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Headache and intracranial hypotension: neuroradiological findings

Abstract The cardinal and classic features of postural headache and low cerebrospinal fluid (CSF) pressure in intracranial hypotension may not dominate the clinical picture of the syndrome and may be associated with additional various neurological symptoms and signs. Reports of unusual clinical presentations continue to appear in the literature. Despite the considerable variability of the clinical spectrum, neuroradiological studies reveal more constant and characteristic features. Brain MRI findings include intracranial pachymeningeal thickening and post-contrast enhancement, subdural fluid collections and downward displacement or "sagging" of the brain. Spinal MRI findings include collapse of the dural sac with a festooned appearance, intense epidural enhancement owing to dilatation of the epidural venous plexus, and possible epidural fluid collections. In fact, spinal studies may demonstrate CSF leakage from spinal dural defects, which are considered the most common cause of the syndrome. Myelo-MR may suggest the possible point of CSF leakage, by demonstrating an irregular root sleeve; myelo-CT and radioisotope myelocisternography (RMC) are often needed to confirm the point of CSF leakage. Neuroimaging studies are, therefore, essential for suggesting and confirming the diagnosis.

Key words Intracranial hypotension • Brain sagging • Dural enhancement • Spinal epidural plexus dilatation

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Introduction

Intracranial hypotension typically presents with postural headache, often associated with one or more of the following symptoms: nausea, vomiting, dizziness, diplopia, photophobia, hearing impairment, neck stiffness and blurred vision [1]. It is the result of low cerebrospinal fluid (CSF) volume caused by either spontaneous or post-traumatic dural laceration. Iatrogenic causes include lumbar puncture or overdraining ventricular or spinal shunts. Spontaneous CSF leaks are uncommon and may manifest themselves as obvious CSF leaks, such as rhinorrhoea in spontaneous CSF fistulas at the level of the skull base, or cause a more insidious syndrome commonly known as spontaneous intracranial hypotension (SIH) [2].

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 disorder [3]. In several cases of SIH, however, CSF leakage is not demonstrated.

The syndrome usually has a benign course; however, cases with stupor or coma caused by diencephalic compression on the skull base or by large subdural haematomas have been reported [4]. Many patients improve spontaneously, some patients require epidural blood patch (EBP); in only well-selected cases is surgical repair necessary [5]. In these patients, precise localisation of the site of the CSF leak is essential.

Clinical considerations

A considerable variability exists in clinical manifestations. In some cases, the headache is not postural and not the dominant symptom; a few patients may have no headache at all [1]. Even the cardinal diagnostic proof of the syndrome, i.e., low CSF pressure at lumbar puncture, may be absent at the time of measurement [6].

Neuroradiological examinations

In SIH, the most informative neuroradiological examination is brain MRI without and with contrast medium administration [7]. The imaging features of SIH may be confused with other neurologic conditions. The herniation of the cerebellar tonsils may be mistaken for a Chiari I malformation [8]. Meningeal, dural enhancement may be confused with meningeal carcinomatosis, which, however, involves the leptomeninges. Infective-inflammatory or granulomatous diseases should also be considered; hypertrophic pachymeningitis may be indistinguishable from SIH but usually has a more focal involvement of the dura with greater thickening [5]. Enlargement of the pituitary gland due to hyperaemia may mimic a pituitary tumour. Enlargement of the spinal epidural venous plexus may be misinterpreted with neoplastic or inflammatory disease, or with engorgement due to a discharging arteriovenous fistula [9].

Once the diagnosis of SIH is established, studies of the spine should be performed to demonstrate the site of the CSF leakage. These studies include: spinal MRI, myelo-MRI, myelo-CT and radioisotope myelo-cisternography (RMC).

Brain CT

It may demonstrate small or slit ventricles, indiscernible basal cisterns, narrowing of the sylvian fissures and subdural collections.

Brain MRI

Brain MRI performed without and with contrast medium reveals the characteristic findings related to the CSF depletion. They include: diffuse dural thickening and enhancement, subdural fluid collections or haematomas and a sagging brain with midbrain caudal displacement, decrease in height of the perichiasmatic cisterns, inferior displacement of the optic chiasm and hypothalamus, which may kink on the dorsum sellae, descent and enlargement of the pons with flattening of the peripontine cisterns, and downward tonsillar displacement (Fig. 1a). Additional findings include: decrease in size of the ventricles, pituitary enlargement and engorgement of the dural venous sinuses [1, 7, 10].

