

Styloidogenic Jugular Venous Compression Syndrome: Diagnosis and Treatment: Case Report

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BACKGROUND AND IMPORTANCE: Intracranial venous hypertension is known to be associated with venous outflow obstruction. We discuss the diagnosis and treatment of mechanical venous outflow obstruction causing pseudotumor cerebri.

CLINICAL PRESENTATION: We report 2 patients presenting with central venous outflow obstruction secondary to osseous compression of the internal jugular veins at the craniocervical junction. The point of jugular compression was between the lateral tubercle of C1 and a prominent, posteriorly located styloid process. In both cases, catheter venography showed high-grade jugular stenosis at the level of C1 with an associated pressure gradient. The dominant jugular vein was decompressed after the styloid process was resected. Postoperative imaging confirmed resolution of the jugular stenosis and normalization of preoperative pressure gradients. In both cases, the symptoms of intracranial hypertension resolved.

CONCLUSION: Intracranial venous hypertension may result from extrinsic osseous compression of the jugular veins at the skull base. Although rare, this phenomenon is important to recognize because primary stenting not only is ineffective but also may actually exacerbate the outflow obstruction. The osseous impingement of the dominant jugular vein can be relieved via a decompressive styloidectomy, and the clinical results can be excellent.

KEY WORDS: Internal jugular vein compression, Pseudotumor cerebri, Styloid process

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Sometimes symptomatic intracranial venous hypertension is associated with venous outflow obstruction. A subset of patients presenting with intracranial venous hypertension may have a central venous outflow obstruction related to osseous compression of the distal cervical internal jugular vein between a prominent, posteriorly located styloid process and the lateral mass of C1. Endovascular stenting (with or without angioplasty) is typically the treatment of choice for vascular stenoses, but this approach is ineffective in this scenario. We report the first 2 such cases in which the jugular venous outflow obstruction was relieved with a decompressive styloidectomy.

ILLUSTRATIVE CASES

Case 1

A 46-year-old man presented with complaints of headache and visual blurriness. The headaches were disabling and required narcotics for management. On

physical examination, he was noted to have papilledema. His neurological examination was otherwise benign. A diagnostic lumbar puncture demonstrated elevated spinal fluid pressure as high as 23 cm H₂O. Computed tomographic (CT) angiography showed occlusion of the left internal jugular vein at the skull base and severe stenosis of the right internal jugular vein where it was compressed between the lateral tubercle of C1 and a prominent, posteriorly located styloid process (Figure 1). A network of enlarged suboccipital veins had developed to provide collateral venous drainage. Catheter venography with venous pressure measurements confirmed a high-grade jugular venous stenosis at the level of C1 and a focal pressure gradient across the stenosis, with pressures of 18 mm Hg measured just above the stenosis and 8 mm Hg just below the stenosis.

A right anterior neck approach for styloidectomy was used, and the carotid sheath was opened. The muscles were stripped from the styloid, and the bone was transected, removing 3.5 cm of styloid process. An additional 1 cm was removed with a high-speed drill until the styloid process was flush with the skull base. The carotid sheath was widely opened until the jugular

