ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Neuro-Oncology,

Editor
Mitsutoshi Nakada, MD
Kanazawa University, Ishikawa, Japan

SNI
Open Access

Case Report

Successful treatment of superior sagittal sinus thrombosis after translabyrinthine resection of metastatic neuroendocrine tumor: A case report and review of literature

Andrew K. Wong[1], Ricky H. Wong[2]

[1]Department of Neurosurgery, Rush University Medical Center, Chicago, [2]Department of Neurosurgery, NorthShore University HealthSystem, Evanston, Illinois, United States.

E-mail: *Andrew K. Wong - andrew.wong8@gmail.com; Ricky H. Wong - wong.ricky@gmail.com

***Corresponding author:**
Ricky H. Wong,
Department of Neurosurgery,
NorthShore University
HealthSystem, Evanston,
Illinois, United States.

wong.ricky@gmail.com

Received : 18 September 2020
Accepted : 07 November 2020
Published : 25 November 2020

DOI
10.25259/SNI_656_2020

Quick Response Code:

ABSTRACT

Background: Postoperative cerebral venous sinus thrombosis (pCVST) after resection of cerebellopontine angle and posterior fossa tumor resections occur almost exclusively in the lateral venous sinuses and are generally asymptomatic. Thrombus extension and involvement of the superior sagittal sinus (SSS) – a serious and potentially devastating complication – are rarely described and, as such, successful treatment for which is still poorly understood. We report a case of pCVST involving the SSS after translabyrinthine approach for resection of a metastatic neuroendocrine tumor (NET), and the first that was successfully treated with anticoagulation therapy.

Case Description: A 40-year-old man presented with headaches, diminished right-sided hearing, and ataxia was found to have a large right-sided cerebellopontine angle (CPA) lesion with extra-axial and possible intraparenchymal invasion. A retrosigmoid craniotomy for debulking and diagnosis was undertaken. Postoperative imaging revealed patent venous sinuses. Pathology confirmed NET. Further imaging revealed a likely pancreatic primary lesion. The patient then underwent subsequent translabyrinthine approach for definitive surgical resection. Postoperative imaging again revealed patent venous sinuses. The patient subsequently developed headaches on postoperative day 10 and was found to have pCVST involving the ipsilateral internal jugular to the SSS. The patient was started on therapeutic heparin with significant improvement in pCVST and symptoms.

Conclusion: Extensive pCVST involving the SSS after CPA and posterior fossa tumor resections is extremely rare. Initial management with anticoagulation can yield promising results and should be initiated early in the clinical course unless otherwise contraindicated.

Keywords: Anticoagulation, Neuroendocrine tumor, Sinus thrombosis, Superior sagittal sinus thrombosis, Translabyrinthine

INTRODUCTION

Cerebral venous sinus thrombosis (CVST) is a known complication after resection of cerebellopontine angle (CPA) and posterior fossa tumors with a reported incidence of 5.2–

38.9%.[1,7] Postoperative CVST (pCVST) is overwhelmingly asymptomatic and treatment is controversial and largely based on evidence available for the management of spontaneous CVST (sCVST).[3,4,7] When symptomatic, pCVST can lead to significant morbidity and mortality secondary to venous congestion including hemorrhagic venous infarction, cerebral edema, and herniation syndromes.[15] This is particularly true when the superior sagittal sinus (SSS) is involved.[9-11,16] While pCVST typically involves the lateral sinuses, reports of SSS involvement are exceedingly rare.[11,16] As such, management of these complications can be particularly challenging. Here, we report the only case, to the best of our knowledge, of a SSS pCVST successfully treated with anticoagulation therapy detailing the presentation, imaging findings, and management to shed further light on its treatment.

CASE DESCRIPTION

A 40-year-old male with a body mass index (BMI) of 37 kg/m^2 but otherwise had no significant medical history presented with symptoms of headaches, diminished hearing on the right, and ataxia. Computed tomography (CT) and subsequent contrasted magnetic resonance imaging (MRI) demonstrated a large right-sided CPA lesion with a circumscribed, extra-axial, contrast-enhancing portion and a diffuse, infiltrating, and sparsely contrast-enhancing portion within the adjacent cerebellum suspicious for intraparenchymal invasion. Patent bilateral codominant lateral venous sinuses were noted on preoperative imaging [Figure 1].

