A Rare Case of Idiopathic Hypertension with Multiple Cranial Palsies: A Case Report and Literature Review

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Abstract

Here is a unique case report of IIH being presented with multiple cranial nerve palsies, an unusual clinical presentation. A 32-year-old female with no prior comorbidities presented to the neurology OPD with headache since last two weeks, followed by double vision since last week along with left eyelid drooping, difficulty swallowing, and drinking from a straw. On evaluation, she was discovered to have left third, right ninth, and tenth cranial nerve palsy with bilateral sixth and seventh cranial nerve palsies. Furthermore, the fundus examination revealed bilateral papilloedema and neuroimaging ruled out any underlying structural or obstructive lesions as possible etiology for her symptoms. The lumbar puncture outcome was not noteworthy except for an increased cerebrospinal fluid (CSF) opening pressure. She was initiated on tablet Acetazolamide 250 mg and tablet Topiramate 50 mg to which she responded dramatically within 10 days. Her headache along with 7th, 9th and 10th cranial nerve palsies improved during her hospital stay. On her six-month follow-up, the papilledema and all other cranial nerve palsies had completely resolved. To conclude IIH should be thought of in every
patient presenting with headache and multiple cranial nerve abnormalities as highlighted in this case report. Neuroimaging and lumbar puncture should be carried out to rule out other alternative causes like infection or structural defects. Diagnosis of unusual IIH requires a high level of clinical suspicion as it can cause permanent loss of vision if left untreated.

**Keywords**

Idiopathic intracranial hypertension, pseudotumor cerebri, multiple cranial nerve palsies, papilledema, headache, opening CSF pressure.

**Introduction**

Idiopathic intracranial hypertension (IIH), previously known as pseudotumor cerebri, is identified by features of raised intracranial pressure (ICP), normal neuroimaging with elevated CSF opening pressures in the background of normal CSF components.[1] Headache, with a characteristic worsening in the morning, is a common presentation amongst 92–94% of patients and is characteristically worse in the morning, followed by other common manifestations such as pulsatile tinnitus (64–70%) and vision abnormalities (72%) of the patients. Single or multiple cranial nerve palsies may be seen in IIH as the elevated intracranial pressure can lead to cranial nerve traction and compression. Sixth cranial nerve palsy, a marker of raised ICP, is one of the most common cranial nerve palsies associated with IIH seen in about 10-50% of children, and 12% of adults. Rarely cranial nerves III, IV, VII, IX, and XII may also be involved.[2] However, the occurrence of six cranial nerve palsies simultaneously (II, III, VI, VII, IX, and X) in the context of IIH as described here is an exception and unusual.

**Case Report**

A 32-year-old Asian woman with no known previous comorbidities and a body mass index of 20.8 kg/m² presented to the outpatient department of Neurology with dull aching headache for two weeks which was holocranial, continuous along with nausea, and vomiting, sans photophobia and phonophobia with no aggravating or relieving factors. After a week from
initial symptoms, the patient developed drooping of the left eyelid with horizontal double vision, predominantly for distant objects. Although, the patient had no difficulty chewing food, swallowing solids, and liquids and sipping liquids using a straw was difficult. The patient had no nasal regurgitation or hoarseness of voice. She had no history of migraine headache, oral contraceptive pills intake, recent illness, fever, seizures, altered sensorium or trauma, and had regular menstrual cycles, with stable hemodynamics at the time of initial assessment.

During the neurological assessment, she was alert and had normal higher mental functions with no signs of meningeal irritation. Her tone and motor system, reflexes, sensory system, coordination, and gait were normal. Cranial nerve examination revealed the left partial third cranial nerve palsy in the form of partial ptosis of the left eyelid (Image 1). Pupils were 3–4 mm in size and reacted sluggishly to light. Bilateral lateral gaze restriction of eye movements was noted with no nystagmus or skew deviation suggestive of bilateral 6th cranial nerve palsy (Image 1). Bilateral facial weakness was observed in the form of weak eye closure, reduced frontal creasing, nasolabial folds, and inability to hold air in the mouth or blow a whistle. Facial sensations were preserved on both sides with normal jaw opening and a midline tongue. The uvula was deviated to the left (Image 1), with an absent gag reflex on the right side suggestive of right 9th and 10th cranial nerve palsies. In addition, the shrugging of her shoulders was bilaterally symmetrical and her hearing was normal and the examination of the fundus revealed bilateral papilledema (Image 2).

