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Does bilateral transverse cerebral venous sinus stenosis exist in patients without increased intracranial pressure?

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Abstract

Objective—Transverse cerebral venous sinus stenosis (TSS) is common among patients with idiopathic intracranial hypertension. TSS likely also exists among individuals with normal ICP but the prevalence is unclear. The goal of this study was to identify patients with incidental TSS and normal ICP and describe their characteristics.

Methods—Among 240 adult patients who underwent brain magnetic resonance imaging (MRI) with magnetic resonance venography (MRV) with contrast at our institution between September 2009 and September 2011, 44 had isolated TSS without further substantial imaging abnormality. Medical records were reviewed for symptoms of increased ICP, papilledema, cerebrospinal fluid (CSF) constituents and opening pressure (OP), and reason for brain imaging. Of these, 37 were excluded for confirmed or possible idiopathic intracranial hypertension. Of the remainder, 5 had CSF-OP ≥ 25 cmH₂O without papilledema, and 2 did not have measured ICP, but had no papilledema or symptoms of increased ICP. Imaging was re-interpreted to assess for signs suggestive of elevated ICP and to characterize the TSS further.

Results—All patients were women (mean age: 41, mean BMI: 37.1). CSF contents were normal, but OPs were at the upper limit of normal (22 – 25 cmH₂O). Indications for MRI/MRV included query pituitary abnormality (1), migraine (4), and anomalous-appearing optic nerves (2). All had bilateral TSS. Six had short TSS and an empty sella; one had long TSS and no empty sella; one had flattening of the posterior sclera; two had prominence of peri-optic nerve CSF.

Conclusion—Asymptomatic bilateral TSS exists in patients with ICP ≥ 25 cmH₂O, but is likely uncommon. CSF-OP was at the upper limit of normal in our patients, who also had other radiologic signs suggestive (but not specific) of chronically-raised ICP. Findings of bilateral TSS on imaging should prompt funduscopy examination for papilledema.

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Keywords

transverse cerebral venous sinus stenosis; magnetic resonance venography; intracranial pressure; intracranial venous sinus anatomy; empty sella; flattening of the posterior sclera; peri-optic nerve cerebrospinal fluid

Introduction

Consider the following real-life scenario: a 24-year-old obese woman presents to an optometrist for a routine eye examination. Funduscopy examination reveals anomalous elevated optic nerves concerning for bilateral disc edema. Brain magnetic resonance imaging (MRI) with head magnetic resonance venography (MRV) performed at an outside institution shows bilateral transverse sinus stenosis (TSS). There is no empty sella, no flattening of the posterior sclera, and no prominence of peri-optic nerve cerebrospinal fluid (CSF). Because of concern for raised intracranial pressure (ICP), a lumbar puncture is performed and shows an opening pressure of 8 cmH₂O with normal CSF constituents. She develops a severe low-pressure headache requiring a blood patch. Neuro-ophthalmologic examination confirms bilateral congenitally crowded discs with peripapillary atrophy but no true disc edema.

Recent advances in non-invasive cerebral vascular imaging such as MRV and computed tomography venography have shown that unilateral or bilateral TSS is common among patients with elevated ICP, particularly those with idiopathic intracranial hypertension (IIH) [1]. In addition, it is not uncommon for neuroradiologists to suggest a diagnosis of elevated ICP when reporting “incidentally found TSS” on routine brain imaging and to recommend “clinical correlation” or even a lumbar puncture to measure the CSF opening pressure (CSF-OP) in the patient. Although it is believed that TSS exists in the general population, and is not specific to elevated ICP, the prevalence of TSS among individuals with normal ICP is unclear. Our patient’s history, along with our neuroradiologists’ experience, led us to believe that bilateral TSS with normal ICP may not be unusual. The goal of our study was to identify patients with TSS and normal ICP and describe their characteristics.

Materials and Methods

This study was approved by our Institutional Review Board. All patients who underwent standardized brain MRI and contrast-enhanced MRV at our institution between September 2009 and September 2011 were identified. All studies were reviewed prospectively to select patients with TSS but no cerebral venous thrombosis. Patients who had a normal MRV or any abnormality other than TSS were excluded. All TSS patients’ medical records were retrospectively reviewed and all patients with definite IIH and those with insufficient clinical information were excluded. Age, race, gender, height and weight were recorded. The reason for the MRI, symptoms of increased ICP, and examination findings, including funduscopy examination, were recorded, as were CSF-OP, when available.