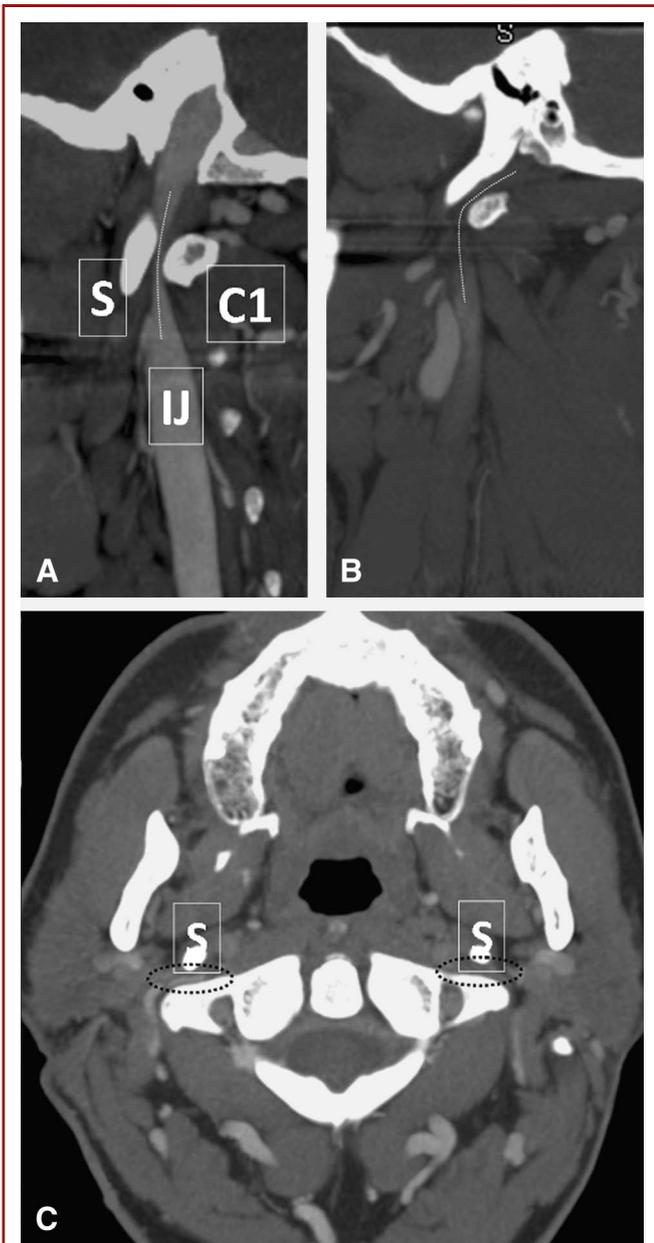


FIGURE 1. *A and B, sagittal CT angiographic views show the narrowing of the jugular vein caused by compression between the right styloid process (S) and the right C1 lateral tubercle. C, axial CT angiographic view shows the same compression (circled area). Note the absence of the left internal jugular vein (IJ). Used with permission from Barrow Neurological Institute.*

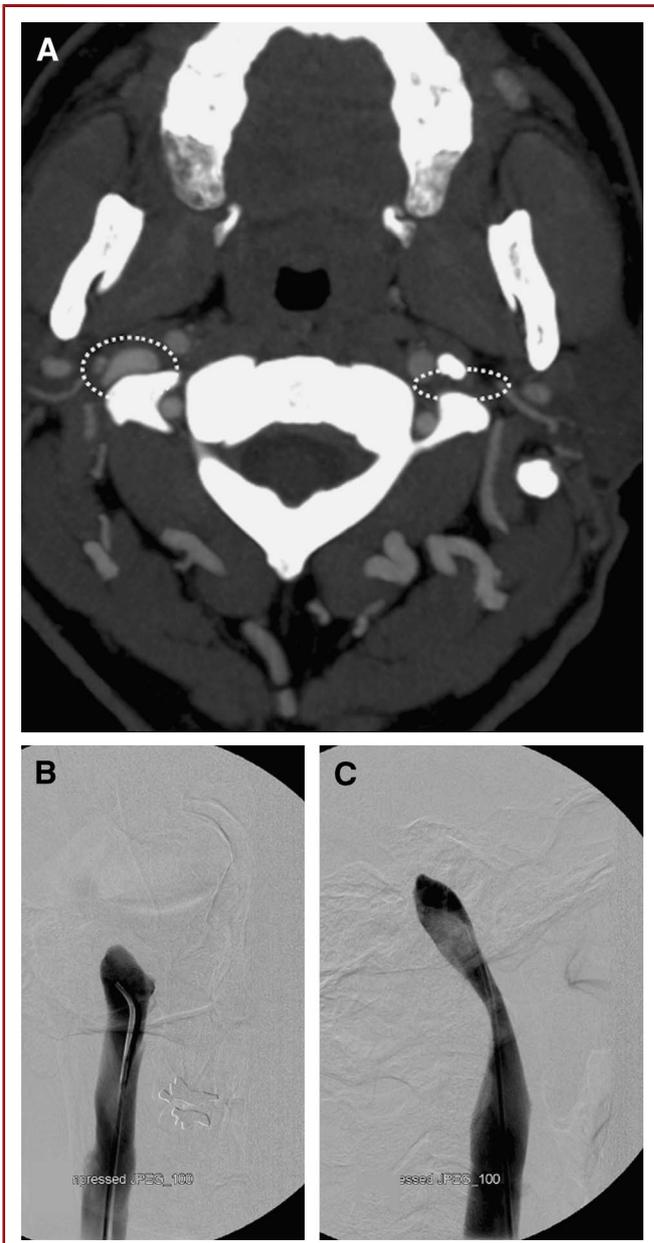


FIGURE 2. *A, postoperative axial CT angiographic view shows decompression of the internal jugular vein in the absence of the styloid process. Lateral (B) and anteroposterior (C) views of the internal jugular vein show that the anteroposterior dimension of the vein is still narrow but wider than before the decompression. There was no pressure gradient across this segment. Used with permission from Barrow Neurological Institute.*

vein was seen to be pulsating freely with both the heartbeat and respiration. The patient did well after surgery. He had no new deficits, and his preoperative headache had resolved.

