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Neuro-ophthalmology Update

Cerebral venous sinus stenting for pseudotumor cerebri A review

Sivashakthi Kanagalingam [a], Prem S. Subramanian [a,b,*]

Abstract

Pseudotumor cerebri is characterized by headaches, visual field changes, papilledema and an elevated cerebrospinal fluid opening pressure without evidence of an intracranial mass. In the setting of failed medical therapy, surgical options such as ventriculoperitoneal shunts and optic nerve sheath fenestrations are considered. Recently, venous sinus stenting has emerged as a new treatment option for patients with pseudotumor cerebri. We review the role of cerebral venous sinus stenting in the management of patients with medically refractory pseudotumor cerebri. Although long- term studies are needed in this field, the current reports indicate a favorable outcome for preventing vision loss and symptom control.

Keywords: Pseudotumor cerebri, Idiopathic intracranial hypertension, Venous sinus stenting, Vision loss, Papilledema

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Introduction

Pseudotumor cerebri (PTC) is a syndrome characterized by raised intracranial pressure (ICP) in the absence of space-occupying intracranial lesions on imaging, elevated CSF opening pressure of >25 cm of water, and normal CSF composition.[1,2] The annual incidence in the general population is approximately 1–2 per 100,000 in North America.[3,4] However, in obese women of ages 22–44 years the incidence surges to 14.9–19.3 per 100,000.[3] Clinically, as many as 90% of patients experience headaches,[5] while 70% experience transient visual obscurations and pulsatile tinnitus.[5,6] Binocular horizontal diplopia can also occur in the setting of unilateral or bilateral sixth nerve palsy, a non-localizing sign secondary to raised intracranial pressure. Papilledema seen in these patients is usually bilateral but may be asymmetric.[7] Associated vision loss can be severe in up to 25% of patients, with blindness reported in 10% of cases.[8–10]

Conventional treatments have been aimed at controlling the headaches and preventing permanent vision loss from ensuing. Given the integral relationship between obesity and PTC, weight loss remains the most important aspect of PTC management, with as little as 5–10% of total body weight loss having been found to be effective in symptom control and papilledema improvement.[11] However, weight loss is a long-term lifestyle modification and an ineffective immediate therapy. Medical treatments that include carbonic anhydrase inhibitors such as acetazolamide and to piramate are frequently used. A recent multi-center, randomized double-masked trial established that acetazolamide in conjunction with weight loss led to better and more rapid improvement in visual fields and papilledema grade than did diet alone. Nonetheless, with acetazolamide and to piramate, patients often report paraesthesias, altered taste sensation, and lethargy.[12]

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[a] Department of Ophthalmology, The Johns Hopkins Hospital, Baltimore, MD, USA
[b] Department of Neurosurgery, The Johns Hopkins Hospital, Baltimore, MD, USA

* Corresponding author at: Wilmer Eye Institute, Johns Hopkins Hospital, Woods 457, 600 N. Wolfe Street, Baltimore, MD 21287, USA. Tel.: +1 410 502 3213; fax: +1 410 502 3214.
e-mail address: psubram1@jhmi.edu (P.S. Subramanian).

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Occasionally, oral steroids are adopted in the treatment of the fulminant variant of PTC.[13] However, its side effect profile includes weight gain, making this a poor treatment choice in obese patients.

Surgical intervention is required for the subset of patients who continue to experience intractable headaches and progressive vision loss despite medical therapy. Customarily, optic nerve sheath fenestration (ONSF) is preferred in patients with vision loss due to severe papilledema with relatively mild or no other symptoms of increased ICP, whereas CSF diversion procedures (e.g., ventriculoperitoneal and lumboperitoneal shunting) are preferred in patients with visual loss, papilledema, and significant systemic symptoms of increased ICP such as headache.[14,15] However, these modalities are not without their pitfalls. ONSF carries a risk of vision loss, pupillary and motility dysfunction, and up to a 32% failure rate with recurrence of visual symptoms in PTC patients.[16] Shunting procedures have been associated with shunt migration, infection, intra-cerebral hemorrhage, and acquired Chiari malformation.[17,18] Shunt failure and revision rates have been reported as being as high as 60% for lumboperitoneal shunts, and 30% for ventriculo-peritoneal shunts.[19]

Anatomic abnormalities of the cerebral venous sinuses have been identified in a number of patients with PTC, and venous sinus stenting has emerged in recent years as an alterative treatment modality for these patients. We review the current literature to assess the role of cerebral venous sinus abnormalities in the pathogenesis of PTC and the potential benefit of interventional treatment.

