Pseudotumor cerebri - Clinical presentation, Surgical management and outcome

Dr. Ibrahim Yassin Hussein F.I.C.M. M.B.CH.B.
Neurosurgeon, Iraq
Corresponding author: almashhadaniibur@gmail.com

Abstract

Pseudotumor cerebri is disorder characterized by increased intracranial pressure without deformity or obstruction of ventricular system, there is predilection to occur in obese women in child bearing age, the pathogenesis of disease is still uncertain but may be due to imbalance between production and absorption of cerebrospinal fluid, papilledema is the hallmark of disease found in every patient, treatment could be medical or surgical depending on patient response.

Objective
To evaluate clinical presentation, possible causes, treatment and incidence of pseudotumor cerebri in patients attending neurosurgical department in GHAZI AL HARRERI teaching hospital for surgical specialities

Patients and Methods
Retrospective study in analysis of 30 patients in neurosurgical department in period between November 2011 and December 2013

Results
30 patient diagnosed with pseudotumor cerebri two males and twenty eight females, 70% obese, papilledema present in all patients, one patient get benefit from medical treatment, 29 patients undergone surgical intervention, only 5 patients needs revision.

Conclusion
Pseudotumor cerebri is disease of unknown etiology in most of patients, the diagnostic methodology of IIH must include LP, MRI and brain venography, CSF diversion is the surgery of choice in patients if there is no benefit from medical treatment.

Keywords: Headache, pseudotumor cerebri, Lumboperitoneal shunt, Optic nerve sheath fenestration, Papilledema, Venous sinus thrombosis, intracranial pressure

Introduction

Definition
The syndrome known as pseudotumor cerebri (PTC) is generally thought as a condition characterized by increased intracranial pressure (ICP) without evidence of dilated ventricles or a mass lesion by imaging, normal cerebrospinal fluid (CSF) content, and papilledema, occurring in most cases in young, obese women without any clear explanation.

Historical view
The terminology used to describe this condition has changed dramatically over time and continues to change as new issues regarding its etiology are raised.

Heinrich Quincke, an early pioneer in the use of lumbar puncture, reported the first recorded cases of intracranial hypertension of unknown cause in what he
described as “meningitis serosa” in 1893; at that time, he posited that inadequate CSF resorption was responsible for the syndrome, a theory that is still entertained by some researchers.\(^{2}\)

The term pseudotumor cerebri appears to have first been used in 1914\(^{1}\).

Subsequently, Foley 1955 suggested calling the condition “benign intracranial hypertension” because it appeared to have a much more benign neurological prognosis than increased ICP caused by a central nervous system mass lesion or infection. Because significant visual morbidity may result from PTC, use of the adjective “benign” is no longer considered appropriate terminology.\(^{1}\)

In the late 1980’s, Corbett et al (1982) altered the name to idiopathic intracranial hypertension, since the syndrome was not benign as once thought\(^{1}\).

Most physicians use the term idiopathic intracranial hypertension (IIH) for cases of PTC that occur in young, obese patients and the term secondary pseudotumor cerebri for the rare cases in which a cause (e.g., drug induced) is identified.\(^{3}\)

It should also be emphasized that some authors use the term pseudotumor syndrome for patients with increased ICP unassociated with ventricular dilation, evidence of cerebral edema, or an intracranial mass lesion but with abnormal CSF content. This condition occurs in some patients with aseptic, carcinomatous, or lymphomatous meningitis.\(^{4}\)

**Intracranial pressure**

Intracranial pressure (ICP) the pressure of the cerebrospinal fluid in the subarachnoid space (the space between the skull and the brain) the normal range is approximately 7 to 13 mm Hg.\(^{5}\)

**Approaches of measuring I.C.P.**

1. Non invasive
   a. Clinical examination
   b. Fundoscopic examination
   c. Imaging
2. Invasive
   a. External ventricular drain placement
   b. Intraparenchymal fibro optic catheter placement
   c. Epidural sensor
   d. Sub arachnoid bolt

**Etiology**

Idiopathic intracranial hypertension (IIH) is a disorder of unknown etiology that predominantly affects obese women of childbearing age. \(^ {1}\)

The primary problem is chronically elevated intracranial pressure (ICP), and the most important neurologic manifestation is papilledema, which may lead to progressive optic atrophy and blindness. \(^ {1}\)

Many theories have been advanced to explain the pathogenesis of IIH. According to the Monro-Kellie rule anything added to the blood, CSF, or brain volume or anything impeding CSF or venous egress would be expected to increase I.C.P.\(^ {12}\)
IIH is a diagnosis of exclusion that occurs primarily in young obese women and, occasionally, in obese men with no evidence of any underlying disease.{4}

In about 10% of cases of PTC are associated with a number of different conditions. Suspicion of a secondary cause of PTC is heightened in prepubertal children, men, nonobese women, and otherwise typical patients with rapidly progressive visual loss that does not respond to treatment.[5]

Secondary causes of PTC include:{4}

(1) impairment or obstruction of cerebral venous sinus drainage by intrinsic or extrinsic lesions.
(2) endocrine and metabolic dysfunction.
(3) exposure to exogenous drugs and other substances.
(4) withdrawal of certain drugs.
(5) systemic illnesses.

Symptoms and signs

Primary PTC (i.e., IIH) and secondary PTC produce identical symptoms and signs.{20}

Occasionally, the condition is asymptomatic and discovered during a routine ophthalmic examination when papilledema is found.{20}

The most common symptom; headache, transient visual loss and diplopia
Papilledema is the diagnostic hallmark of PTC and is present in almost all patients, the papilledema of PTC is identical with that in patients with other causes of increased ICP. [3]

Diagnosis

The diagnosis of both IIH and secondary PTC requires that there be no intracranial or spinal mass, no evidence of hydrocephalus, documented increased ICP, and normal CSF contents. Thus, the diagnosis cannot and should not be made without neuroimaging and a lumbar puncture (LP).[23]

Treatment

a. Medical therapy
   1. Medication to lower ICP
   2. Headache prophylaxis
   3. Corticosteroids
   4. Repeated L P

B. Surgical treatment
   1. Lumboperitonial shunt
   2. Optic nerve sheath fenestration
   3. Venous sinus stenting.

Patients and Methods

This is a study of thirty patient whose ages range between (19-46) years old, present to neurosurgical department in Ghazi Al- Harriery teaching hospital for surgical speciality in period between November 2011- December 2013 as typical cases of I.I.H. presented with headache, visual disturbance, papilledema….etc.

The radiological investigation has failed to demonstrate intracranial pathology responsible for the raised I.C.P. except in three cases were venous sinus thrombosis.

Twenty eight were female (93.3%) and two were males (6.66%).

C.S.F. pressure was measured through lumber puncture at L4-L5 level in left lateral position with horizontal plane of the body and head to reduce errors as possible.

Visual acuity and visual field was assessed in twenty patients in our hospital by ophthalmologist using Nilson chart and Goldsmann visual field.

Neuroradiological evaluation as computer tomography (C.T.) and M.R.I. was done for all patients.

Ten patients wasdiagnosied in Neuromedicine department in Baghdad teaching hospital and referred to neurosurgical department after failure of medical management and repeated LP to resolve papilledema.

The period of time between diagnosis and surgical intervention range between 6 months and one year.

Twenty nine of our patient treated surgically after receiving medical treatment, and only one female patient undergo repeated LPs and surgical intervention without medical treatment.

The Results

All of our patient have elevated I.C.P. greater than 20 cm H2O consistent with the diagnosis of pseudotumor cerebri
The gender of patient

The majority of our patients were females (28 females and 2 males) 93.33% females and 6.6% males.

The age distribution

The range of age distribution is between (18 and 46 years) with the mean of 28 years old.

The majority of our patients' ages were in the age group between (20-29 years) and (30-39 years) equally 10 patients in each age group which form 33.3% age from the total percentage, and six patients in the age group below 20 years old 20% and four cases above 40 years old 13.4%, as shown in the following table and figure:

<table>
<thead>
<tr>
<th>Age</th>
<th>Below 20</th>
<th>20-29</th>
<th>30-39</th>
<th>Above 40</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>6</td>
<td>10</td>
<td>10</td>
<td>4</td>
<td>30</td>
</tr>
<tr>
<td>percentage</td>
<td>20%</td>
<td>33.3%</td>
<td>33.3%</td>
<td>13.4%</td>
<td>100%</td>
</tr>
</tbody>
</table>
Figure (1) showing age distribution of the study

Associated factors

Obesity is one of the main associated factors in pseudotumor cerebri was present in 21 patient . Two patient was found to have history of oral contraceptive pills intake in time of diagnosis 6.66% , no history of tetracycline intake or hypervitaminosis A , no pregnancy nor miscarriage, No Addison’s disease as shown in the following tables and figures:

Table (2) showing the associated factors of pseudotumor cerebri

<table>
<thead>
<tr>
<th>The associated factor</th>
<th>Number of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>obesity</td>
<td>21</td>
<td>70%</td>
</tr>
<tr>
<td>Menstrual irregularity</td>
<td>9 (28 female patients)</td>
<td>32.14%</td>
</tr>
<tr>
<td>History of oral contraceptive pills</td>
<td>2(28 female patients)</td>
<td>7.14%</td>
</tr>
<tr>
<td>pregnancy</td>
<td>0</td>
<td>0%</td>
</tr>
<tr>
<td>miscarriage</td>
<td>0</td>
<td>0%</td>
</tr>
<tr>
<td>No Addison’s disease</td>
<td>0</td>
<td>0%</td>
</tr>
</tbody>
</table>
Presenting symptoms

The commonest presenting feature is headache which was present in 26 patients from thirty patients (86.6%), the second presenting symptom in our study is visual disturbance which was found in nineteen patient (70%), in the form of decrease visual acuity, blurring of vision, transient visual obscuration (T.V.O.), the other compliance was vomiting which was found in seven patients (23.3%), dizziness was found in four patients (13.3%), as shown in the following table and figure:

Table (3) showing presenting symptoms presented in our study

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Number of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>headache</td>
<td>26</td>
<td>86.6%</td>
</tr>
<tr>
<td>Visual disturbance</td>
<td>19</td>
<td>63.3%</td>
</tr>
<tr>
<td>vomiting</td>
<td>7</td>
<td>23.3%</td>
</tr>
<tr>
<td>dizziness</td>
<td>4</td>
<td>13.3%</td>
</tr>
<tr>
<td>Menstrual disturbance</td>
<td>9</td>
<td>32.15%</td>
</tr>
<tr>
<td>Cognitive disturbance</td>
<td>0</td>
<td>0%</td>
</tr>
</tbody>
</table>
Figure (3) showing presenting symptoms presented in our study

**Presenting signs**

In our study the most neurological finding is papilledema which was found in all the patients (100%), followed by obesity in 21 patient , decrease visual acuity in five patients (16.66%), enlarge blind spot in 3 patients 10% abducent nerve palsy was observed only in one case (3.3%) same as central scotomas

Table (4) showing the signs founded in our study

<table>
<thead>
<tr>
<th>signs</th>
<th>Number of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>papilledema</td>
<td>30</td>
<td>100%</td>
</tr>
<tr>
<td>Decrease visual acuity and field</td>
<td>5</td>
<td>16.66%</td>
</tr>
<tr>
<td>obesity</td>
<td>21</td>
<td>70%</td>
</tr>
<tr>
<td>6\textsuperscript{th} nerve palsy</td>
<td>1</td>
<td>3.3%</td>
</tr>
<tr>
<td>Enlargement of blind spot</td>
<td>3</td>
<td>10%</td>
</tr>
<tr>
<td>Central scotoma</td>
<td>1</td>
<td>3.3%</td>
</tr>
</tbody>
</table>
Figure (4) showing signs founded in our study

Neuro- Ophthalmological findings in our study was papilledema at least in one eye in all our cases (100%) with enlarge blind spot in three cases (10%), decrease visual acuity in five patients (16.66%), central scotomas in one patient only 3.3%, as shown in the following table and figure.

Table (5) showing ophthalmological findings in our study

<table>
<thead>
<tr>
<th>Neuro-ophthalmological finding</th>
<th>Number of cases</th>
<th>percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>papilledema</td>
<td>30</td>
<td>100%</td>
</tr>
<tr>
<td>Enlarge blind spot</td>
<td>3</td>
<td>10%</td>
</tr>
<tr>
<td>Decrease visual acuity</td>
<td>5</td>
<td>16.66%</td>
</tr>
<tr>
<td>Central scotomas</td>
<td>1</td>
<td>3.3%</td>
</tr>
<tr>
<td>6th nerve palsy</td>
<td>1</td>
<td>3.3%</td>
</tr>
</tbody>
</table>
All the patients in our study were undergone lumbar puncture for diagnostic and therapeutic purposes either in our department or neuromedical department in Baghdad teaching hospital except five patients one patient referred to our department who already diagnosed as venous sinusous thrombosis and the other four refuse the procedure of lumbar puncture.