Given the unclear extent of local invasion, the patient underwent a keyhole retrosigmoid craniotomy for diagnostic tumor debulking and subtotal resection. Intraoperative frozen pathology returned as meningioma while final pathological specimen revealed a well-differentiated neuroendocrine tumor (NET). Postoperative contrasted MRI demonstrated patent venous sinuses. CT of the chest, abdomen, and pelvis was inconclusive for a possible primary source. Subsequent Dotatate positron emission tomography scan utilizing gallium-68 revealed increased uptake in the tail of the pancreas.

The posterior fossa tumor was initially followed with repeat MRI at 3-month intervals. At 6 months, some growth was noted. After a multidisciplinary discussion, the patient underwent a translabyrinthine approach for definitive surgical resection. While the patient had serviceable hearing, given the tumor extent and risk of hearing loss even with a retrolabyrinthine approach, a translabyrinthine approach was chosen for increased surgical freedom and improved resection. Intraoperative findings were significant for a well-circumscribed extra-axial component that was resected as well as an intraparenchymal portion largely homogenous with surrounding normal brain tissue not amenable for

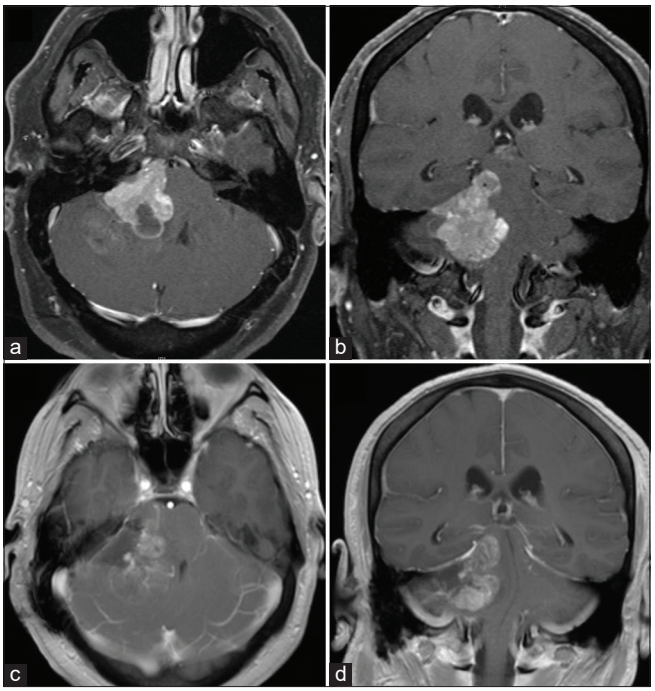

Figure 1: (a and b) Axial and coronal T1 postcontrast preoperative magnetic resonance imaging (MRI) before retrosigmoid craniotomy. (c and d) Axial and coronal T1 postcontrast postoperative MRI after retrosigmoid craniotomy.

resection. Total operative time was 481 min. No sinus injury was encountered and no fixed retractors were used. Postoperative contrasted MRI again revealed patent venous sinuses [Figure 2]. Prophylactic subcutaneous heparin (5000 U every 8 h) was started and the patient was mobilized on postoperative day 1. He was continued on prophylactic heparin until discharge on postoperative day 5. The patient did well after surgery with mild gait instability and expected ipsilateral hearing loss.

The patient was discharged to an inpatient rehabilitation unit where on postoperative day 10, he developed significant headaches. CT demonstrated hyperdense signal throughout the venous sinuses. CT venogram confirmed the presence of thrombosis within the ipsilateral internal jugular (IJ), sigmoid sinus (SS), transverse sinus (TS), SSS, and superficial cortical veins but without associated hemorrhage [Figure 3]. Hypercoagulable workup including prothrombin time, partial thromboplastin time (PTT), international normalized ratio, platelet function, protein C and S levels, and factor V Leiden mutation was negative.

Given the relative proximity to surgery, the patient was started on therapeutic anticoagulation in the form of intravenous heparin with a PTT goal of 50–70 s to preserve reversibility should hemorrhage occur. The patient's symptoms resolved after treatment was initiated and was ultimately transitioned to apixaban before discharge, which

he remained on for 6 months. The patient received adjuvant external beam radiation therapy and started on lanreotide for the treatment of his NET. Magnetic resonance venogram at 6 months demonstrated complete recanalization of the SSS with partial recanalization of the IJ, SS, and TS. The patient remained asymptomatic at 6 months follow-up [Figure 4].

