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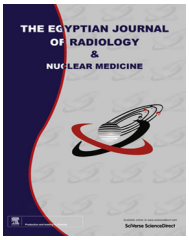

ORIGINAL ARTICLE

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

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KEYWORDS

Pseudotumor cerebri;
Venous sinus stenosis;
Headache;
MRV;
Digital subtraction angiography (venous phase)

Abstract *Objectives:* To study the prevalence of intracranial venous stenosis in Pseudotumor cerebri patients.
Patients and methods: Thirty patients were diagnosed having PTC according to Dandy criteria. All underwent general and neurological assessment. Radiological assessment included CT scan brain ±MRI brain without contrast, MRV. All underwent digital subtraction angiography (DSA) (venous phase) to confirm the validity of filling gaps seen at the level of MRV.
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.
Conclusion: Studying the intracranial venous system in patients with PTC is an important step in understanding the pathophysiology of the disease. Detection of venous sinus stenosis opens the way to a novel therapeutic option for refractory patients like venous sinus stenting.

1. Introduction

Pseudotumor cerebri (PTC) is a neurological disorder presenting with symptoms of increased intracranial pressure (headache, visual disturbances, papilledema) without localizing neurological findings in an alert patient.

Various pathogenic mechanisms have been considered to explain the raised intracranial pressure in those patients. These included increased cerebral volume, bio-hormonal

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mechanisms, disturbed normal CSF production/absorption balance, and increased cerebral venous pressure (1,2). None appears satisfactory on its own; however, it is well known that symptoms of PTC can be mimicked by cerebral venous sinus thrombosis and venous outflow obstruction (3). However, in practice not enough attention is paid to the role of venous disease in the etiology of PTC (4).

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 (5). 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). King et al. (5) using venography, reported narrowing of the transverse sinuses with either smooth tapering of uncertain cause or intraluminal filling defects suggestive of a mural thrombus in some patients with PTC (7).

In our study we aimed at evaluating the prevalence of the venous sinus disease in the etiology of PTC using MRV and digital subtraction cerebral angiography (venous phase).

2. Patients and methods

2.1. Subjects

This study was conducted on 30 Egyptian patients with symptoms and signs of PTC. Patients were recruited from the neurology, neurosurgery departments and outpatient clinics of Ain-Shams University Hospitals. Ethics Committee approved the study and patients were informed about the details of the study and written consents were obtained.

2.1.1. Inclusion criteria were:

Modified Dandy criteria (7).

2.1.2. 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.

2.2. Methods

All patients included in this work were subjected to the following:

1. Complete general and neurological assessment.
2. Lumbar puncture (LP).
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. 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 angiography (DSA) (venous phase).
5. Statistical methodology: Analysis of data was done by IBM computer using SPSS (statistical program for social science) (version 10).

2.3. 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).

3. Discussion

Pseudotumor cerebri (PTC), also known as benign intracranial hypertension (BIH), is a syndrome seen more frequently in young obese women characterized by symptoms and signs of increased intracranial pressure in the absence of an intracranial mass lesion, infection, or hydrocephalus in otherwise healthy and alert patient. The exact etiology of benign intracranial hypertension is still unknown. However, various pathological conditions may be associated with this syndrome. These include endocrine and metabolic disorders, intracranial venous sinus thrombosis, drugs and toxins, hematological and connective tissue disorders, high cerebrospinal fluid (CSF) protein content, "meningism" with systemic bacterial or viral infections, and empty sella syndrome (11).

The mechanism of increased intracranial pressure in these disorders is still unclear. Involvement of the venous flow was

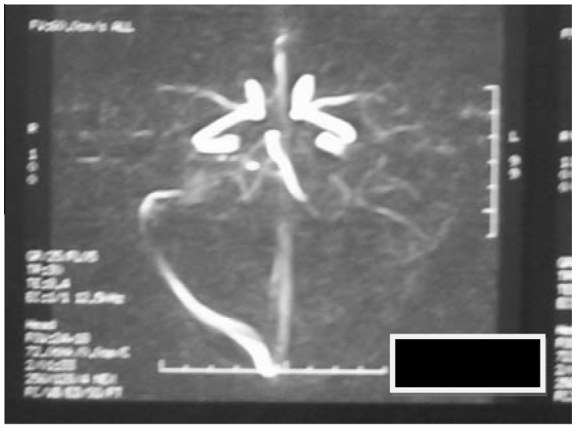

Fig. 1 (A) MRV with a filling defect in the Rt transverse sinus suggestive of stenosis with aplasia of the left transverse and sigmoid sinus.

Fig. 2 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.

presumed to be one of the causative mechanisms as any disorder causing a rise in venous pressure will secondarily have an effect on CSF absorption (5,6,12,13). In line with this, there is ample evidence from infusion and perfusion studies that PTC is associated with an impairment of CSF outflow (14).

Elevated intracranial intravenous pressure seems to be of importance in Pseudotumor cerebri syndromes, either as a cause (secondary intracranial hypertension) or as a consequence of increased intracranial pressure with compression of dural sinus (idiopathic intracranial hypertension) (5). Lesions which increase venous sinus pressure (dural arteriovenous malformations), or impede venous drainage (for example, venous sinus thrombosis) are known to give rise to the same syndrome as PTC (3).