CSF Finding of 29 patients in our study

Table (6) shows C.S.F. findings in our patients

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>appearance</th>
<th>Pressure cm H2O</th>
<th>Protein mg/dl</th>
<th>Sugar mg/dl</th>
<th>Cell count</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>clear</td>
<td>25.5</td>
<td>17</td>
<td>64</td>
<td>1-2</td>
</tr>
<tr>
<td>2</td>
<td>clear</td>
<td>26</td>
<td>23</td>
<td>58</td>
<td>3-5</td>
</tr>
<tr>
<td>3</td>
<td>clear</td>
<td>27</td>
<td>40</td>
<td>70</td>
<td>0-3</td>
</tr>
<tr>
<td>4</td>
<td>clear</td>
<td>27.5</td>
<td>35</td>
<td>45</td>
<td>-</td>
</tr>
<tr>
<td>5</td>
<td>clear</td>
<td>28</td>
<td>25</td>
<td>62</td>
<td>1-4</td>
</tr>
<tr>
<td>6</td>
<td>clear</td>
<td>28</td>
<td>39</td>
<td>46</td>
<td>5</td>
</tr>
<tr>
<td>7</td>
<td>clear</td>
<td>290</td>
<td>20</td>
<td>55</td>
<td>2-4</td>
</tr>
<tr>
<td>8</td>
<td>clear</td>
<td>29.5</td>
<td>40</td>
<td>57</td>
<td>0</td>
</tr>
<tr>
<td>9</td>
<td>clear</td>
<td>31</td>
<td>33</td>
<td>36</td>
<td>1-4</td>
</tr>
</tbody>
</table>
All our patients under gone medical treatment by receiving corticosteroid (dexamethasone) diuretics (furosemide and acetazolamide) and repeated lumber puncture, low molecular weight heparin was giving to three patient in which venous sinous thrombosis was the diagnosis.

Diamox (acetazolamide) 250 mg X 3 alone was given to six patients 20%, Lasix (furosemide) 40 mg X 2 was given to two patients 6.6 %, corticosteroid never been gave alone, while combination of two diuretics was given to nine patients 30%, combination of two diuretics and corticosteroid in dose of 8mg X 3 was given to 3 patients 10%, combination of acetazolamide and dexamethasone was given to ten patients 33.3%

Twenty nine of our patients performed repeated lumber puncture for diagnostic and therapeutic purposes, five patients did not performed it (16.66%), eight patients done the lumber puncture once (26.66%), thirteen patient performed it twice (43.3%) , four patient performed it three times (13.33%) as shown in the following figure and table.
Table (7) showing the number of cases who performed lumber puncture in our study

<table>
<thead>
<tr>
<th>Lumber puncture trials</th>
<th>No. of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>No L.P.</td>
<td>5</td>
<td>16.6%</td>
</tr>
<tr>
<td>Performed once</td>
<td>8</td>
<td>26.66%</td>
</tr>
<tr>
<td>Performed twice</td>
<td>13</td>
<td>43.3%</td>
</tr>
<tr>
<td>Performed three times</td>
<td>4</td>
<td>13.33%</td>
</tr>
</tbody>
</table>

Figure(6) showing the number of cases who performed lumber punctures

The outcome of our patients

Only one female patient get benefit from the medical treatment and lumber puncture and there was no need for further surgical intervention and the papilledema and headache resolve after five to seven days of medical treatment and two lumber puncture sessions.

The other twenty nine patients undergone surgical intervention in the form of lumber peritoneal shunt and show improvement in their vision and headache, except two patient did not improve because both diagnosis was so delay so they were present to our department with only slight light perception.

Five patients developed complication of surgery in the form of include spontaneous obstruction of the distal end and immigration of the distal end of the catheter from the peritoneal cavity, three cases shows spontaneous obstruction (10.34%), two patient (6.89%) showed migration of distal end and required shunt revision.

None of our patient developed infection as complication, perforation of abdominal viscus nor ascites.

