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Mini-Review

Headache and neuroimaging findings in conditions of cerebrospinal fluid (CSF) circulation disorders: in hydrocephalus, *pseudotumor cerebri*, and CSF hypotension syndrome

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Abstract

The authors wish in this narrative minireview show and comment on some neuroimaging findings encountered in patients with conditions of cerebrospinal fluid (CSF) circulation disorders, such as in the hydrocephalus, pseudotumor cerebri, and CSF hypotension syndrome. The MRI of a young woman with a clinical diagnosis of post-dural puncture headache, performed on the fourth postpartum day after cesarean delivery, evolving with headache and diplopia, is shown. Non-contrast-enhanced sagittal T1 magnetic resonance imaging shows that the cerebellar tonsils are at the level of the foramen magnum, therefore still within normal limits, but, despite that, the opening of the cerebral aqueduct in the third ventricle is below the imaginary line connecting the anterior clinoid to the vein of Galen, therefore considered abnormally lower than the expected anatomical position. The axial T1-weighted images with post-contrast fat suppression also show impregnation and thickening of the dura mater. There is also mild engorgement of the cerebral venous sinuses, best demonstrated on T1 with post-contrast fat suppression, which is also identified on post-contrast magnetic resonance angiography, with no signs of venous thrombosis. We conclude that the diagnosis of a patient with intracranial hypotension syndrome can be suspected or confirmed with typical neuroimaging findings.

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Introduction

In the intracranial cavity we find three distinct compartments: the brain parenchyma (≅1,300 ml), blood (≅120 ml), and cerebrospinal fluid (CSF, ≅120 ml) in normal physiological conditions. According to the Monro-Kellie doctrine, the sum of the volumes of these three compartments is constant.[1-5] The production of CSF is about 0.35 ml/min (≅500 ml/day), and there is dynamic blood in and outflow (approximately 700 ml/min) regarding the intracranial space.[6] The only real constant is the brain parenchyma volume, which is inelastic. Recent studies have observed changes in these compartments in abnormal situations.[6-10]

During the cardiac cycle (systole-diastole), the volume of the other two compartments varies, with an inverse relationship between intracranial blood volumes and intracranial CSF volume. A certain amount of CSF has to exit at each systole via the foramen magnum to the dural sac to allow arterial blood to enter the cranial cavity; without it, there would be no space for more blood inside the cranial cavity.

The authors wish in this narrative minireview show and comment on some neuroimaging findings encountered in patients with conditions of circulation disorders, such as in the hydrocephalus, *pseudotumor cerebri*, and CSF hypotension syndrome.

Hydrocephalus

Patients with hydrocephalus often complain of headache. The headache pattern will depend on several variables, such as intracranial hypertension or hypotension, type of hydrocephalus, associated conditions (*e.g.*, tumor, infection, trauma), age, the position of the head in relation to the body, among other factors.[11-14]

Hydrocephalus can be defined as CSF accumulation within the ventricular cavities. It can have several causes such as inflammation, neoplasms, congenital malformations, and consequent obstruction of CSF outflow pathways in the ventricles or subarachnoid space, leading, in most cases, to increased intracranial pressure.

This increased intracranial pressure causes headache, vomiting, lowered consciousness, seizures, coma, and even death if left untreated. Differential diagnosis of the cause of the pain can be difficult in patients with hydrocephalus. However, patients with hydrocephalus treated either by a shunt (ventriculoperitoneal shunt) or third ventriculostomy may have headache of another etiology. The possibility of shunt dysfunction has to be considered, but migraine may be present in a patient with hydrocephalus, especially if there is a strong family history of migraine.

An intriguing form of hydrocephalus when the patient has a headache is the hydrocephalus associated with a colloid cyst of the third ventricle.[15] Although the association is rare, one of the authors (MMV) saw a few patients with headache complain who went to his office. The neuroimage investigation displayed a colloid cyst of the third ventricle without hydrocephalus or associated with a unilateral dilatation of the lateral ventricle, and the fundus examination was normal. Is the colloid cyst of the third ventricle an incidentaloma in these patients, and is the headache of the primary type (e.g., migraine)?