Pathophysiology

The mechanisms that underlie PTC are poorly understood and have been subject to long standing debate and speculation. Prior theories have proposed increased CSF production or decreased CSF absorption as an underlying etiology. Recently, intracranial venous hypertension associated with venous sinus stenosis has been implicated as a possible mechanism for PTC.[20–22] Cerebral venous sinus thrombosis represents the extreme variant of such a phenomenon and will not be discussed in this article, as its treatment is quite dissimilar (anticoagulation rather than physical relief of the obstruction). Rather, severe narrowing of one or more venous outflow channels would increase the pressure gradient across which CSF is resorbed at the arachnoid granulations and cause increased ICP. Narrowing occurs most commonly at the distal transverse sinus or transverse/sigmoid sinus junction, either unilaterally or bilaterally. Anatomic variability of the cerebral venous sinuses is well described, and venous sinus stenosis is distinct from these normal findings. The confluences of the sinuses at the torcular herophili drain into each transverse sinus asymmetrically. The right transverse sinus is usually larger and drains the superior sagittal sinus, whereas the smaller left transverse sinus usually drains the straight sinus.[22] Anatomical studies with cadaver dissections have observed the presence of septa, especially larger septa, which could be an etiological factor in the development of an intrinsic pattern of venous stenosis resulting in PTC.[22] The venous sinuses are also a site for arachnoid granulations, which are extensions of arachnoid mater and subarachnoid space through the wall of the dural venous sinuses.[22,23] They increase in number with advancing age[24–26] and are thought to play a role in the resorption of CSF.[27,28] They are observed in the transverse and sigmoid sinuses[28] and can cause focal intra-luminal filling defects in 24% of CT examinations and 13% of contrast-enhanced MR studies in normal populations.[29] When arachnoid granulations are enlarged, they can obstruct either one or both transverse sinuses. It is thought that this obstruction creates a resistance to flow, with ensuing decreased absorption of the CSF and a concomitant increase in intracranial pressure.[28,29] As discussed below, relief of obstruction by a hypertrophic arachnoid granulation has been shown to relieve PTC symptoms and signs.

There is also evidence to suggest that transverse sinus stenosis may occur as a secondary phenomenon in response to elevated intracranial pressure. King et al.[30] described the reduction of the venous pressure in the superior sagittal and transverse sinus, disappearance of the pressure gradient across the transverse sinus, and resolved stenosis with CSF drainage. De Simone et al.[31] presented a case of bilateral transverse sinus stenosis without any evidence of flow gaps. Reversal of the stenosis was noted just 24 h after the removal of 20 mL of CSF. Another reported patient[32] with an opening pressure of 50 cm H20 had magnetic resonance venography (MRV) immediately before and 15 min after a lumbar puncture (LP). Partial resolution of the transverse sinus narrowing was detected with CSF drainage. A subsequent third LP reduced the pressure to 8 cm H20 and showed complete resolution of the stenosis. These findings have been reproduced by others[33–35] who have reported a similar pattern of reversal of the venous sinus stenoses either by means of lumbar puncture or by CSF shunting.

Some authors[36,37] have suggested the presence of a positive feedback mechanism in linking these physiological processes. Remodeling of the transverse sinus wall in response to sustained external compression from elevated ICP may lead to fibrosis and formation of a fixed narrowing that cannot reverse even with normal ICP. Changes in the bony groove occupied by the transverse sinus have been reported in patients with chronic ICP elevation and venous sinus stenosis.[38] Regardless of the initial precipitating cause of the focal stenosis, it is suggested that a cyclical mechanism of sinus stenosis and venous hypertension further reduces CSF absorption, raises ICP and causes worsening venous stenosis.

Irrespective of these conflicting reports, it is apparent that a venous stenosis is somehow an important element in the PTC progression, be it causative or resultant. Developments and refinements in MRV imaging have now revealed that a majority of PTC patients have transverse sinus narrowing. Farb et al.[39] utilized a high resolution, auto triggered elliptic-centric-ordered (ATECO) 3-dimensional, gadolinium enhanced MRV to detect venous stenosis. The conventional use time-of-flight MR venography (TOF MRV), a 2-dimensional system, frequently suffers from artifacts in the region of the distal transverse sinus because of in-plane, turbulent, and tortuous flow.[40] These artifactual signal losses may be a reason as to why the role of venous stenosis in PTC was not identified or recognized in earlier literature. Higgins et al.[41] reanalyzed the MRVs of twenty PTC patients that were initially interpreted as normal. Bilateral lateral sinus flow gaps were identified in 13 of 20 patients with PTC, and in none of 40 controls. Kumpe et al.[42] reported that although