King et al. (5), Sugerman et al. (8), Karahalios et al. (6) have proposed that increased intracranial venous pressure is the major mechanism of raised intracranial pressure in PTC. King et al. (5) performed their study in PTC patients and they found abnormalities in the venous phase of cerebral angiograms. Venography showed narrowing of the transverse

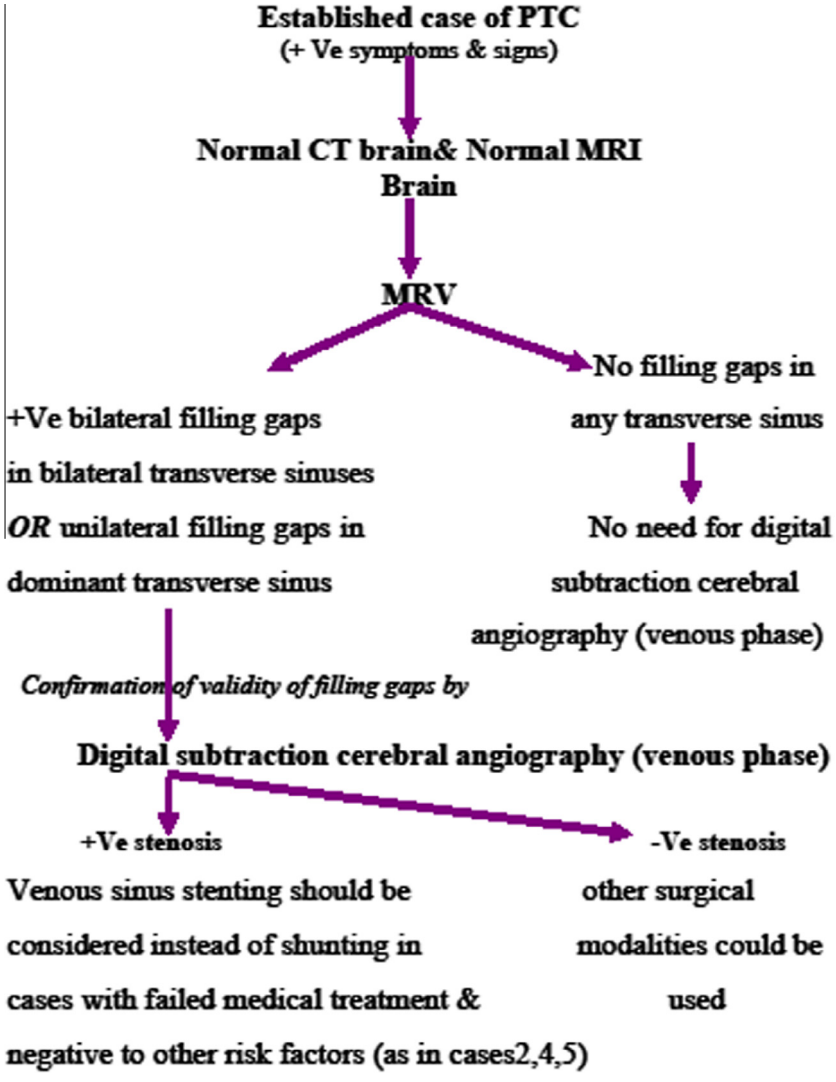

Fig. 3 New protocol could be designed in the management of Pseudotumor cerebri patients.

sinuses with either smooth tapering of uncertain cause, or intraluminal filling defects suggestive of mural thrombosis. In addition, manometry had documented raised pressures in the superior sagittal sinuses and proximal transverse sinuses, with a drop in pressure in the distal transverse sinuses (9,10).

In our study, MRV brain showed 24 patients (80%) with filling gaps. Digital subtraction cerebral angiography (venous phase) showed 9 patients (30%) had stenosis in their dural sinuses.

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%, lesions detected should be confirmed with digital subtraction cerebral angiography (venous phase) particularly those involving the transverse and sigmoid sinus (see Fig. 2).

Therefore, in our study we classified the patients into 2 main groups depending on the results of digital subtraction cerebral angiography (venous phase). Twenty-one patients (70%) were considered to be true idiopathic intracranial hypertension since they had normal intracranial sinuses. On the other hand, the remaining 9 patients (30%) with transverse/sigmoid sinus stenosis were considered to have symptomatic intracranial hypertension according to the new classification of Corbett (12).

This emphasizes the importance of studying the intracranial venous system in patients with PTC as it may help in understanding the pathophysiology of the disease in certain cases. Also digital subtraction cerebral angiography (venous phase) could help in determining the cause of stenosis as in one of our cases where there was irregular narrowing seen in the right transverse and sigmoid sinus suggestive of partial recanalization of an old sinus thrombosis, although the patient's past history was negative to sinus thrombosis (see Fig. 3).

In conclusion, studying the intracranial venous system in patients with PTC is important as it 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.

4. Recommendation

1. The role of venous sinus stenosis should be strongly considered in the circumstance of PTC.
2. Digital subtraction cerebral angiography (venous phase) is a corner stone investigation in PTC patients especially in patients with bilateral or unilateral dominant sinus filling gaps at the level of MRV.

Conflict of interest

None.

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