

# Atypical Presentation of Idiopathic Intracranial Hypertension

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Dear Sir,

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a disorder of elevated intracranial pressure in the absence of localizing neurological signs, intracranial mass lesions or cerebrospinal outflow obstruction (1).

The usual presentation is headache, pulsatile tinnitus, papilledema, visual disturbances, diplopia and vomiting (2,3).

However, in some cases, involvement of cranial nerves can accompany and make the diagnosis difficult. We report a female case presented with headache, nausea, vomiting, diplopia, unilateral abducens nerve palsy with papilledema and diagnosed with IIH.

## CASE

A 32-year-old woman was admitted to our outpatient clinic with a month history of severe, daily, throbbing bitemporal headache and persistent nausea, sometimes vomiting, and also diplopia for 4 days. The headache was very different from her previous ones and was the worst she had ever experienced. It worsened in the morning and exacerbated with the valsalva maneuver.

On neurological examination, she had an entire right lateral rectus immobility and funduscopy showed bilateral papilledema. Her past medical history was

unremarkable and she had not been taking any drugs known to influence intracranial pressure. She was not obese.

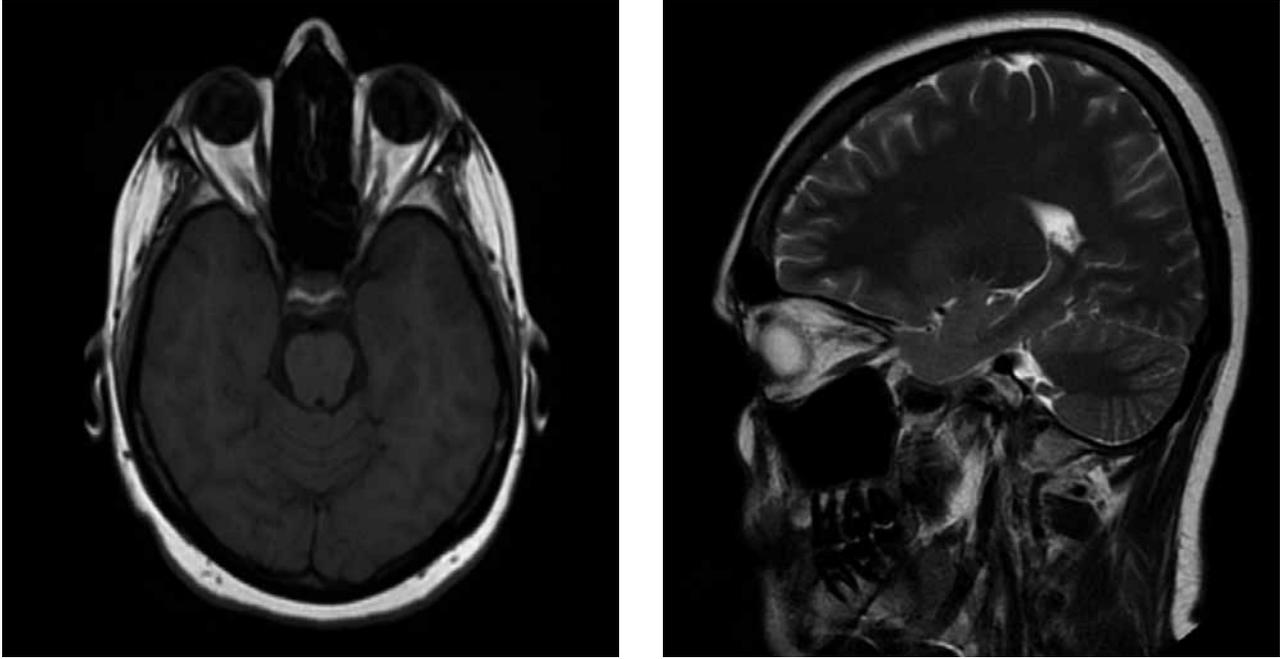
Her biochemical profile, including renal and hepatic functions, urine analysis, thyroid hormones, and vitamin B12 level were all normal.

Brain magnetic resonance imaging (MRI) showed an incidental finding of left frontal arachnoid cyst with no evidence of edematous brain (Figure 1). MR angiography and venography studies confirmed no intracranial lesion or thrombosis. Lumbar puncture from the lateral position showed an opening pressure of 38 cm of water with normal biochemical and cytological composition. All the secondary causes of IIH were ruled out (Table 1).

Based on the features described above, the diagnosis of IIH with unilateral abducens nerve palsy was made. The patient was treated with prednisone (1 mg/kg/day) and acetazolamide (15 mg/kg/day). Her papilledema, diplopia and abducens nerve palsy were ameliorated

**Table 1: Causes of Intracranial Hypertension**

a- Idiopathic
b- Symptomatic
Dural venous sinus thrombosis
Exogenous substances or certain drugs
Withdrawal from corticosteroids
Systemic diseases
Endocrine disturbances
Disorders of cerebral venous drainage
Obesity



**Figure 1: Brain magnetic resonance imaging (MRI) showed no marked abnormality**

two weeks after the treatment with prednisone and acetazolamide. Prednisone was gradually reduced after the 15<sup>th</sup> day of treatment and discontinued two weeks later. With the neuro-ophthalmologic examination follow-up, acetazolamide was reduced and stopped after 3 months of treatment. She has been asymptomatic with no recurrence for 6 months.

Some patients who are nonrespondent to maximum medical therapy or who have progressive visual loss appear to benefit from surgical intervention. The two main surgical procedures are ventriculoperitoneal or lumboperitoneal shunting and optic nerve sheath fenestration (4).

## DISCUSSION

In a case-control study, the most common symptoms reported by IHH 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%) (5).

Abducens nerve palsy is the most commonly

described abnormality seen in association with IHH, although oculomotor, trochlear, trigeminal, facial, glossopharyngeal and hypoglossal nerve palsies have also been reported in isolated case reports (6).

Recent studies have shown that abducens nerve involvement can occur in 25-30% of the cases, usually in combination with the other symptoms and signs of IHH.

The pathophysiology of the cranial nerve palsies accompanying IHH is poorly understood. The probable mechanism of the sixth nerve involvement seems to be by stretching of the nerve across the petrous apex (7).

Previous diagnostic criteria for IHH required the condition of no localising neurological signs, but currently proposed criteria accept abducens nerve palsy, either unilateral or bilateral, as a false localising sign in IHH.

Finally, we present this patient due to the rare coexistence of IHH and sixth nerve palsy and because only limited cases of such patients have been presented in the literature, and we believe that neurologists should consider this coexistence while dealing with atypical presentation of IHH.

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