Styloidogenic Jugular Venous Compression Syndrome: Clinical Features and Case Series

Xiaochun Zhao, MD
Claudio Cavallo, MD
Randall J. Hlubek, MD
Michael A. Mooney, MD
Evgenii Belykh, MD
Sirin Gandhi, MD
Leandro Borba Moreira, MD
Ting Lei, MD
Felipe C. Albuquerque, MD
Mark C. Preul, MD
Peter Nakaji, MD

Department of Neurosurgery, Barrow Neurological Institute, St. Joseph's Hospital and Medical Center, Phoenix, Arizona

Correspondence:
Peter Nakaji, MD,
c/o Neuroscience Publications,
Barrow Neurological Institute,
St. Joseph's Hospital and Medical Center,
350 W. Thomas Rd.,
Phoenix, AZ 85013.
Tel: 602.406.3593; Fax: 602.406.4104
Email: Neuropub@barrowneuro.org

Received, June 5, 2018.
Accepted, February 7, 2019.

BACKGROUND: Styloidogenic jugular venous compression syndrome (SJVCS) is a rare cause of idiopathic intracranial hypertension (IIH).

OBJECTIVE: To elucidate the pathophysiology and the hemodynamics of SJVCS.

METHODS: We conducted a retrospective review of medical records, clinical images, dynamic venography, and manometry for consecutive patients with SJVCS undergoing microsurgical decompression from April 2009 to October 2017. Patients with IIH with normal venography and manometry findings served as controls.

RESULTS: Data were analyzed for 10 patients with SJVCS who presented with headaches. Neck flexion exacerbated headaches in 7 patients. Eleven patients with IIH provided control data for normal intracranial venous pressure and styloid process anatomy. Patients with SJVCS had bilateral osseous compression of venous outflow. The styloid processes were significantly longer in patients with SJVCS than in those with IIH (mean [standard deviation (SD)] distance, 31.0 [10.6] vs 19.0 [14.1] mm; $P < .01$). The styloid process–C1 lateral tubercle distance was shorter in patients with SJVCS than in those with IIH (mean [SD] distance, 2.9 [1.0] vs 9.9 [2.8] mm; $P < .01$). Patients with SJVCS had significantly higher global venous pressure and a higher pressure gradient across the stenosis site than controls (mean [SD] pressure, 2.86 [2.61] vs 0.13 [1.09] cm H_2O; $P = .09$). All 10 patients with SJVCS experienced venous pressure elevation during contralateral neck turning (mean [SD] pressure, 4.29 [2.50] cm H_2O). All 10 patients with SJVCS underwent transcervical microsurgical decompression, and 9 experienced postoperative improvement or resolution of symptoms. One patient had transient postoperative dysphagia and facial drooping, and another patient reported jaw numbness.

CONCLUSION: SJVCS is a novel clinical entity causing IIH. Patients should be evaluated with dynamic venography with manometry. Surgical decompression with removal of osseous overgrowth is an effective treatment in select patients.

KEY WORDS: Idiopathic intracranial hypertension, Jugular venous compression, Styloidectomy

Operative Neurosurgery 0:1–8, 2019 DOI: 10.1093/ons/opz012

Idiopathic intracranial hypertension (IIH), which is also known as pseudotumor cerebri, is a neurological disorder associated with elevated intracranial pressure in the absence of an intracranial mass lesion.[1] IIH typically occurs in obese women[2] and is associated with headaches and visual changes. Venous outflow obstruction, which occurs most commonly in the transverse sinus, is present in some patients with a diagnosis of IIH.[3,4] Several investigators have documented the efficacy of venous sinus stenting for the treatment of IIH associated with venous outflow obstruction.[5-8] Previously, we reported 2 cases of styloidogenic jugular venous compression syndrome (SJVCS) and described an alternative mechanism of venous outflow obstruction caused by subcranial compression of the internal jugular vein (IJV) between the lateral tubercle of C1 and the styloid process.[9]

An elongated styloid process can cause Eagle syndrome, which occurs when the elongated styloid process compresses adjacent

