Prevalence of venous sinus stenosis in Pseudotumor cerebri (PTC) using digital subtraction angiography (DSA)

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Throughout history, many terms have been used to describe the same disease interchangeable as *Pseudotumor cerebri* (PTC), *benign intracranial hypertension* (BIH), *serous meningitis and Otitic hydrocephalus* and *Idiopathic intracranial hypertension* (IIH).6
What is PTC?

According to Modified Dandy Criteria: (1985) Most famous and Used:

1. Symptoms and signs of increased intracranial pressure.
2. No Localizing in neurological examination (Except sixth cranial nerve lesion or rarely other false localizing signs).
3. Normal neuroimaging (CT brain, MRI brain).
4. Increased intracranial pressure as measured by lumbar puncture (250 mmH2O).
5. Normal CSF constituents.
6. Awake alert patient.
7. No other Causes of increased intracranial pressure present.
8. Benign clinical course apart from visual deterioration.
Most recently, Friedman and Jacobson (2002):

1. If symptoms present, they may only reflect those of generalized intracranial hypertension or papilledema.
2. If signs present, they may only reflect those of generalized intracranial hypertension or papilledema.
3. Documented elevated intracranial pressure measured in lateral decubitus position.
4. Normal CSF composition
5. No evidence of brain lesion by MRI AND MRV
6. No other cause of I.I.H.
Is it really Benign?

It is benign as regard the etiology, but 30% of cases ends with visual loss.
• Is it really idiopathic?

• Elevated intracranial venous pressure seems to be of importance in PTC either as a cause (secondary intracranial hypertension) or as a consequence (idiopathic intracranial hypertension) of increased intracranial pressure. 8
Incidence

- **Rhadharkrishnan et al (1993)** estimated annual incidence in Benghazi, Libya in a study conducted between 1982 and 1989 as 2.2 per 100,000 populations.
- Female to male ratio is 8:1.
- 21.4 per 100,000 obese women aged 15-44.

- **Corbett et al., (1988):** Reported that age group ranging from 1 to 67 years with peak incidence in the *third* decade.
**Pseudotumor cerebri (PTC)** is a neurological disorder presenting with symptoms of increased intracranial pressure (headache, visual disturbances, papilledema) without localizing neurological findings in awake alert patient.

- Various pathogenic mechanisms have been considered to explain the raised intracranial pressure in those patients.

- *The role of such venous disease* in PTC has been revisited as several groups using invasive monitoring, have documented high pressure in the venous sinuses in typical cases.
It appears to be the result of focal stenotic lesions in the dural sinuses obstructing the venous outflow. This has led to the suggestion that undetected intracranial venous hypertension may after all be the substrate for PTC.6

Aim of the study

In our study we aimed at evaluating the prevalence of venous sinus stenosis along patients of PTC.
**Materials and Methods**

This study was conducted on THIRTY patients with symptoms and signs confirming the diagnosis of PTC.

- Patients were recruited from the neurology, neurosurgery departments and outpatient clinics.

- **Inclusion criteria were:** *(Modified Dandy Criteria)*

- **Exclusion criteria:**

  1. Patients with true localizing findings on examination denoting focal brain dysfunction.

  2. Patients with traumatic, neoplastic, infectious, structural or iatrogenic causes of intracranial hypertension.

  3. Patients with clinical and neuroimaging evidence of acute primary dural sinus thrombosis or cortical vein thrombosis.
Methods:
All patients included in this work were subjected to the following:
1) Complete general and neurological assessment.
2) Measurement of body mass index: (BMI)
3) Full ophthalmologic assessment included:
   A. Visual acuity measurement: using Snellen chart.
   B. Direct and indirect ophthalmoscopic fundus examination:
      To assess and grade papilledema.
   C. Automated perimetry:
4) Full Laboratory investigations
5) Lumbar puncture (LP).
Radiological investigations:

a. **CT scan brain +/- MRI brain without contrast.**

b. **Magnetic resonance venography (MRV)** of the intracranial venous system by time of flight (TOF) or phase contrast techniques.

c. **Digital subtraction cerebral Angiography (DSA) (venous phase):**

7) **Statistical methodology:** Analysis of data was done by IBM computer using SPSS (statistical program for social science) (version 10)
Results

- MRV brain showed that 24 patients (80%) showed filling gaps. Digital subtraction cerebral angiography (venous phase) showed 9 patients (30%) had stenosis in their dural sinuses.