Postoperative CT angiography confirmed complete decompression of the osseous impingement (Figure 2A). Postoperative catheter venography showed resolution of the obstruction (Figure 2B and 2C). Venous pressure

measurements showed that the preoperative central venous hypertension and focal pressure gradient across the affected segment had resolved completely and was 10 mm Hg at the sigmoid sinus, jugular bulb superior to C1, and lower cervical internal jugular vein. At follow-up, the patient remained asymptomatic with complete resolution of his symptoms of pseudotumor cerebri and papilledema. Follow-up magnetic resonance

venography performed 17 months later showed continued wide patency of the venous sinuses and of the right internal jugular vein.

Case 2

A 39-year-old man presented to an outside hospital with a severe headache and was found to have superior sagittal sinus thrombosis. He was treated successfully by lacing the clot with 2.5 mg tenecteplase, followed by mechanical thrombectomy with a 4F Fogarty catheter. Soon thereafter, his sinuses reoccluded despite full anticoagulation therapy. Repeat mechanical thrombectomy was successful, at which time complete occlusion of the left internal jugular vein and severe stenosis of the right internal jugular vein were found. These occlusions were considered the anatomic factor that likely predisposed the patient to repeat venous thrombosis. Therefore, the patient underwent primary stenting of his right internal jugular vein. After this procedure, the patient was still experiencing moderate headaches and a new pain extending over the right side of his neck and scapula. Four months after his initial procedure, the patient presented to our institution with recurrent severe headaches and papilledema.

Poststenting CT venography showed significant residual stenosis at the midpoint of the stent within the right internal jugular vein (Figure 3). A CT venography showed that the persistent stenosis was the result of extrinsic compression of the jugular vein and stent between the right styloid process and the lateral tubercle of C1. Central venous pressure measurements showed a 5-mm Hg gradient across the stented vein.

The patient’s anticoagulation therapy was switched from Coumadin to heparin. He then underwent lateral neck dissection for removal of the styloid process and the lateral tubercle of C1. At the conclusion of surgery, the internal jugular vein appeared to be widely patent.

Postoperative cerebral venography showed that the stent in the right internal jugular vein was completely open after the decompression, without residual stenosis (Figure 4). The pressure gradient across the stent had dissipated, measuring 9 mm Hg both above and below the stented segment. Postoperatively, the patient’s headache, neck pain, and shoulder pain resolved completely. The patient was treated with clopidogrel for 3 months

and with 325 mg aspirin daily for an indefinite period. At his 5-month follow-up examination, CT venography confirmed continued patency of the decompressed stented segment of the internal jugular vein. Since surgery, the patient has been headache free and his papilledema has resolved.

DISCUSSION

Intracranial hypertension is known to be associated with intracranial venous obstruction related to thrombosis or stenosis.¹⁻¹⁶ We report 2 novel cases presenting with symptoms of increased central venous pressure related to jugular venous outflow obstruction caused by osseous impingement at the level of the craniocervical junction. The point of compression was between the lateral tubercle of C1 and a large, posteriorly located styloid process. In both cases, this anatomic impingement was bilateral.

The styloid process is a bony outgrowth of the temporal bone that functions as the point of attachment of the stylohyoid muscle, which acts to elevate the hyoid bone.¹⁷ Symptoms caused by an elongated styloid are rare but, when present, usually manifest as Eagle syndrome. Two different types are now recognized.¹⁸⁻²² The classic form of Eagle syndrome is caused by various degrees of impingement on cranial nerves V, VII, IX, or X by the styloid process. These patients often become symptomatic with facial pain, ear pain, voice changes, dysphagia, or the sensation of a foreign body in the throat. The second type of Eagle syndrome is related to carotid compression by an elongated styloid process. Patients with this type of Eagle syndrome can experience transient ischemia when they turn their head.¹⁸ Symptomatic jugular vein obstruction, as seen in our cases, has never been reported in association with Eagle syndrome.

Pseudotumor cerebri caused by compression of the internal jugular vein by the styloid process should arise only in the context

