Case Report

Sinonasal schwannoma in a patient with Eisenmenger syndrome

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ABSTRACT

Background: Sinonasal schwannoma is a rare tumor of the nasal cavity and paranasal sinuses, often presenting with recurrent epistaxis and nasal obstruction. Eisenmenger syndrome is a life-threatening cardiopulmonary condition resulting from uncorrected congenital heart disease. **Purpose:** To report a rare case of sinonasal schwannoma in a patient with Eisenmenger syndrome, and to explore therapeutic considerations. **Case report:** A 23-year-old male presented with recurrent nasal bleeding and progressive dyspnea. Imaging revealed a lobulated mass in the right nasal cavity. Histopathological evaluation confirmed sinonasal schwannoma. The patient also had an unrepaired secundum atrial septal defect with Eisenmenger physiology, rendering surgical excision high-risk. **Clinical question:** In patients with sinonasal schwannoma and Eisenmenger syndrome, when is surgical excision appropriate? Can conservative therapy provide adequate control in high-risk cases? **Method:** A literature search was conducted on PubMed, Medline, and Google Scholar using relevant keywords. Articles were selected based on the last 10 years of publication and full-text availability. **Result:** No studies specifically addressed this dual pathology. However, available literature emphasized the need for individualized management, with surgery being curative in healthy patients, and conservative observation preferred in high-risk cardiac cases. **Conclusion:** While surgical excision remains the "In patients with sinonasal hemangiopericytoma (HPC), conservative management may be justified in patients with Eisenmenger syndrome due to the prohibitive cardiopulmonary risks.

Keywords: sinonasal schwannoma, Eisenmenger syndrome, pulmonary hypertension, conservative therapy, surgical risk

ABSTRAK

Latar belakang: *Schwannoma sinonasal merupakan tumor langka pada rongga hidung dan sinus paranasal, yang sering kali muncul dengan gejala mimisan berulang dan sumbatan hidung. Sindrom Eisenmenger adalah kondisi kardiopulmoner yang mengancam jiwa akibat penyakit jantung bawaan yang tidak dikoreksi.* ***Tujuan:*** *Melaporkan kasus langka schwannoma sinonasal pada pasien dengan sindrom Eisenmenger, dan mengeksplorasi pertimbangan terapeutik yang relevan.* ***Laporan kasus:*** *Seorang pria berusia 23 tahun datang dengan keluhan mimisan berulang dan sesak napas yang progresif. Pemeriksaan pencitraan menunjukkan massa berbenjol di rongga hidung kanan. Evaluasi histopatologi menegakkan diagnosis schwannoma sinonasal. Pasien juga diketahui memiliki defek septum atrium sekunder yang belum diperbaiki, dengan fisiologi Eisenmenger, sehingga tindakan pembedahan menjadi berisiko tinggi.* ***Pertanyaan klinis:*** *Pada pasien dengan schwannoma sinonasal*

dan sindrom Eisenmenger, kapan eksisi bedah sebaiknya dilakukan? Apakah terapi konservatif dapat memberikan kontrol yang memadai pada kasus dengan risiko tinggi? ***Metode:*** *Pencarian literatur dilakukan melalui basis data* PubMed*,* Medline*, dan* Google Scholar *dengan kata kunci yang relevan. Artikel dipilih berdasarkan publikasi dalam 10 tahun terakhir, dan ketersediaan teks lengkap.* ***Hasil:*** *Tidak ditemukan studi yang secara spesifik membahas patologi ganda ini. Namun, literatur yang ada menekankan pentingnya pendekatan penatalaksanaan individual, yaitu pembedahan bersifat kuratif pada pasien dengan kondisi sehat, sedangkan observasi konservatif lebih disarankan pada pasien dengan risiko kardiovaskular tinggi.* ***Kesimpulan:*** *Meskipun eksisi bedah tetap menjadi terapi definitif untuk schwannoma sinonasal, penatalaksaan konservatif dapat dibenarkan pada pasien dengan sindrom Eisenmenger karena tingginya risiko kardiopulmoner.*

Kata kunci: *schwannoma sinonasal, sindrom Eisenmenger, hipertensi pulmonal, terapi konservatif, risiko pembedahan.*

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INTRODUCTION

Sinonasal schwannoma is a rare benign neoplasm arising from Schwann cells of peripheral nerve sheaths. Although schwannomas are commonly encountered in the vestibulocochlear nerve and spinal roots, their occurrence in the sinonasal tract is exceedingly uncommon, accounting for less than 4% of all head and neck schwannomas, and an even smaller fraction of sinonasal tumors. These lesions typically present as slow-growing, expansile masses that may cause nasal obstruction, epistaxis, or facial pressure depending on their size and location.[1]

Histologically, sinonasal schwannomas are characterized by a biphasic pattern consisting of Antoni A and Antoni B areas. The Antoni A areas show compact spindle-shaped cells with palisading nuclei (Verocay bodies), while the Antoni B regions are more myxoid and loosely arranged. Immunohistochemically, these tumors strongly express S100 protein, reflecting their neural crest origin.[1,2]

Sinonasal schwannomas are typically indolent lesions with a slow, expansile growth pattern. Although they are histologically benign and non-vascular in nature, their progressive enlargement within the confined sinonasal space can lead to compressive symptoms such as nasal obstruction, facial fullness, hyposmia, or headache. In some cases, particularly when the tumor exerts pressure on adjacent mucosa or disrupts fragile mucosal vessels, episodic epistaxis may occur, although it is generally less profuse compared to highly vascular tumors. Local recurrence is uncommon following complete excision, and distant metastasis is virtually nonexistent.[1,3]

Sinonasal schwannomas can occur across a wide age range but are most frequently diagnosed in adults, with no consistent sex predilection reported in the literature. Diagnosis typically involves a combination of nasal endoscopy, cross-sectional imaging such as computed tomography (CT) or magnetic resonance imaging (MRI), and definitive confirmation through histopathological evaluation. Immunohistochemical staining, particularly strong diffuse positivity for S100 protein, is a key feature supporting the diagnosis. Complete surgical excision remains the primary treatment of choice, given the benign nature of the tumor and its low recurrence rate following adequate resection. The role of adjuvant therapy is minimal and generally not indicated in the absence of malignant transformation or

incomplete excision.[1,2,4]

In patients with significant cardiovascular comorbidities such as reversed shunt-associated pulmonary hypertension, surgical decision-making becomes notably more complex. This condition, often resulting from uncorrected congenital heart defects, leads to irreversible pulmonary vascular remodeling and right-to-left intracardiac shunting. Consequently, affected individuals experience chronic hypoxemia, reduced cardiopulmonary reserve, and an elevated risk of perioperative complications such as paradoxical embolism, right heart failure, or sudden circulatory collapse.[5]

To date, there is a paucity of literature addressing the perioperative challenges and therapeutic decision-making in the management of sinonasal tumors such as schwannoma in patients with reversed shunt-associated pulmonary hypertension. Although schwannomas are generally benign and non-vascular, their location within the sinonasal cavity, combined with mass effect and the potential need for general anesthesia, introduces considerable complexity when coexisting with severe cardiopulmonary compromise. The interplay between a space-occupying tumor and an underlying cyanotic congenital heart disease demands careful coordination across disciplines, as the hemodynamic fragility and hypoxemia inherent to such patients pose substantial risks during surgical planning and anesthetic exposure.[6]

This report aimed to present a rare case of sinonasal schwannoma in a young adult patient with reversed shunt-associated pulmonary hypertension, highlighting the clinical presentation, diagnostic workup, and the complexity of therapeutic decision-making in the setting of severe cardiopulmonary compromise due to uncorrected congenital heart disease.

CASE REPORT

A 23-year-old male presented to the Emergency Department with complaints of recurrent nasal bleeding and progressively worsening shortness of breath over the preceding three months. He also reported intermittent palpitations, easy fatigability with minimal exertion, and occasional headaches. Episodes of epistaxis had become more frequent and severe over the past two weeks, prompting further evaluation. The patient denied any history of facial trauma, nasal instrumentation, or recent infections. There were no accompanying systemic symptoms such as fever, weight loss, or night sweats.

The patient had a known history of an uncorrected atrial septal defect (ASD) diagnosed in early childhood but had been lost to follow-up, and had never undergone corrective intervention. According to his family, he began experiencing intermittent cyanosis and progressive exercise intolerance during adolescence, but declined further cardiologic evaluation and treatment. He denied any history of smoking, alcohol consumption, or anticoagulant use.

On physical examination, the patient appeared centrally cyanotic with digital clubbing, and peripheral oxygen saturation measured 66% on room air, improving to 95% with high-flow oxygen (15 L/min) delivered via a non-rebreathing mask (Figure 1). