Internal jugular venous aneurysm—A report of two cases with literature review

Tamas Kovacs, MD, Salah El Haddi, MD, *and* W. Anthony Lee, MD, *Boca Raton, Fla*

ABSTRACT

Internal jugular venous aneurysm (IJVA) is a rare entity that usually remains asymptomatic with only rare complications. We report two cases of IJVA. Both patients presented with a palpable soft tissue mass in the neck and were found to have IJVA on imaging with associated lymphadenopathy. In both cases, the aneurysms and involved lymph nodes were resected, with the jugular vein being primarily reconstructed. There are only a few case reports involving IJVA, and treatment guidelines are not well established. Whereas nonoperative management is frequently chosen, the most common indication for surgery is cosmetic; both management options have favorable outcomes. (J Vasc Surg Cases and Innovative Techniques 2020;6:326-30.)

Keywords: Internal jugular vein; Venous aneurysm; Surgery

Aneurysms of the internal jugular veins are typically asymptomatic and are manifested as a palpable soft tissue mass of the neck. They are often misdiagnosed because of the wide range of differential diagnoses that include benign or malignant head and neck neoplasms, such as lymphoma, lymphadenopathy, lipoma, lymphangioma, laryngocele, branchial cyst, cystic hygroma, hemangioma, and carotid body tumor.[1,2] The exact cause remains unknown; however, histopathologic findings suggest a localized degenerative process due to an increase in matrix metalloproteinases (MMPs).[3,4]

We present two cases of internal jugular venous aneurysm (IJVA) with associated lymphadenopathy that underwent resection and primary repair. The patients agreed with and consented for the publication of their cases.

CASE REPORTS

Case 1. An 81-year-old man with history of atrial fibrillation, hypertension, and diabetes presented with a palpable soft tissue mass in the left submandibular area (Fig 1). He was referred to an oncologist by his primary care physician. He initially underwent imaging studies that included duplex ultrasound and computed tomography (CT) venography (Fig 2). The duplex ultrasound examination demonstrated a large, blood-containing structure in the left mandibular region, with mixed intraluminal arterial and venous flow, suggestive of underlying arteriovenous fistula with a venous aneurysm. CT venography confirmed a jugular aneurysm with multiple feeding collaterals arising from it but also described arterial flow within the mass concerning for an arteriovenous fistula. To rule out a fistulous connection, internal jugular venography and carotid angiography were performed, and both were negative for any arterial connection to the aneurysm (Fig 3). The patient was referred for exploration and resection for cosmetic reasons per the patient's wishes and to rule out any associated pathologic processes. Intraoperatively, the aneurysm was pedunculated and connected to the internal jugular vein by a slender venous pedicle and attached to a benign-appearing, enlarged lymph node. The aneurysm and the lymph node were resected en bloc with ligation of multiple feeding branches and simple ligation of the pedicle to the internal jugular vein (Fig 4). The aneurysm measured 3.5 × 1.7 cm, and histologic evaluation showed a smooth lined endothelium and thinned wall without malignant changes and a 0.7 × 1.2 cm lymph node with no pathologic findings. The patient had an uneventful postoperative course. Anticoagulation for atrial fibrillation was restarted, and the patient was discharged on postoperative day 1.

Case 2. A 50-year-old man with no significant medical history presented with a palpable left-sided neck mass. Initial workup was performed by an otorhinolaryngologist. Duplex ultrasound revealed a 3.6 × 2.2 × 1.4 cm mass without any apparent flow. A non-contrast-enhanced CT scan was consistent with a saccular IJVA with an associated 2 cm mass. Core needle biopsy of the mass was nondiagnostic. The patient underwent a combined procedure with the otorhinolaryngology surgery service during which the venous aneurysm was resected with the adjacent mass along with an en bloc level IIA and III lymph node dissection (Fig 5, *A-C*) to ensure clear margins in the event of a malignant neoplasm. The internal jugular vein was then repaired primarily with a lateral venorrhaphy. Pathologic

From the Department of Surgery, Charles E. Schmidt College of Medicine, Florida Atlantic University.

