Idiopathic Intracranial Hypertension in Pediatrics; Clinical Characteristics and Management Options

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Abstract

Background: Idiopathic intracranial hypertension is a well known entity in adults. It is rare in children, with clinical characteristics that may mimic or not that of adults.

Objective: Is to highlight the clinical characteristics and surgical view of idiopathic intracranial hypertension in pediatrics.

Patients and Methods: This is a retrospective study of the children below 18 years old admitted at the department for management of increased intracranial pressure matched with the modified Dandy Criteria. Review of the clinical presentations, imaging studies, opening pressure, and management options was done.

Results: There were 10 patients, 6 males, and 4 females. Age ranged from 9 years to 17 years, papilledema was a feature of all, opening pressure ranged from 28.5cm to 60cm. There were 6 patients obese and 4 patients were normal. Five patients responded to medical treatment and frequent lumbar punctures, and five patients required surgical intervention in the form of lumbo-peritoneal shunt. One patient of the five operated required revision of the shunt.

Conclusion: Idiopathic intracranial hypertension in pediatrics is an important entity in the differential diagnosis of papilledema, and is different in some clinical features from the adults'. Surgical intervention is reserved for non-responding patients with rapid deterioration of vision.

Key Words: Idiopathic – Intracranial – Hypertension – Children (IIH).

Introduction

IDIOPATHIC intracranial hypertension (IIH) is a disease characterized by increased intracranial pressure without a space occupying lesion or hydrocephalus, and normal CSF composition [1]. The incidence of the disease in the general population is 0.9: 100,000 [2]. It is rare in children, and there is increasing tendency among adolescents (12-15 years) than in younger children (2-12 years) [3]. Headache, visual symptoms, papilledema, and visual deterioration are the main presenting clinical features [4,5,6]. Initial pressure release followed by medical treatment is associated by high response rate [7]. Surgical intervention in the form of lumbo-peritoneal shunt is reserved for patients who are non adherent to medical treatment or progressive papilledema in spite of lumbar puncture and medical treatment [8]. Review of our records of the patients admitted to the hospital aged below 18 years old was done demonstrating their clinical characteristics and management options.

Patients and Methods

Review of the charts of the hospital for the patients who presented with papilledema, and matched with the modified dandy criteria was done. Search included the children during the period of the last five years from 2007-2011. There were 12 patients matched with the criteria based on CT imaging, but with reviewing the charts 2 patients were excluded. One patient was diagnosed as cerebral sinus thrombosis, and the other was diagnosed as Lupus Cerebritis (Based on MRV and MRI with contrast). The remaining 10 patients, their data included: Clinical history data: Age, sex, body mass index, complaint, duration of symptoms, signs, fundus examination, visual acuity, visual field, opening pressure of cerebrospinal fluid, medical treatment, and surgical intervention. Opening pressure was measured at the lateral decubitus position with the legs extended. No sedation was required. Lumbo-peritoneal shunt was decided in cases of failure of medical treatment with progressive visual deterioration. Patients were followed-up for a period at least 1 year and maximum of 3 years. Patients were followed up for recurrence of symptoms or papilledema. Data is presented in Tables.
Results

The study included 10 patients. There were 6 males and 4 females. The age ranged from 9 years to 17 years. There were 2 female patients 12 years or less: 9 years and 12 years, and 2 female patients above 12 years: (13 and 17 years). There were 3 males 12 years or less: (10, 11, and 12 years), and three patients above 12 years; (13, 15, and 17 years old). The duration of symptoms ranged from 1 month to 7 months. Headache and blurring of vision were the presenting symptoms in all the patients. Three patients had 6th nerve palsy at presentation. Papilledema with different grades confirmed the diagnosis with normal CT and MRI with contrast. None of our patients presented with optic atrophy. Visual field by perimetry was done for all the patients at diagnosis and at follow-up. Opening pressure ranged from 28.5cm to 60cm H\(_2\)O. Lumbar puncture is tried for more than once. None of our patients presented with vision loss.

Medical management is started immediately after completing the diagnosis. Acetazolamide is given in a dose of 25mg/kg/day in two divided doses. Steroids are reserved for cases of severe papilledema at initial presentation. Five patients out of the ten responded to medical treatment and frequent lumbar puncture (conservative group), and five patients required lumbo-peritoneal shunt insertion (surgical group). The opening pressure of the conservative group ranged from 28 to 39cm H\(_2\)O, whereas at the surgical group it is ranged from 35 to 60cm H\(_2\)O. There was no difference between the two groups regarding obesity; there were 2 patients with normal BMI and three patients were obese for each group. For all the patients; 6 out of 10 patients were obese. Three obese patients were below 12 years old and the other three were above this age. The obese patients; three of them were males and the other three were females. One patient, at the follow-up, period required shunt revision after 6 months duration from surgery. Fundus examination showed improvement of the papilledema at both groups (conservative and surgical group) with no recurrence except at the patient that had shunt revision.