Classically, third ventricle colloid cyst headache is associated with the development of acute hydrocephalus caused by a change in head position. In these cases, neurosurgeons interpret the absence of hydrocephalus in the image as a regression of hydrocephalus because the cyst has moved, clearing the Monro's orifice. They almost always indicate an emergency surgery for fear of having intermittent hydrocephalus.

One of the authors (MMV) recently became aware of a case of a woman who had continuous pain in the right frontal region, and neuroimaging showed the presence of a colloid cyst in the third ventricle. This patient was immediately operated on for the cyst. Days later, vesicles appeared in the territory of the trigeminal branch. The pain was due to ophthalmic herpes zoster.

Pseudotumor Cerebri

Idiopathic intracranial hypertension, also known as *pseudotumor cerebri*, is an idiopathic syndrome in which increased intracranial pressure is observed without hydrocephalus or associated expansive lesion. It most often affects middle-aged obese women.[16-18] Other associations have been described with the syndrome, such as some use of specific drugs, endocrinopathies, and nutritional disturbances.[18] The classic clinical presentation is headache associated with visual loss, photophobia, and tinnitus.[19-22]

Some conditions associated with the development of idiopathic intracranial hypertension, such as vitamin A intake[23-25], are described.[26]

The most frequent radiological findings are ectasia and tortuosity of the cerebrospinal fluid sheaths of the optic nerves, papilledema, cerebrospinal fluid insinuation into the sellar cavity with compression of the pituitary parenchyma (partially empty *sella*), enlargement of Meckel's cavities, and stenosis of the lateral portions of the transverse sinuses.[27-34]

Cerebrospinal Fluid Hypotension

Cerebrospinal fluid hypotension is characterized by orthostatic headache associated with low CSF pressure, usually resulting from CSF leakage due to lumbar punctures, trauma, great physical exertion, severe dehydration, or spontaneous tear of the dura mater.[35] Peak incidence is among people between 30-40 years of age.[35]

The patient characteristically presents with severe headache (which may be orthostatic, pulsatile, with or without neck stiffness), abducens nerve palsy, and visual disturbances.[35] The headache is self-limiting because the dura mater has self-repairing properties, as observed in experiments with human dura mater from adult cadavers.[36] Most cases resolve spontaneously in one week.[35]

Typical imaging findings are[2, 37-38]:

- Smooth enhancement of the pachymeninge (dura mater);
- Engorgement of venous structures (dural venous sinuses, cerebral veins, epidural venous plexuses);
- Pituitary swelling;
- Subdural hematomas and effusions;
- Herniation of structures (tonsils);
- Rectification of the anterior surface of the pons;
- Point of communication between the 3rd ventricle and the aqueduct of Sylvius below the line between the anterior clinoid process and the vein of Galen, and
- Reduction of the interpeduncular cistern of the pons.