DISCUSSION

NETs are a rare and heterogeneous group of slow-growing tumors derived from enterochromaffin cells that are widely distributed throughout the body. While NETs are known to have malignant potential, they infrequently metastasize to the brain – doing so in only 1.5–5% of cases – and with only two reported cases of neurological symptoms being the first manifestations of metastatic NET disease. Median survival after such diagnosis is 7–10 months.[8,14] In the largest series of NET brain metastasis, Hlatky *et al.* reported better overall survival in patients who underwent surgical resection and adjuvant whole brain radiation therapy (WBRT) than those who underwent surgery or WBRT alone. NETs represent 1.3–1.4% of metastatic brain lesions while malignancy overall has been associated with a hypercoagulable state, an association with metastatic intracranial NET lesions and CVST has not been delineated or described.[8]

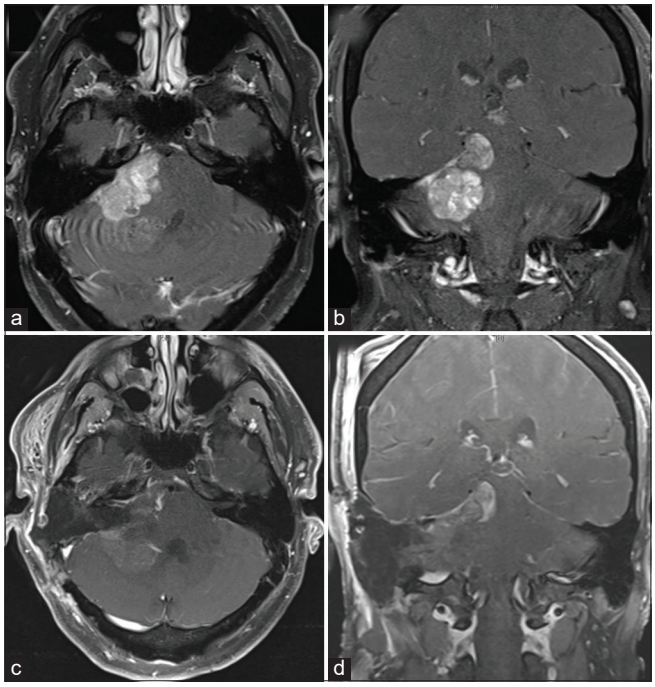

Figure 2: (a and b) Axial and coronal T1 postcontrast preoperative magnetic resonance imaging (MRI) before translabyrinthine craniotomy. (c and d) Axial and coronal T1 postcontrast postoperative MRI after translabyrinthine craniotomy.

CVST in the lateral venous sinuses is a known complication after resection of CPA and posterior fossa tumors. Surgical risk factors for pCVST, particularly in the translabyrinthine approach, include extensive exposure of the sinuses, desiccation from prolong exposure under the operative microscope lamp, direct and thermal injury during bony drilling, mechanical fixed or dynamic retraction, and migration of bone wax used on emissary veins.[3,13] Early retrospective studies reported pCVST incidence between 5.2% and 11.6% with more recent prospective studies revealing much higher rates between 32.4% and 38.9%.[1,4,7,12] This discrepancy is, in part, a result of the overwhelmingly asymptomatic nature of pCVST where capturing the diagnosis is largely dependent on the standardization of postoperative imaging.

Evidence-based treatment paradigms for pCVST remain lacking, controversial, and largely based on evidence available for the treatment of sCVST. Some advocate diagnostic vigilance and therapeutic anticoagulation or antiplatelet therapy regardless of clinical symptoms as supported by trials involving sCVST.[5,6,12] However, others choose to treat only if it is symptomatic noting that therapeutic anticoagulation in the immediate postoperative period is not without risk of significant intracranial hemorrhage and that available evidence point toward pCVST being a largely benign and

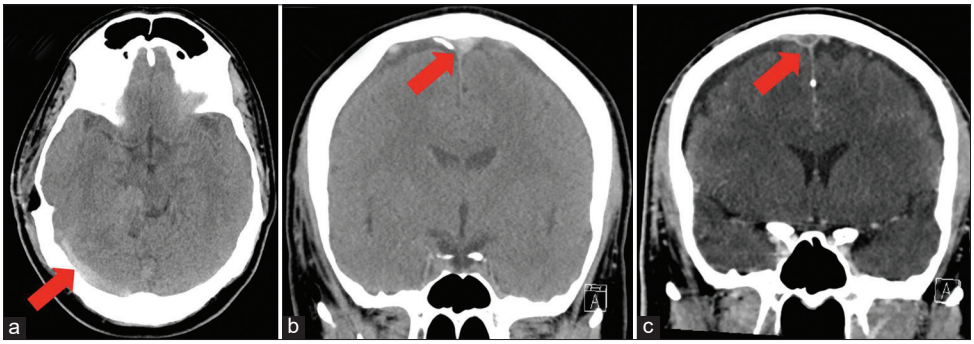

Figure 3: (a) Axial noncontrast computed tomography (CT) demonstrating a hyperdense right transverse sinus (arrow). (b) Coronal noncontrast CT demonstrating a hyperdense superior sagittal sinus (SSS) (arrow). (c) Coronal CT venogram demonstrating thrombosis of the SSS (arrow).