Three patient required revision due to obstruction of the peritoneal end of the catheter, and two patient required two revision surgeries due to migration of distal end from peritoneal cavity as shown in the following table and figure.
Table (8) showing the complication and number of cases that develop the complication

<table>
<thead>
<tr>
<th>Complication</th>
<th>Number of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Revision</td>
<td>5</td>
<td>17.25%</td>
</tr>
<tr>
<td>Obstruction of distal end</td>
<td>3</td>
<td>10.34%</td>
</tr>
<tr>
<td>Migration of distal end</td>
<td>2</td>
<td>6.89%</td>
</tr>
<tr>
<td>Infection</td>
<td>0</td>
<td>0%</td>
</tr>
<tr>
<td>Viscus perforation</td>
<td>0</td>
<td>0%</td>
</tr>
<tr>
<td>Ascites</td>
<td>0</td>
<td>0%</td>
</tr>
<tr>
<td>Death</td>
<td>0</td>
<td>0%</td>
</tr>
</tbody>
</table>

Figure (7) showing the complication of surgery in our study

Discussion

Pseudotumor cerebri is an idiopathic disorder defined by the modified Dandy criteria as the following:{28}

1) Signs and symptoms of raised ICP (headache, nausea, vomiting, transient obscuration of vision, and papilledema).
2) Normal neurological examination, except for a sixth nerve palsy.
3) Elevated CSF pressure (>25cm H2O) with normal constituents.
4) Modern neuroimaging, CT with and without contrast, or MRI demonstrating normal to small symmetrical ventricles and excluding a mass lesion or other cause of raised ICP.

It may be associated with menstrual irregularity or amenorrhoea and variously been reported as complication of Addisons disease with improvement after replacement therapy and as being more likely to occur when steroid dosage is reduced and relative adrenal insufficiency is present.{65}
The results as shown in our study that patient are usually females (28 from 30) 93.33%, similar results found in a study made by (Vincent Giuseffi, MD, Michael Wall, MD, Paul Z. Siegel, MD and Patricio B. Rojas, PhD) in which (90%) 45 of 50 patients study was female. {53}

Only Johnston and Paterson in 1974 shows both genders are equally affected {54}.

The peak of age in our study was found to be in the second and 3rd decade of life, ten female patient was in the age between (20-29yrs) 33.3% from the total number and percentage of our study, and nine females and one male between the (30-39) years old 33.3% from the total number and percentage of our cases, compared to the study made by Brian in 1985 which shows that the peak incidence was found in the 4th decade of age. {55}

Also in our study we found 21 patient were complaining from obesity (70%), the same result found by Rowe FJ, Sarkies were 70.5% twenty four patient from thirty four patients were obese. {56}

Also Contreras-Martin Y, Bueno-Perdomo JH found similar result in a study of sixty one patient 72.13% of the patients were obese. {57}

Two female patients from twenty eight in our study (7.14%) was found using oral contraceptive pills which is important predisposing factor in pseudotumor cerebri in study of five cases by Horst A, & Rutten were receiving low dose contraceptive pills lead to aseptic dural sinus thrombosis. {58}

In our study the major complain was headache, found in 26 patients (86.6%), while González-Hernández A, Fabre-Pio Headache was present in 85.4% of their studyof fifty five patient. {59}

Also in retrospective study of twenty patient done by (Merle H, Smadja D, Ayeboua L, Cabre P, Gerard M, Alliot E, Rapoport P, Jallot-Sainte-Rose N, Richer R, Poman G.) sixteen patient 80% were complaining of headache as main presenting feature of pseudotumor cerebri. {60}

The second major complain in our study was visual disturbance which include decrease in visual acuity, blurring of vision, transient visual obscuration (T.V.O). These complains affected 19 (63.3%) patients in our study comparing in study performed by (Wall M, George D), in which 50% consecutive newly diagnosed patients complain of transient visual obscurations affecting 36 patient (72%). {61}

Also in another retrospective study of twenty patient done by (Merle H, Smadja D, Ayeboua L, Cabre P, Gerard M, Alliot E, Rapoport P, Jallot-Sainte-Rose N, Richer R, Poman G.) fifteen patient show a loss of visual acuity (75%) and five patient shows transient visual loss (25%). {60}

Vomiting and dizziness were the third complian found in our patients (23%) and (13%) respectively, these symptoms are not specific and its related to elevated intracranial pressure.

CSF pressure via lumbar puncture was the investigation of choice for the diagnosis, and it shows that CSF pressure was elevated above 22cm H2O, and this explains the papilledema formation and severity of the complains, papilloedema either directly or indirectly is the cause of visual loss in pesudotumor cerebri, the higher the grade of the papilledema, the worse the visual loss.

Papilledema due to increased intracranial pressure, is the cardinal sign of pseudotumor cerebri and found in all the patients in our study, similar results were found in prospective study of fifty patients done by Friedman DI, Jacobson DM. {3}

Two patients present to our department with severe visual disturbance only slight perception to light at time of presentation and one of the patient was male referred to our department already diagnosed with cerebral sinus thrombosis and both did not improve even after surgical intervention.

Computerized tomography scan (CT scan) was found to be very useful in defining the extent of brain normality and excluding any pathological abnormality, although small size ventricles was a common feature among our patients.

Computerized tomography scan (CT scan) and magnetic resonant image (MRI) was done for all the patient in our study to exclude space occupying lesions, three of our patients was further diagnosed by the use of magnetic resonant venogram (MRV) and magnetic resonance angiography (MRA) found to have venous sinus thrombosis.

Three patients in our study (10%) was found to have venous sinus thrombosis which undergo medical treatment with low molecular weight heparin and LP shunt, several case reports done by Gary Y. Shaw and
Stephanie K. Million notice that patients with venous sinous thrombosis present with headache and papilledema with normal (CT scan) findings but (MRV) shows venous sinous thrombosis an extensive work out shows these patient has Factor V Leiden mutation.{64}

The empirical therapy of elevated ICP due to pseudotumour cerebri include acetazolamide (Diamox), loop diuretics (furosemide), corticosteroids, repeated lumbar punctures, and lumboperitoneal shunting, which has the advantage of being completely extracranial surgical management, minimizing the intracranial complications, but in our study only one patient shows improvement in severity of headache and gradual disappearance of papilledema without the need of surgical intervention by lumboperitoneal shunt.{20}

Only one patient in our study improve using medical treatment (acetazolamide and furosemide corticosteroids and two repeated lumber punctures and papilledema resolve in period between two days to one week.

Twenty nine patients in our study performed lumber peritoneal shunt , twenty four patients had shown dramatic response in improving headache and visual outcome, five patients showed complications which is higher than expected from medical management, these complications include spontaneous obstruction of the distal end and immigration of the distal end of the catheter from the peritoneal cavity, in our study three cases shows spontaneous obstruction10% comaired with other study done by (Waleed F. El-Saadany, Ahmed Farhoud, Ihab Zidan) in study of twenty two patient six of them(27%) developed shunt obstruction and required shunt revision.{62}

In other study performed by Eggenberger ER, Miller NR, Vitale Sa retrospective study of 27 patients with pseudotumor cerebri (PTC) treated with at least one lumboperitoneal shunt (LPS) Twelve patients (44%) required no revisions. The number of revisions among the 15 patients (56%) who required revisions due to failure of the shunt.{63}

The most important postoperative complain is headache, that is found to occur upon mobilization , this type of headache is a low pressure headache (over shunting)and was related to the fall of CSF pressure after shunting which is attributed to continued leakage of CSF through the catheter to the peritoneum.

The severity of headache reduced by tilting the patient’s head down to the supine posture , thereby reducing CSF hydrostatic pressure in the subarachnoid space and thus reducing amount of CSF leakage to the peritoneum.

The use of intravenous fluid in the form of normal saline 9% was tried in some patients and show no significant difference in the headache after shunting.

**Conclusion**

Idiopathic intracranial hypertension is a disease with complex pathophysiological structure, which until now has not been fully clarified.

The plurality of possible etiologies is the reason why many different treatments have been developed with a variety of response from patient to patient. The diagnostic methodology of IIH must include LP, MRI and brain venography.

Treatment always begins with instructions to the patient for exercise, life style modification and weight loss especially for obese people.

Treatment by medication to reduce intracranial pressure with or without repeated lumber puncture is of benefit in some patients.

CSF diversion is the surgery of choice in patients with refractory headache with or without papilledema and when vision is threatened optic nerve sheath fenestration must be performed.

while stent placement in venous sinuses should be the last resort when all previous treatment options for patients with radiologic and manometric confirmation of venous sinus stenosis have failed.

**Recommendations**

Potential agents that might cause or worsen P.T.C. (eg, tetracycline derivatives) should be discontinued, and treatment provided for comorbid diseases such as anemia if present.

We recommend counseling and/or treatment for weight loss in all obese patients with P.T.C.

