ENDOVASCULAR TREATMENT OF IDIOPATHIC INTRACRANIAL HYPERTENSION

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ABSTRACT

BACKGROUND: Idiopathic intracranial hypertension (IIH) causes increased intracranial pressure without a mass lesion or hydrocephalus. It is caused by venous sinus obstruction in an unknown percentage of cases. In these cases, high intracranial venous sinus pressures appears to be the result of a focal venous sinus lesion causing partial or complete obstruction to cranial venous outflow. **CASE DESCRIPTION:** We describe an alternative treatment of venous sinus obstruction using angioplasty and stenting in a young, obese woman with refractory idiopathic intracranial hypertension. Magnetic resonance venography (MRV) was suggestive of Transverse sinus stenosis, which was confirmed by venography and manometry that showed raised pressures across the venous stenosis. A self-expanding stent was deployed across the venous stenosis immediately reducing the pressure gradient, with a striking clinical improvement. **CONCLUSIONS:** Angioplasty and stent placement should be considered as a treatment option to refractory idiopathic intracranial hypertension associated with dural sinus stenosis. The long-term effectiveness of sinus stenting in IIH seems to be good, but still remains uncertain.

Key Words: Idiopathic intracranial hypertension; venous sinus obstruction; stenting; endovascular treatment

Introduction

The syndrome of increased intracranial pressure without hydrocephalus or mass lesion and normal Cerebrospinal fluid (CSF) composition, previously referred as pseudo tumor cerebri or benign intracranial hypertension, is a diagnosis of exclusion now termed idiopathic intracranial hypertension (IIH).[1] The diagnostic criteria was first described by Dandy in 1937[2] and later modified by Smith in 1985[3] and Friedman et al in 2002.[1] The disease has predominance for obese women, in childbearing years. Classic symptoms and signs include headache, nausea, vomiting, papilledema, visual obscurations and abducens nerve palsy. Vision loss begins peripherally and may not be noticed by patients until late in the course. Permanent vision loss is the most serious consequence.[4]

Although the pathophysiology of IIH is uncertain, various mechanisms have been proposed: brain swelling, increased CSF production, decreased CSF absorption, undiagnosed venous thrombosis and venous outflow obstruction.[5]

Disorders like Addison's disease, hypoparathyroidism, iron deficiency anemia, renal failure, sleep apnea, corticosteroid withdrawal and Vitamin A toxicity have also been associated with IIH. Dural sinus thrombosis and anomalous venous drainage patterns, described in some forms of craniosynostosis, can cause clinical presentation very similar to if not identical to IIH and, as a consequence, diagnostic studies of venous system have become important in the workup of these patients to exclude underlying venous sinus pathologies.[1,6]

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Treatment is aimed at reducing pressure and relieving symptoms. Weight loss, Acetazolamide, NSAIDs and symptomatic drugs used are effective measures in a high percentage of cases. If vision deteriorates despite treatment, optic nerve sheath fenestration, shunting (lumboperitoneal or ventriculoperitoneal) may be indicated. Farb et al conducted a prospective controlled study with 29 patients with established IIH and 59 control patients, using auto-triggered elliptic-centric-ordered three-dimensional gadolinium-enhanced MR venography. He found substantial bilateral venous stenosis in 27 of 29 patients with IIH and in only 4 of 59 control patients.[7] Karahalios et al. studied 10 patients with IIH with conventional venography and manometry and showed that half of the patients had angiographic venous outflow obstruction, and all had elevated venous pressures, collaborating with the hypothesis that venous hypertension is related with IIH.[4] Malek et al demonstrated in an early report that sinus recanalization with angioplasty with stenting was effective to treat refractory venous thrombosis.[8] In the selected cases that outlet obstruction is demonstrated, endovascular venous stenting may be indicated.

Case Report

A 23-year-old woman (body-mass index 30.5 kg/m2) was referred with a history of chronic headache and visual changes. Neurologic examination was normal, apart from bilateral papilledema (Fig. 1).

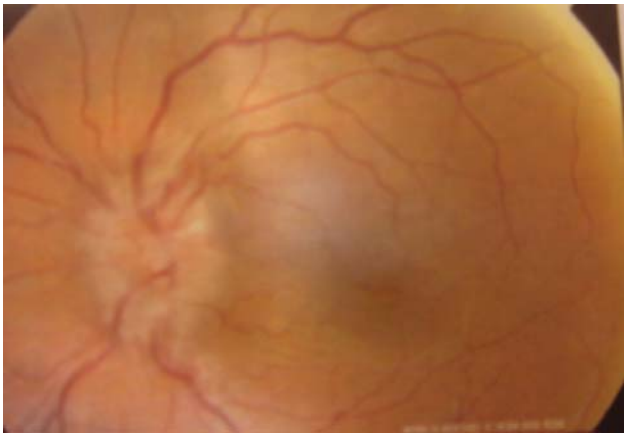

Figure 1: Fundoscopic examination showing papilledema.

