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Nasal Cavity Glomangiopericytoma: A Diagnostic Challenge and Surgical Management Insights

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Abstract: ***Background: Glomangiopericytoma, a rare sinonasal tumor arising from pericytes, represents less than 0.5% of sinonasal neoplasms. Its nonspecific symptoms, such as epistaxis and nasal obstruction, often lead to misdiagnosis as sinusitis or nasal polyps. Diagnosis relies on imaging (CT/MRI) and histopathological analysis with immunohistochemistry (SMA, beta-catenin positivity). Endoscopic surgical resection is the primary treatment, with recurrence rates of 10–40% necessitating long-term follow-up. Case Report: A 43-year-old woman presented with recurrent nasal bleeding and vomiting, initially managed as sinusitis and inflammatory arthritis. Prior septoplasty provided no relief. CT revealed a right nasal cavity mass with septal erosion, and functional endoscopic sinus surgery (FESS) identified a polypoidal lesion. Histopathology and immunohistochemistry confirmed glomangiopericytoma (SMA, beta-catenin positive). MRI showed a 2.7 cm lesion with ethmoid sinus involvement. Definitive endoscopic resection achieved clear margins, with no postoperative complications. Follow-up endoscopy showed no recurrence. Conclusion: This case highlights the diagnostic challenges of glomangiopericytoma due to its atypical presentation mimicking sinusitis and arthritis. Thorough imaging, histopathological confirmation, and multidisciplinary management are critical for accurate diagnosis. Endoscopic resection proved effective, offering minimal morbidity and a favourable prognosis. Long-term surveillance is essential due to recurrence risks, emphasizing the importance of comprehensive surgical and follow-up strategies in managing this rare sinonasal tumor.***

Keywords: Glomangiopericytoma, Sinonasal Tumor, Endoscopic Surgery, Epistaxis, Diagnostic Challenge, Histopathology

1. Introduction

Glomangiopericytoma is a rare sinonasal tumor, comprising less than 0.5% of sinonasal neoplasms, originating from Zimmermann's pericytes that regulate capillary blood flow. Distinct from hemangiopericytomas, it exhibits borderline or low- grade malignancy with potential for local invasion and recurrence. Predominantly affecting adults, particularly females in their fifth to seventh decades, its etiology remains unclear, with possible hormonal, traumatic, or genetic influences. Symptoms like epistaxis or nasal obstruction often mimic common nasal pathologies, complicating diagnosis. Imaging (CT/MRI) and histopathological analysis with immunohistochemistry (SMA, beta-catenin positivity) are crucial for confirmation. Endoscopic surgical resection is the primary treatment, with recurrence rates of 10–40% necessitating long-term follow-up. This case highlights diagnostic and management challenges.

2. Case Presentation

A 43-year-old woman presented with recurrent bilateral nasal bleeding (10–15 mL) and vomiting, initially misdiagnosed as sinusitis and inflammatory arthritis following a prior septoplasty. CT imaging revealed a right nasal cavity polypoidal mass with septal erosion. Functional endoscopic sinus surgery (FESS) identified a lesion extending into the right middle meatus. Histopathology and immunohistochemistry (SMA, beta-catenin positive) confirmed sinonasal glomangiopericytoma. MRI showed a 2.7 cm heterogeneously enhancing mass with ethmoid sinus involvement. Definitive endoscopic resection achieved clear margins, with adjacent bone drilling ensuring complete removal. Postoperative recovery was uneventful, and follow-up endoscopy revealed no recurrence, highlighting successful surgical management.

