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Headache in intracranial hypotension

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Abstract We describe the headache characteristics of two patients with intracranial hypotension (IH) and correlate the magnetic resonance imaging (MRI) pattern to the clinical aspects of this type of headache. The first case was that of a patient with spontaneous IH, whereas the second patient had IH following rhinorrhoea. Cerebral computed tomography (CT) performed at presentation of symptoms was normal in both patients. Cerebral MRI in the

first case showed meningeal contrast enhancement and an MRI pattern consistent with a subdural hygroma, while in the second case there was only a mild meningeal thickening. The symptoms improved spontaneously in both cases in about 2–3 months, confirming that invasive manoeuvres are not mandatory in these patients.

Key words Intracranial hypotension • Postural headache • MRI • CSF

Introduction

Headache is one of the most common symptoms observed in patients with abnormal intracranial pressure. Modification of intracranial pressure may be due to variations in flow, production or absorption of cerebrospinal fluid (CSF) [1].

Intracranial hypertension may be caused by increased CSF production, decreased CSF absorption, increased venous pressure, obstruction of normal CSF flow, mass lesion, cerebral oedema, and increased pressure in dura.

Common causes of secondary intracranial hypotension (IH) are lumbar puncture head and spine trauma, neurosurgery, spontaneous or post-traumatic rhinorrhoea, and pituitary tumour. In contrast, spontaneous IH is by definition not preceded by any event and therefore its interpretation and diagnosis are more difficult. In the past, it was thought that spontaneous IH was due to a low production of CSF, but since the 1990s the most common theories regard an increased absorption of CSF or CSF leakage due to dural

tearing. Headache is present in almost all patients with IH, but in 30%–80% of patients with intracranial hypertension. This difference may be explained by the lack of direct correlation between the degree of pressure elevation and the presence of headache, and by the variable effect on pain-sensitive structures [2].

Patients with IH complain of pain in the frontal and occipital regions or of diffuse pain. The pain worsens with standing and improves with lying down, i.e. it is typically orthostatic. The pain is severe, dull or throbbing; it is not relieved by analgesics and it worsens with the Valsalva manoeuvre. Accompanying symptoms are nausea, vomiting, cervical or interscapular pain, tinnitus, diplopia, photophobia, visual field defects or radicular symptoms in the upper limbs. Clinical examination is generally normal, although mild neck stiffness and slow pulse rate may be observed. CSF pressure is generally low (0–60 mmH₂O), but in a few cases it is normal, in particular after recumbency. CSF analysis may show a mild increase in protein concentration or in cell number, with mild xanthochromia [3, 4].

Magnetic resonance imaging (MRI) reveals diffuse meningeal thickening, gadolinium enhancement, and subdural fluid collections (hygromas and rarely haematomas), at times associated with diminished size of the subarachnoidal cisterns, with the descent of the cerebellar tonsils or with flattening of the optic chiasm [4–9]. In such cases, differential diagnosis with systemic diseases like rheumatoid arthritis, mucopolysaccharidosis, sarcoidosis and meningeal carcinomatosis must be performed [10–12].

In cases of suspected spontaneous IH, if the site of CSF leakage is not found on MRI, isotope cisternography and computed tomography (CT) or MRI myelography may be performed. The latter two tests may reveal single or multiple meningeal diverticula at cervical or thoracic spinal level, which may lead to dural tearing [4–8]. Still, the correct localisation of dural tearing may not be shown because of the limits of the current imaging techniques. In some cases, invasive tests are not necessary due to spontaneous recovery of symptoms [13].

We describe two patients with IH: the first with primary IH and the second following rhinorrhoea. Headache characteristics, the spontaneous clinical recovery and the improvement of neuroradiology patterns are described.

Case reports

Case 1

A 40-year-old woman was admitted in the Neurology Ward of the Regional Hospital of Aosta, Italy, in January 2001 because of persistent headache that was diffuse and dull. There were no main neurovegetative signs, but intermittent

pain in the right upper limb was reported. Medical history was positive only for menstrual migraine. Clinical and neurological examination were normal, as were blood tests. No pathological findings were seen at brain CT. During admission, we observed that headache worsened in erect position and during Valsalva manoeuvres, whereas it improved in recumbency.

Three weeks after onset of symptoms, the patient underwent brain MRI that showed mild meningeal thickening, in particular over the convexity, associated with subdural fluid collection, consistent with hygroma. Gadolinium administration showed diffuse meningeal enhancement in supratentorial and infratentorial regions (Fig. 1).

The clinical and neuroradiological findings were consistent with IH syndrome. However, lumbar puncture, rheumatic and autoimmune tests, paraneoplastic marker analysis, serum levels of angiotensin-converting enzyme, thoracic radiography and an abdominal ultrasound were performed to rule out a diffuse inflammatory disease or a tumoral process.

The patient underwent a second MRI examination brain and of the spine (cervical and thoracic) forty days after the first one. There was an improvement of the pattern, with partial absorption of the subdural hygroma and reduction of the meningeal thickening and contrast enhancement. Meningeal thickening and contrast enhancement were present in the cervical spine and down to T2 (Fig. 2). The improvement of the neuroradiological pattern was accompanied by the regression of symptoms with complete spontaneous remission over a 3-month period. No treatment was offered, but rest.