Author conflict of interest: none.

Presented at the Vascular Techniques & Technology Thinktank (VAST[3]), Saratoga Springs, NY, August 1-3, 2019.

Correspondence: Tamas Kovacs, MD, Florida Atlantic University, Charles E. Schmidt College of Medicine, Department of Surgery, 777 Glades Rd, Boca Raton, FL 33431 (e-mail: tdrkovacs@health.fau.edu).

The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

https://doi.org/10.1016/j.jvscit.2020.04.004

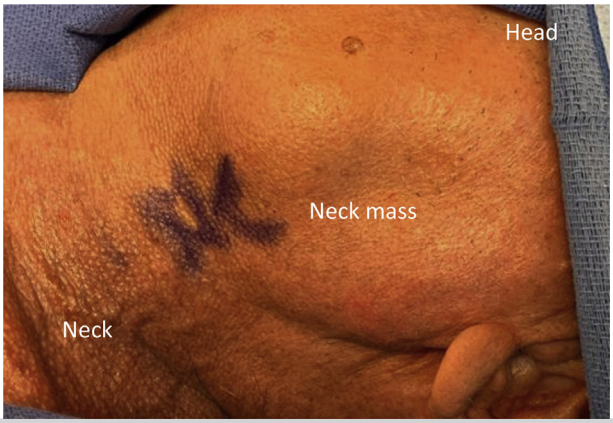

Fig 1. Palpable neck mass.

examination revealed an unremarkable venous aneurysm with an associated small benign hemangioma and reactive lymph nodes (Fig 5, *D*). The patient suffered no complications with no evidence of recurrence at 6 months of follow-up. He received no perioperative anticoagulation because of the low reported risk of thromboembolic complications.[5-8]

DISCUSSION

Aneurysms are abnormal dilation of blood vessels that are usually found in arteries. Venous aneurysms, although rare, have been described and can involve the extremities, the head and neck, and the abdomen. IJVAs are defined as an area of dilation two or three times the adjacent jugular vein.[9] Venous aneurysms can be divided into primary (congenital) or secondary (acquired) cases. Congenital aneurysms are more common in children and caused by inherited weakness of the vessel wall, as in neurofibromatosis type 1 and Ehlers-Danlos syndrome. Acquired IJVAs can arise from trauma, inflammation, mechanical stress, invasive procedures such as central venous catheterization, positive pressure ventilation, neck operations, and other degenerative processes.[10-13] There are only scarce case reports in the literature, with only 247 cases identified to date in a recent systematic review.[14] IJVA most commonly is manifested as a unilateral, asymptomatic, typically right-sided, intermittent or persistent neck mass that enlarges with increased intrathoracic pressure, such as a Valsalva maneuver. The right to left ratio is 4:1. The mechanism for this apparent laterality has been speculated to be secondary to the proximity of the right brachiocephalic vein to the apical pleura, which could result in increased intrathoracic pressure being transmitted preferentially to the right (vs left) internal jugular vein, making it susceptible to aneurysm formation.[15,16] Only a few bilateral cases have been described. They are more commonly found in children than in adults with an incidence of 84% vs 16%. In adults, women are more likely to be affected, whereas there is a male predominance in children.

The most common presentation of an IJVA is a persistent or intermittent neck mass that is typically found incidentally on physical examination. Size may increase on talking, coughing, or swallowing. Other presenting signs and symptoms include voice changes, dysphagia, Horner syndrome, and ear pain due to the proximity to the vagus nerve and other cranial nerves. The average diameter in adults is 3.9 cm on the right and 4.7 cm on the left.[14] Duplex ultrasound is the most commonly used diagnostic study, followed by contrast-enhanced CT, venography, angiography, and magnetic resonance imaging.

