International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

Case Report: A Tumor with a Twist: Navigation the Complexities of Sinonasal Glomangiopericytoma

Dr. Arpitha H. C., Dr. Sudhir M Naik

Department of Otorhinolaryngology, The Oxford Medical College Hospital and Research Centre, Bangalore, India
Email ID: arpitha120397[at]gmail.com

Abstract: ***Background: Sinonasal glomangiopericytoma (SGP) is a rare, low-grade mesenchymal tumor arising from pericytes of Zimmerman, primarily affecting the nasal cavity and paranasal sinuses. It commonly presents with nasal obstruction and recurrent epistaxis, making diagnosis challenging. Due to its histological resemblance to other vascular tumors, immunohistochemistry (IHC) is essential for confirmation. Surgical excision is the mainstay of treatment, with a low risk of metastasis but potential for local recurrence, necessitating long-term follow-up. Case report: A 50-year-old female presented with recurrent right-sided nasal bleeding for one year, worsening over the past three days. Nasal endoscopy revealed a vascular mass, and CT imaging showed a soft tissue lesion extending into the nasopharynx. The patient underwent endoscopic surgical excision. Histopathology confirmed SGP, supported by IHC findings-positive for SMA and β-catenin, negative for CD34, ERG1, CD31, S100, CK, and D240. Postoperative recovery was uneventful, with no recurrence observed. Conclusion: Sinonasal glomangiopericytoma is an uncommon vascular tumor requiring a high index of suspicion. Accurate diagnosis relies on clinical, radiological, and histopathological correlation. Endoscopic surgical resection is the preferred treatment, with careful follow-up to monitor recurrence. Increasing awareness and case reports will aid in optimizing diagnosis and management.***

Keywords: Sinonasal glomangiopericytoma, mesenchymal tumor, nasal cavity, paranasal sinuses, nasal obstruction**, epistaxis,** immunohistochemistry, endoscopic surgical excision, histopathology, smooth muscle actin, β-catenin, local recurrence, vascular tumor, differential diagnosis, follow-up monitoring

1.Introduction

Sinonasal glomangiopericytoma (SGP) is a rare, low-grade mesenchymal tumor arising from pericytes of Zimmerman, primarily affecting the nasal cavity and paranasal sinuses[1]. Clinically, it presents with nasal obstruction and recurrent epistaxis, often mimicking other vascular tumors such as hemangiopericytoma and angiofibroma[2]. Diagnosis relies on histopathology and immunohistochemistry (IHC), with markers like smooth muscle actin (SMA) and β-catenin aiding differentiation [3]. Surgical excision is the mainstay of treatment, with a low metastatic potential but possible local recurrence (Barnes et al., 2020). Here, we present a case of a 50-year-old female with SGP, highlighting its clinical presentation, diagnostic challenges, and management.

History of Present Illness

A 50-year-old female presented to the ENT outpatient department with complaints of recurrent right-sided nasal bleeding for the past year, which had worsened in frequency and intensity over the last three days. She also reported intermittent nasal obstruction but denied any facial pain, headaches, visual disturbances, or anosmia. There was no history of trauma, prior nasal surgeries, systemic bleeding disorders, or recent infections. The patient had no significant past medical history or relevant family history.

Clinical examination revealed a well-circumscribed, vascularized mass in the right nasal cavity. Given the progressive nature of her symptoms, further imaging and histopathological evaluation were performed, leading to the diagnosis of sinonasal glomangiopericytoma.

Investigations

1. Nasal Endoscopy

- Revealed a well-circumscribed, vascularized mass in the right nasal cavity without signs of ulceration or necrosis.
- No evidence of bony erosion or significant extension into adjacent structures.

2. Imaging Studies

- Computed Tomography (CT) Scan: Showed a well-defined, enhancing soft tissue mass in the right nasal cavity extending into the nasopharynx without bony destruction.
- Magnetic Resonance Imaging (MRI) (if performed): Demonstrated a hyperintense lesion on T2-weighted images with strong post-contrast enhancement, suggestive of a vascular tumor.

3. Histopathological Examination

- Biopsy of the mass revealed spindle-shaped tumor cells arranged around thin-walled, branching blood vessels, consistent with glomangiopericytoma.

