Venous Sinus Stenting in the Management of Patients with Intracranial Hypertension Manifesting with Skull Base Cerebrospinal Fluid Leaks

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BACKGROUND: **A subset of patients with skull base cerebrospinal fluid (CSF) leaks are found to have elevated intracranial pressure (ICP). In these patients, elevated ICP is thought to contribute to both the pathophysiology of the leak and postoperative leak recurrences. Current strategies for postoperative ICP control include medical therapy and shunting procedures. The aim of this study is to report the use of venous sinus stenting (VSS) in the management of patients with skull base CSF leaks caused by elevated ICP.**

METHODS: **We performed a retrospective investigation of 2 patients who underwent surgical repair of skull base CSF leaks and were found to have elevated ICP associated with venous sinus stenosis and subsequently treated with VSS.**

RESULTS: **Two patients underwent successful surgical repair of skull base CSF leaks with perioperative ICP monitoring via temporary lumbar catheters. Postoperative CSF pressure measurement demonstrated elevated ICP. Both patients were found to have venous sinus stenosis on further workup and subsequently underwent VSS for treatment of intracranial hypertension. Both patients had improvement in their symptoms with no evidence of recurrent CSF leak at follow-up.**

CONCLUSIONS: **Patients with skull base CSF leaks of unknown etiology should undergo CSF pressure monitoring postoperatively and, if found to be elevated, be treated for intracranial hypertension. In patients unresponsive to, or intolerant of, medical therapy, VSS can provide an alternative option to medical and surgical shunting procedures for treatment of intracranial hypertension in patients with skull base CSF leaks and venous sinus stenosis.**

INTRODUCTION

There are numerous classification schemes of cerebrospinal fluid (CSF) leaks. Cairns[1] divided CSF rhinorrhea into 4 groups, including acute traumatic, delayed posttraumatic, iatrogenic secondary to cranial or sinus surgery, and spontaneous. Coleman and Troland[2] further defined primary or spontaneous CSF rhinorrhea as discharge of CSF from the nose without definite demonstrable cause. More specifically, the authors defined such leaks as those that occurred in the absence of trauma (acute, delayed, or operative), infection of the bones of the paranasal sinuses, tumors eroding the base of the cranium, prolonged causes of increased intracranial pressure (ICP) (cerebral tumors or congenital or acquired hydrocephalus), or congenital etiologies.[2] Noting that transient or continued alteration in cerebrospinal pressure and dynamics may initiate leakage, Ommaya[3] classified nasal CSF leaks into traumatic and nontraumatic and recommended classifying spontaneous CSF leaks as a variety of the nontraumatic category. That same year, O'Connell[4] subdivided spontaneous CSF leaks into secondary and primary. In secondary cases, an obvious cause for the leak is present; in the

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- Cerebrospinal fluid leak
- Encephalocele
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- Idiopathic intracranial hypertension
- Meningoencephalocele
- Venous sinus stenosis
- Venous sinus stenting

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
CT: Computed tomography
ICP: Intracranial pressure
IIH: Idiopathic intracranial hypertension
MR: Magnetic resonance
VSS: Venous sinus stenting

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primary group, no such cause is identified. Ommaya et al.[5] further classified nontraumatic leaks into high- and normal-pressure leaks. Because there are various operational definitions and many spontaneous leaks are caused by elevated ICP, this term remains confusing even in contemporary literature.

Despite this confusing terminology, the relationship between elevated ICP, and notably idiopathic intracranial hypertension (IIH), and skull base CSF leaks is well established. Between 1970 and the late 1990s, there were scattered reports of patients with IIH who developed skull base CSF leaks.[6-11] More recent studies have objectively measured ICP in patients with CSF leaks. In 2003, Schlosser et al.[12] demonstrated elevated ICP in 10 of 10 patients with skull base CSF leaks who underwent postoperative lumbar puncture with CSF pressure measurement during clinically indicated CT cisternograms. The same group in 2004 investigated CSF pressure measurements postoperatively in patients with both spontaneous and traumatic CSF leaks and reported that 7 out of 8 patients with spontaneous leaks demonstrated elevations in ICP compared with none of the patients with traumatic CSF leaks.[13] Recently, Xie et al.[14] measured ICP through lumbar drain catheters in patients with spontaneous CSF leaks treated with endoscopic endonasal repair and found that 36% of patients required either medical or surgical intervention for elevated ICP. Numerous additional studies have also highlighted the association between skull base CSF leaks and elevated ICP.[15-18]