Whereas the cause of venous aneurysms remains unclear, histopathologic findings suggest a localized degenerative process resulting in thinning of the elastic and muscular layers that could be inherited, such as neurofibromatosis type 1, or acquired by trauma, mechanical stress, or inflammation. MMPs like MMP-2, MMP-9, and MMP-13 have been found to have increased expression in endothelial cells of venous aneurysms.[4] On microscopic examination, they are characterized by fragmentation of the elastic lamellae, loss of smooth muscle cells, and attenuation of the venous wall compared with normal saphenous veins; however, most cases show no significant changes compared with the normal venous walls. Some studies suggest a relationship between elevated estrogen levels and venous aneurysms, which can explain the increased formation of venous aneurysm during pregnancy and the presence of estrogen receptors in venous tissue, but direct correlation has never been established.[17-20]

Complications such as spontaneous rupture, thrombophlebitis, thrombosis, and pulmonary embolism are rare.[5-8] When thromboses occur, they are more frequent in adults than in pediatric patients (17% vs 1.5%, respectively).[14] There has been only one case report that described a pulmonary embolism in the presence of IJVA thrombus with no other source.[21] Because of the rarity of this condition, treatment guidelines for IJVAs are not well established. The most frequently reported treatment in the pediatric population is nonoperative management. However, in adults, both surgery and conservative management have been chosen equally with similar favorable outcomes. When the diagnosis is made, the most common indication for surgery is cosmetic and for symptoms including pain, swelling, or tenderness from local tissue compression. Saccular aneurysms have higher tendency for development of mural thrombosis compared with those that are fusiform.[22] Some authors suggest that pain or tenderness is related

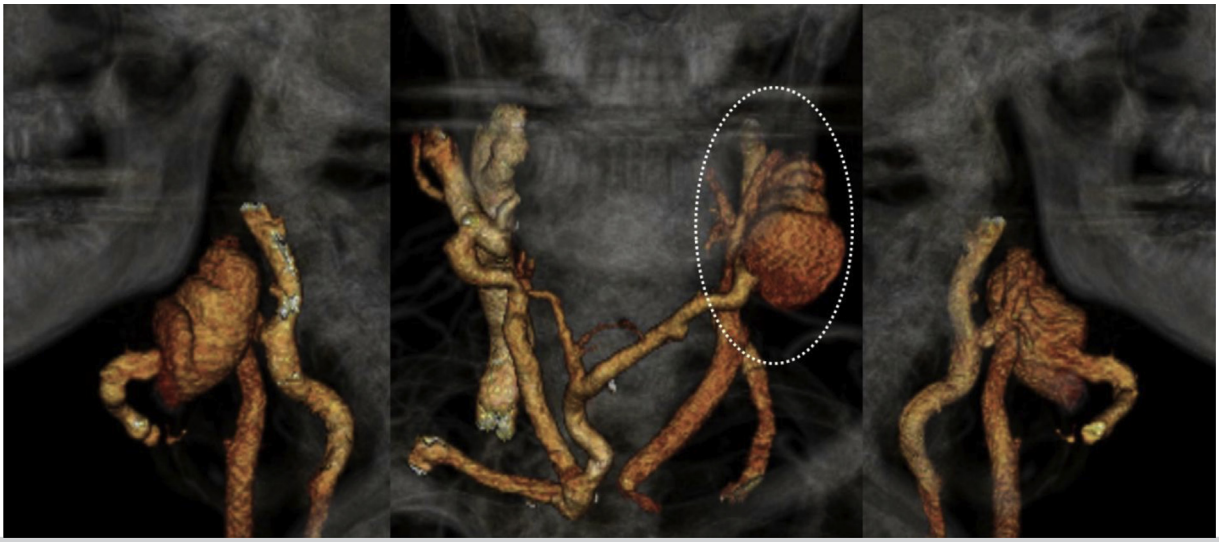

Fig 2. Three-dimensional computed tomography (CT) venography images showing right lateral, anteroposterior, and left lateral projections of the internal jugular venous aneurysm (IJVA).