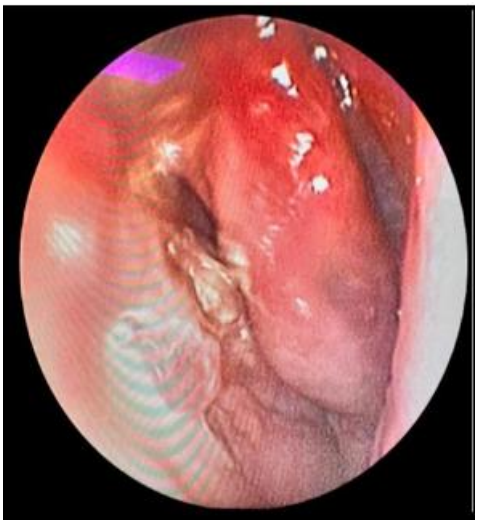

Figure 1: A polypoidal mass in the right nasal cavity with posterior septal erosion

Investigation

Initial CT imaging of the 43-year-old patient revealed right ethmoid sinusitis and left inferior turbinate hypertrophy, prompting a misdiagnosis of sinusitis. A repeat CT after recurrent nasal bleeding showed a polypoidal mass in the right nasal cavity with posterior septal erosion. Functional endoscopic sinus surgery (FESS) facilitated mass debridement, and histopathological examination suggested a low-grade neoplasm, likely glomangiopericytoma. Immunohistochemistry confirmed the diagnosis, showing positivity for SMA and beta-catenin, focal CD34 positivity, and negativity for ERG, CD3, S100, CK, and D2-40. MRI further delineated a 2.7 x 1.5 x 1.3 cm heterogeneously enhancing lesion with ethmoid sinus roof and septal destruction, guiding definitive surgical planning.

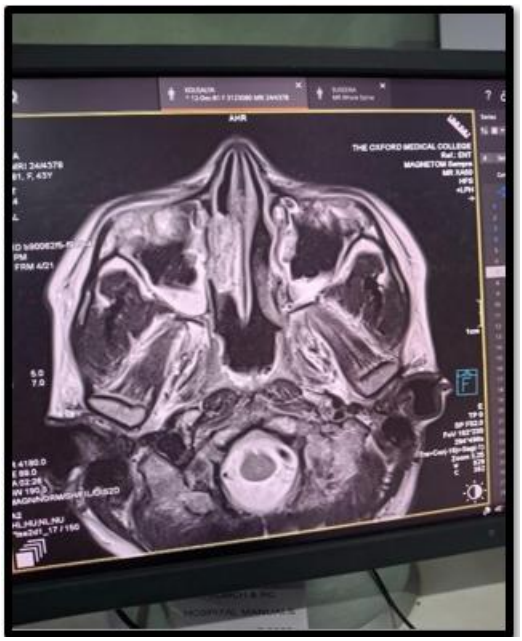

Figure 2 A

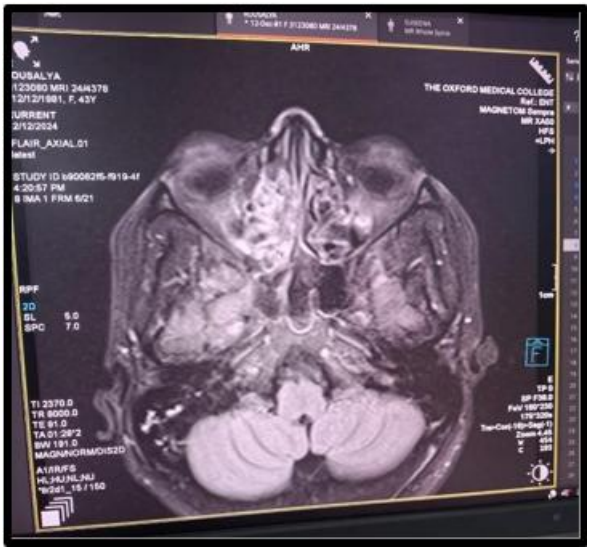

Figure 2 B

Fig 2(A-B): MRI scan showing a well defined heterogenous enhancing lesion on right nasal cavity measuring around 2.7x1.5x1.3 cms. Superiorly its causing destruction of the right ethmoidal sinus roof. No obvious intracranial extension seen.

Management:

The patient underwent definitive endoscopic endonasal resection under general anesthesia for the sinonasal glomangiopericytoma. Given the tumor's vascularity, meticulous surgical planning was employed to minimize intraoperative bleeding. The procedure utilized microdebriders and angled instruments to excise the lesion completely, achieving clear margins. Adjacent bone drilling was performed to ensure thorough removal of the tumor, particularly where it involved the ethmoid sinus roof and superior nasal septum. Hemostasis was secured with nasal packing, and no preoperative embolization was required. Postoperative care included regular endoscopic surveillance to monitor for recurrence, with an uneventful recovery and no further nasal bleeding reported.