### Table (2): Data of the patients had conservative management.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>BMI</th>
<th>Opening pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11</td>
<td>Male</td>
<td>Obese</td>
<td>28</td>
</tr>
<tr>
<td>2</td>
<td>10</td>
<td>Male</td>
<td>Normal</td>
<td>39</td>
</tr>
<tr>
<td>3</td>
<td>13</td>
<td>Female</td>
<td>Normal</td>
<td>28.5</td>
</tr>
<tr>
<td>4</td>
<td>13</td>
<td>Male</td>
<td>Obese</td>
<td>29</td>
</tr>
<tr>
<td>5</td>
<td>12</td>
<td>Female</td>
<td>Obese</td>
<td>37</td>
</tr>
</tbody>
</table>

### Table (3): Data of the patients required surgical intervention.

<table>
<thead>
<tr>
<th>Patient</th>
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<th>Sex</th>
<th>BMI</th>
<th>Opening pressure</th>
</tr>
</thead>
<tbody>
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<td>Obese</td>
<td>45</td>
</tr>
<tr>
<td>2</td>
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<td>Male</td>
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<td>50</td>
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<td>Female</td>
<td>Obese</td>
<td>60</td>
</tr>
<tr>
<td>5</td>
<td>17</td>
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<td>Obese</td>
<td>48</td>
</tr>
</tbody>
</table>

Discussion

Pseudotumor cerebri is a disease characterized by increased intracranial pressure without brain tumor that was first described by Quincke [9]. This definition is then modified by Dandy and Smith [10]. There are other conditions are associated with these criteria but with known causes, so the name Idiopathic Intracranial Hypertension is proposed [11] and modification of Dandy criteria is adopted [1,12].

The diagnosis of idiopathic intracranial hypertension in pediatrics based on these criteria is
challenging. The age of presentation is different. Some reports state that it is commoner in post-pubertal age than pre-pubertal age [3,4]. In pre-pubertal age it is reported from age as young as 1.9 year [13]. In contrast, other studies as of Tibussek et al., more than 60% of patients were at the pre-pubertal age (thirty two patients out of 53 patients) [7]. Ten patients out of 12 patients of Distelmaier et al., were below 12 years [14]. In study of Phillips et al., included 35 patients, the age of the patients ranged from 3 years to 17 years and seventeen patients (about 50%) were younger than 11 years [8]. The same percentage at the study of Soiberman et al., meta analysis of their patients, revealed that the female/male ratio in children young-er than age 11 years seems to be fairly equal, with relatively low rate of obesity, contrasting to a majority of females in the group of adolescents at high risk to become obese [21]. Balcer et al., at their review found that 43% of patients aged 3 to 11 years were obese, whereas 81% of those 12-14 years age group and 91% of those in the 15-17 years age group met the criteria for obesity [24]. Others are contradicting these results and show no relation between obesity, sex, age, and the disease [5,23].

Headache and papilledema were constant features at our patients. There is different variation at presentation at the literature. Patients may be asymptomatic and only diagnosed by papilledema [7,25], at the other hand patients may be symptomatic (headache, neck pain, abducens palsy...) but without papilledema [7,26]. Lower motor neuron facial paresis [27] or fourth cranial nerve palsy [28], are rare presentations of IIH.

Elevated opening pressure is one of the diagnostic criteria of IIH. Diagnostic opening pressures are: In neonates >76 mmH2O, patients aged less than 8 years with papilledema: >180 mmH2O, and patients aged 8 years or above or less than 8 without papilledema: >250 mmH2O [29]. In our study patients were more than 8 years, and the opening pressures were between 280mmH2O and 600mm H2O. We tried to find a correlation between the opening pressure and the management methods (conservative versus surgical), and we found that at the conservative group of patients the pressures were lower in values than the surgical group, which was statistically significant. The highest opening pressures were not predictors of outcome at Soiberman et al., series [8]. The rational for surgical intervention is failure of conservative measures to prevent progressive visual deterioration [8,14,24,30]. In our study, in contrary to the literature, there were high incidence of lumbo-peritoneal shunt insertion; five patients out of ten. This can be explained by the persistence of high opening pressure at repeated lumbar punctures, the ineffective medical treatment of them, and the rapid visual deterioration. Shunt revision is required in one patient during the follow-up period. All patients had good visual outcome at their follow-up.

**Conclusion:**

Idiopathic intracranial hypertension in pediatrics is an important differential diagnosis in pediatric population complaining of headache or visual complaints. From these results, we found no sex, age, and obesity association with the disease, and larger number of patients at our community is required to have sound figures of the disease. The surgical intervention is mandatory in case of rapid visual deterioration and failure of medical treatment.
References