The relationship between skull base CSF leaks and elevated ICP underscores that this association should be considered and formally evaluated in leaks that are not congenital, iatrogenic, traumatic, or secondary to tumors or infections. Suspicion for elevated ICP can often be heightened from the patient's history and physical examination and from diagnostic imaging. For example, obese women of childbearing age fit the classic phenotype of a patient with IIH. These individuals may present with characteristic symptoms, including headaches, transient visual obscurations, and pulsatile tinnitus.[19,20] Ophthalmologic evaluation may reveal papilledema of varying grades, decreased visual acuity, diminished visual fields, or widened blind spots on perimetry testing.[20,21] Imaging studies, which are typically performed in patients with CSF leaks to identify the site(s) of the leak(s), may demonstrate radiographic features associated with elevated ICP, including an empty sella, optic nerve sheath distention or tortuosity, or skull base bony remodeling with meningocele formation.[22-24]

Treatment of IIH involves a combination of weight loss, medical treatment, and surgical options. Surgical intervention with optic nerve sheath fenestration or shunting is generally reserved for medically refractory cases involving vision deterioration or intractable headaches.[20,25] More recently, venous sinus stenting (VSS) has been demonstrated as an effective alternate treatment modality for patients with IIH and imaging evidence of transverse sinus stenosis.[26-34] VSS has been shown to improve the venous sinus pressure gradient and clinical symptoms associated with IIH, in addition to quantitatively reducing postprocedure ICP.[30,33-35] Treatment of intracranial hypertension in patients with skull base CSF leaks has included medical therapy and shunting procedures.[12-14,16] In this report, we describe 2 patients with skull base CSF leaks who underwent surgical repair and were found to have intracranial hypertension associated with venous sinus stenosis. VSS was successfully performed in both patients, with no clinical evidence of CSF leak recurrence or elevated ICP at last follow-up.

METHODS

We performed a retrospective investigation of 2 patients who underwent surgical repair of skull base CSF leaks at our institution between 2013 and 2014 that were found to have elevated ICP associated with venous sinus stenosis, subsequently treated with VSS. Data collected included age, sex, medical history, diagnostic imaging, lumbar puncture opening pressure, operative procedures, perioperative ICP monitoring data, manometric testing during angiography, and clinical follow-up. This study was performed in accordance with institutional review board guidelines.

VSS Procedure

The endovascular procedures were performed under general anesthesia with endotracheal intubation in a biplane neuroangiography suite (Siemens, Erlangen, Germany). Both patients were started on daily aspirin (325 mg) and clopidogrel (75 mg) 1 week prior to treatment and received perioperative intravenous heparin (activated clotting time, 250–300 seconds). After a 4-vessel diagnostic cerebral angiogram performed via 4-French femoral arterial access confirmed the absence of other neurovascular pathologies, an 8-French guiding catheter (Neuron [Penumbra, Alameda, California, USA]) was advanced into the internal jugular vein, ipsilateral to the dominant transverse sinus, via femoral venous access. A 2.4-French catheter (Renegade HI-FLO [Boston Scientific, Marlborough, Massachusetts, USA]) was then navigated over a 0.16 guidewire (Fathom-16 [Boston Scientific]) across the torcula into the opposite internal jugular vein under roadmap guidance. The guidewire was then exchanged for a 300-cm 0.014 microwire (Luge [Boston Scientific]), the catheter hub was connected to a manometer, and the catheter tip was slowly withdrawn across both transverse and sigmoid sinuses, where pressure measurements were obtained. After confirmation of the existence of a pressure gradient across the transverse sinus stenoses, the dominant sinus was stented using a 6- × 40-mm self-expandable carotid stent (PRECISE PRO Rx, off-label use [Cordis, Fremont, California, USA]) advanced over the 0.014 microwire. The stent delivery system was withdrawn, and manometry was repeated to confirm the venous pressure correction. Both patients were kept heparinized and observed in an intensive care setting overnight; the arterial and venous femoral sheaths were pulled the next morning, with hemostasis being achieved by manual compression. Both patients were discharged on poststenting day 1.

RESULTS

Case Illustration 1

History and Examination. A 61-year-old man presented for evaluation of beta-2 transferrin positive left-sided rhinorrhea of 7 months' duration. His body mass index was 26.6 kg/m^2, and there was no clinical history of known obstructive sleep apnea, meningitis, trauma, or sinonasal/cranial surgery. The patient had no neurologic deficits on examination; leaning forward elicited watery drainage