Intraoperative Findings

During surgery, a polypoidal, vascular mass was identified in the right nasal cavity, extending into the middle meatus and involving the ethmoid sinus. The lesion caused bony destruction of the ethmoid sinus roof and superior nasal septum, consistent with preoperative MRI findings. The tumor was well-defined but adherent to surrounding structures, requiring careful dissection to avoid damage to adjacent tissues. No intracranial extension was observed, and the use of microdebriders and angled instruments facilitated precise excision with clear margins. The procedure confirmed the tumor's vascular nature, but bleeding was effectively controlled intraoperatively.

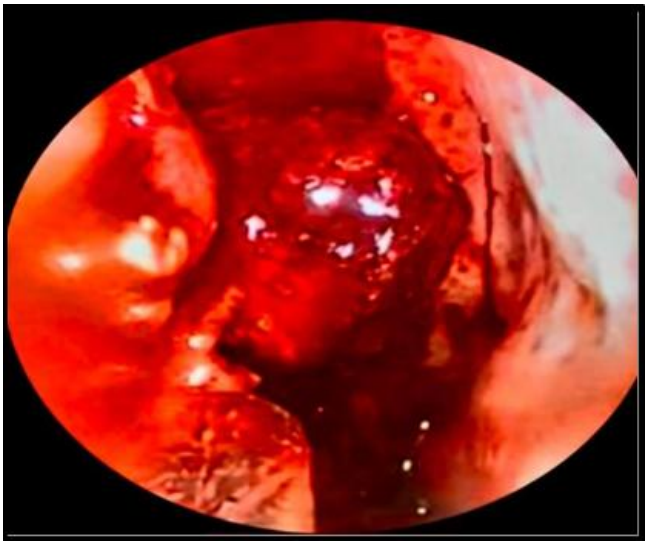

Figure 3: Intraoperative Findings: a polypoidal, vascular mass was identified in the right nasal cavity, extending into the middle meatus and involving the ethmoid sinus

Post-Treatment Endoscopic Findings & Follow-Up

The patient experienced an uneventful recovery following endoscopic endonasal resection of the sinonasal glomangiopericytoma. Nasal packing was removed without complications, and no postoperative nasal bleeding or other symptoms, such as vomiting or joint pain, were reported. Regular endoscopic surveillance conducted during follow-up visits revealed a well-healed surgical site with no evidence of tumor recurrence. The nasal cavity and ethmoid sinus showed no residual mass or abnormal tissue growth. The patient remained asymptomatic, with restored nasal function and no signs of complications, indicating a successful surgical outcome. Long-term follow-up with periodic endoscopy and imaging was recommended to monitor for potential recurrence.

Histopathology;