ABBREVIATIONS: 3D, 3-dimensional; **BMI,** body mass index; **CT,** computed tomography; **IIH,** idiopathic intracranial hypertension; **IJV,** internal jugular vein; **SD,** standard deviation; **SJVCS,** styloidogenic jugular venous compression syndrome; **STROBE,** Strengthening of Reporting of Observational Studies in Epidemiology

structures.[10,11] Two subtypes of Eagle syndrome have been described. The classic form is associated with facial pain, voice changes, dysphagia, and throat paresthesia caused by an elongated styloid process impinging the cranial nerves. The other type is associated with a transient ischemic attack caused by an elongated styloid process compressing the carotid artery when the head is turned.[12] Although SJVCS does not fit neatly into either category, it may represent an alternative condition resulting from an elongated styloid process. In this report, we describe the clinical features of patients who underwent microsurgical decompression for SJVCS, elucidate the pathophysiology and the hemodynamics of this condition, and guide diagnosis and treatment for this novel condition.

METHODS

The STROBE (Strengthening of Reporting of Observational Studies in Epidemiology) guidelines for an observational case-control study were applied to this study and report.

Patients

Between April 2009 and October 2017, a total of 10 patients received a diagnosis of SJVCS and were treated at our institution (Barrow Neurological Institute, St. Joseph's Hospital and Medical Center, Phoenix, Arizona). These patients underwent surgical treatment with microsurgical transcervical decompression, which included a styloidectomy and a C1 tuberculectomy. We retrospectively reviewed the medical records and imaging studies of this patient population. None of the patients who received a diagnosis of or treatment for SJVCS were excluded from this analysis. A total of 21 patients with a diagnosis of IIH underwent venography, with or without manometry, for evaluation as part of their work-up for possible styloidogenic stenosis (including the 10 patients with SJVCS; Figure 1). The 11 patients whose findings did not reveal any stenosis at the level of styloid process and C1 tubercle were selected as a control group. Institutional review board approval was obtained for this retrospective analysis of data. All patients had previously consented to the use of their medical records for research purposes, and the need for informed consent was waived because of the retrospective nature of the study.

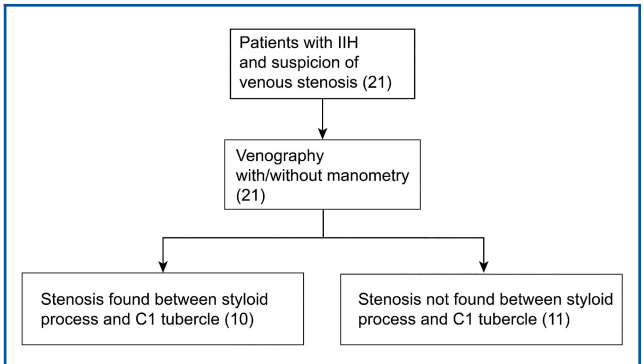

FIGURE 1. *Flow of participants in a study of 21 patients with idiopathic intracranial hypertension (IIH) and suspected venous stenosis. Used with permission from Barrow Neurological Institute.*

Preoperative Clinical Characteristics

In reviewing the institutional medical records of patients with SJVCS and IIH, we extracted information about characteristics such as age, sex, and body mass index (BMI, calculated as weight in kilograms divided by height in meters squared).

Radiological Features

We reviewed preoperative computed tomography (CT) with 3-dimensional (3D) reconstruction for all patients in both the SJVCS group and the control group. CT was used to measure the length of the styloid process, the shortest distance between the styloid process and the C1 tubercle, and the relative position of the styloid tip along the Chamberlain line, which connects the posterior edge of the hard palate and the opisthion. Preoperative neck CT with 3D reconstruction was not available for 1 patient in the SJVCS group and 4 patients in the control group. We measured both sides in the control group, but we measured only the decompression side in the SJVCS group.

Venography and Manometry

We reviewed the operative reports to collect venous pressures at different sites in the jugular venous system. We then calculated the pressure gradient between the jugular bulb and the jugular vein proximal to the C1 lateral tubercle. We compared the mean venous pressure at each site and the mean pressure gradient across the stenosis site, which is at the level of the C1 tubercle in both the SJVCS group and the control group. We included the pressure readings at the sagittal sinus, torcular, transverse sinus, sigmoid sinus, jugular bulb, and proximal IJV on both sides in the control group, whereas we included only the pressure readings at the decompression side in the SJVCS group. One patient in the SJVCS group did not undergo preoperative venography. The results for 5 patients in the control group were excluded from the venous pressure measurements, including 4 who had a history of transverse sinus thrombosis and 1 who underwent venography but not manometry because of an intraprocedural hemorrhagic event. We also compared the pressure taken at the jugular bulb before and after turning the head to the contralateral side in the SJVCS group.

