Sinonasal Glomangiopericytoma with Prolonged Postsurgical Follow-Up

Alex J. Gordon¹ Michael R. Papazian¹ Michael Chow² Aneek Patel¹ Dimitris G. Placantonakis³ Seth Lieberman² Babak Givi²

¹ Grossman School of Medicine, New York University (NYU) Langone Health, New York, New York, United States
² Department of Otolaryngology-Head and Neck Surgery, New York University (NYU) Langone Health, New York, New York, United States
³ Department of Neurosurgery, New York University (NYU) Langone Health, New York, New York, United States

J Neurol Surg Rep 2022;83:e87–e89.

Address for correspondence Alex J. Gordon, BS, New York University Grossman School of Medicine, 550 1st Avenue, New York, NY 10016, United States (e-mail: Alex.Gordon@nyulangone.org).

Abstract
Sinonasal glomangiopericytoma is a rare vascular tumor of the respiratory epithelium. Treatment consists mainly of surgical resection, though there is no consensus regarding the use of adjuvant therapies or preoperative endovascular embolization. The postsurgical prognosis is favorable, though there is a high risk of delayed recurrence. Here, we present the case of a patient who underwent endoscopic resection of a sinonasal glomangiopericytoma and a review of the literature.

Keywords
► sinonasal glomangiopericytoma
► endoscopic surgery
► surveillance
► literature review

Introduction
Sinonasal glomangiopericytoma (SNGPC) is a rare, indolent vascular tumor of the respiratory epithelium, accounting for 0.5 to 1.0% of all sinonasal tumors.¹ Standard therapy consists of total surgical resection. Limited evidence supports the use of adjuvant therapies.²,³ Though its prognosis is favorable, this tumor has a propensity for delayed recurrence.⁴

Case Report
A 73-year-old male with known pituitary microadenoma presented with an incidental, asymptomatic right sinonasal mass on imaging. Computed tomography and magnetic resonance imaging (MRI) showed opacification of right ethmoid bulla extending to the superomedial nasal cavity, with extension through the cribiform plate and fovea ethmoidalis (►Fig. 1). Biopsy revealed SNGPC. After workup the tumor was staged as T3N0M0. The patient underwent uncomplicated endoscopic transnasal resection, with planned dural resection by the neurosurgery team. The tumor was found medial to the right middle turbinate, transgressing the cribiform plate and dura mater. Negative margins were confirmed on frozen section, and MRI confirmed absence of residual disease (►Fig. 2). The defect was repaired with acellular dermal matrix and a nasoseptal flap.⁵ The patient has returned for routine follow-up for 6 years, with no evidence of locoregional or distant recurrence.

Literature Review
Previously known as a sinonasal-type hemangiopericytoma, SNGPC was reclassified by the World Health Organization in 2005 due to its pathologic resemblance to glomus tumors. It is distinguished from soft tissue hemangiopericytomas or...
solitary fibrous tumors by positive immunostaining for smooth muscle actin.\textsuperscript{5,7}

SNGPC presents most commonly during the sixth and seventh decades of life, but can present throughout the lifespan.\textsuperscript{8} The etiology has not been fully elucidated; however, trauma, hypertension, and corticosteroid use are predisposing factors. SNGPC most commonly presents with epistaxis, nasal obstruction, and headache, though advanced disease can cause facial pain, bulging, and proptosis.\textsuperscript{9–11} Cases of misdiagnosis as benign nasal polyps have been previously reported.\textsuperscript{12}

The management of SNGPC consists mainly of total surgical resection. This is typically performed endoscopically,\textsuperscript{13} but open approaches via medial maxillectomy have also been reported in advanced cases.\textsuperscript{14} Chemotherapy and radiotherapy are less effective and are reserved for recurrent

---

**Fig. 1** Preoperative magnetic resonance imaging (MRI) (A) and computed tomography (CT) (B) images of sinonasal glomangiopericytoma (star) showing opacification of right ethmoid bulla and extension through cribriform plate.

**Fig. 2** Postoperative magnetic resonance imaging (MRI) (A, B) images showing absence of residual tumor.
disease or palliative therapy. Among these, Adriamycin-based regimens have shown the most promising results.

Due to the vascularity of this tumor, several cases of preoperative endovascular embolization have been reported. In two cases, embolization was achieved with transnasal injection of liquid polymeric agents. This is not currently standard-of-care, and strict criteria for embolization have not been established.