The intracranial post-contrast examination demonstrates diffuse involvement of the thickened dura along both cerebral convexities, the falx, the tentorium and, often, the clivus [10] (Fig. 1b). There is no abnormal leptomeningeal

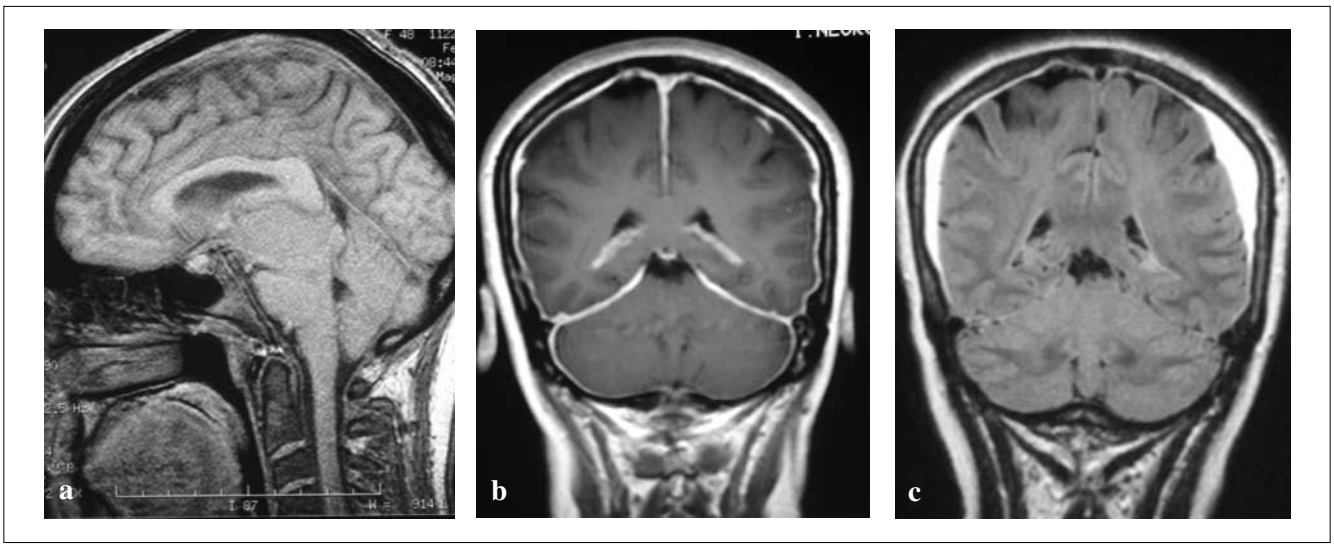

Fig. 1 Mid-line sagittal T1-weighted image (**a**) demonstrates caudal displacement of the pons, effacement of the pre-pontine cistern and decrease in height of the perichiasmatic and interpeduncolar cisterns. Caudal displacement of the tonsils is also noted. Contrast-enhanced coronal T1-weighted image (**b**) in a different patient shows diffuse intense enhancement and thickening of the dura of the convexities and tentorium. Coronal FLAIR image (**c**) in a third patient shows bilateral subdural fluid collections

enhancement on the surface of the brainstem or in the depth of the cortical sulci. In rare cases, no meningeal abnormalities are seen on MRI.

The diffuse and intense pachymeningeal enhancement may result either from thickening of the dura and tearing of small, dilated intradural blood vessels owing to the traction from the sagging brain, or from vasodilatation of the meningeal vessels in an attempt to compensate for the low CSF volume, in agreement with to the Monro-Kellie hypothesis. The dural hyperaemia also explains the engorgement of the cerebral venous sinuses and the pituitary enlargement [10].

The subdural fluid collections may be bilateral or unilateral and are located over the cerebral convexity (Fig. 1c) [1, 7]. They are often thin, without mass effect and often isointense or hyperintense with respect to the CSF in T1- and T2-weighted images, depending upon presence of blood, time from bleeding and protein concentration. Subdural haematomas that compress and shift the brain may sometimes form because of rupture of the bridging veins [7].

