



DOI: 10.5137/1019-5149.JTN.15846-15.1

Received: 27.08.2015 / Accepted: 13.11.2015

Published Online: 25.05.2016

Original Investigation

Normal Pressure Pseudotumor Cerebri: A Series of Six Patients

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ABSTRACT

AIM: Pseudotumor cerebri (PTC) is a condition characterized by symptoms and signs of increased intracranial pressure (ICP) with no intracranial mass or hydrocephalus, and with normal cerebrospinal fluid (CSF) composition. A variant of PTC known as “Normal Pressure Pseudotumor Cerebri” has the same features of PTC except for normal opening pressure.

MATERIAL and METHODS: This is a series of 6 patients with typical symptoms and signs of increased ICP and normal neuroimaging, but with normal CSF opening pressure. A second lumbar puncture (LP) was done to re-measure the CSF opening pressure.

RESULTS: The mean age of the patients at diagnosis was 25 years (ranging from 19 to 31 years). All patients were female. All patients were obese with a mean body mass index (BMI) of 32.8 (ranging from 30 to 35). The mean duration of symptoms before presentation was 2.8 weeks (ranging from 1 to 8 weeks). The mean CSF opening pressure during the first LP was 11 cm H₂O (ranging from 9 to 15 cm H₂O). The CSF chemistry and culture of all patients were normal. The mean CSF opening pressure in the second LP was 7.3 cm H₂O (ranging from 6 to 10 cm H₂O). All patients were symptom free after one week of treatment, but the medications were continued till the complete disappearance of papilledema and the normalization of the visual field. The mean duration of treatment was 8 weeks (ranging from 5 to 12 weeks).

CONCLUSION: Normal pressure PTC should be treated by medical treatment to avoid visual loss.

KEYWORDS: Pseudotumor cerebri, Papilledema, Acetazolamide

INTRODUCTION

Pseudotumor cerebri (PTC) is a condition characterized by increased intracranial pressure (ICP) with no evidence of intracranial mass or hydrocephalus, and with normal cerebrospinal fluid (CSF) composition (6). PTC patients usually present with typical symptoms and signs of increased ICP, such as headache, vomiting, blurred vision, and papilledema (3).

The neuroimaging abnormalities suggestive of PTC are empty sella, flattening of the posterior aspect of the globe, distension of the perioptic subarachnoid space with or without a tortuous optic nerve, and transverse venous sinus stenosis (3).

ICP is usually measured by lumbar puncture (LP) and a CSF pressure of above 25 cm H₂O is one of the diagnostic criteria

of PTC (3). Green et al. (4) reported a case of PTC with normal ICP, and introduced the term “Normal Pressure PTC” to describe this variant of PTC. Normal Pressure PTC is rare and written in the literature as case reports (1,4,8).

MATERIAL and METHODS

This is a series of 6 consecutive patients with normal pressure pseudotumor cerebri whom I experienced from 2007 to 2014 out of 162 patients with PTC. A detailed medical history was taken with general, neurological, and ophthalmological examinations.

Routine labs, brain magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) were done for the six patients.



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Table I: Patients' Demographic Characteristics

Patients	Age (years)	Sex	Marital state	Pregnant	Body mass index
1	24	Female	Married	In 7 th month	33
2	29	Female	Divorced	Not	34
3	19	Female	Married	Not	30
4	31	Female	Married	Not	35
5	22	Female	Married	In 8 th month	32
6	25	Female	Married	Not	33

Table II: Drug Intake

Patients	Excess Vitamin A	Hormonal contraception
1	No	No
2	No	No
3	No	Yes
4	Yes	Yes
5	No	No
6	Yes	No

LP was done for all patients in the lateral decubitus position using a yellow 20 G spinal needle under local anaesthesia. The CSF opening pressure (OP) was measured. Only 5 cc CSF were withdrawn. CSF chemistry and culture were done. A second LP was done for all patients few days after the first LP to recheck the CSF opening pressure.