4. Immunohistochemistry (IHC) Profile

- Positive Markers: Smooth muscle actin (SMA), β-catenin.
- Negative Markers: CD34, ERG1, CD31, S100, Cytokeratin (CK), D2-40.
- Findings confirmed the diagnosis of sinonasal glomangiopericytoma, differentiating it from other vascular tumors like hemangiopericytoma or angiofibroma.

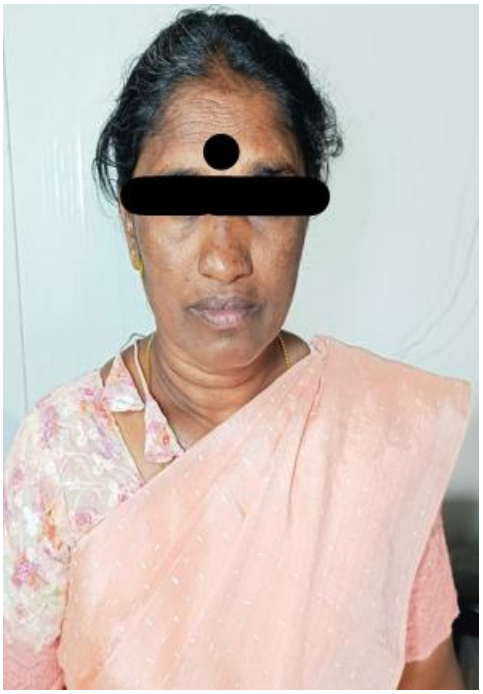

Figure 1: Patient

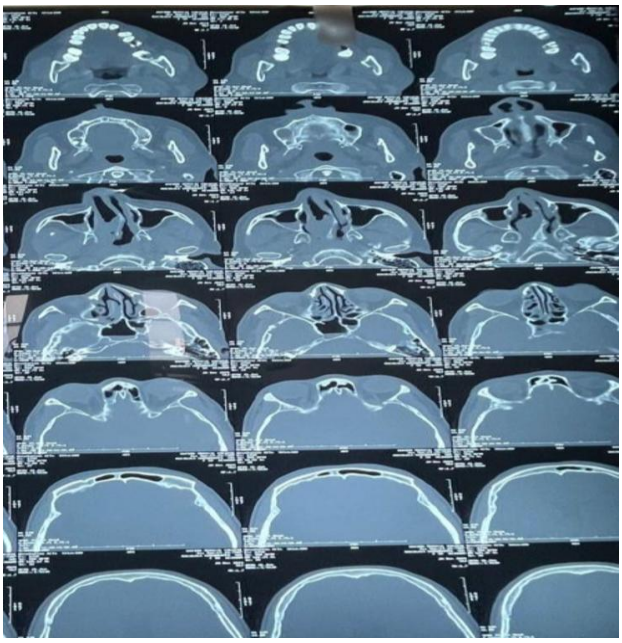

Figure 2: CT PNS

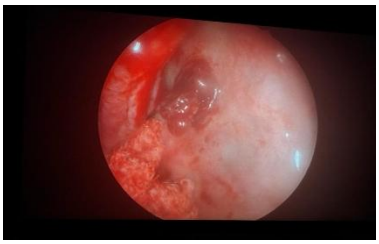

Figure 3 (a)

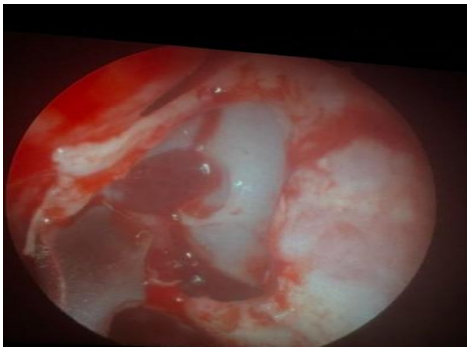

Figure 3 (b)

Figure 3 (a): Showing DNE findings well-circumscribed, vascularized mass in the right nasal cavity, **(b):** showing intraoperative findings

2.Discussion

Sinonasal glomangiopericytoma (SGP) is a rare mesenchymal tumor arising from pericytes of Zimmerman, accounting for less than 0.5% of all sinonasal neoplasms[1]. It primarily manifests with nasal obstruction and recurrent epistaxis, often mimicking other vascular tumors such as angiofibroma, hemangiopericytoma, and hemangioma[2]. While the exact etiology remains unclear, proposed contributing factors include trauma, hormonal influences, and genetic predisposition[4].

Imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) help in assessing the extent of the lesion, but definitive diagnosis requires histopathology and immunohistochemistry (IHC). SGP is characterized by perivascular spindle cells with a branching staghorn vascular pattern. IHC typically demonstrates **positive staining for smooth muscle actin (SMA) and β-catenin**, while markers such as **CD34, ERG1, CD31, S100, CK, and D2-40** are negative, differentiating it from other vascular and spindle-cell tumors[3].

Surgical resection, preferably via an endoscopic approach, is the mainstay of treatment. While SGP has low metastatic potential, local recurrence rates range from **10-20%**, emphasizing the need for complete resection and long-term surveillance[5].

3.Conclusion

Epistaxis, or nasal bleeding, is a common clinical presentation with a wide range of aetiologies, including local, systemic, environmental, and neoplastic causes. In this case, the chronicity and progressive nature of the nasal bleeding, along with associated nasal obstruction and headache, raised suspicion for an underlying mass lesion. The differential diagnoses considered included benign nasal tumors (such as nasal polyps, hemangiomas, and inverted papillomas) and malignant lesions (such as sinonasal carcinomas and lymphomas). Inflammatory conditions such as granulomatous diseases or chronic rhinosinusitis with polypoid changes were also considered.

Diagnostic evaluation included a CT scan of the paranasal sinuses, which revealed a soft tissue mass in the right nasal cavity with bony remodelling, suggestive of a neoplastic

lesion. Subsequent diagnostic nasal endoscopy (DNE) confirmed the presence of a mass lesion, and a biopsy was performed for histopathological examination (HPE). Given the findings, the patient underwent functional endoscopic sinus surgery (FESS) with excision of the right nasal mass under general anaesthesia. Intraoperative findings included a well-encapsulated, friable mass with significant vascularity, requiring meticulous haemostasis and nasal packing.

Postoperative recovery was uneventful, with the patient hemodynamically stable and no signs of recurrent bleeding. He was managed with supportive care, including antibiotics, nasal decongestants, and topical antiseptics. Histopathological evaluation was done and confirmed Sinonasal glomangiopericytoma further oncological evaluation was done.

This case underscores the importance of early evaluation of chronic nasal bleeding, as it can be a harbinger of serious underlying pathology. Nasal endoscopy and imaging play crucial roles in the diagnosis, guiding appropriate surgical or medical interventions. Furthermore, histopathological confirmation remains the gold standard for differentiating benign from malignant lesions. Multidisciplinary collaboration between otolaryngologists, radiologists, and pathologists is essential in optimizing patient outcomes.

Sinonasal glomangiopericytoma is a rare vascular tumour requiring a high degree of clinical suspicion for accurate diagnosis. A **multimodal approach incorporating clinical, radiological, and histopathological assessment** is crucial for differentiating it from other sinonasal neoplasms. Endoscopic surgical excision remains the **preferred treatment**, with postoperative monitoring essential for detecting recurrences. Increased case reporting and research will aid in optimizing diagnostic and therapeutic strategies for this uncommon entity.

References

[1] Thompson LD, Miettinen M, Wenig BM. Sinonasal-type hemangiopericytoma: a clinicopathologic and immunophenotypic analysis of 104 cases. **Mod Pathol.2**003; 16 (6): 505-514.
[2] Fletcher CDM, Bridge JA, Hogendoorn P, Mertens F. **WHO Classification of Tumours of Soft Tissue and Bone.4**th ed. IARC Press; 2013.
[3] Chen Y, Wang J, Bai M, Xu H. Clinicopathological characteristics and treatment outcomes of sinonasal glomangiopericytoma: A case series and literature review. **Int J Clin Exp Pathol.2**019; 12 (2): 584-590.
[4] Barnes L, Eveson JW, Reichart P, Sidransky D. **Pathology and Genetics of Head and Neck Tumours.** World Health Organization Classification of Tumours. IARC Press; 2020.
[5] Gleason BC, Fletcher CD. Glomangiopericytoma: analysis of 50 cases emphasizing histologic predictors of outcome. **Am J Surg Pathol.2**021; 45 (7): 945-953