The last MRI examination was performed at 4 months after the first one and showed complete absorption of the subdural hygroma and resolution of the abnormal meningeal thickening and contrast enhancement (Fig. 3).

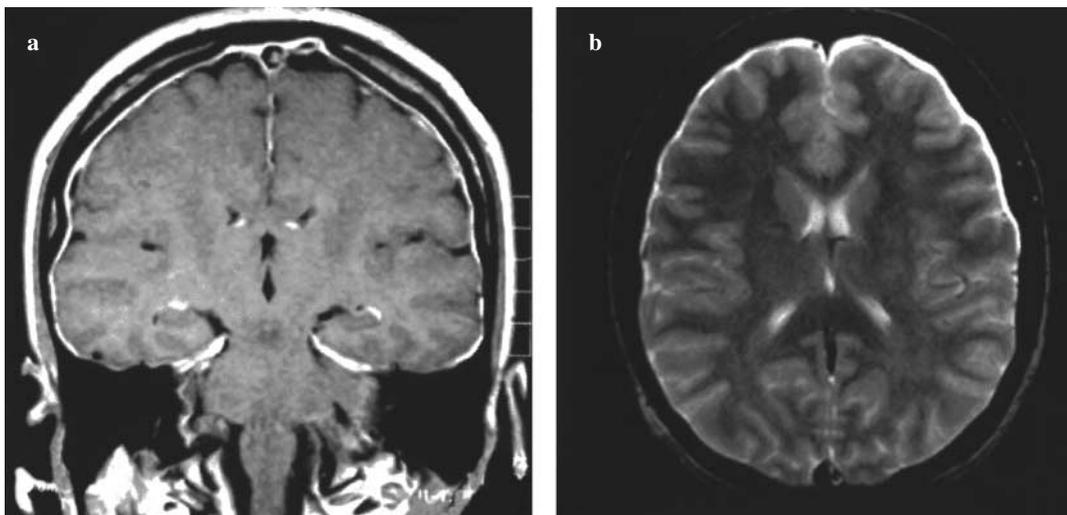


Fig. 1a,b Brain MRI performed three weeks after onset of symptoms. **a** T1-weighted coronal image after administration of gadolinium. **b** T2-weighted axial image

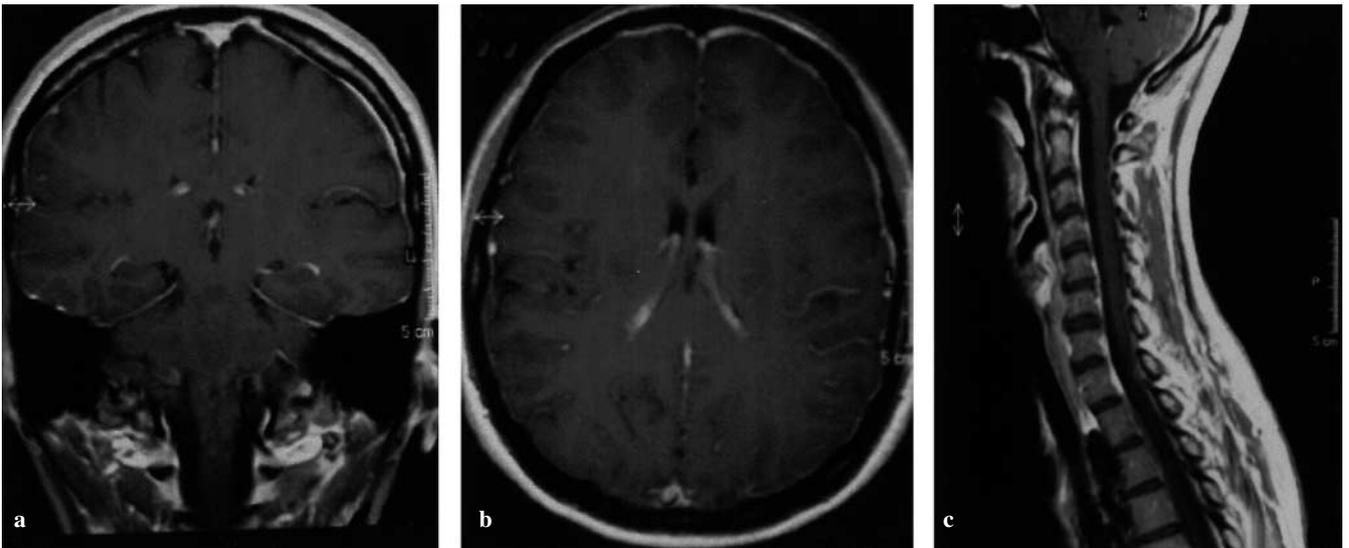


Fig. 2a-c Brain and cervical spinal cord MRI performed forty days after the first MRI examination. **a** Brain T1-weighted coronal image after administration of gadolinium. **b** Brain T1-weighted axial image after administration of gadolinium. **c** T1-weighted sagittal image of cervical spinal cord