MR imaging was performed using a standardized protocol at either 3.0-Tesla (Siemens Trio, Erlangen, Germany) or 1.5-Tesla (Siemens Avanto, Erlangen, Germany or GE Signa, Milwaukee, Wisconsin) using a standard head coil. All patients underwent contrast-enhanced MRI along with contrast-enhanced MRV. The MRI/MRV protocol included routine pre-contrast axial diffusion-weighted, T1-weighted, T2-weighted gradient recalled echo (GRE), and sagittal T1-weighted images. An axial precontrast MRV mask was obtained (TR of 4–6 ms, TE of 1–2 ms, and flip angle of 22–30 degrees with slice thickness of 0.8–1.4 mm). A standard dose (0.1 mmol/kg) intravenous gadolinium based contrast agent (Multihance,

Bracco Diagnostics Inc.) was administered at 2.0 cc/second, and the axial MRV sequence was repeated 60 seconds following contrast administration. Post-contrast axial T2-weighted and T1-weighted, and sagittal volumetric T1-weighted GRE images of the brain were then acquired. The pre-contrast MRV dataset was subtracted from the post-contrast dataset, and multiple oblique maximum intensity projections (MIPs) were generated from this subtracted dataset with rotation around the craniocaudal axis (“spin”) or the transverse axis (“nod”) at 6 degree increments.

All MRIs and MRVs were re-interpreted by an experienced neuroradiologist (AMS). Each patient’s full set of images was evaluated for the following imaging findings: presence of empty sella, flattening of the posterior sclera, prominence of peri-optic nerve CSF and bone changes at the level of the stenosed sinus. MRV source data and MIP images were evaluated for location and extent of any of dural venous sinus stenosis, according to the description of Farb et al [1]. A combined conduit score (CCS) was calculated for each patient (Table). Only patients with TSS considered “typical of IIH” were included in this study. One patient had prominent arachnoid granulations in addition to TSS. None had any prior history of cerebral venous stenosis.

Results

Two hundred and forty consecutive patients underwent standardized brain MRI/MRV with contrast at our institution during the inclusion period. One hundred and ninety-six patients were excluded: 140 with normal MRV, 36 with venous sinus thrombosis, 14 with other intracranial abnormalities and 6 with venous sinus stenosis and additional parenchymal abnormalities. Medical records of the remaining 44 patients were reviewed for symptoms of increased ICP, evidence of papilledema, CSF constituents and opening pressure. Twenty-nine patients with definite IIH diagnosed using the most recent diagnostic criteria [2] were excluded. Eight patients with incomplete clinical information in whom IIH was not definitively ruled out were also excluded.

The characteristics of the 7 remaining patients are summarized in the table (the patient presented in the introduction did not have her MRV performed at our institution, and therefore was not included in our study). All patients were women (5 African-American, 2 Caucasian). The mean age was 41 years old (range 30–60). All were overweight or obese (mean BMI 37.1 kg/m²). One patient had obstructive sleep apnea, one had anemia and five had systemic hypertension. Five patients had episodic headaches consistent with migraine. None of the seven patients had papilledema. Five of the 7 patients had a lumbar puncture in the lateral decubitus position. All five of these patients had normal CSF constituents and a CSF-OP of ≤ 25 cmH₂O, but all had a CSF-OP that was at the upper limit of normal (22, 23, 24, 24, 25 cmH₂O). The other two patients did not have a lumbar puncture performed, but neither had papilledema nor symptoms indicative of elevated ICP.

Indications for MRI/MRV were: a query of pituitary abnormality (1), migraine (4) and anomalous-appearing optic nerves (2). By definition, all patients had bilateral TSS (Table, Figures 1 and 2). In addition, one patient had three other radiologic signs suggestive of increased ICP (empty sella, flattening of the posterior sclera and prominence of peri-optic nerve CSF), one patient had both an empty sella and prominence of peri-optic nerve CSF and 4 patients had an empty sella. No patient had bone changes at the level of the stenosed sinus.

Discussion

Among 240 patients who had a high-quality contrast-enhanced MRV specifically looking for TSS performed at our institution over a two-year period, only 7 patients had isolated TSS without symptoms or signs of elevated ICP. Although this number is likely an underestimate because of our very strict inclusion criteria and the retrospective nature of our study, this suggests that normal ICP is unlikely to be found in patients with TSS. This is in concordance with prior studies which have evaluated this question from the opposite direction: that is, determining the frequency of normal MRVs among patients with possible IIH or for so-called “IIH without papilledema” [3, 4]. Farb et al’s [1] original MRV study demonstrated bilateral TSS in 27 of 29 patients with IIH and in only 4 of 59 controls, suggesting that bilateral TSS is highly suggestive of raised ICP in the correct clinical setting. However, although the authors mentioned that none of the controls was being evaluated for IIH and that they had no symptoms or signs of neurologic disease, the CSF-OPs were not provided and it is unclear whether these patients had fundoscopic examinations looking for papilledema.

Although our patient described in the introduction with obvious bilateral TSS on MRV had a CSF-OP of only 8 cmH₂O, all 5 patients included in our study who had a lumbar puncture performed had CSF-OPs that were at the upper limit of normal. None of these 5 patients had symptoms or signs of raised ICP, but they all had episodic headache suggestive of migraine. These observations raise the question as to whether the bilateral TSS observed in our patients (who do not have IIH) are truly incidental or might be somehow related to mildly elevated ICP responsible for migraine-like headaches. Interestingly, as previously described [5], other MRI findings commonly found in patients with chronically raised ICP [3, 6, 7] were present in most of our patients, including an empty sella in 6, prominence of the peri-optic nerve CSF in two, and flattening of the posterior sclera in one. A recent study [6] suggested that the presence of at least two radiologic signs suggestive of increased ICP (e.g., empty sella and TSS, or prominence of peri-optic nerve CSF and empty sella) is very suggestive of intracranial hypertension in that patient, whereas one sign only (e.g., isolated empty sella or isolated TSS) was commonly incidentally found in control subjects with normal ICP.

Interestingly, other authors have reported bilateral TSS in presumably primary headache patients (mostly with migraine) in whom CSF-OPs were above 20 cmH₂O [8]. None of these patients had papilledema and the authors suggested the diagnosis of “IIH without papilledema”, given their “borderline” CSF-OP. However, the same study included 193 patients with CSF-OPs less than 20 cmH₂O, among whom 26 (13.4%) had evidence of bilateral TSS on MRV, confirming that incidentally found bilateral TSS might not be so uncommon, even in patients with normal ICP. Clearly, a radiologic finding of bilateral TSS is not sufficient to make the diagnosis of IIH in patients with headaches and no papilledema. However, the same group [4] suggested a possible correlation between CSF-OP and the presence of bilateral TSS on MRV. They described bilateral TSS in 49 of 724 (6.7%) patients diagnosed with migraine. Although none of these patients had papilledema, 28 had a lumbar puncture: CSF-OP was greater than 20 cmH₂O in 19 patients, and less than 20 cmH₂O in 9 patients. These authors concluded that headache patients with bilateral TSS should undergo a lumbar puncture to assess for elevated ICP. However, these results must be interpreted with caution. Indeed, although it seems that bilateral TSS is more commonly observed in patients with CSF-OP greater than 20 cmH₂O, most authors consider CSF-OP less than 25 cmH₂O within normal limits, and the most recent diagnostic criteria for IIH uses 25 cmH₂O as the lower limit for increased ICP [2]. This limit of 25 cmH₂O is based on studies showing that there is an overlap between “normal” and “elevated” ICP ranging between 20 and 25 cmH₂O in adults [9, 10] and up to 28 cmH₂O in children [11].

It is reasonable to postulate that there is a continuum of changes that occur in the brain and cerebral venous sinuses when the intracranial pressure becomes elevated. The spectrum may range from an individual with normal ICP, absence of TSS and lack of radiologic signs commonly found in patients with raised ICP to the other extreme in which there is elevated ICP, bilateral TSS and one or more radiologic signs such as empty sella, prominence of the peri-optic nerve CSF and flattening of the posterior sclera. An individual may present anywhere along this spectrum, including the scenario, although uncommon, in which there is bilateral TSS but ICP is normal. Normal ICP in the setting of bilateral TSS may occur due to compensatory mechanisms, such as via collateral venous drainage through enlarged dural veins, the vein of Labbe or the occipital sinus [12–16].

The anatomy of intracranial venous sinuses varies greatly among normal subjects, particularly at the level of the transverse and sigmoid sinuses, making the interpretation of an “abnormal transverse sinus” very difficult [12–16]. Additionally, MRI and MRV techniques vary, and older time-of-flight MRVs largely overestimated the prevalence of TSS, mistaking flow-related artifacts. Newer MRV techniques, particularly the auto-triggered elliptic centric-ordered (ATECO) MRV used by Farb [17], similar to the contrast-enhanced MRV used in our study, have allowed better visualization of the transverse sinuses and allow accurate description of transverse sinus anomalies on most MRV studies. Hypoplasia of a transverse sinus (most often the left) is found in about 20% of cases [16]; asymmetry of transverse sinuses is observed in at least 50% of cases (with the right TS being most often dominant) [12, 14, 16], and arachnoid granulations, sometimes large enough to result in partial sinus obstruction, are also common [18]. Additionally, partial obstruction or sinus asymmetry can result from a fibrous septation of the lumen of the transverse sinus, usually near the torcula [14]. These “anomalies” are found so commonly that a firm association with headache or other symptomatology suggesting increased ICP must be guarded. However, it is well established that a stenosis in a dominant sinus, or stenoses that are bilateral can cause partial obstruction of venous flow, resulting in venous hypertension and worsening of intracranial hypertension; extrinsic long smooth stenosis of the transverse sinus can also be a consequence of intracranial hypertension itself. These observations emphasize how difficult interpretation of modern MRVs has become and reinforce the fact that the clinical presentation prevails over any “incidental” radiologic finding.

There are several limitations of this study. One is the fact that the patient charts were reviewed retrospectively. However, the use of a standardized neuro-ophthalmologic clinic form maximizes consistency of data collected at each patient encounter. In addition, 6 patients were re-examined immediately before the study to obtain updated information and clinical examination. Furthermore, all diagnostic imaging was reinterpreted prospectively. Another limitation is that 2 of the 7 patients did not have a lumbar puncture. However, since these patients had no papilledema and no symptoms suggestive of elevated ICP, lumbar puncture was not justifiable.

Conclusion

Asymptomatic bilateral TSS does exist in patients with a normal ICP, but its presence in a patient with headaches should raise the possibility of intracranial hypertension. We remain skeptical regarding making (or even suggesting) a diagnosis of so-called “IIH without papilledema” in any headache patient with bilateral TSS or TSS of a dominant sinus. However, an incidental finding of bilateral TSS on MRV should prompt a funduscopic examination looking for papilledema, especially in headache patients. Whether such patients should undergo a lumbar puncture in the absence of papilledema remains to be proven,

although other factors associated with IHH, such as female gender and obesity, should obviously influence such decisions.

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Figure 1. Magnetic resonance venography (MRV), two views, patient 4. A, Craniocaudal projection maximum intensity projection (MIP) demonstrating bilateral transverse sinus stenosis (TSS) (arrows) with a combined conduit score of 3 (2 + 1). B, Oblique lateral projection of MIP demonstrating bilateral TSS (arrows).

Figure 2. Magnetic resonance venography (MRV), two views, patient 7. A, Craniocaudal projection maximum intensity projection (MIP) demonstrating bilateral transverse sinus stenosis (TSS) (arrows) with a combined conduit score of 2 (1 + 1). B, Oblique lateral projection of MIP showing the right-sided TSS in profile (arrow).

Table

Characteristics of patients with transverse sinus stenosis and no intracranial hypertension

Patient	Age (y)/race/sex	BMI (kg/m ²)	Medical conditions	Symptoms	CSF OP (cm H ₂ O)	Reason for MRI/MRV	Side and severity (CCS) of stenosis	Length of stenosis*	Empty sella	Flattening of the posterior sclera	Prominence of peri-optic nerve CSF
1	45 C W	39.9	OSA, HTN, DM	Headache	25	Query pituitary abnormality	Bilateral 3	Short	+	-	-
2	41 AA W	30.3	HTN, DM	Headache	24	Migraine	Bilateral 5	Short	+	-	+
3	40 AA W	27.3	None	Headache	24	Migraine	Bilateral 4	Long	-	-	-
4	34 AA W	48.6	Anemia, HTN	Headache	23	Migraine	Bilateral 3	Short	+	-	-
5	60 C W	39.3	HTN	Headache	22	Migraine	Bilateral 4	Short	+	-	-
6	30 AA W	31.2	None	None	Not done	Optic nerve anomaly	Bilateral 2	Short	+	+	+
7	37 AA W	43.3	HTN	None	Not done	Optic nerve anomaly	Bilateral 2	Short	+	-	-

y = years; BMI = body mass index; CSF = cerebrospinal fluid; OP = opening pressure; MRI = magnetic resonance imaging; MRV = magnetic resonance venography; CCS = combined conduit score (ranges from 1 to 8, with 1 corresponding to severe transverse sinus stenosis and 8 corresponding to no transverse sinus stenosis based on Farb et al [1] classification. Each transverse sinus is graded from 0 to 4 [0: aplastic segment; 1: severe stenosis with less than 25% of residual lumen; 2: moderate stenosis with between 25 and 50% of residual lumen; 3: mild stenosis between 50 and 75% of residual lumen; 4: no stenosis]. The sum of the right and left transverse sinus grade provides the combined conduit score). C = Caucasian; AA = African-American; W = woman; OSA = obstructive sleep apnea; HTN = hypertension; DM = diabetes; + = yes; - = no

* Short transverse sinus stenoses were focal stenoses, with the remainder transverse sinus having a normal caliber, whereas long stenoses involved most or the whole portion of the transverse sinus.