We suggest the use of acetazolamide as the initial treatment of patients with P.T.C. due to documented CSF-pressure lowering effect, and little serious toxicity in patients,Furosemide or other diuretics may
provide an additional benefit to acetazolamide in patients who experience continuing symptoms on acetazolamide.

We recommend against prolonged corticosteroid treatment for treatment of IIH (we also suggest not using serial lumbar punctures for more than three attempts as a primary treatment modality for P.T.C. However, both short-term use of corticosteroids and serial lumbar punctures have been successfully used as short term temporizing measures in patients with rapidly progressive symptoms who are waiting more definitive surgical therapy.

For patients with progressing visual loss, we recommend surgical intervention with CSF shunting procedure and/or optic nerve sheath fenestration (ONSF). The choice of surgical procedure is individualized based upon available expertise and patient preference. We prefer CSF shunting procedure rather than optic nerve sheath fenestration for most patients because of better documentation of efficacy and a lower rate of severe side effects. However, headache response may be superior with shunting.

Patients require regular follow-up visits with serial examinations including visual acuity, formal visual field testing and a fundoscopic examination.

References

2) Foley J: Benign forms of intracranial hypertension: “toxic” and “otic hydrocephalus.”. Brain 1955 78:1-41
7) Suarez J. Critical Care Neurology and Neurosurgery. 1. Humana Press; 2004 {medline}
8) Hedges TR, papilledema, its recognition and relation to increase I.C.P. suryophthalmology 2009{medline}
10) Friedman DI, Jacobson DM, diagnostic criteria for IIH. 2002 PAGE 1492
23) Panagiotis kerezoudis, Evangelos anagnostou, Evangelia kararizou: idiopathic intracranial hypertension: update on the pathogenesis, clinical features and therapy 2013
36) Baker RS, Baumann RJ, Buncie JR. Diagnosis and management of benign intracranial hypertension 1998 [MEDLINE]
40) Mark S Gans, Robert A Egan, MD, Robert A Egan Idiopathic Intracranial Hypertension Workup Apr 1, 2013 [Medscape]
41) D Soler, T Cox, P Bullock, D M Calver, R O Robinson Diagnosis and management of benign intracranial hypertension 1998 updated on January 5, 2014 [british medical journal(BMJ)]


49) Schmidek and Sweet Operative Neurosurgical Techniques: Indications, Methods, and Results 6th edition, by Alfredo Quiñones-Hinojosa


53) Vincent Giuseffi, MD, Michael Wall, MD, Paul Z. Siegel, MD and Patricio B. Rojas, PhD) [official journal of American academy of neurology feb 1991


55) Brain K Owler(benign intracranial hypertension ,disease of nervous system 9th edition London walenr oxford medical publication Brain page 363 1985

56) The relationship between obesity and idiopathic intracranial hypertension. Rowe FJ, Sarkies NJ1999 Jan;23.[PUBMED]

57) Idiopathic intracranial hypertension: Descriptive analysis of 61 patients done by Contreras-Martin Y, Bueno-Perdomo JH performed 2013 Dec 01[pubmed]


62) WAleed F. El-Saadany, Ahmed Farhound, Ihab Zidan (Lumboperitoneal shunt for idiopathic intracranial hypertension: patients’ selection and outcome) Neurosurgical Review April 2012 [springer link] {IVSL}


64) Gary Y. Shaw and Stephanie K. Million :Case Report Benign Intracranial Hypertension: A Diagnostic Dilemma 12 July 2012[pubmed]


66) Exogenous Substances Whose Exposure, Ingestion, or Withdrawal Has Been Associated with Pseudotumor Cerebri table 150-4 chapter 150 youman’s 6th edition page 1707

67) Systemic Illnesses Associated with Pseudotumor Cerebri table 150-5 chapter 150 youman’s 6th edition page 1707

68) Endocrine and Metabolic Disturbances Associated with Pseudotumor Cerebri table 150-3 chapter 150 youman’s 6th edition page 1706


70) Panagiotis kerezoudis,Evangelos anagnostou, Evangelia kararizou:idiopathic intracranial hypertension:update on the pathogenesis, clinical features and therapy 2013

71) Papilledema Grading System (Frisén Scale) TABLE 150-1 chapter 150 youman’s 6th edition page 1702

72) Stages of papilledema according to the Frisén grading scale FIGURE 150-3 chapter 150 youman’s 6th edition page 1703
How to cite this article:
DOI: http://dx.doi.org/10.22192/ijarbs.2017.04.09.011