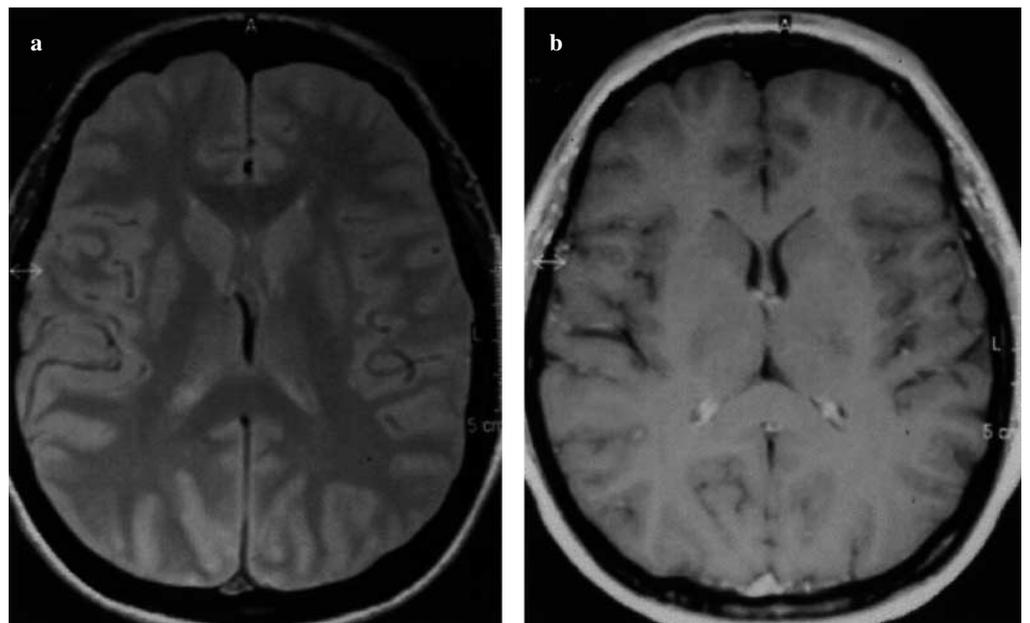


Fig. 3a,b Brain MRI performed four months after the first examination. **a** T1-weighted axial image. **b** T1-weighted axial image after administration of gadolinium

Case 2

In July 2002, a 45-year-old woman with negative medical history had an episode of throat infection. She consulted her general practitioner because a few hours after a violent sneeze she experienced rhinitis with experienced loss of liquid material and headache. Her physician suggested bed rest and antibiotics, however, two days later the patient complained of severe postural headache associated with mild cervical pain. Brain CT performed in the hospital's emergency department was normal and no treatment was offered.

In August 2002, because of the persistency of the symptomatology, she consulted a neurologist. Clinical examination was normal and brain MRI showed mild meningeal thickening over the convexity. This finding, associated with the persistent loss of liquid material and postural headache, was consistent with a diagnosis of IH. Because of the clinical history, we performed CT of the lamina cribrosa and sphenoid bone and an ear-nose-throat (ENT) examination. Both were normal. Two months after onset, there was complete remission of symptoms, without treatment except for rest.

Discussion

In 1938, Schaltenbrand described intracranial hypotension (IH) for the first time, applying the term “spontaneous aliquorrhea”. Clinical presentation was similar to a post-lumbar puncture syndrome [14]. It is known that after lumbar puncture, patients may complain of postural headache, possibly due to CSF leakage through the dura caused by the needle. Postural headache appears when the total volume of CSF is reduced by more than 10%. This is why some authors describe IH as a syndrome due to a reduction of volume rather than of CSF pressure [15, 16]. According to this theory, a volume reduction permits descent of the brain with traction on its pain-sensitive, suspending structures, causing headache especially in the upright position [17]. In the majority of cases of spontaneous IH, the headache has an acute or subacute onset. Therefore, although reduced production or hyperabsorption of CSF may be considered in the pathogenesis of IH, the acute onset of symptoms suggests that spontaneous dural tearing is the most frequent cause

[13]. MRI findings confirm that spontaneous CSF leakage is the main cause of primary IH. It is thought that even minimal but diffuse tearing of the deep layer of the dura may result in a pattern of thickening at MRI. When the tearing is more severe, the MRI pattern may suggest a haematoma or subdural hygroma [13].

In the first case described in this paper, MRI did not show meningeal diverticula. More invasive techniques were not performed because of the improvement in symptoms.

Hydration, administration of caffeine or high doses of steroids and, if these fail, injection of epidural blood patch or saline solution to stop the CSF leakage have been proposed as possible treatments [17, 18]. However, the majority of patients described in the literature recover spontaneously with rest.

The present cases are consistent with those already described in the literature: both showed spontaneous recovery of symptoms and contemporaneous regression of neuro-radiological abnormalities, possibly due to spontaneous repair of the dura.

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