Conventional magnetic resonance imaging (MRI) was normal; MR Venography (MRV) was suggestive of left transverse sinus stenosis (Fig. 2).

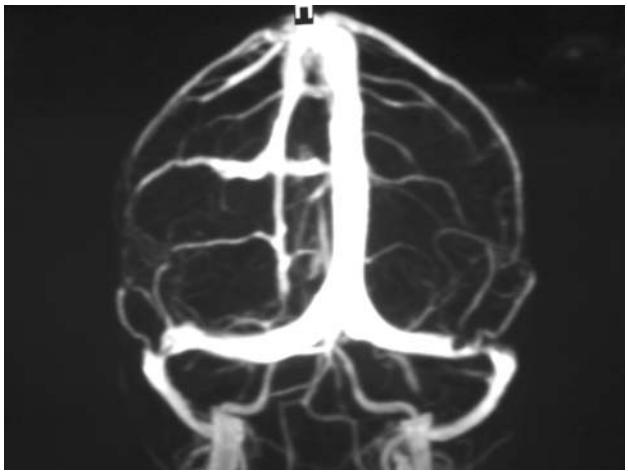

Figure 2: MRV suggesting a stenotic lesion in the left transverse sinus. The right transverse sinus filling defect was considered as a Pacchionian granulation, and showed no pressure gradient.

Cerebrospinal fluid (CSF) pressure was elevated at 50-60cm H2O in several occasions, consistent with benign intracranial hypertension. Symptoms remained after medical treatment with Acetazolamide and weight reduction. As medical measures and serial lumbar punctures gave only temporary symptomatic relief, endovascular treatment was considered.

Venous catheterization confirmed a stenotic lesion in the left transverse sinus (Fig. 3) and venous pressure gradients measurements were obtained.

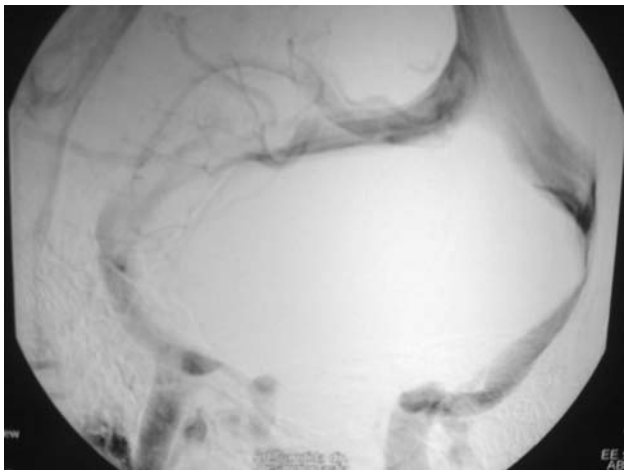

Figure 3: Digital subtraction angiography, venous phase, demonstrating a stenosis in the left transverse sinus.

A 23mmHg pressure gradient was found (mean 51mmHg pre-stenosis and 28 mm Hg post-stenosis) (Fig. 4).

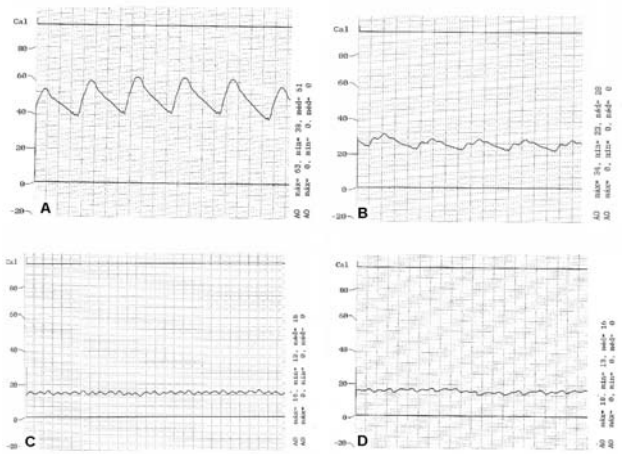

Figure 4(a)**:** Pre-procedure, left transverse sinus - pressure measurement distal to the stenosis (systolic pressure 68mm Hg/Diastolic pressure 38mmHg/mean pressure 51mmHg); (b) Pre-procedure left transverse sinus - pressure measurement proximal to the stenosis (systolic pressure 34mm Hg/Diastolic pressure 23mmHg/mean pressure 28mmHg); (c) Post-procedure, left transverse sinus - pressure measurement (systolic pressure 16mm Hg/ Diastolic pressure 13mmHg/mean pressure 15mmHg); (d) Right transverse sinus pressure measurement (systolic pressure 18mm Hg/Diastolic pressure 13mmHg/mean pressure 16mmHg)

