

Benign Intracranial Hypertension: A case report and Review of Literature

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Abstract

This is a case report of benign intracranial hypertension. Her CT scan head was normal and cerebrospinal fluid (CSF) study at the time of admission only revealed high CSF pressure (47 cm of water). She improved with conservative management and papilledema was resolved at the time of discharge. A brief review of literature is also included.

INTRODUCTION

Benign intracranial hypertension was first described by Quincke in 1896, but the cause of this disorder continues to be a source of speculation. The prevalence of benign intracranial hypertension has been estimated to range from 1 to 19 cases per 100,000 population.¹ Women are affected eight times more frequently than men.² The pathophysiologic basis of benign intracranial hypertension remains unclear,³ but is associated with the conditions such as aberrations in intracranial volume regulation, metabolic diseases, medication-associated abnormalities and miscellaneous disorders. Diagnosis is primarily clinical and requires radiographic exclusion of an intracranial mass and measurement of cerebrospinal fluid pressure. Treatment is directed at reducing intracranial pressure in idiopathic cases or correcting associated conditions.

CASE REPORT

A 14 years old girl presented to National Institute of Neurological and Allied Sciences, Kathmandu with complains of headache and vomiting for 5 days. There was no history of fever, seizures, loss of consciousness. On examination, the child was conscious and well oriented to time, place and person. Her vitals along with higher mental functions were normal and cranial nerves intact. Her systemic examinations were normal and no evidence of focal neurological deficit except bilateral papilledema. Haematology and biochemistry were within normal limits. CT scan head revealed no abnormality. Cerebrospinal fluid (CSF) study at the time of admission only revealed high CSF pressure (47 cm of water). She was kept in the hospital with the diagnosis benign intracranial hypertension and managed

with acetazolamide and tapering dose of steroids. She improved with conservative management and papilledema was resolved at the time of discharge.

DISCUSSION

Benign intracranial hypertension (BIH) also known as idiopathic intracranial hypertension (IIH) or pseudotumor cerebri, is a cause of progressive visual loss in children and young adults. It is a neurological disorder that is characterized by increased intracranial pressure, in the absence of a tumor or other diseases affecting the brain or its lining. The diagnosis of benign intracranial hypertension is in large part clinical, but radiologic and laboratory studies have a role in confirming the diagnosis. Pediatric ICP very often is found in association with an underlying causative factor, such as ear infection, dural sinus thrombosis, steroid withdrawal, malnutrition associated with refeeding, hypervitaminosis A, minocycline, and others.⁴

Headache is the most common symptom⁵ and most frequently described as throbbing, episodic and without localization. It is exacerbated by the Valsalva maneuver and head movement and is most severe in the morning. Visual changes are a frequent clue to the diagnosis. The most frequently reported visual changes are episodic horizontal diplopia or tangential visual obscuration.⁶ Patients may report pulsatile tinnitus and pain in the shoulders, neck, back and arms. Finally, children may be entirely asymptomatic and present only with papilledema during a routine eye examination.⁷ Loss of visual acuity more commonly is a late finding in IIH. Uni- or bilateral sixth nerve palsy is frequent (40% to 48%) in children who have IIH,⁸ and the incidence

seems to be higher in children than in adults.

The original criteria for IIH were described by the American neurosurgeon Walter E. Dandy in 1937.⁹ They were modified by Smith in 1985 to become the “modified Dandy criteria”¹⁰ (Table:1).

Figure 1

Table 1: Modified Dandy criteria

1.	Signs & symptoms of increased ICP – CSF pressure >25 cmH ₂ O
2.	No localizing signs with the exception of abducens nerve palsy
3.	Normal CSF composition
4.	Normal to small (slit) ventricles on imaging with no intracranial mass

Magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) are, at present, the imaging studies of choice for detecting IIH. Neuroimaging is mandatory not only to exclude the possibility of herniation prior to a lumbar puncture but to identify potential secondary causes of elevated ICP. The upper normal limit of CSF opening pressure in children is approximately 180 to 200 mm H₂O, but precise cutoff values remain unknown.¹¹ Analysis of the cerebrospinal fluid (CSF) reveals normal cell count, glucose and sterile culture in the absence of other disease. CSF protein is usually low and not diagnostically helpful. Management of IIH is based almost entirely on clinical experience, due largely to the absence of randomized prospective trials that allow for evidence-based recommendations.¹¹

The goal of treatment is to relieve symptoms and to normalize ICP to preserve vision.¹¹ Despite the putative role of steroids in the genesis of benign intracranial hypertension, it has been postulated that dexamethasone decreases vasogenic edema, and this agent has been recommended for short-term (less than four weeks) treatment of benign intracranial hypertension. Patients who relapse during dexamethasone tapering should be treated with prednisone.¹³ Carbonic anhydrase inhibitors such as acetazolamide or methazolamide may be given. Both lumboperitoneal and

ventriculoperitoneal shunts have been recommended in the treatment of IIH.¹⁴ Bariatric surgery (gastroplasty or gastric bypass) appears to be a safe and efficient method of losing weight and diminishing comorbidities related to obesity, such as IIH, hypertension, and diabetes.¹⁵

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