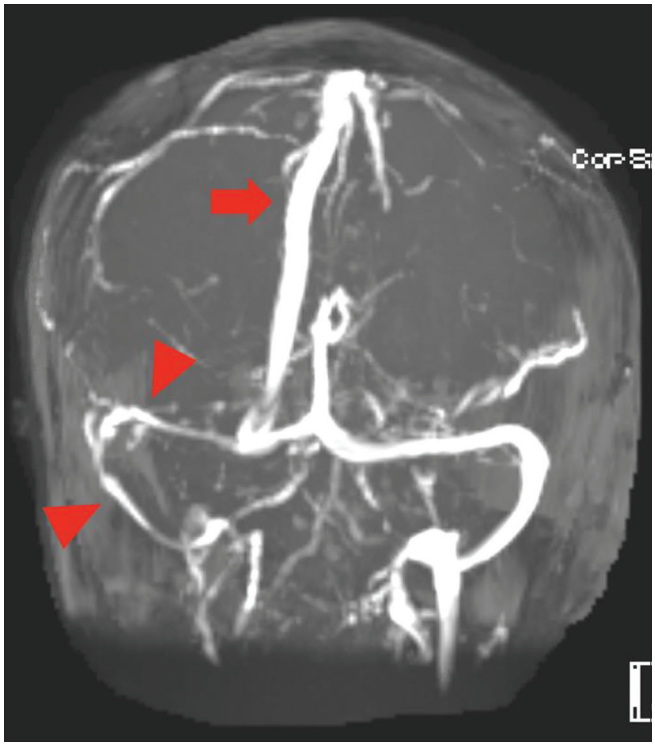

Figure 4: Magnetic resonance venogram demonstrating recanalization of the superior sagittal sinus (arrow) and partial recanalization of the right transverse and sigmoid sinuses (arrowheads).

asymptomatic process with up to 61% rate of recanalization at 1 year follow-up without any intervention.[2,3,7] Indeed, in two large prospective studies, Benjamin *et al*. and Guazzo *et al*. reported 100% asymptomatic rates of pCVST in 24 and 21 patients, respectively, though both studies did note increased rates of cerebrospinal fluid leak in such patients.[4,7] Quantifying the risks as well as the potential benefits are paramount in developing definitive management guidelines for pCVST overall.

As opposed to the lateral sinuses, pCVST involving the SSS after CPA or posterior fossa tumor resection is exceedingly rare and almost always symptomatic with only two reported cases in the literature. Manzoor *et al.* reported a case of pCVST involving the ipsilateral IJ, SS, TS, and SSS after translabyrinthine resection of an acoustic schwannoma.[11] The patient presented with headaches, diplopia, tinnitus, and emesis and, after failing initial anticoagulation therapy, the patient was successfully treated with endovascular mechanical thrombectomy. In a separate report, Sawarkar *et al*. described a patient who presented with isolated SSS thrombosis and hemorrhagic venous infarction after a retrosigmoid resection of a large acoustic schwannoma.[16] They elected to withhold anticoagulation therapy due to the hemorrhage. The patient underwent a decompressive hemicraniectomy but continued to deteriorate and ultimately died.

Our patient represents the third reported case of pCVST involving the SSS after resection of CPA or posterior fossa lesion and the first that was successfully treated with anticoagulation therapy. While the patient had an elevated BMI, he otherwise had no identifiable underlying predisposition for hypercoagulability and was not a smoker. It remains unclear if the nature of the skull base metastatic NET played a role in the extensive thrombosis given the unique presentation of both the tumor and thrombosis as well as the paucity of reported cases. Of note, the patient developed symptoms 10 days after surgery while this was outside the immediate postoperative period, making the decision to initiate anticoagulation therapy simpler, it highlights the importance of maintaining a high level of suspicion for CVST in the extended postoperative period as well. Nevertheless, this case provides evidence that anticoagulation therapy can continue to play a central role in the frontline management of extensive pCVST.

