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ORAL COMMUNICATION

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Thunderclap headache caused by spontaneous intracranial hypotension

Abstract We report a group of 4 patients with thunderclap headache as the initial manifestation of spontaneous intracranial hypotension.

Key words Thunderclap headache • Orthostatic headache • Spontaneous intracranial hypotension • Meningeal enhancement

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Introduction

An excruciating headache of instantaneous onset is known as a thunderclap headache (TH). A subarachnoid haemorrhage (SAH) is the prototypical cause, but other serious disorders may also present with a TH, including cerebral venous sinus thrombosis, carotid artery dissection and pituitary apoplexy. We report on 4 patients with TH as the initial manifestation of spontaneous intracranial hypotension (SIH).

Materials and methods

In the period 1992–2004, we observed 24 patients affected by SIH. One patient was affected by Marfan's syndrome. The diagnosis of SIH was confirmed by brain magnetic resonance imaging (MRI) in all patients.

Results

Among the group of 24 patients, four (16%) had experienced an excruciating headache of instantaneous onset (Table 1). The mean age of these three women and 1 man was 31 years (range, 24–45 years). Excruciating pain duration range was 10 seconds to a few minutes. The pain suffered was described as head swelling or as a hard stroke on the head, followed by gravative occipito-nuchal and frontal orthostatic headache. The delay between the onset of headache and diagnosis of SIH ranged from 9 days to 1 month. Mild neck stiffness was present in 2 patients. SIH was clearly demonstrated on brain MRI (Fig 1). Spinal MRI and MRI myelography (3 patients) did not show cerebrospinal fluid (CSF) leak level. Radioisotope cisternography (RC) (2 patients) showed indirect signs of CSF leak. Only in 1 patient was CSF leak demonstrated with CT myelogram at cervical level (Fig. 2). Opening CSF pressure was low (3 patients). CT scan, cere-

Table 1 Characteristics of four patients with TH and SIH

Patient no.	Age, years/sex	Headache	Associated symptoms	Neurological examination findings	MRI findings	Lumbar puncture findings	Location of CSF leak
1 RM	43/F	Acute, severe, occipital, nuchal	Nausea, vomiting, diplopia	Normal	Meningeal enhancement, brain sagging	OP: unmeasurable	Unidentified
2 DR	26/F	Acute, severe, occipital	Nausea, diplopia	Mild nuchal rigidity	Meningeal enhancement, subdural collection	OP: unmeasurable	Unidentified
3 SR	25/M	Acute, severe, generalized	Nausea, vomiting	Normal	Meningeal enhancement	Not performed (patient treated with warfarin)	Probable at sacral meningeal diverticula
4 MP	31/F	Acute, severe, occipital, nuchal	Nausea, vomiting	Mild nuchal rigidity	Meningeal enhancement	OP: unmeasurable	Cervical

CSF, cerebrospinal fluid; *MRI*, magnetic resonance imaging; *OP*, opening pressure

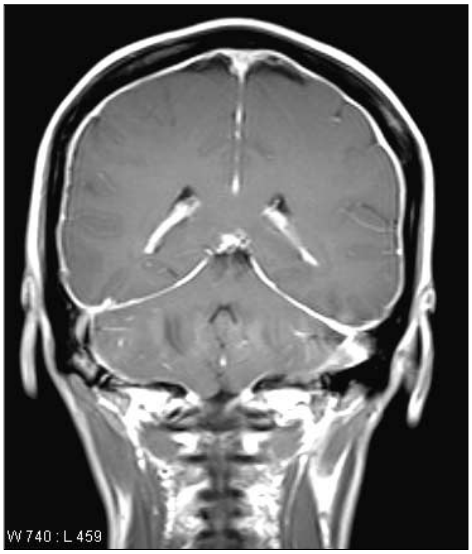

Fig. 1 Patient 4. Coronal T1-weighted gadolinium-enhanced MRI shows diffuse pachymeningeal enhancement

bral angiography (1 patient) and MRI angiography (1 patient) were normal.

Three patients received supportive measures only (bed rest, analgesic and hydration); one patient with cervical CSF leak underwent epidural blood patch.

Discussion

SIH is increasingly recognised as a cause of postural headaches [1]. The diagnosis is made by lumbar puncture revealing low opening CSF pressure, diffuse pachymeningeal enhancement or brain sagging on MRI, or the demonstration of a spinal CSF leak on CT myelography, RC, or spinal MRI or MRI myelography.

Treatment may include bed rest, epidural blood patching or surgical repair of the CSF leak [2].

SIH was initially mistaken for an SAH in these patients. Headache was severe and instantaneous in onset

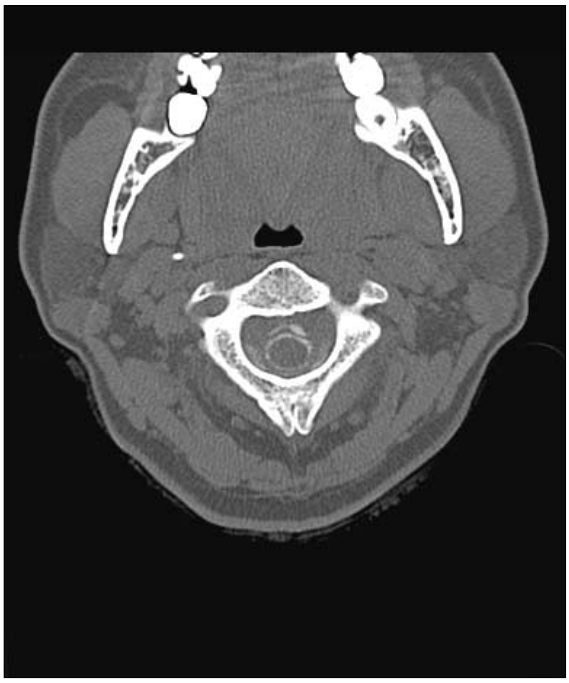

Fig. 2 Patient 4. CT myelogram of the cervical spine shows extrathecal contrast

but nuchal rigidity was found in 2 patients. Meningeal irritation is a cardinal feature common to both SIH and SAH. Other clinical features include the common occurrence of physical exertion preceding the onset of headache, such as sexual or sporting activities, and an association with generalised connective tissue disorders [3].

Additional potential pitfalls in patients with SIH are related to the frequent occurrence of cranial nerves dysfunction.

To further complicate the diagnosis, CSF examination in SIH sometimes may reveal xanthochromia. In addition, because of the very low opening pressure, a traumatic bloody tap is common in patients with SIH. Careful measurement of opening pressure is useful for diagnosis.

The instantaneous onset of headache in the patients described here is likely to have occurred at the time that the leak first developed, probably because of a great CSF leak through spinal dural hole.

We suggest that SIH should be included in the differential diagnosis of TH even when meningismus is present.

References

1. Mokri B (1999) Spontaneous cerebrospinal fluid leak, from intracranial hypotension to cerebrospinal fluid hypovolemia: evolution of a concept. Mayo Clin Proc 74:1113–1123
2. Schievink WI, Meyer FB, Atkinson JLD, Mokri B (1996) Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. J Neurosurg 84:598–605
3. Ferrante E, Citterio A, Savino A, Santalucia P (2003) Postural headache in a patient with Marfan's syndrome. Cephalalgia 23:552–555