Endovascular Stenting for Idiopathic Intracranial Hypertension

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ABSTRACT

Transverse sinus stenosis (TSS) is often observed in patients with idiopathic intracranial hypertension (IIH). Studies show that symptoms of IIH can be resolved by transverse sinus stenting. We report a case of a 39-year-old woman who presented with several weeks of progressive headaches and visual disturbances. Ophthalmologic examination demonstrated papilledema and imaging features confirmed the diagnosis of IIH. The need for interventional management was necessitated by the preceding failure of several months of medical treatment. Her vascular imaging demonstrated stenosis of the transverse sinus and her intracranial venous pressure measurements indicated elevation with a high pressure gradient across the stenosis. She underwent transverse sinus stent placement across the stenotic segment. After this intervention, her symptoms improved and her intracranial pressure normalized. The imaging follow-up revealed efficacious patency of the stent. Based on a mathematical model, we suggest that a Starling-like resistor demonstrating a collapsible transverse sinus can permanently be replaced by a rigid-walled sinus upon employment of an endovascular stent in the stenotic transverse sinus, preventing the recurrence of IIH. This novel method of transverse sinus stenting provides a less invasive, effectual alternative to other high-risk surgical procedures currently used for IIH patients.

RÉSUMÉ

La sténose du sinus transverse (SST) est souvent observée chez les patients souffrant d'hypertension intracrânienne idiopathique (HII). Des études démontrent que les symptômes de l'HII peuvent être résolus avec l'installation d'endoprothèses (stents) dans le sinus transverse. On signale le cas d'une femme de 39 ans qui souffrait depuis plusieurs semaines de céphalées progressives, accompagnées de troubles de la vue. L'examen ophtalmique a démontré de l'œdème papillaire et l'imagerie a confirmé le diagnostic d'HII. L'échec du traitement médical durant plusieurs mois a nécessité une prise en charge interventionnelle. Son imagerie vasculaire a démontré une sténose du sinus transverse. La mesure de la pression veineuse intracrânienne indiquait une hypertension démontrant un gradient de pression à travers de la sténose. On a installé une endoprothèse dans le segment sténosé du sinus transverse. Après l'intervention, ses symptômes se sont améliorés et sa pression intracrânienne s'est normalisée. L'examen de suivi par imagerie a démontré la perméabilité efficace de l'endoprothèse. En fonction d'un modèle mathématique, nous suggérons qu'une résistance de type Starling démontrant un sinus transverse collabable pourrait être remplacée en permanence par un sinus rigide en installant une endoprothèse vasculaire dans le sinus transverse sténosé, prévenant ainsi la récurrence de l'HII. Ce nouveau modèle d'installation d'une endoprothèse dans le sinus transverse est un choix moins invasif et plus efficace que les autres méthodes chirurgicales à risque élevé qui sont utilisées actuellement auprès des patients souffrant d'HII.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a syndrome of elevated spinal fluid pressure, in the absence of any tumors or other diseases [1]. Numerous associations related to the occurrence of IIH have been established [1,2], including stenosis of the transverse sinus [3]. The transverse sinus, located at the base of the brain, is one of the venous sinuses that form the major drainage pathway from the brain to the internal jugular vein. Since the cerebrospinal fluid (CSF) drains into the transverse sinus, narrowing or 'stenosis' of this venous channel causes fluid backup and elevates the CSF pressure (intracranial pressure - ICP). This increased ICP, if left untreated can lead to damage of the optic nerve, resulting in blindness. The diagnosis of IIH typically includes swelling of the optic disc (papilledema) and an increased CSF opening pressure during the lumbar puncture, in the absence of mass lesions [1]. Although the disease does occur in men and older women, most of the IIH patients are obese women of childbearing age presenting with symptoms such as nausea, vomiting, headache, and visual disturbances. While the prognosis is generally good, the principal morbidity associated with this disease is vision loss [4,5]. Although the pathophysiology of raised ICP in IIH remains unclear, numerous mechanisms have been proposed, including augmented rate of CSF production, prolonged increase in intracranial venous pressure, decreased rate of CSF absorption by arachnoid villi, and an increase in brain volume [6,7].

