Sinonasal Glomangiopericytoma with Prolonged Postsurgical Follow-Up

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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 cribriform 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 cribriform 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

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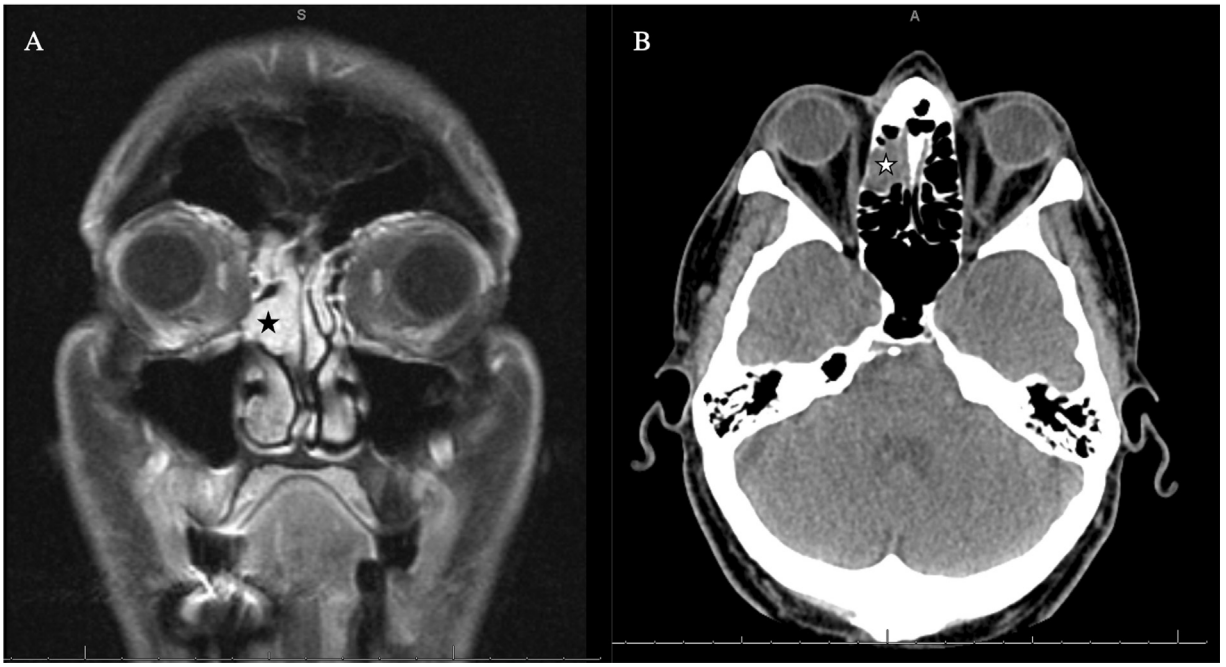

Fig. 1 Preoperative magnetic resonance imaging (MRI) (**A**) and computed tomography (CT) (**B**) images of sinonasal glomangiopericytoma (star) showing opacification of right ethmoid bulla and extension through cribriform plate.

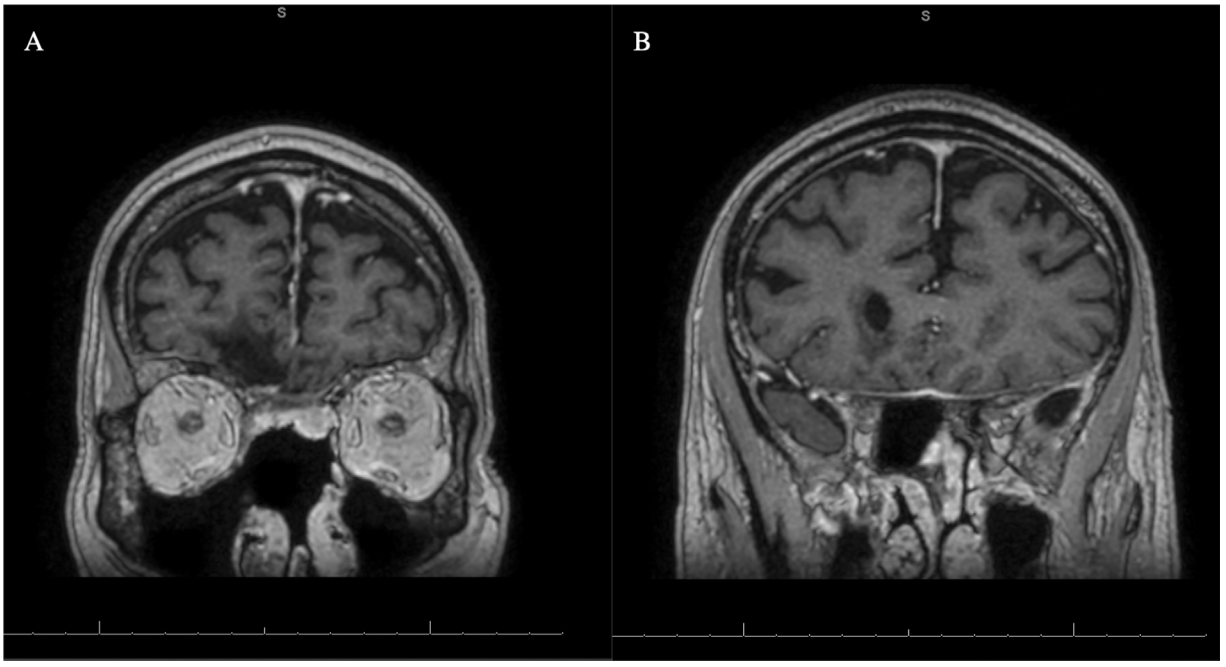

Fig. 2 Postoperative magnetic resonance imaging (MRI) (**A**, **B**) images showing absence of residual tumor.

solitary fibrous tumors by positive immunostaining for smooth muscle actin.[6,7]

SNGPC presents most commonly during the sixth and seventh decades of life, but can present throughout the lifespan.[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.[9–11] Cases of misdiagnosis as benign nasal polyps have been previously reported.[12]

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

disease or palliative therapy.[2] Among these, Adriamycin-based regimens have shown the most promising results.[3]

Due to the vascularity of this tumor, several cases of preoperative endovascular embolization have been reported.[15,16] In two cases, embolization was achieved with transnasal injection of liquid polymeric agents.[17,18] 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.[19] 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[4]; 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.

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