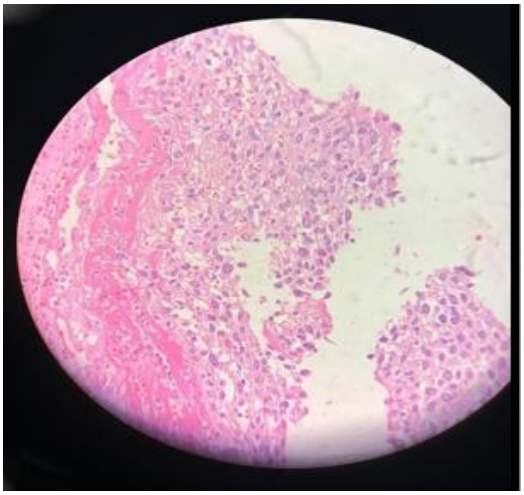

Figure 4 A

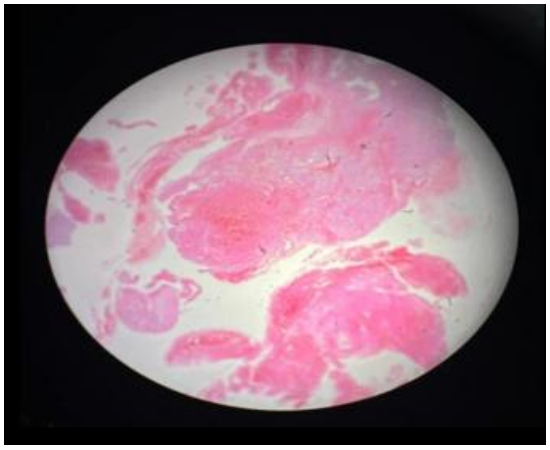

Figure 4 B

Fig 4 (A-B): Histopathology slide showing features favoring a low-grade/borderline soft tissue neoplasm possible Sinonasal glomangiopericytoma

3. Discussion

Sinonasal glomangiopericytoma is a rare and often misdiagnosed tumor due to its variable clinical presentation. The differential diagnosis includes a wide range of benign and malignant nasal tumors, such as nasal polyps, hemangiomas, schwannomas, juvenile nasopharyngeal angiofibroma's, and sinonasal carcinomas. Unlike aggressive sinonasal malignancies, glomangiopericytomas are typically slow-growing and have a low metastatic potential. However, local recurrence can occur, particularly if surgical excision is incomplete. Histopathological examination is crucial for definitive diagnosis, as glomangiopericytomas exhibit a characteristic perivascular growth pattern with spindle-shaped cells. Immunohistochemistry further aids in distinguishing it from other soft tissue tumors, with positivity for SMA and beta-catenin being key diagnostic markers. The absence of markers such as S100, CD3, and ERG helps rule out neural, lymphoid, and endothelial tumors, respectively. Surgical excision remains the gold standard treatment, with endoscopic techniques offering a minimally invasive approach with reduced morbidity and better visualization of tumor margins. In cases with extensive vascularity or skull base involvement, preoperative embolization and adjunctive radiotherapy may be considered. However, the role of adjuvant therapy remains controversial and is usually reserved for cases with incomplete resection or recurrence. Given the risk of local recurrence, long-term follow-up with periodic nasal endoscopy and imaging is essential. Recurrence rates range from 10% to 40%, emphasizing the need for thorough surgical clearance. In this case, despite the initial diagnostic challenge, prompt surgical intervention and regular postoperative surveillance have ensured a favorable outcome with no evidence of recurrence to date.

For tumor resection, an endoscopic approach has some advantages over more traditional open approaches, including clear visualization of each wall of the nasal cavity and minimum invasion of intact tissues reducing postoperative facial deformity or abnormal subsequent deformity of the nose and paranasal sinuses [1]. Even though intranasal endoscopic treatment can be effective in most cases, highly vascularized tumors or large tumors can make it difficult. Preoperative embolization can be considered to reduce the volume of bleeding during surgery, especially for large or highly vascular tumors. Post-operative management should include long-term surveillance with nasal endoscopy at regular intervals considering the local recurrence of the tumor and/or MRI and CT [2].

4. Conclusion

Glomangiopericytoma typically presents as a painless, slow-growing vascularized nasal mass, often leading to nasal obstruction or epistaxis. However, this case was unusual due to the patient's recurrent acute nasal bleeding, vomiting, and symptoms mimicking inflammatory arthritis, leading to an initial misdiagnosis. This case underscores the need for thorough imaging, histopathological confirmation, and a multidisciplinary approach in managing sinonasal tumors to ensure timely and accurate diagnosis. Endoscopic endonasal resection remains a safe and effective treatment modality with a favourable prognosis. Among sinonasal tumors, sinonasal glomangiopericytoma is quite uncommon. Because glomangiopericytomas often behave in a lame manner and seldom require harsh therapy beyond local excision, it is crucial to identify them in the differential diagnosis of tumors of the nasal cavity or paranasal sinuses. Although tissue sampling is still required for the final diagnosis of GPC, CT and MRI can help with differential diagnosis, staging of the disease's severity, and pre-operative planning for anatomic variations of the paranasal sinus that may make surgical resection more difficult. Our case supports the current assessment and management of GPC, which is still underrepresented in the medical literature, as well as the literature evaluation of imaging findings and treatment.[1]

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