The primary focus of the treatment of IIH is uphold-

Keywords: idiopathic intracranial hypertension (IIH), transverse sinus stenosis (TSS), endovascular, stent, intracranial pressure (ICP), headache, papilledema, cerebrospinal fluid (CSF), Starling-like Resistor, neuroradiology

ing optic nerve function while normalizing the ICP, initially with drugs that reduce CSF production (such as Acetazolamide), combined with dietary counseling for weight loss. In patients with severe symptoms, or intolerance/poor response to standard medical therapy, CSF diversion procedures are used. These surgical procedures comprise of implantation of a shunt that allows excess CSF to be drained into another part of the body. Lumboperitoneal (LP) shunts divert the CSF from the spinal space of the lower back into the abdomen. Ventriculoperitoneal (VP) shunts channel the CSF from the brain ventricles into the abdomen [8]. Nevertheless, just as any surgical procedure, shunts can lead to many complications including shunt dislocation, infection, and intracerebral hemorrhage. In addition, up to 64% of VP/LP shunts fail within 6 months and re-operation is common for recurrence of papilledema due to high CSF pressure. Despite the poor surgical results, these procedures have endured due to lack of alternatives for the treatment of IIH [9,10].

Recently, numerous studies have demonstrated that a great percentage of IIH patients have transverse sinus stenosis (TSS). Moreover, direct manometry of the stenotic transverse sinus has shown elevated venous pressures in the superior sagittal and proximal transverse sinuses, and a significantly reduced venous pressure in the distal transverse sinus, with a pressure gradient across the TSS [11-13]. Normal transverse sinuses are collapsible segments susceptible to stenosis, either due to extrinsic compression (smooth steady narrowing), an intrinsic obstruction resulting from intraluminal filling defects (more focal eccentric narrowing), or a combination of both [14]. Irrespective of the source of the stenosis, findings suggest that TSS can be both a cause and a consequence of IIH. Although removing or diverting the CSF resolves the stenosis [15], TSS can persist in IIH patients even after normalization of CSF pressure. Furthermore, intrinsic obstructions such as arachnoid granulations can be seen in the transverse sinus on computerized tomography venography (CTV) and magnetic resonance venography (MRV) scans of patients without IIH [16]. IIH has been predicted on the basis of a mathematical model (see discussion) to compress the collapsible transverse sinus, resulting in venous outflow obstruction and causing additional venous hypertension, which then minimizes CSF absorption and promotes further increase in the ICP. This supplementary increase in the ICP constitutes a positive feedback loop causing further compression of the transverse sinus [17].

A new surgical procedure known as transverse sinus stenting for IIH patients has been reported in a few studies, with the largest one conducted by Ahmed and colleagues in 2008 [14]. In this study, 49 out of the 52 IIH patients who were unresponsive to maximum medical treatment were cured of their symptoms as a result of transverse sinus stenting. Placing a stent into the stenotic transverse sinus abolished the TSS pressure gradient, normalized the ICP, and resolved IIH symptoms. The risks associated with this procedure were much lower than other surgical options, suggesting a safer and more efficient alternative to the current CSF shunting procedures [14]. Based on the literature discussed, we hypothesized that, as an alternative to LP/VP shunting, precisely positioning an expandable stent across the narrowed portion of the transverse sinus will allow blood to drain more freely, relieve the CSF pressure, and alleviate IIH. Here, we report a patient with IIH successfully treated by an endovascular stent placement in the stenotic transverse sinus, and discuss the mathematical model that highlights the rationale for this new method of treatment.

CASE

A 39-year-old woman presented to her family doctor with several weeks of progressive headaches and visual troubles. Her ophthalmologic examination demonstrated papilledema. Further imaging and lumbar puncture were performed confirming the diagnosis of IIH. Her CSF opening pressure measured by lumbar puncture was severely increased (41mmHg) as compared to a normal pressure of below 20-25mmHg. Since the patient started but was not able to tolerate Acetazolamide for various reasons, interventional management procedures such as LP/VP shunting, as well as transverse sinus stenting, had to be considered. Her vascular imaging demonstrated focal stenosis of the right transverse sinus and an underdeveloped, or hypoplastic, left transverse sinus. She underwent retrograde intracranial venous manometry that revealed abnormal pressures in the venous sinuses and a high-pressure gradient of more than 8mmHg across the stenosis (Figure 1). After balancing the risks and benefits for all therapeutic options, it was decided that the patient was a suitable candidate for transverse sinus stenting across the stenotic segment. Two wall stents measuring 9x40mm and 7x40mm were placed as shown in Figure 2. The procedure was uneventful and the patient recovered well. After this intervention, there was a significant improvement in her headaches, papilledema, and CSF pressure. Moreover, her long term imaging follow-up with dynamic CT venogram showed efficient patency of the stent (Figure 3). The patient required dual antiplatelets therapy for 3 months to avoid in-stent thrombosis or stenosis.

