IDIOPATHIC INTRACRANIAL HYPERTENSION WITHOUT PAPILLEDEMA: A CASE REPORT

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Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is characterized by headache, papilledema, and normal cerebrospinal fluid (CSF) findings, all occurring in the absence of enlarged ventricles or an intracranial mass on computerized tomographic (CT) scan or magnetic resonance imaging (MRI). Papilledema is a cardinal feature of IIH, however, in rare cases, IIH may occur without papilledema or even with pseudopapilledema. We describe an overweight woman with long-standing headache and mild right abducent nerve paresis with a normal neurological examination including fundoscopy, Snellen visual acuity, and perimetry. After normal findings on brain MRI and laboratory tests, a lumbar puncture was performed, which was normal except for a CSF opening pressure of 400 mmH2O. However, after repeated lumbar punctures and the diagnosis of IIH, the patient's headache gradually subsided and her diplopia improved. In obese patients with chronic headaches and cranial nerve paresis, the diagnosis of IIH should be considered, even in the absence of papilledema. A lumbar puncture is helpful both diagnostically and therapeutically.

Keywords: Chronic headache • idiopathic intracranial hypertension • lumbar puncture • normal optic disc • papilledema

Introduction

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri,1, 2 is characterized by headache, papilledema, minimal or absent focal neurological signs, and normal cerebrospinal fluid (CSF) findings, all occurring in the absence of enlarged ventricles or an intracranial mass on CT scan or magnetic resonance imaging (MRI).1, 3 Pseudotumor cerebri has a number of causes and pathogenetic associations,4 however, the most common form is idiopathic and generally referred to as IIH.2

Case Report

A 45-year-old woman was referred for a headache of 5 months duration. The pain was described as a dull and “filling” pressure lasting between a few hours and a full day and recurring 3 – 4 times per week. The pain was mainly localized in the occipital region and radiated occasionally to the frontal region. It was nonpulsatile and was accompanied by occasional nausea without vomiting. There was no photophobia or phonophobia and no other complaints, except for transient diplopia. Past medical history was negative for systemic diseases such as hypo- or hyperthyroidism, hyperparathyroidism, and Cushing’s disease. Medications including ibuprofen (Brufen®), propranolol (Inderal®), trimipramine (Surmontil®), amitrypilin (Triptysol®), and sodium valproate had been prescribed for the headache in with no improvement.

Physical findings on admission were as follows: blood pressure (BP) =130/90 mmHg, respiratory rate (RR) =18/min, pulse rate (PR) = 78/min, and weight (Wt) = 105 kg. Systemic physical examination was normal. On neurological examination mental status, cranial nerves, fundoscopy, Snellen visual acuity, and...
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confrontation perimetry were all normal except for right abducent nerve paresis.

A detailed ophthalmologic examination, including checking the stereoscopic view, revealed nothing except for right abducent nerve palsy. Brain MRIs, with and without gadolinium, were normal. A lumbar puncture was performed which revealed: CSF opening pressure = 400 mmH₂O, white blood cells (WBC) = 0, red blood cells (RBC) = 5, protein = 15 mg/100dL, and sugar = 55 mg/100dL (concomitant blood sugar = 95mg/dL). Gram stain and sedimentation for malignant cells were negative. Laboratory tests including complete blood count (CBC), peripheral blood smear, ESR, thyroid function tests, antinuclear antibody (ANA), antineutrophil cytoplasm antibody (ANCA), rapid plasma reagin (RPR), and purified protein derivative (PPD) were all normal.

The patient was diagnosed with IIH and received a lumbar puncture every other day. The patient’s headache gradually subsided and her diplopia improved. After five successive lumbar puncture (LP) sessions, the CSF pressure fell to 200mmH₂O (Table 1) and the patient was discharged with acetazolamide 250 mg three times a day. Two weeks later, the CSF pressure was 240 mmH₂O and after three alternate-day lumbar punctures, the CSF pressure fell to 190 mmH₂O and the patient was continued on acetazolamide (Table 2).

Table 1. CSF pressure during 5 consecutive LP.

<table>
<thead>
<tr>
<th>Days</th>
<th>First day</th>
<th>Third day</th>
<th>Fifth day</th>
<th>Seventh day</th>
<th>Ninth day</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSF pressure</td>
<td>400 mmH₂O</td>
<td>340 mmH₂O</td>
<td>290 mmH₂O</td>
<td>230 mmH₂O</td>
<td>190 mmH₂O</td>
</tr>
</tbody>
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Table 2. CSF pressure during another 3 consecutive LP (second visit).

<table>
<thead>
<tr>
<th>Days</th>
<th>First day</th>
<th>Third day</th>
<th>Fifth day</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSF pressure</td>
<td>240 mmH₂O</td>
<td>240 mmH₂O</td>
<td>200 mmH₂O</td>
</tr>
</tbody>
</table>

Discussion

IIH, in its classic form, presents as a headache and papilledema raising suspicion of hydrocephalus or tumors. However, when brain imaging shows normal-size ventricles without evidence of mass lesions, and subsequent lumbar puncture shows only raised CSF pressure, a diagnosis of pseudotumor cerebri or IIH is made. Although papilledema is a cardinal feature of IIH, in rare cases it may occur without papilledema, or pseudopapilledema. It is, therefore, advised that in the presence of obesity and chronic headache, a spinal tap may be indicated to exclude the presence of IIH without papilledema.

References