Acetazolamide 500 mg per day was started after the first LP and continued till the complete disappearance of papilledema and the normalization of the visual field.

Patients were followed for at least 12 months after the normalization of the visual field to detect any recurrence of symptoms.

■ RESULTS

Six patients are included in this study with a variant of PTC known as "Normal Pressure Pseudotumor Cerebri". This variant comprises 3.7% from the total number of PTC patients seen in 7 years in Ain-Shams University hospitals. All patients were females in the childbearing years with a mean age at presentation of 25 years (ranging from 19 to 31 years). At the time of presentation, two patients were pregnant in their third trimester, one in the 7th month and the other in the 8th month of gestation.

All patients were obese with a mean body mass index (BMI) of 32.8 (ranging from 30 to 35). Patients' demographic characteristics are shown in Table I. Two patients were taking excess vitamins and especially vitamin A for few months as a hair tonic. Two patients were on hormonal contraception (Table II).

One patient had a history of idiopathic intracranial hypertension 7 years ago and the opening pressure was high (42 cm H₂O) and resolved on repeated LPs within 6 months.

The six patients presented to the outpatient clinic in Ain-Shams University hospitals with typical symptoms of increased ICP. The mean duration of symptoms before presentation was 2.8 weeks (ranging from 1 to 8 weeks). All patients had headache due to increased ICP, nausea, and blurring of vision.

Three patients were complaining of horizontal diplopia. One patient complained of rapidly progressive diminution of vision within three days (Table III).

On examination, all patients were fully conscious with normal blood pressure. Three patients had bilateral abducens palsy. All patients had papilledema on fundus examination.

Regarding the visual acuity; two patients were 6/6 in both eyes, three were 6/9 in both eyes, and one was hand motion at 30 cms by both eyes.

Visual field examination using Goldmann perimetry revealed that five patients had enlarged blind spot (Table IV).

All patients had iron deficiency anemia with a mean hemoglobin of 9.5 gm/dl (ranging from 8 to 11 gm/dl). All patients had normal glycosylated hemoglobin and normal coagulation profile.

The brain MRI and MRV of all patients were normal except for empty sella that was present in the six patients.

LP was done for all patients by yellow 20 G spinal needle under local anaesthesia. The CSF OP was measured and only 5 cc CSF was withdrawn. The mean CSF OP was 11 cmH₂O (ranging from 9 to 15 cmH₂O). CSF chemistry and cultures were normal for all patients.

Acetazolamide 500 mg orally daily was started after LP for all patients. All patients complained of low tension positional headache in the first day after LP and improved on bed rest. A second LP was done few days after the first lumbar puncture and the CSF OP was re-checked. The mean CSF OP in the second LP was 7.3 cm H₂O (ranging from 6 to 10 cm H₂O) (Table V).

All patients were symptom free after one week of treatment, but acetazolamide was continued till the complete disappearance of papilledema and the normalization of the visual field.

Table III: Symptoms of the Patients

Patients	Duration of symptoms	Headache	Nausea	Blurring of vision	Diplopia	Decreased vision
1	4 weeks	Present	Present	Present	No	No
2	1 week	Present	Present	Present	Present	Yes
3	8 weeks	Present	Present	Present	Present	Yes
4	1 week	Present	Present	Present	Present	Yes
5	2 weeks	Present	Present	Present	No	No
6	1 week	Present	Present	Present	No	Yes

Table IV: Signs of the Patients

Patients	Papilledema	Visual acuity in right eye	Visual acuity in left eye	Visual field in right eye	Visual field in left eye	Sixth nerve palsy
1	Present	6/6	6/6	Enlarged blind spot	Enlarged blind spot	No
2	Present	HM	HM	Not applicable	Not applicable	Bilateral
3	Present	6/9	6/9	Enlarged blind spot	Enlarged blind spot	Bilateral
4	Present	6/9	6/9	Enlarged blind spot	Enlarged blind spot	Bilateral
5	Present	6/6	6/6	Enlarged blind spot	Enlarged blind spot	No
6	Present	6/9	6/9	Enlarged blind spot	Enlarged blind spot	No

HM: Hand motion.