- MRV showed to be a good screening tool since it had 100% sensitivity and negative predictive value.

- However, since it has a moderate specificity (62%) with a positive predictive value (PPV) of only 35%, then lesions detected should be confirmed with digital subtraction cerebral angiography (venous phase) particularly those involving the transverse and sigmoid sinus (see Fig. 1A).
Table 5 Validity of MRV in relation to DSA in detection of sinus stenosis.

<table>
<thead>
<tr>
<th>Variables</th>
<th>%</th>
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<tbody>
<tr>
<td>Sensitivity</td>
<td>100%</td>
</tr>
<tr>
<td>Specificity</td>
<td>62%</td>
</tr>
<tr>
<td>PPV</td>
<td>35%</td>
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<tr>
<td>NPV</td>
<td>100%</td>
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• MRV was found to be a good screening tool since it had 100% sensitivity and negative predictive value. Therefore, if MRV is normal no further investigations are needed.

• However, since it has a moderate specificity (62%) with a positive predictive value (PPV) of only 35%, then lesions detected should be confirmed with digital subtraction cerebral angiography (venous phase) particularly those involving the transverse and sigmoid sinus.
Fig. (1): (A) MRV with a filling defect in the Rt transverse sinus suggestive of stenosis with aplasia of the left transverse and sigmoid sinus.

(B) Digital subtraction cerebral angiography (venous phase) oblique view showing the tight stenosis (99%) of the distal part of the right dominant transverse sinus with aplasia of the left transverse and sigmoid sinus.
Figure (49): MRV of case number 1.
Figure (50): Digital subtraction cerebral angiography (Venous phase)
Anteroposterior view in case 1.
Studying the intracranial venous system in patients with PTC is an important helps in understanding the pathophysiology of the disease, expecting response to medical and surgical treatment.

Also detection of venous sinus stenosis may open the way to a novel therapeutic option for refractory patients.
1. The role of venous sinus stenosis should be strongly considered in the etiology of PTC.

2. Although modified DANDY criteria, considered being standard criteria for diagnosis of PTC, yet it is criticized in two main points. It missed the atypical types of PTC, secondly it underestimates the role of venous sinus pathology especially stenosis. So it is possible to add additional point to these criteria, which is "Normal venous phase of digital subtraction cerebral angiography". For proper diagnosis of PTC.
3. Digital subtraction cerebral angiography (venous phase) could be a standard investigation in PTC patients especially in patients with bilateral or unilateral dominant sinus filling gaps at the level of MRV.

4. A new protocol could be designed in the management of pseudotumor cerebri patients.
Established case of PTC
(+ Ve symptoms & signs)

Normal CT brain & Normal MRI Brain

MRV

+Ve bilateral filling gaps in bilateral transverse sinuses
OR unilateral filling gaps in dominant transverse sinus

No filling gaps in any transverse sinus

No need for digital subtraction cerebral angiography (venous phase)

Confirmation of validity of filling gaps by

Digital subtraction cerebral angiography (venous phase)

+Ve stenosis

Venous sinus stenting should be considered instead of shunting in cases with failed medical treatment & negative to other risk factors (as in cases 2, 4, 5)

- Ve stenosis

other surgical modalities could be used

Fig (62): Proposed protocol for management of PTC
THANKS
References


It has been published in THE EGYPTIAN JOURNAL OF NEUROLOGY AND NEUROSURGERY, VOL. 45 (1) - JAN 2008.


It will be represented on 16 th EFNS in Stockholm and re-published in EUROPEAN NEUROLOGY JOURNAL.