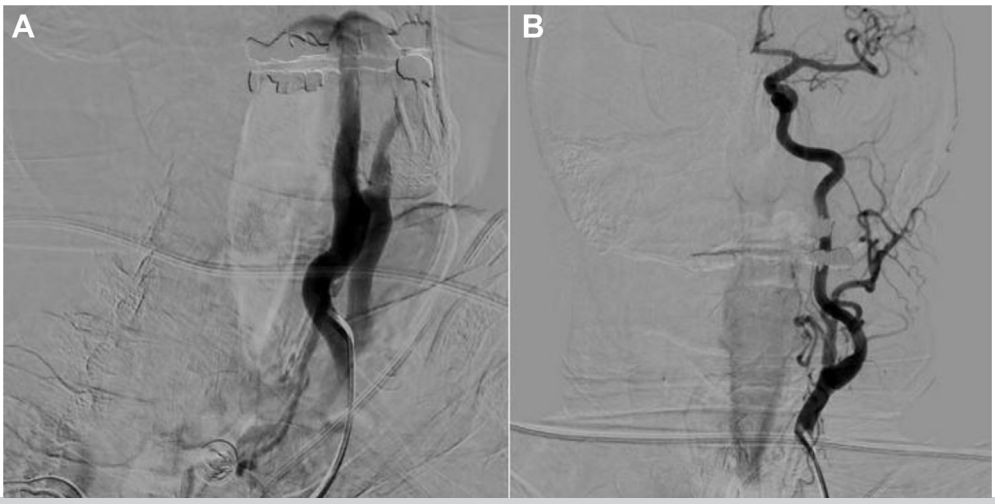

Fig 3. Venogram of the left internal jugular vein aneurysm **(A)** and left carotid angiogram demonstrating no fistulous connection **(B)**.

to underlying mural thrombus formation, and therefore resection should be considered more often for a saccular aneurysm than for fusiform dilations.[23]

If resection is chosen, ligation of the feeding vessels with aneurysmectomy and lateral venorrhaphy or rarely with a vein patch angioplasty is frequently the operation of choice. Other surgical options include aneurysm resection with primary anastomosis or interposition grafting. Endovascular options have also been successfully implemented in rare circumstances.[24,25]

Whereas hemangiomas are often included in the differential diagnosis for a neck mass, there has been no cases of a hemangioma within the wall of a venous aneurysm as was described in our second case. Hemangiomas have not been known to be classically associated with IJVAs or to cause lymphadenopathy. However, IJVAs can form secondary to inflammation or mechanical stress that could have been induced by the presence of the associated hemangioma in our second case. The reactive lymph nodes found in the histologic specimen were likely to be the result of a localized inflammatory reaction due to tissue compression.

CONCLUSIONS

IJVA is a rare, benign condition that is often treated conservatively but may be resected with favorable results and low morbidity.