Table V: Opening Pressures (in cm H₂O)

Patients	In First Lumbar puncture (cm H ₂ O)	In Second Lumbar puncture (cm H ₂ O)
1	12	8
2	10	7
3	9	7
4	11	6
5	15	10
6	9	6

The mean duration of treatment was 8 weeks (ranging from 5 to 12 weeks).

Weight reduction was strongly recommended for all patients. Excess vitamin A was stopped and anemic patients had iron supplement. Changing the method of contraception from hormonal to another one was advised.

No recurrence of symptoms after 12 months of follow up.

■ DISCUSSION

PTC, and its variant "Normal pressure PTC", is an avoidable cause of visual loss. Few diagnostic measures are usually sufficient to determine the correct diagnosis. Most patients

will have complete resolution of symptoms without persistent deficits, if they have regular follow-up visits (6).

Repeat lumbar puncture or 24 hours ICP monitoring is suggested in patients who are suspected of having PTC but have a normal ICP by lumbar puncture (2). All patients in this study had repeated LP to re-check the OP. Only 5 cc CSF was withdrawn during the first LP and the second LP was done days later.

If the OP in the second LP was equal or higher than that in the first LP, this means that the OP in the first LP was not the real ICP pressure and this occurred in some cases that were later diagnosed as PTC with high CSF pressure by continuous ICP monitoring. In this study, the OP in the second LP was much lower than that in the first LP in all patients which confirms that the OP in the first LP was correct and so, there was no need for continuous ICP monitoring.

I suggest to reserve continuous ICP monitoring for those cases that had a normal OP in the first LP and a higher OP in the second LP but still lower than 25 cm H₂O.

The CSF opening pressures in this study does not correlate with the severity of affection of visual acuity or visual field, as the highest CSF OP in the first LP was 15 cm H₂O in a patient without affection of visual acuity and with only enlarged blind spots in the visual field, and the patient who presents with severe affection of visual acuity had an OP of 10 cm H₂O.

Although increased ICP, defined as an opening pressure of over 25 cm H₂O, is one of the diagnostic criteria of PTC,

it should be kept in mind that some patients may have optic discs that are more susceptible to lower ICP than others. Even when a patient's ICP is within the normal range, the possibility of PTC should always be considered in a patient with typical clinical features of PTC, such as, papilledema and blind spot enlargement (8).

In this study, two patients were pregnant and two patients were on hormonal contraception, which suggests an underlying hormonal etiology.

The exact cause of the normal CSF pressure at LP in patients with normal pressure PTC despite the symptoms and signs suggestive of increased ICP is not yet known. The presence of papilledema in these patients indicates increased ICP, which due to a certain cause failed to be transmitted to the spinal subarachnoid space.

Obesity has long been associated with the development of PTC. One theory proposes that obesity predisposes patients to having elevated intraabdominal pressure, elevated intrathoracic pressure, and thus elevated central venous pressure as the final common pathway leading to PTC (7). In this study, all patients at presentation were obese with a BMI ranging from 30 to 35.

Various systemic diseases and especially anemia have been associated with PTC (2). In this study all patients had iron deficiency anemia.

Johnston et al. reported a series of atypical PTC patients, one of which was a 13-year-old boy whose disc edema rapidly resolved after lumboperitoneal shunt insertion even though his initial CSF pressure was normal (5).

Surgical intervention was not offered to the patients in this study as they rapidly improved with lumbar puncture and acetazolamide, and this was especially significant in the patients with severe visual deficit.

The importance of knowing this variant of PTC is to start the medical treatment soon after lumbar puncture to save vision.

■ CONCLUSION

Normal pressure PTC is a variant of PTC that should be treated by medical treatment to avoid visual loss.

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