symptoms without the disappearance of neuroradiological findings. It has to be remarked that the clinical picture in these patients became similar to the chronic tensive headache syndrome without the postural changes typical of the SIH and pain was highly refractory to drugs.

Discussion and conclusion

Some topics from our experience in the reported series are remarked

- All patients at hospital admission were highly invalidated and after the blood patch procedures 87% returned to normal life in few weeks.
- None of our patients needed open surgery to repair CSF leak.
- The pathophysiological mechanism of the epidural lumbar blood patch is probably related to raise the spinal epidural pressure and to balance this pressure with the pressure of epidural veins. EPB was effective independently from severity of disease and the site of the CSF leak, so confirming that the negative epidural pressure plays a major role in the pathogenesis of SIH.

We support the hypotheses that the CSF leak is not the primary cause of the disease but the result of abnormally negative epidural pressure within the spinal canal, which acts as aspiration on the dural surface and particularly at the site of the radicular pockets where the meningeal sheaths are thinner. Standing and walking may contribute to the origin and maintenance of negative pressure within the spinal canal via the epidural venous drainage toward the inferior vena cava [2]. The presence of the fibrin glue within the blood patch mixture is motivated to reinforce the compression on the large epidural lumbar veins to limit the steal of venous blood from the epidural space by the inferior cava system.

In conclusion, we think that in case of postural headache or of the described radiological findings (subdural haematomas, descent of cerebellar tonsils, brain sagging), SIH

must be suspected and investigated by contrast enhanced MRI. Epidural lumbar blood patch is the procedure of choice when conservative treatments failed. Finally a long history of the disease and a late delayed diagnosis may lead to chronic headache syndromes.

Conflict of interest The authors declare that there is no actual or potential conflict of interest in relation to this article.

References

1. Mea E, Chiapparini L, Savoiaro M, Franzini A, Bussone G, Leone M (2009) Clinical features and outcomes in spontaneous intracranial hypotension: a survey of 90 consecutive patients. *Neurol Sci.* 30(Suppl 1):S11–S13
2. Franzini A, Messina G, Nazzi V, Mea E, Leone M, Chiapparini L, Broggi G, Bussone G (2010) Spontaneous intracranial hypotension syndrome: a novel speculative physiopathological hypothesis and a novel patch method in a series of 28 consecutive patients. *J Neurosurg* 112(2):300–306
3. Savoiaro M, Minati L, Farina L, De Simone T, Aquino D, Mea E, Filippini G, Bussone G, Chiapparini L (2007) Spontaneous intracranial hypotension with deep brain swelling. *Brain* 130(Pt 7):1884–1893
4. Schievink WI, Gordon OK, Tourje J (2004) Connective tissue disorders with spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension: a prospective study. *Neurosurgery* 54:65–70