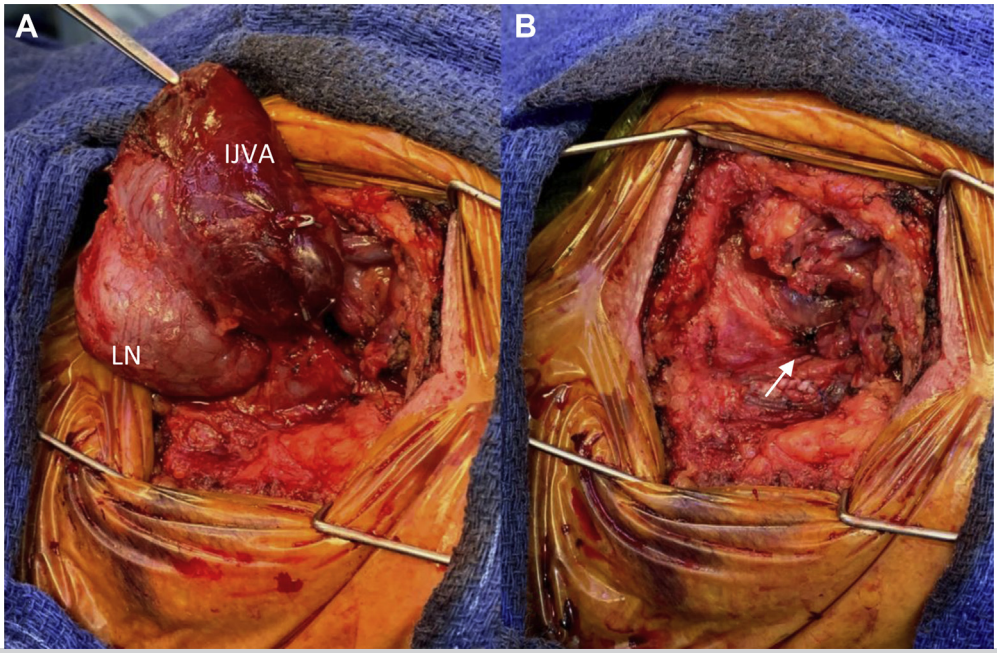

Fig 4. A, Internal jugular venous aneurysm (*IJVA*) in situ with an adjacent enlarged lymph node (*LN*). **B,** After en bloc excision with ligation of the inflow pedicle (*arrow*).

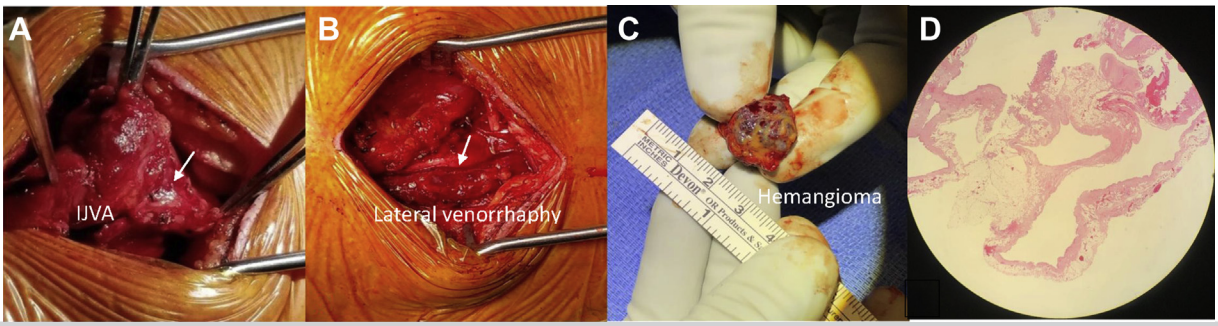

Fig 5. A, Internal jugular venous aneurysm (*IJVA*) in situ with small adjacent mass (*arrow*). **B,** After resection with lateral venorrhaphy (*arrow*). **C,** Small adjacent hemangioma. **D,** IJVA on histologic evaluation with attenuated wall.

REFERENCES

1. Kloppenburg GT, de Vries JP, Schuurman JP, Koelemij R, Wille J. Current perspectives on management of congenital jugular vein aneurysms. Vasc Endovasc Surg 2011;45:237-40.
2. Bora MK. Internal jugular phlebectasia: diagnosis by ultrasonography, Doppler and contrast CT. J Clin Diagn Res 2013;7:1194-6.
3. Gloviczki P. Handbook of venous and lymphatic disorders: guidelines of the American Venous Forum. 4th ed. Boca Raton, Fla: CRC Press; 2017.
4. Irwin C, Synn A, Kraiss L, Zhang Q, Griffen MM, Hunter GC. Metalloproteinase expression in venous aneurysms. J Vasc Surg 2008;48:1278-85.
5. Alshehri WM, Alothmani SK, Alshamrani AM, Almadhari RI, Alqahtani AS, Alrasheedi Saud D. Internal jugular vein aneurysm: a case report. J Surg Case Rep 2019;2019:rjz159.
6. Khashram M, Walker PJ. Internal jugular venous aneurysm. J Vasc Surg Venous Lymphat Disord 2015;3:94.
7. Chua E, Udom V, Huang DY. Internal jugular vein aneurysm in an adult: diagnosis on non-invasive imaging. BMJ Case Rep 2018;2018. bcr-2017-223593.
8. Refson JS, Wolfe JH. An aneurysm of the internal jugular vein. Eur J Vasc Endovasc Surg 1996;11:371-2.
9. Gillespie DL, Villavicencio JL, Gallagher C, Chang A, Hamelink JK, Fiala LA, et al. Presentation and management of venous aneurysms. J Vasc Surg 1997;26:845-52.
10. Chakraborty S, Dey PK, Roy A, Bagchi NR, Sarkar D, Pal S, et al. Internal jugular vein phlebectasia presenting with hoarseness of voice. Case Rep Vasc Med 2013;2013:386961.
11. Lubianca-Neto JF, Mauri M, Prati C. Internal jugular phlebectasia in children. Am J Otolaryngol 1999;20:415-8.
12. Nakayama M, Fujita S, Kawamata M, Namiki A, Mayumi T. Traumatic aneurysm of the internal jugular vein causing vagal nerve palsy: a rare complication of percutaneous catheterization. Anesth Analg 1994;78:598-600.