She underwent magnetic resonance imaging (MRI) of the brain with magnetic resonance venography which revealed tortuous bilateral optic nerves with a mild bulge of the optic nerve head at the optic disc, these findings were consistent with raised ICP (Image 3). Lumbar puncture (LP) revealed a CSF opening pressure of 320mm H2O in the lateral decubitus position, which was more than the standard reference interval. The cytological and chemical results of LP were within the normal range: white blood cells 2 (lymphocytes 100%), protein 20.1mg/dL,
and glucose 79mg/dL. No clinical or radiological features were suggestive of Addison’s or Cushing’s syndrome, and her serology was negative. As she fulfilled modified Dandy’s criteria for definite IIH she was started on a cerebral decongestant, 250 mg of Acetazolamide twice daily along with tablet Topiramate 50 mg once at night for headache.

During her hospital stay, the patient showed dramatic improvement in headache along with the 7th, 9th, and 10th cranial nerve palsies. At her six-month routine follow-up, her papilledema and all the cranial nerve palsies had completely resolved.

**Discussion**

We present the case of a 32-year-old woman complaining of dull aching holocranial continuous headache and features of multiple cranial nerve palsies. Neuroimaging involving MRI with MRV brain ruled out secondary causes and showed only features suggestive of raised intracranial pressure. Thus, the most likely diagnosis for the patient’s presentation was IIH according to modified Dandy’s criteria.[3] This diagnosis was further supported by the markedly elevated CSF opening pressure and improvement upon receiving acetazolamide therapy.

IIH affects both children and adults, with a yearly incidence of approximately 1 to 2 cases per 100,000 population.[4] Thus far this condition is more prevalent among obese women of childbearing age wherein the incidence rises to 4-21 cases per 100,000 population, however, age is no bar to presentation.[5]

IIH can be described by many imaging finding such as perioptic nerve sheath distention, vertical buckling of the optic nerve, globe flattening, optic nerve head protrusion, and an empty sella turcica.[2]

Other differential diagnoses include neurosarcoidosis, Lyme disease, and Bell’s palsy. The definite pathophysiology of IIH is not clear, however, it is suggestive that it involves CSF production and absorption and cerebral venous pressure elevation.[2]

Headache, pulsatile tinnitus, brief visual obscurations, and vision loss are the most typical symptoms of IIH.[1] Also, in a case-control study, the most common symptoms reported by IIH
patients were headache (94%), transient visual obscuration (68%), pulsatile tinnitus (58%), photopsia (54%), diplopia (38%), vision loss (30%), and retrobulbar pain on eye movements (22%). In addition to these classical signs of IIH patients may present with multiple cranial nerve palsies as a false localizing sign. The most common being unilateral or bilateral 6th cranial nerve palsy. Rare cases of association of IIH with involvement of the right second, third and sixth cranial nerves and left seventh cranial nerve has been reported. However, the association of IIH with bilateral multifocal cranial nerve palsies as observed in our case report has not been published to date to the best of our knowledge. Awareness that signs may be false localizing has implications for diagnostic investigations.

Ophthalmological evaluation for IIH should include fundoscopy, optical coherence tomography (OCT), and perimetry as it aids in early diagnosis and in monitoring response to therapy on follow-up. Neuroimaging with lumbar puncture cerebrospinal fluid (LP CSF) opening pressure further aids in confirming the diagnosis.

According to Rezazadeh and Rohani, a patient with IIH and third nerve palsy requires attention, in case of the presence of normal ancillary investigations such as angiography and demonstration of pupillary involvement. Compression in the subarachnoid space in the setting of elevated ICP appears to be the most likely mechanism of third nerve (and other cranial nerves) involvement. The reversal of oculomotor palsy after serial lumbar punctures establishes its link to elevated ICP.

A prospective study of 724 patients with recurrent headaches used MR venography. Of these, 6.7% of migraine patients had bilateral transverse sinus stenosis, and 67.8% of these patients had idiopathic intracranial hypertension without papilledema (IIHWOP) after lumbar puncture. These findings suggest that migraine patients with bilateral transverse sinus stenosis on cerebral MR venography should have a lumbar puncture to rule out IIHWOP. The main goal of therapy is to protect vision and reduce disabling symptoms. The IIH is
commonly managed with acetazolamide, a carbonic anhydrase inhibitor. Vomiting, diarrhea, renal stones, and aplastic anaemia are the most common adverse effects associated with acetazolamide. Topiramate, another carbonic anhydrase inhibitor is used in the management of headache and also reduces intracranial pressure. Topiramate also additionally suppresses appetite and helps in promoting weight loss in obese women. Furosemide, a loop diuretic can also be considered. Methyl prednisone, 1 g per day along with acetazolamide may be used in the short-term management of IIH, especially in fulminant disease-threatening vision loss. Due to the adverse effects of methyl prednisone such as weight gain and fluid retention it is not recommended for long-term management of IIH.