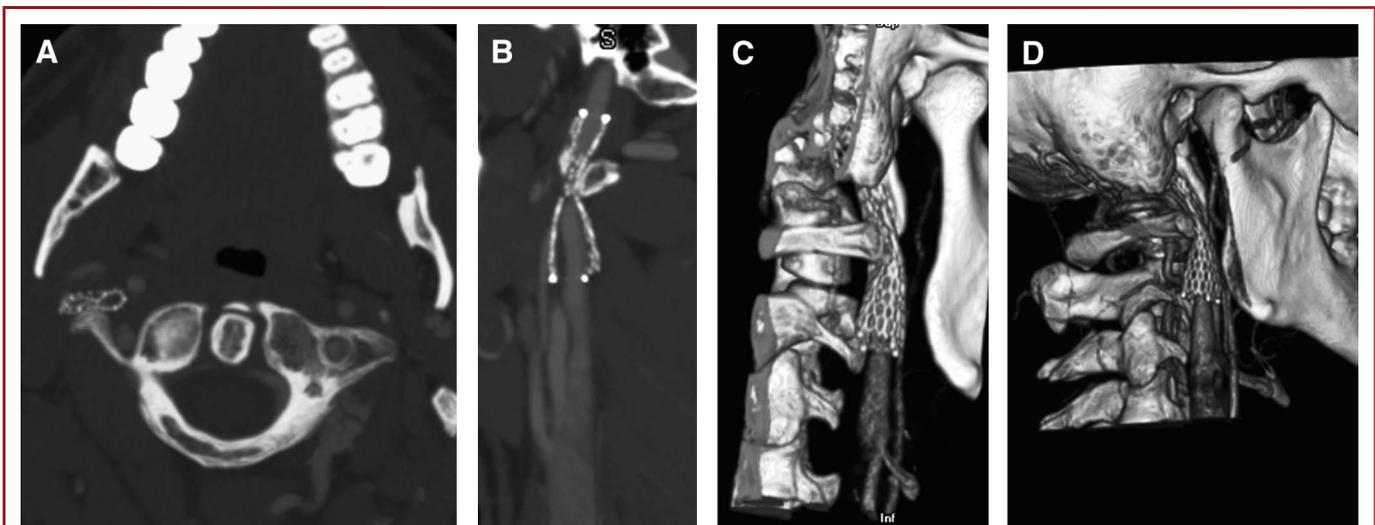


FIGURE 3. A CT venogram, axial (A) and sagittal (B) views, performed 4 months after stenting shows extrinsic compression of the stented internal jugular vein with significant residual stenosis at the midpoint of the stent. C and D, 3-dimensional reconstructions of the stented vein. Used with permission from Barrow Neurological Institute.



FIGURE 4. Right internal jugular vein venograms obtained before (A) and after (B) styloidectomy show increased stent patency. There is evidence of intimal hyperplasia narrowing the stent. Used with permission from Barrow Neurological Institute.

of contralateral venous occlusion or bilateral styloid compression. In the presence of a normal contralateral jugular vein and “balanced” intracranial venous outflow (ie, a patent torcula in communication with both the superficial and deep venous systems and adequate bilateral transverse-sigmoid patency), there is no reason for styloid compression of the internal jugular vein to become symptomatic.

Eagle¹⁹⁻²¹ performed a transoral approach for resection of the styloid process. Drawbacks of this approach are contamination with oral flora, pharyngeal edema, increased risk of neurovascular injury, and poor visibility. We favor a transcervical approach, which allows access to the styloid process and the C1 tubercle through a small submandibular lateral approach.¹⁷ The transcervical approach, however, is associated with a risk of postoperative lip weakness from damage to the marginal mandibular branch of the facial nerve. Nonetheless, if bony obstruction can be relieved, the clinical results of both approaches can be excellent.

Endovascular angioplasty and stenting are ineffective treatments when vascular stenosis is related to rigid bony compression. In fact, the implantation of a stent into an extrinsically fixed vascular stenosis can even be detrimental. The collapsed stent mass constrained within a stenosis functions as a foreign body, which not only exacerbates the obstruction of outflow but in effect represents a potentially thrombogenic intravascular foreign body. For example, in patient 2, the fixed osseous compression of the internal jugular vein clearly restricts expansion of the stent.

Consequently, it is important to recognize this uncommon entity as the cause of bilateral jugular stenosis to avoid an ineffective and potentially detrimental intervention. Computed tomographic venography represents an optimal modality with which to assess patients for styloidogenic jugular compression syndrome because this modality effectively demonstrates the regional osseous structures in relation to the venous vascular anatomy.

After the styloidectomy in patient 2, the self-expanding stent spontaneously opened postoperatively. Thus, in patients who fail to respond to decompression alone (with persistent angiographic stenosis and a retained pressure gradient), we hypothesize that stenting may be a useful adjunctive intervention to augment flow.

CONCLUSION

Styloidogenic jugular compression syndrome is caused by osseous impingement of the jugular vein(s) between the lateral condyle of C1 and a prominent, posteriorly located styloid process. When this impingement is present bilaterally (as in our cases) or when it affects the dominant jugular vein, patients may experience symptomatic central venous hypertension and may be predisposed to venous stasis and cerebral venous sinus thrombosis. This anatomic impingement is best diagnosed with CT venography. It is important to recognize this entity prospectively so that jugular venous stenting, which is an ineffective (and potentially

detrimental) procedure in this condition, can be avoided. Styloid resection is a technically straightforward procedure that can result in excellent clinical outcomes.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENT

The article describes and documents quite clearly the management of 2 patients with extracranial compression of the internal jugular vein by the styloid process. The authors are to be congratulated for making the diagnosis and applying surgical management that resolves the patients' problems. I am particularly impressed with the second case, which also demonstrates the futility of using stents in similar cases. The hope is that this article will deter others from wasting valuable time and resources in similar cases.

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