Hospital Course

For the SJVCS group, we reviewed the surgical reports and follow-up medical records to determine the length of the removed styloid process in the surgery. We also identified any postoperative complications and determined clinical outcomes.

Statistical Analysis

The statistical analysis was performed using SPSS 18.0.0 (IBM Inc, Armonk, New York). As the data were not normally distributed, the Mann-Whitney *U* test was used for all comparisons of numerical variables, and χ^2 tests were used for categorical variables. Means and standard deviations are reported. Differences were considered significant if the *P* value was less than .05.

TABLE 1. Clinical Characteristics of 10 Patients With SJVCS

Variable	No. (%)[a]
Age, mean (range), yr	35.8 (21-59)
Sex	
Female	3 (30)
Male	7 (70)
BMI, mean (SD)	26.9 (6.5)
Symptoms[b]	
Headache	10 (100)
Aggravation with neck flexion	7 (70)
Blurry vision	5 (50)
Nausea and vomiting	1 (10)
Pressure elevation during neck turning, mean (SD), cm H_2O	4.29 (2.50)
Operation side	
Left	3 (30)
Right	7 (70)
Length of styloid process removed, mean (SD), mm	18.4 (10.6)
C1 tubercula drilling	10 (100)
Hospital stay, mean (range), d	2.1 (1-6)
Follow-up duration, mean (range), mo	11.8 (1-42)
Complications	
Difficulty swallowing	1 (10)
Facial droop	1 (10)
Jaw numbness	1 (10)
Outcome	
Resolved	3 (30)
Improved	6 (60)
Unchanged	1 (10)
Relapsed	1 (10)

BMI, body mass index, calculated as weight in kilograms divided by height in meters squared; SJVCS, styloidogenic jugular venous compression syndrome.
[a]Values are number (percentage) unless indicated otherwise.
[b]Percentages total more than 100% because some patients presented with more than 1 symptom.

RESULTS

Preoperative Clinical Characteristics

General characteristics of patients are summarized in Table 1, and each case is briefly reviewed in Table 2. Seven of the 10 patients were male, and the mean (SD) patient age was 35.8 (12.9) yr (range, 21-59 yr). The mean (SD) BMI of the SJVCS group was 26.9 (6.5). Only 1 of 10 patients was obese, with a BMI $\geq$ 30. All patients reported headache as part of their clinical presentation. Seven patients experienced a gradual exacerbation of headaches with neck flexion or upon turning the head to the contralateral side. Five patients presented with decreased visual acuity (ie, blurry vision), and 1 patient presented with nausea and vomiting. In the control group, the mean age was 37.7 $\pm$ 13.4 (range 19-40) yr, 9 of 11 patients were female, and 8 patients had a BMI $\geq$ 30 (mean [SD] BMI, 35.3 [9.6]). All control patients reported headaches that were not positional in nature. A comparison of the rate of obesity and the rate of positional headache is summarized in Table 3. The SJVCS group had a lower obesity rate (10% vs 73% for controls; $P < .01$) and a higher rate of positional headaches (70% vs 0% for controls; $P < .01$).

Radiological Features

Table 4 summarizes our comparison of the radiological features of the 2 groups. The SJVCS group had a significantly longer styloid process than the control group (mean [SD] distance, 31.0 [10.6] vs 19.0 [14.1] mm; $P < .01$). The distances between the styloid process and the C1 lateral tubercle in the SJVCS group were also significantly different than those in the control group (mean [SD] distance, 2.9 [1.0] vs 9.9 [2.8] mm; $P < .01$). With the Chamberlain line used as a reference, the difference in the relative position of the styloid tip between the 2 groups was not significant (mean [SD] difference, −14.5 [12.7] vs −7.0 [13.3] mm; $P = .09$). Although we measured only the decompression side in the SJVCS group, we noticed that the compression was usually bilateral and that a dilated occipital sinus can provide compensatory drainage, as shown in Figure 2 (patient 4).