13. Spiro SA, Coccaro SF, Bogucki E. Aneurysm of the internal jugular vein manifesting after prolonged positive pressure ventilation. Head Neck 1991;13:450-2.
14. Figueroa-Sanchez JA, Ferrigno AS, Benvenutti-Regato M, Caro-Osorio E, Martinez HR. Internal jugular phlebectasia: a systematic review. Surg Neurol Int 2019;10:106.
15. LaMonte SJ, Walker EA, Moran WB. Internal jugular phlebectasia. A clinicoroentgenographic diagnosis. Arch Otolaryngol 1976;102:706-8.
16. Paleri V, Gopalakrishnan S. Jugular phlebectasia: theory of pathogenesis and review of literature. Int J Pediatr Otorhinolaryngol 2001;57:155-9.
17. Burnley HM, McCormick D, Hurren J, Gallagher PJ. Primary venous dissecting aneurysm arising during pregnancy: a case report and review of the literature. J Clin Pathol 2003;56:634-5.
18. Haas E, Meyer MR, Schurr U, Bhattacharya I, Minotti R, Nguyen HH, et al. Differential effects of 17β-estradiol on function and expression of estrogen receptor α, estrogen receptor β, and GPR30 in arteries and veins of patients with atherosclerosis. Hypertension 2007;49:1358-63.
19. Hodges YK, Tung L, Yan XD, Graham JD, Horwitz KB, Horwitz LD. Estrogen receptors α and β: prevalence of estrogen receptor β mRNA in human vascular smooth muscle and transcriptional effects. Circulation 2000;101:1792-8.
20. Pang Y, Dong J, Thomas P. Progesterone increases nitric oxide synthesis in human vascular endothelial cells through activation of membrane progesterone receptor-α. Am J Physiol Endocrinol Metab 2015;308:E899-911.
21. Sevuka U, Ayaza F, Demirdas E, Erkul A. An extremely rare source of pulmonary embolism: internal jugular vein aneurysm. J Med Cases 2016;7:195-6.
22. Hopsu E, Tarkkanen J, Vento SI, Pitkäranta A. Acquired jugular vein aneurysm. Int J Otolaryngol 2009;2009:535617.
23. Zhang C, Li H, Guo X, Civilini E, Malik K, Wang F. Clinical diagnosis and treatment of internal jugular venous aneurysms. Ann Vasc Surg 2020;62:497.e7-12.
24. Chua W, Xu G, Cheng SC. Hybrid surgical management of a saccular aneurysm of the internal jugular vein. Singapore Med J 2012;53:e90-1.
25. Zhang HY, Chai DZ, Fu WG, Wang YQ, Yang J, Shi Y, et al. Endovascular stent-protected resection of a large jugular vein aneurysm containing thrombus. Eur J Vasc Endovasc Surg 2012;44:601.

Submitted Nov 12, 2019; accepted Apr 15, 2020.