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Editorial

Headache attributed to spontaneous intracranial hypotension and associated possible causes

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Bem Junior and coworkers[1] published a very interesting and opportunely case report on spontaneous intracranial hypotension associated with cerebrospinal fluid (CSF) fistula in this issue. In recent decades, many publications have been addressing the subject.[2-6] Headache attributed to spontaneous intracranial hypotension (ICHD 7.2.3.)[7] is little known among physicians, and the diagnosis is difficult even considering the group of neurologists. Many patients progress without the correct diagnosis for weeks or even months. The clinical expression is classically similar to that found in post-dural puncture headache syndrome, an entity prevalent after spinal anesthesia.[8-10] The most frequent symptom presentation is orthostatic headache, which worsens in the upright position and subsides after lying down.[7, 11-13]

Radiological signs seen on brain MRI are very suggestive of a CSF hypotension syndrome.[14] Although the MRI of the intracranial structures may be considered normal in up to 20% of the individuals.

The correct localization of the CSF leak in individuals with headache attributed to spontaneous intracranial hypotension is often tricky and inconsistently attained.[15] As procedures for dealing with patients with spontaneous intracranial hypotension, we can comment on the results of a study[15] with a series of cases, using brain and spine MRI combined with digital subtraction myelography for CSF leak localization and the presence of a spinal longitudinal extradural collection.[15] Digital subtraction myelography was performed in those with spinal longitudinal extradural CSF collection primarily in the prone position and, in patients negative for spinal longitudinal extradural CSF collection, in the lateral decubitus positions.[15] The site of CSF leakage was located in 27/31 (87%) patients. Of these, 21 were positive for spinal longitudinal extradural CSF collection and categorized as having a ventral (type 1, 15 [48%]) or lateral dural tear (type 2; 4 [13%]). Ten patients were negative for spinal longitudinal extradural CSF collection and were categorized as having a CSF-venous fistula (type 3, 7 [23%]) or distal nerve root sleeve leak (type 4, 1 [3%]). The locations of leakage of 2 patients with spinal longitudinal extradural CSF collection remain undefined due to the resolution of spontaneous intracranial hypotension before the repetition of digital subtraction myelography. The leakage site could not be localized in 2/31 patients negative

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for spinal longitudinal extradural CSF collection. In 9/21 (43%) individuals with spinal longitudinal extradural CSF collection, the epidural blood patch successfully treated them, and 12 required surgery. None of the ten patients negative for spinal longitudinal extradural CSF collection were effectively treated with an epidural blood patch.[15] The authors concluded that patients with CSF-venous fistula may forgo further epidural blood patch treatment and go on to surgical repair.[15]

In diagnosing the possible location of the fistula or dural-arachnoid rupture, we must consider the possibility of misdiagnosing the fistula site by interpreting the leakage of contrast through the puncture hole used in myelography.

Sulioti and coworkers[16] reported an interesting case of a woman with symptomatic idiopathic intracranial hypertension who developed a thoracic spinal nerve root sleeve tear, probably while popping a balloon. Therefore, a Valsalva maneuver may lead to a CSF leak in susceptible subjects.[7] She was treated with epidural blood and a fibrin glue patch. A comment registered at ICHD-3[7] about the headache attributed to spontaneous intracranial hypotension added that a history of a trivial increase in intracranial pressure, such as during vigorous coughing, may be referred by the patient. I have already diagnosed a case of a woman with headache attributed to spontaneous intracranial hypotension that developed after three days of having pedaled a bicycle for more than an hour on a very bumpy path causing frequent tremors throughout the body due to the sudden changes in the pathway levels. In this way, we must look for some possible causes for "idiopathic" etiology. Associated conditions, such as idiopathic intracranial hypertension, must also be examined.[16]

Nevertheless, another patient I saw was a man with headache attributed to spontaneous intracranial hypotension who had undergone bariatric surgery a few years earlier. In this concern, a study[17] examined 338 patients with spontaneous intracranial hypotension compared with 245 patients with unruptured intracranial aneurysms. Eleven of the 338 (3.3%) patients with spontaneous intracranial hypotension had a history of bariatric surgery versus only 2/245 (0.8%) of the patients with intracranial aneurysms (p=0.02).[17] Among the subjects with spontaneous intracranial hypotension after bariatric surgery, the mean age at the time of bariatric surgery was 40.8 years (26-53 years), and the mean age at the time of start of spontaneous intracranial hypotension was 45.6 years (31-59 years).[17] The mean weight loss from bariatric surgery to the onset of spontaneous intracranial hypotension was 52.5 kg (25-98 kg).[17] The time interval from bariatric surgery to the onset of symptoms of spontaneous intracranial hypotension ranged from 3 to 241 months (mean, 56.5 months), indicating that bariatric surgery is a potential risk factor for spontaneous intracranial hypotension.[17]

An underlying connective tissue disorder may also cause dural weakness and propensity to CSF leak.[7,18] Cases of spontaneous intracranial hypotension in patients with Marfan syndrome have been reported.[19]

We conclude that the physician should look for a possible clinical cause that predisposes to a rupture of the dura mater/arachnoid or a precipitating factor for the rupture, such as unusual physical activity, particularly those that involve a Valsalva maneuver.

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