Venography and Manometry

The results of venous pressure measurements at different sites in the intracranial venous system are demonstrated in Table 5. The intracranial venous pressures were significantly higher in the SJVCS group at all sites above the stenosis site (all $P \leq .02$). There was no significant difference found at the proximal IJV, which is proximal to the stenosis site. We reported a significantly higher gradient across the stenosis site in the SJVCS group (mean [SD] pressure, 2.86 [2.61] cm H_2O) than in the control group (mean [SD] pressure, 0.13 [1.09] cm H_2O). In the SJVCS group, an elevated pressure (mean [SD], 4.29 [2.50] cm H_2O) was also found when the head was turned to the contralateral side.

Microsurgical Decompression

Nine of the 10 patients with SJVCS underwent decompression with a C1 tuberculectomy and a styloidectomy. In one case (patient 9), the styloidectomy was aborted because decompression from the C1 lateral tuberculectomy and the transection of the styloid ligaments were sufficient to relieve the venous outflow obstruction. Because of the universally bilateral nature of the venous outflow obstruction, the preference at our institution is to perform decompression of the nondominant jugular vein first. This strategy is used to minimize the likelihood of injury to the dominant venous drainage, which could further intensify the patient's IIH-like pathology. The mean (SD) length of styloid process that was removed was 18.4 (10.6) mm.

Hospital Course

The mean hospital stay was 2.1 d (range, 1-6 d), and the mean duration of follow-up was 11.8 mo (range, 1-42 mo). The complications after microsurgical decompression were minimal. One patient had transient postoperative dysphagia and facial droop, and another patient reported postoperative jaw numbness. Nine of the 10 patients with SJVCS improved after surgery; among these, 3 patients reported complete resolution of preoperative

TABLE 2. Summary of 10 Cases of SJVCS

Pt. No.	Sex	Age, yr	BMI	Symptoms	Positional headache	Side of operation	Styloid process removed, mm	Hospital stay, d	Complications	Outcome
1	M	23	23.0	Headache	Yes, with neck flexion	R	42	1	None	Improved
2	F	30	22.3	Headache, blurry vision	Yes, with neck flexion	R	20	6	Facial droop and dysphagia	Resolved
3	M	59	29.9	Headache, blurry vision	Yes, with neck flexion	R	17	1	None	Improved
4	F	44	23.4	Headache	Yes, with right neck flexion	R	11	3	Numbness of jaw, neck, and ear	Improved
5[a]	M	51	42.9	Headache, blurry vision	NR	L	15	3	None	Resolved
6	M	38	26.3	Headache, blurry vision	Yes, with neck flexion	R	20	2	None	No change
7	M	32	23.5	Headache	Yes, with left neck flexion	R	20	1	None	Improved
8	F	21	26.7	Headache, blurry vision	Yes, with neck flexion	L	24	2	None	Improved, returned in 6 mo
9	M	21	24.0	Headache	NR	L	0	1	None	Improved
10[b]	M	39	NR	Headache	NR	R	15	1	None	Resolved

BMI, body mass index, calculated as weight in kilograms divided by height in meters squared; F, female; L, left; M, male; NR, not reported; Pt., patient; R, right; SJVCS, styloidogenic jugular venous compression syndrome.
[a]Died 6 mo after surgery due to myocardial infarction.
[b]Underwent previous internal jugular vein stent.

symptoms immediately after surgery. Symptoms were postoperatively unchanged in 1 patient. One patient, who had significant postoperative improvement, reported a headache relapse 6 mo after surgery.

DISCUSSION

SJVCS is a novel clinical entity with the anatomical features of Eagle syndrome and the symptomatic features of IIH. We report and discuss the clinical features, diagnosis, treatment, and outcomes of transcervical decompression for SJVCS at our institution.

Clinical Features

To identify the distinct clinical characteristics of this group of patients, we compared data about BMI and the quality of headache with those of the control group. We found a significantly lower obesity rate in the SJVCS group than in the control group (10% vs 73%; $P < .01$), which suggests that patients with SJVCS tend to have normal weight compared with patients with IIH. Positional headache associated with neck flexion was a unique feature of the patients with SJVCS. Indeed, neither IIH nor intracranial venous sinus stenosis is associated with positional headache.[8]

TABLE 3. Comparison of Clinical Characteristics of Patients With SJVCS and Controls

Variable	No. (%) of patients with SJVCS (n = 10)	No. (%) of controls (n = 11)	*P* value
Obesity	1 (10)	8 (73)	<.01
Positional headache	7 (70)	0 (0)	<.01

SJVCS, styloidogenic jugular venous compression syndrome.