The anatomic landmarks to measure, on midsagittal images, the downward displacement of the brain, which is a very specific finding in SIH, include: (1) the position of the opening of the aqueduct of Sylvius (iter) relative to the incisural line (a straight line drawn from the tuberculum sellae to the point of confluence of the vein of Galen into the straight sinus); (2) the position of the cerebellar tonsils relative to the Chamberlain's line (the line drawn from the hard palate to the posterior lip of the foramen magnum), and (3) the position of the fastigium of the fourth ventricle relative to Twining's line (from the tuberculum sellae to the internal occipital protuberance). The normal position of the iter is –0.2±0.8 mm below the incisural line. Significant displacement is a measurement greater than two standard deviations: for the iter, 1.8 mm below the incisural line. A fourth measurement is also used: the normal tonsillar position is –0.1±2.1 mm below the foramen magnum line (i.e., the line drawn from the inferior tip of the clivus to the posterior tip of the foramen magnum); significant displacement is >4.3 mm below the foramen magnum line [7]. Descent of the iter and cerebellar tonsils may be seen singularly or coexist. Downward displacement of the iter is usually more marked than tonsillar herniation.

Spinal studies

Spinal neuroimaging is performed to determine the exact location of the CSF leak when targeted EBP or surgical repair of the dural laceration are considered.

Spinal MRI studies may reveal 3 types of characteristic findings: (1) epidural fluid collections, (2) collapse of the dural sac accompanied by engorgement of the epidural venous plexus, and (3) abnormalities of the root sleeves [9].

The epidural collections usually extend along several spinal levels and, probably, never reveal the exact site of the CSF leak. They represent the epidural accumulation of the CSF leaking out of the subarachnoid space. On axial images, collapse of the spinal dural sac assumes a festooned appearance due to the points of adhesion or anchoring of the spinal dura mater to the posterior longitudinal ligament and to the nerve roots [9]. Axial and coronal T2-weighted images may demonstrate extradural fluid extravasation in the paraspinal soft tissue pointing to the site of the leak.

Myelo-MR reveals meningeal diverticula or Tarlov's cysts that may represent the site of the leak (Fig. 2).

If no suggestions about the point of leakage are obtained by MR and myelo-MR, RMC should be performed. It may

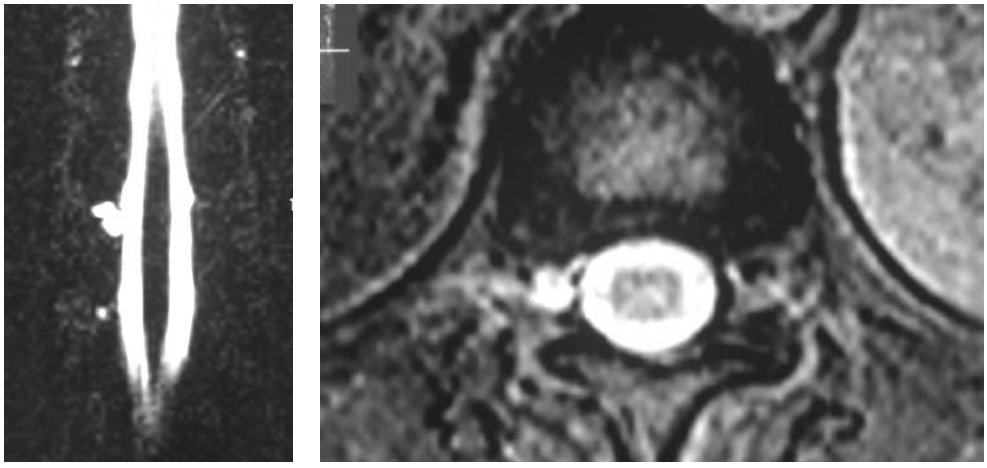

Fig. 2 MR-myelography and axial T2-weighted images at lower dorsal level demonstrate an irregularly dilatated root sleeve on the right, suggesting the probable point of CFS leakage

demonstrate accumulation of radioactivity outside the subarachnoid space suggesting the approximate site of the leak.

If MRI or RMC strongly suggests the point of CSF leakage, a confirmation may be obtained by myelo-CT, which gives fine details and has a better localising power than RMC [9]. We consider myelo-CT necessary when a direct surgical approach is planned, whereas the less precise localisation of the leak provided by RMC (and sometimes even by MRI) may be sufficient when EBP is considered.

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