The endovascular procedure was done by femoral route using a 6Fr sheath to deploy a self-expanding stent (Easy Wallstent, Boston Scientific, Watertown, USA) across the venous stenosis. After stenting, no significant pressure gradient was noticed (Fig. 5). Patient was heparinised only during the procedure and kept on aspirin after an early discharge. The symptoms rapidly subsided, and the papilledema resolved in serial clinical controls.

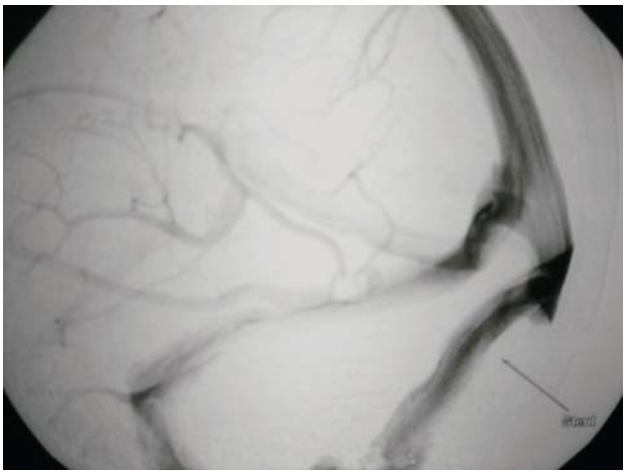

Figure 5: Immediate angiographic control after stenting showing complete resolution of the stenosis of the left transverse sinus.

Discussion

Raised intracranial pressure (ICP) has been associated with partial venous outflow obstruction in the lateral sinuses.[5,9] There has been a discussion over cause and effect of these phenomena. Scoffings et al reported a case where the stenosis resolved immediately after CSF withdrawal.[10] However, patients treated with CSF diversion procedures not always experience a relief in symptoms.[7] CSF diversion procedures are considered after failure of medical treatment but are associated with substantial morbidity and often are only partly effective; their goal is to reduce intracranial pressure. If the cause of IIH is venous hypertension, the main problem is not being accessed.

Not long past, digital subtraction angiography was performed to exclude other pathologies of intracranial disease and not specifically to interrogate the venous system, thus, subtle venous outflow findings were likely overlooked.[11,12] MR is the modality of choice for analyzing the venous system in patients suspected of having IIH to exclude venous sinus thrombosis or stenosis.[13,14] TOF MR angiography is the most currently used technique of MR venography despite the documented limitations of the technique,[15] specifically artifactual signal losses that occurs at certain locations due to in-plane flow and turbulence.[7] TOF MRV has difficulty evaluating the distal transverse sinus: whether normal, stenosed, or thrombosed. This pitfall of earlier MRV technique has been a source of great difficulty in trying to define the role of MRV in IIH.

We believe venous sinus stenosis plays important role in symptoms. Stenting of the affected venous sinuses has been proposed as an alternative treatment to CSF diversion procedures.[16] Higgins et al reported 12 patients with refractory IIH associated to venous stenosis treated with stenting, without procedure-related complications. Of 12 patients treated, five became asymptomatic, two improved but had residual headache, and five remained unchanged.[7]

Our patient has been followed for over 1 year, with complete resolution of the symptoms and papilledema. An immediate reduction in the pressure gradients after stenting was observed. This indicates a direct relation between the procedure and disappearance of clinical symptoms, implying that venous outflow obstruction

can, at least sometimes, be responsible for IIH. The need of invasive methods to make an accurate diagnosis of sinus stenosis raises the question that we might have underestimated this pathology as causative of IIH.[7]

In the view of these findings, angioplasty and stent placement should be considered as a treatment option to refractory idiopathic intracranial hypertension associated with dural sinus stenosis. The long-term effectiveness of sinus stenting in IIH seems to be good, but still remains uncertain.

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