A 40-year-old Hispanic woman with a history of obesity and hypertension presented with a 3-day bifrontal headache, nausea, vomiting, transient visual disturbances, and a picture of right-sided cranial nerve VII palsy. Her neurologic examination was otherwise normal, including other cranial nerves, but a fundus examination revealed bilateral grade II papilledema. Imaging studies ruled out structural and obstructive lesions as potential causes of her symptoms, and lumbar puncture results were unremarkable with the exception of increased opening pressure. Prednisone and acetazolamide were then administered to her. She reported a significant improvement in both her headache and facial nerve palsy two days later. As our patient showed a significant response to acetazolamide alone, steroids were not considered. The other case reports of IIH with facial nerve palsy involvement are shown in table 1.

The benefits of octreotide and somatostatin analogues in IIH are still being evaluated. Patients who do not tolerate or improve with medical management may undergo surgical management. Optic nerve sheath fenestration and CSF diversions (lumboperitoneal or ventriculoperitoneal shunting) are surgical treatment options for IIH. Bilateral papilloedema is a feature of IIH, and OCT imaging provides an objective, noninvasive, and reproducible technique for diagnosing and monitoring papilloedema. Transverse venous sinus stenting is one of the cutting-edge
Conclusion

In the context of a young woman with headaches and multiple cranial nerve palsies, IIH should be strongly considered in the differential diagnosis. IIH as we all know is an exclusionary diagnosis and neuroimaging should be carried out to obliterate any secondary causes for raised ICP. Here we sought to raise clinicians’ awareness of the potential for IIH in presentation of multiple cranial nerve palsies, a presentation that has very rarely been described in IIH. High index of suspicion and targeted investigations are key for early diagnosis and prompt treatment to prevent irreversible vision loss.

Disclosures

The author(s) declare(s) that they have no conflicts of interest to disclose.

References


Tables:

Table 1: Idiopathic Hypertension with Multiple Cranial Palsies: Literature Review

<table>
<thead>
<tr>
<th>No.</th>
<th>Author Name, Year</th>
<th>Gender</th>
<th>Age</th>
<th>Cranial nerve involvement</th>
<th>Symptoms</th>
<th>Treatment</th>
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<tr>
<td>2.</td>
<td>Chutorian et al., 1977 [12]</td>
<td>M</td>
<td>12</td>
<td>Unilateral CN VII</td>
<td>Bifrontal headache, drooping of the left side of the face, tearing from the left eye, and nausea and vomiting</td>
<td>Lumbar puncture</td>
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<td>Transfrontal headache, facial asymmetry</td>
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<td>14</td>
<td>Unilateral CN VII</td>
<td>Intermittent bitemporal headache, right side face drooping, right brow could not elevate</td>
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<td></td>
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<td>Gender</td>
<td>Age</td>
<td>Symptomatology</td>
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<td></td>
<td>Pressure like, bifrontal headaches and decreased peripheral vision</td>
<td>Lumbar puncture, prednisone, acetazolamide, and lumboperitoneal shunt</td>
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<td>6.</td>
<td>Agarwal et al., 1989 [16]</td>
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<td>29</td>
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<td>Headaches, numbness of the right side of the face, altered taste sensation, and tingling and weakness of the right arm and right leg</td>
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<td>Lumbar puncture, prednisone, and acetazolamide</td>
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<td>Bifrontal headaches, right-sided facial drooping</td>
<td>Lumbar puncture</td>
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<td>Capobianco et al., 1997 [20]</td>
<td>F</td>
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<td>Unilateral CN VII</td>
<td>Severe throbbing headaches, pulsatile tinnitus, blurred vision in the left eye, painless horizontal diplopia, and right-sided facial pain</td>
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<td>Bifrontal headaches, right-sided facial drooping</td>
<td>Prednisone, and serial LPs</td>
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<td>Gender</td>
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<td>Symptom</td>
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<td>Soroken et al., 2015 [22]</td>
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<td>Diplopia and horizontal strabismus of the left eye</td>
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<td>Headache (holocranial, continuous), nausea, and vomiting</td>
<td>Acetazolamide, Topiramate</td>
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</table>

**Images:**

![Images](image-url)

Image 1: A Partial ptosis of left eyelid in primary gaze, B and C lateral gaze restriction; Right: Uvula deviated to left side due to right X CN palsy
Image 2: Fundoscopy showing bilateral papilledema

Image 3: MRI Brain showing tortuous bilateral optic nerves with mild bulge of optic nerve head at optic disc with hypoplastic left transverse sinus