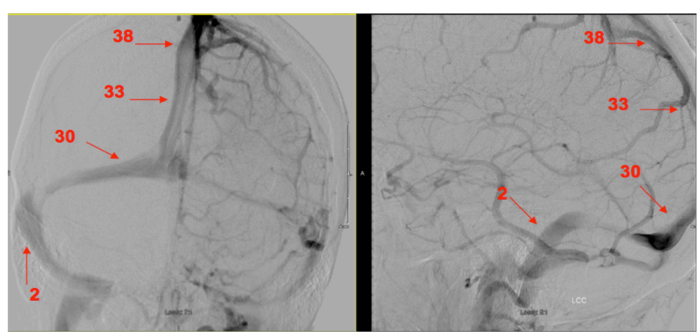

Figure 1: Antero-posterior and lateral view on venous phase cerebral angiogram demonstrating left TS hypoplasia and right TS stenosis. Pressure values (shown in red) obtained via retrograde intracranial venous manometry in superior sagittal sinus, proximal and distal TS are shown. Note the significant pressure gradient (30 mmHg to 2mmHg) across the right TSS. TS; transverse sinus. TSS; transverse sinus stenosis.

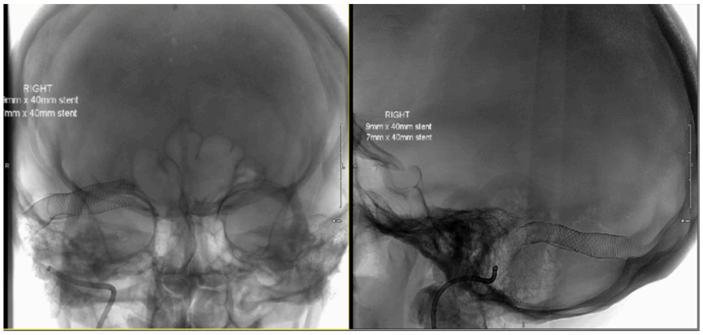

Figure 2: Crane X-Ray antero-posterior and lateral view showing placement of stents within the right transverse sinus.

DISCUSSION

Patients with IIH are frequently diagnosed with papilledema and increased CSF without any mass lesions [1], and they often have stenosis of either both or only the dominant transverse sinus, along with venous hypertension [3,11]. The emphasis during treatment is on perpetuating optic nerve function while stabilizing the ICP. Surgical procedures such as CSF shunting, despite their high risks, are habitually employed on IIH patients with severe symptoms. We used a novel method called transverse sinus stenting as an efficient and a minimally invasive alternative to improve IIH symptoms in a patient. As proposed in Figure 4, a mathematical model of intracranial pressure dynamics provides a basis for sinus stenting in IIH. This model consists of three compartments: proximal transverse sinus, brain/CSF, and distal transverse sinus. A key component of the model is a Starling-like resistor that consists of a flexible tube (collapsible transverse sinus) inside a pressurizable chamber (brain/CSF compartment). As the pressure inside the chamber (intracranial pressure) elevates, the tube (sinus) compresses, and the resistance of fluid (venous blood/CSF) flow through the tube increases (Figure 4A). The tube will collapse at a point where the pressure in the tube becomes critically less than the pressure in the chamber.

In the presence of a Starling-like resistor (a collapsible transverse sinus) under normal pressure conditions, alterations in the intracranial environment such as an upsurge of cerebral blood flow or an infusion of CSF, elevates the ICP and leads to the collapse of the resistor, in other words, stenosis of the transverse sinus. This ultimately generates a transition from a disease-free condition to a higher-pressure IIH condition. Similarly, changes such as a decline in CSF production or removal of excess CSF by shunting lead to a transition from a disease state back to a normal ICP state. However, the stenosis of the collapsible sinus can return by further perturbations in the intracranial environment. We suggest that the incorporation of an endovascular stent in the stenotic transverse sinus is modeled by elimination of the collapsible Starling-like resistor and the constitution of a rigid-walled sinus (Figure 4B). This rigid-walled (stented) transverse sinus is now no longer prone to collapse by further distresses in the intracranial environment and thus prevents the recurrence of IIH by permanently interrupting the positive feedback loop discussed earlier.

This procedure is performed in the angiography suite, using X-ray and iodine contrast (radiographic dye). When placement of an intravascular device, such as a stent, is necessary, the patient requires several months of blood thinner therapy (usually dual antiplatelets therapy). Contraindications of this procedure are related to the presence of an intravascular device and contrast intolerance. Thus the main contraindications are pregnancy, previous history of venous thrombosis and X-ray contrast allergy. Patients are screened for inclusion criteria and contraindications before having the intervention.

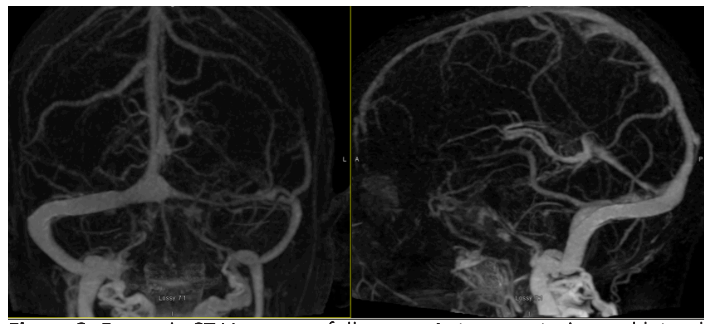

Figure 3: Dynamic CT Venogram follow-up. Antero-posterior and lateral view demonstrating normal caliber of the right transverse sinus after stent placement with no in-stent stenosis.

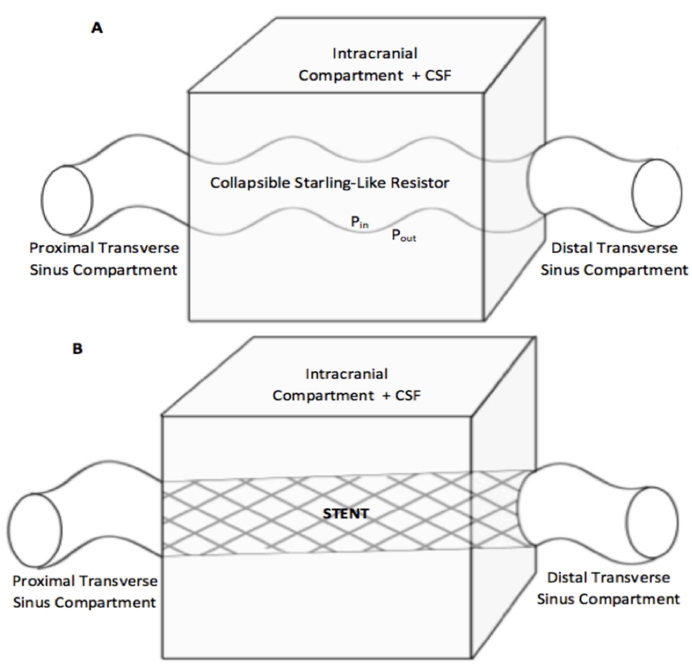

Figure 4: Model of Intracranial Pressure Dynamics for Transverse Sinus Stenting in IIH. **(A)** Three compartments described as proximal sinus, intracranial structures and CSF (boxed), and distal sinus, as well as a collapsible sinus as Starling-like resistor is shown. **(B)** Endovascular stent in the transverse sinus modeled by eliminating the Starling-like resistor resulting in a rigid-walled sinus unsusceptible to further perturbations.

CONCLUSION

We propose that if an IIH patient is being considered for a CSF diversion procedure, and has a TSS with significant pressure gradient, then transverse sinus stenting could be considered as an alternative. The results of this case study, if validated through larger future prospective studies, have the potential to provide interventional neuroradiologists with a primary, low-risk interventional method for IIH treatment.

CONSENT

Both oral and written informed consent were obtained from the patient for publication of this case presentation.

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