The prognosis of SNGPC is favorable, with 5-year overall and disease-free survival rates of 88.1 and 74.2%, respectively. Though SNGPC most frequently recurs within 1 year of surgery, delayed recurrence is common. One systematic review of 337 cases found that up to 40% of recurrences occur more than 5 years after surgery; thus, regular follow-up is essential in patients with SNGPC.

**Conclusion**

SNGPC has a favorable prognosis after primary surgical resection, but delayed recurrence is common. The current evidence suggests long-term follow-up would benefit most patients, but this paradigm has not been tested prospectively.

**Conflict of Interest**

None declared.

**References**

1. Anzai T, Saito T, Tsuyama S, Toh M, Ikeda K, Ito S. A Case of glomangiopericytoma at the nasal septum. Head Neck Pathol 2018;12(04):572–575
2. Carew JF, Singh B, Kraus DH. Hemangiopericytoma of the head and neck. Laryngoscope 1999;109(09):1409–1411
3. Wong PP, Yagoda A. Chemotherapy of malignant hemangiopericytoma. Cancer 1978;41(04):1256–1260
4. Park ES, Kim J, Jun SY. Characteristics and prognosis of glomangiopericytomas: a systematic review. Head Neck 2017;39(09):1897–1909
5. Hadad G, Bassagasteguy L, Carrau RL, et al. A novel reconstructive technique after endoscopic expanded endonasal approaches: vascular pedicle nasoseptal flap. Laryngoscope 2006;116(10):1882–1886
6. Lasota J, Felisiak-Golabek A, Aly FZ, Wang ZF, Thompson LD, Miettinen M. Nuclear expression and gain-of-function β-catenin mutation in glomangiopericytoma (sinonasal-type hemangiopericytoma): insight into pathogenesis and a diagnostic marker. Mod Pathol 2015;28(05):715–720
7. Kono M, Bandoh N, Matsuoka R, et al. Glomangiopericytoma of the nasal cavity with CTNNB1 p.S37C mutation: a case report and literature review. Head Neck Pathol 2019;13(03):298–303
8. Saito Y, Ohta N, Konosu-Fukaya S, et al. Endoscopic treatment of sinonasal glomangiopericytoma: a case report in light of the literature. Yonago Acta Med 2019;62(02):236–239
9. Asimakopoulos P, Syed MI, Andrews T, Syed S, Williams A. Sinonasal glomangiopericytoma: is anything new? Ear Nose Throat J 2016;95(02):E1–E5
10. Roy NP, Desai DP, Jain SA. Glomangiopericytoma of nasal cavity. Indian J Pathol Microbiol 2015;58(04):554–556
11. Ghallou SK, Dhanani R, Pasha HA, Waisf M, Fatima S, Ikram M. Glomangiopericytoma: a rare tumour of sinonasal cavity. J Pak Med Assoc 2020;70(12(8)):2469–2471
12. Sharma N, Mandlik D, Patel P, et al. A rare case of sinonasal glomangiopericytoma post operative accidental diagnosis and management-a case report. Int J Surg Case Rep 2019;62:54–57
13. Tsesma B, Eloy JA, Folbe AJ, et al. Endoscopic management of sinonasal hemangiopericytoma. Otolaryngol Head Neck Surg 2012;146(03):483–486
14. Dandekar M, McHugh JB. Sinonasal glomangiopericytoma: case report with emphasis on the differential diagnosis. Arch Pathol Lab Med 2010;134(10):1444–1449
15. Oliveira VM, Neto Almeida G, Silva DR, Escada PA. Endoscopic resection of invasive glomangiopericytoma following preoperative embolisation. BMJ Case Rep 2016;2016:bcr2015213423
16. Psoma E, Karkos PD, Dova S, et al. Sinonasal glomangiopericytoma treated with preoperative embolisation and endoscopic sinus surgery. Ecanernetmedicalscience 2016;10:692
17. Ciceri EF, Plebani M, Augelli R, et al. Transnasal devascularisation of a sinonasal hypervascular tumour (glomangiopericytoma) with direct injection of liquid polymer agent (Squid®). Interv Neuroradiol 2019;25(02):230–233
18. Ledderose GJ, Gellrich D, Holtmannspötter M, Leunig A. Endoscopic resection of sinonasal hemangiopericytoma following preoperative embolisation: a case report and literature review. Case Rep Otolaryngol 2013;2013:796713–796713
19. Thompson LD, Miettinen M, Wenig BM. Sinonasal-type hemangiopericytoma: a clinicopathologic and immunophenotypic analysis of 104 cases showing perivascular myoid differentiation. Am J Surg Pathol 2003;27(06):737–749