Given the prevalence of headaches secondary to IIH and the complexity of this diagnosis, we hypothesize that SJVCS may be an underdiagnosed cause of headaches in this patient population. Notably, bilateral venous outflow obstruction is found incidentally in a significant portion of patients who undergo CT angiography (10 of 108 [9.3%]; in one series).[13]

Anatomical Considerations

The IJV is located anterior to the C1 lateral tubercle and posterior to the styloid process at the craniocervical junction. The styloid process is a bony outgrowth from the lower surface of the

TABLE 4. Mann-Whitney *U* Test of the Anatomy of the Styloid Process in Patients With SJVCS and Controls

Variable	SJVCS group, mean (SD), mm (n = 9 × 1 side)	Control group, mean (SD), mm (n = 7 × 2 sides)	*P* value
Length of the styloid process	31.0 (10.6)	19.0 (14.1)	<.01
Shortest distance between the styloid process and the C1 tubercle	2.9 (1.0)	9.9 (2.8)	<.01
Relative position of the styloid process tip along the Chamberlain line	−14.5 (12.7)	−7.0 (13.3)	.09

SJVCS, styloidogenic jugular venous compression syndrome.
Computed tomography and 3-dimensional reconstruction images were used to measure both sides in the control group and the decompression side in the SJVCS group.

temporal bone, and it is the point of attachment for the stylohyoid muscles.[14] A normal styloid process is 1.5 to 4 cm long.[15-17] The patients with SJVCS had a longer styloid process and a shorter distance between the styloid process and the C1 lateral tubercle than the patients in the control group.

Neck flexion may cause greater compression of the jugular vein as the styloid process moves posteriorly toward the C1 lateral tubercle, as demonstrated in Figure 3. We piloted performing neck CT imaging with 3D reconstruction for the neutral position, with neck extension and with neck flexion. We found that CT imaging with multiple neck positions can have a higher diagnostic specificity than CT imaging with the patient only in the neutral position.

An elongated styloid process, an enlarged C1 lateral tubercle, or an abnormal juxtaposition of these 2 structures might compress the jugular vein, resulting in the pathophysiological development of SJVCS. In our cohort, some patients with SJVCS had styloid processes that were relatively short but benefited from decompression (patient 4). Ho et al[18] proposed that the juxtaposition of a styloid process of normal length with the C1 lateral tubercle may result in Eagle syndrome. We hypothesize that the positional relationship of the styloid process in relation to the C1 lateral tubercle could be more important in the pathophysiology of SJVCS than the length of the styloid process itself.

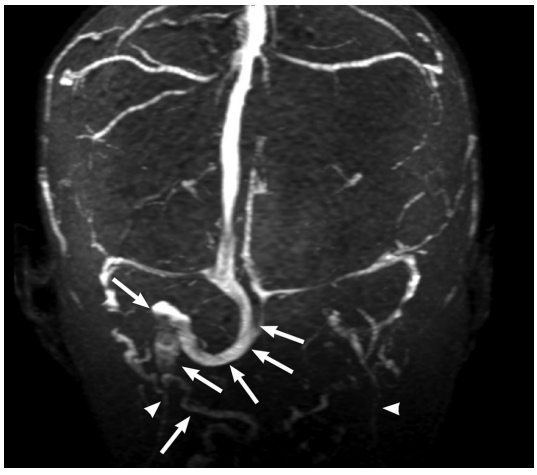

FIGURE 2. *Preoperative magnetic resonance venography illustrating the stenosis of both internal jugular veins (arrowheads) and the dilated occipital sinus providing compensatory venous drainage (arrows). Used with permission from Barrow Neurological Institute.*

Venography With Manometry as a Diagnostic Tool

High venous pressure can cause elevated intracranial pressure, and several authors have described IIH caused by venous sinus stenosis, which can be treated with dural venous sinus stenting.[19,20] We found a significantly higher global venous pressure distal to the stenosis site in the patients with SJVCS than in the patients with IIH, but we found no significant difference at the proximal IJV. Moreover, we found a mean (SD) pressure gradient of 2.86 (2.61) cm H_2O between the jugular bulb and the proximal IJV, which was significantly different from that of the control group (mean [SD] pressure, 0.13 [1.09] cm H_2O; $P = .03$). This finding indicates that elevated intracranial pressure was associated with stenosis. In our series, one patient (patient 10) had undergone a previous stenting treatment, but it was not effective because the compression was osseous in nature.