CONCLUSION

Although CVST is a known and likely frequent occurrence after resection of CPA and posterior fossa tumors, especially in approaches that require extensive exposure of the venous sinuses, involvement of the SSS is exceedingly rare. Strong evidence of best treatment options in pCVST involving the SSS is lacking and likely unattainable given its extremely low incidence. This report further highlights the importance of maintaining a high level of suspicion for pCVST after such approaches and demonstrates the feasibility in treating extensive thrombosis with therapeutic anticoagulation.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Abou-Al-Shaar H, Azab MA, Karsy M, Guan J, Alzhrani G, Gozal YM, *et al*. Assessment of costs in open surgery and stereotactic radiosurgery for vestibular schwannomas. J Neurosurg 2018;131:561-8.
2. Abou-Al-Shaar H, Gozal YM, Alzhrani G, Karsy M, Shelton C, Couldwell WT. Cerebral venous sinus thrombosis after vestibular schwannoma surgery: A call for evidence-based management guidelines. Neurosurg Focus 2018;45:E4.

3. Apra C, Kotbi O, Turc G, Corns R, Pagès M, Souillard-Scémama R, *et al*. Presentation and management of lateral sinus thrombosis following posterior fossa surgery. J Neurosurg 2017;126:8-16.
4. Benjamin CG, Sen RD, Golfinos JG, Sen C, Roland JT, McMenomey S, et al. Postoperative cerebral venous sinus thrombosis in the setting of surgery adjacent to the major dural venous sinuses. J Neurosurg 2019;2019:1-7.
5. Capecchi M, Abbattista M, Martinelli I. Cerebral venous sinus thrombosis. J Thromb Haemost 2018;16:1918-31.
6. Coutinho J, de Bruijn SF, Deveber G, Stam J. Anticoagulation for cerebral venous sinus thrombosis. Cochrane Database Syst Rev 2011;2011:CD002005.
7. Guazzo E, Panizza B, Lomas A, Wood M, Amato D, Alalade A, *et al*. Cerebral venous sinus thrombosis after translabyrinthine vestibular schwannoma-a prospective study and suggested management paradigm. Otol Neurotol 2020;41:e273-9.
8. Hlatky R, Suki D, Sawaya R. Carcinoid metastasis to the brain. Cancer 2004;101:2605-13.
9. Keiper GL Jr., Sherman JD, Tomsick TA, Tew JM Jr. Dural sinus thrombosis and pseudotumor cerebri: Unexpected complications of suboccipital craniotomy and translabyrinthine craniectomy. J Neurosurg 1999;91:192-7.
10. Kowoll CM, Kaminski J, Weiß V, Bösel J, Dietrich W, Jüttler E, *et al*. Severe cerebral venous and sinus thrombosis: Clinical course, imaging correlates, and prognosis. Neurocrit Care 2016;25:392-9.
11. Manzoor NF, Ray A, Singer J, Nord R, Sunshine J, Megerian CA, *et al*. Successful endovascular management of venous sinus thrombosis complicating trans-labyrinthine removal of vestibular schwanomma. Am J Otolaryngol 2016;37:379-82.
12. Moore J, Thomas P, Cousins V, Rosenfeld JV. Diagnosis and management of dural sinus thrombosis following resection of cerebellopontine angle tumors. J Neurol Surg B Skull Base 2014;75:402-8.
13. Paulo DL, Lopez AM, Jermakowicz WJ, Yu H, Shah H, Konrad PE, *et al*. Microvascular decompression for trigeminal neuralgia in patients with multiple sclerosis: Predictors of treatment success. World Neurosurg 2020;136:e165-70.
14. Pavel M, Grossman A, Arnold R, Perren A, Kaltsas G, Steinmüller T, *et al*. ENETS consensus guidelines for the management of brain, cardiac and ovarian metastases from neuroendocrine tumors. Neuroendocrinology 2010;91:326-32.
15. Puppa AD, Tosatto L, Amistà P, Munari M, Scienza R. Transverse sinus thrombosis after posterior fossa surgery for cerebellar tumor treated by endovascular thrombectomy. A case report. Neuroradiol J 2007;20:562-5.
16. Sawarkar DP, Verma SK, Singh PK, Doddamani R, Kumar A, Sharma BS. Fatal superior sagittal sinus and torcular thrombosis after vestibular schwannoma surgery: Report of a rare complication and review of the literature. World Neurosurg 2016;96:607.e619-24.

How to cite this article: Wong AK, Wong RH. Successful treatment of superior sagittal sinus thrombosis after translabyrinthine resection of metastatic neuroendocrine tumor: A case report and review of literature. Surg Neurol Int 2020;11:410.