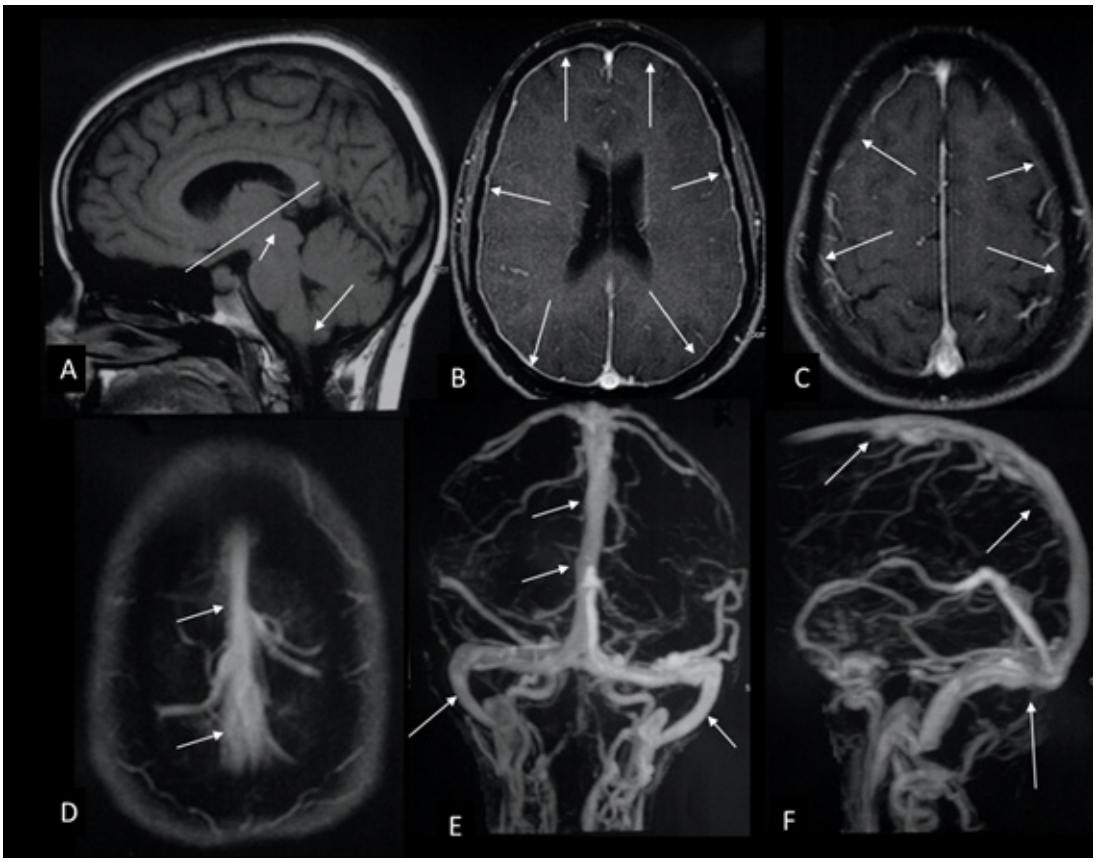

Figure 1. A 29-year-old woman on the fourth postpartum day after cesarean delivery is evolving with headache and diplopia in the right eye. Non-contrast-enhanced sagittal T1 magnetic resonance imaging (A) shows that the cerebellar tonsils are at the level of the foramen magnum (arrows) therefore still within normal limits (there is no previous MRI to compare), but that the opening of the cerebral aqueduct in the third ventricle is below the imaginary line connecting the anterior clinoid to the vein of Galen, therefore lowered (short arrow). The axial T1-weighted images with post-contrast fat suppression (A and B) show impregnation and thickening of the dura mater (arrows). There is also mild engorgement of the cerebral venous sinuses, best demonstrated on T1 with post-contrast fat suppression (arrows, D), which is also identified on post-contrast magnetic resonance angiography (E and F), with no signs of venous thrombosis. This case was courtesy of Dr. Jader Aretakis Cordeiro Filho, Recife, Brazil.

Comment

A neuroimaging investigation of the patient, particularly those individuals with red flags such as postpartum headache[39], may reveal the cause of the headache. In the example of postpartum headache, we have to think of several possibilities.[40-42] The first is to know whether it is not just another migraine attack in patients who have been migraineur for years, with a pattern of frequent attacks. However, it is known that migraine attacks decrease or even disappear during pregnancy[43]; they even remain with low frequency during the period that the mother breastfeeds her child.[44] Obviously, the characteristic of the headache being precipitated when the woman gets out of bed strongly suggests that it is a headache caused by spinal anesthesia, if any. Post-dural puncture headaches are prevalent after spinal anesthesia, mainly when the anesthetist uses needles with a larger gauge. Needles used to remove CSF for diagnosis are also usually of larger gauge; therefore, the frequency of post-dural puncture headaches is higher. Remember that individuals who have already had post-dural puncture headaches have a greater chance of having the same type of headache again.[45] Also, patients with post-dural puncture headache may develop subdural hematomas[46], and neuroimaging evaluation may show this type of complication of hypotension.

We also have to think about cerebral venous thrombosis in the postpartum period.[47]

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