We found a mean (SD) venous pressure elevation of 4.29 (2.50) cm H_2O at the jugular bulb while the patient's head was turned to the contralateral side. This finding may explain the associated positional headache during neck flexion. Venography with manometry for different neck positions can be a useful supplementary diagnostic step for this disease. As an effective tool for diagnosis, venography with manometry can evaluate high global venous pressure, the gradient across the stenosis site, and any elevation of the venous pressure during neck flexion.

Surgical Consideration

As our results indicate, microsurgical decompression of the jugular vein is effective. However, venous stenting is less effective when compression is osseous. The surgical steps are similar to a styloidectomy, which is used to treat Eagle syndrome,[21] but a C1 tuberculectomy is an essential and unique technique to relieve the compression of the jugular vein between the styloid process and the C1 tubercle. In one patient (patient 9), we performed only a C1 tuberculectomy with transection of styloid ligaments. Styloidectomy was not performed, as we believed that the decompression would have been already sufficient, and the patient's symptoms improved postoperatively.

TABLE 5. Mann-Whitney *U* Test of the Parameters of Venography and Manometry of Patients With SJVCS and Controls

Location	Pressure, mean (SD), cm H_2O		
	SJVCS group (n = 10)[a]	Control group (n = 6)[b]	*P* value
At posterior superior sagittal sinus	14.11 (20.9)	9.14 (3.34)	<.01
At torcular	13.44 (2.06)	9.29 (3.40)	.02
At transverse sinus	12.22 (2.64)	8.67 (3.28)	.02
At sigmoid sinus	11.33 (2.74)	6.41 (2.87)	<.01
At jugular bulb	11.00 (3.24)	5.75 (3.33)	<.01
At proximal jugular vein	8.43 (3.51)	5.58 (3.02)	.07
Gradient across the stenosis site	2.86 (2.61)	0.13 (1.09)	.03

SJVCS, styloidogenic jugular venous compression syndrome.
[a]Ten of the 11 pressure readings were taken only on the decompression side in the SJVCS group.
[b]Five of the 11 patients in the control group were excluded from the venous pressure measurements because of a history of severe transverse sinus thrombosis. Pressure readings were taken on both sides in the control group for transverse sinus, sigmoid sinus, jugular bulb, and jugular vein; thus, 12 readings were used in the comparison. One reading was taken per patient for the superior sagittal sinus and torcular pressure in the control group; thus, 6 readings were used in the comparison.

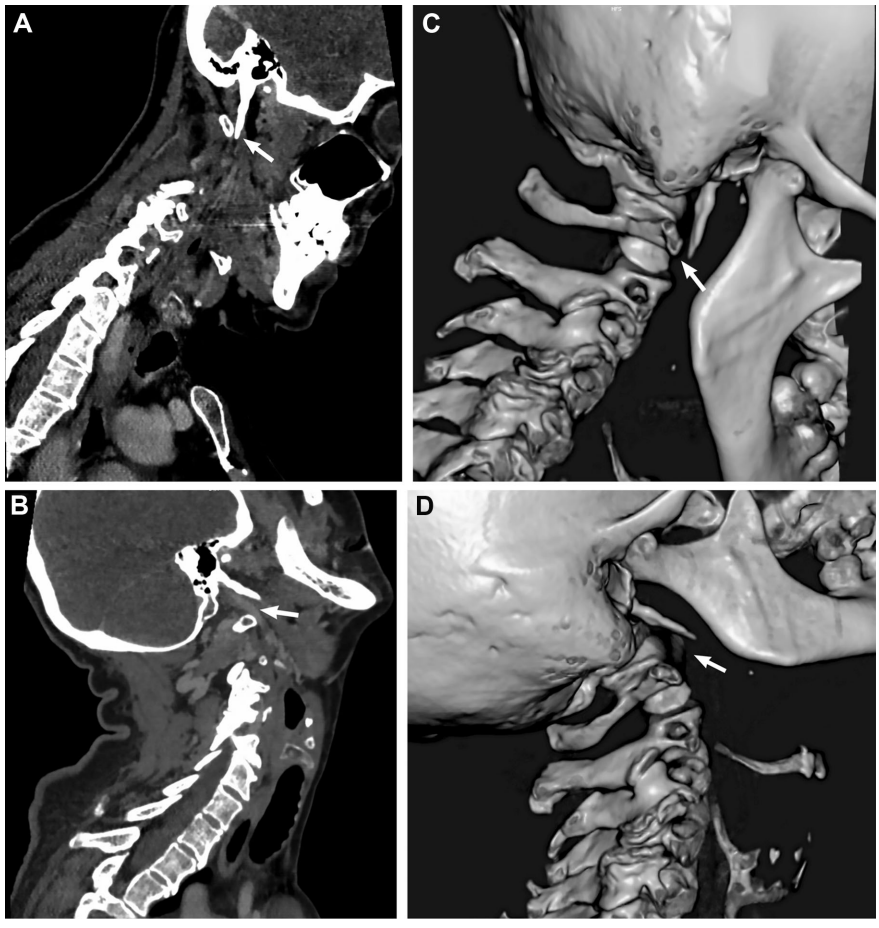

FIGURE 3. *Computed tomography (CT) of the right side of the neck during* **A**, *neck flexion and* **B**, *extension, and 3-dimensional reconstruction neck during* **C**, *neck flexion and* **D**, *extension. The space (arrows) between the styloid process and the C1 lateral tubercle was much narrower during neck flexion than during neck extension. Used with permission from Barrow Neurological Institute.*

Study Limitations

Our case series contained only a small number of patients, and the retrospective and observational nature of this study means that it is limited by possible inaccuracy in the recording of data. Missing data for the subjects were excluded, which can present a risk of bias.

CONCLUSION

SJVCS is a novel clinical entity that is similar to IIH, and patients with these conditions often present with the same symptoms. Jugular venous compression is caused by the C1 lateral tubercle and the styloid process. Preoperative CT 3D reconstructions, with the neck in different positions, can reveal the abnormal positional relationship, and venography with manometry, with and without the head turned, can evaluate the severity of the compression and the intracranial hypertension. Surgical decompression with resection of the osseous overgrowth is an effective treatment with a good outcome for select patients.

Disclosure

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

REFERENCES

1. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology*. 2002;59(10):1492-1495.
2. Rowe F, Sarkies N. The relationship between obesity and idiopathic intracranial hypertension. *Int J Obes*. 1999;23(1):54-59.
3. Farb R, Vanek I, Scott J, Mikulis D, Willinsky R, Tomlinson G. Idiopathic intracranial hypertension: the prevalence and morphology of sinovenous stenosis. *Neurology*. 2003;60(9):1418-1424.
4. Johnston I, Kollar C, Dunkley S, Assaad N, Parker G. Cranial venous outflow obstruction in the pseudotumour syndrome: incidence, nature and relevance. *J Clin Neurosci*. 2002;9(3):273-278.
5. Gross BA, Albuquerque FC, Moon K, McDougall CG. New frontiers in venous sinus stenting: illustrative cases. *J Clin Neurosci*. 2016;33(Nov):241-244.
6. Asif H, Craven CL, Siddiqui AH, et al. Idiopathic intracranial hypertension: 120-day clinical, radiological, and manometric outcomes after stent insertion into the dural venous sinus. *J Neurosurg*. 2017;129(3):723-731.
7. Ahmed R, Wilkinson M, Parker G, et al. Transverse sinus stenting for idiopathic intracranial hypertension: a review of 52 patients and of model predictions. *AJNR Am J Neuroradiol*. 2011;32(8):1408-1414.
8. Albuquerque FC, Dashti SR, Hu YC, et al. Intracranial venous sinus stenting for benign intracranial hypertension: clinical indications, technique, and preliminary results. *World Neurosurg*. 2011;75(5-6):648-652.
9. Dashti SR, Nakaji P, Hu YC, et al. Styloidogenic jugular venous compression syndrome. *Neurosurgery*. 2012;70(3):E795-E799.
10. Fusco DJ, Asteraki S, Spetzler RF. Eagle's syndrome: embryology, anatomy, and clinical management. *Acta Neurochir*. 2012;154(7):1119-1126.
11. Eagle W. The symptoms, diagnosis and treatment of the elongated styloid process. *Am Surg*. 1962;28(Jan):1-5.
12. Chuang W, Short J, McKinney A, Anker L, Knoll B, McKinney Z. Reversible left hemispheric ischemia secondary to carotid compression in Eagle syndrome: surgical and CT angiographic correlation. *Am J Neuroradiol*. 2007;28(1):143-145.
13. Jayaraman M, Boxerman J, Davis L, Haas R, Rogg J. Incidence of extrinsic compression of the internal jugular vein in unselected patients undergoing CT angiography. *AJNR Am J Neuroradiol*. 2012;33(7):1247-1250.
14. Martin TJ, Friedland DR, Merati AL. Transcervical resection of the styloid process in Eagle syndrome. *Ear Nose Throat J*. 2008;87(7):399.
15. Kaufman SM, Elzay RP, Irish EF. Styloid process variation: radiologic and clinical study. *Arch Otolaryngol*. 1970;91(5):460-463.
16. Monsour PA, Young WG. Variability of the styloid process and stylohyoid ligament in panoramic radiographs. *Oral Surg Oral Med Oral Pathol*. 1986;61(5):522-526.
17. Balcioglu H, Kilic C, Akyol M, Ozan H, Kokten G. Length of the styloid process and anatomical implications for Eagle's syndrome. *Folia Morphol (Warsz)*. 2009;68(4):265-270.
18. Ho S, Luginbuhl A, Finden S, Curry JM, Cognetti DM. Styloid/C1 transverse process juxtaposition as a cause of Eagle's syndrome. *Head Neck*. 2015;37(11):153-156.
19. Satti SR, Leishangthem L, Spiotta A, Chaudry MI. Dural venous sinus stenting for medically and surgically refractory idiopathic intracranial hypertension. *Interv Neuroradiol*. 2017;23(2):186-193.
20. Liu KC, Starke RM, Durst CR, et al. Venous sinus stenting for reduction of intracranial pressure in IIH: a prospective pilot study. *J Neurosurg*. 2016;127(5):1126-1133.
21. Jalisi S, Jamal BT, Grillone GA. Surgical management of long-standing eagle's syndrome. *Ann Maxillofac Surg*. 2017;7(2):232.

Acknowledgment

The authors thank the staff of Neuroscience Publications at Barrow Neurological Institute for assistance with manuscript preparation.

COMMENT

The authors present a case series in which they describe a novel entity, "styloidogenic jugular venous compression syndrome" (SJVCS). The authors aimed to show that a subset of patients diagnosed with idiopathic intracranial hypertension (IIH) in fact suffer from an entity similar to Eagle Syndrome, in which the jugular veins are compressed bilaterally between the styloid process and the C1 tubercle, which leads to decreased venous outflow and consequently, intracranial hyperemia and increased ICP. The authors go on show that surgical removal of the styloid process and a portion of the C1 tubercle relieves the stenosis and restores normal venous outflow from the intracranial compartment. The cohort of patients with SJVCS was compared to matched IIH patients without venous outflow compression. The authors revealed that, in the SJVCS group, craniocervical metrics related to the styloid processes and C1 tubercle differed significantly between groups. In addition, the SJCVS group suffered high global venous pressure and mean pressure gradient of 2.86 cm H_2O across the stenosis, which was exacerbated during contralateral neck turning. Overall, the authors should be congratulated for their description of this rare condition. If nothing else, it should be kept in the back of every neurosurgeon's mind when evaluating patients with IIH. Our colleagues further describe their surgical results with styloidectomy and demonstrate the procedure to be safe within the confines of this small cohort. While these data are certainly interesting and well presented, questions remain given the SJCVS group consisted of only 10 patients and the true prevalence and safety of surgical treatment require further investigation. For instance, why was mean pressure gradient in this cohort approximately 3 cm H_2O, while that for patients who are candidates for transverse sinus stenting for IIH closer to 10 cm H_2O or more? Given the presumed rarity of this condition, a meaningful prospective study aimed at determining the prevalence and surgical outcomes may not be imminently forthcoming, but collaborative effort across institutions might pool enough patients with presumed IIH to better evaluate the frequency with which these patients have

craniocervical metrics consistent with SJCVS and to complete multivariate analyses to determine if particular factors might suggest to the treating surgeon that he or she should look more carefully for SJCVS and those that are more indicative of "run of the mill IIH". Much as venous sinus stenting has proven effective in treating a subset of patients with intracranial hypertension due to venous outflow stenosis, so too may styloidectomy in patients with SJCVS, while avoiding an indwelling CSF shunt with its inherent risks. If these data can be corroborated, time will tell.

Brian M. Howard
Atlanta, Georgia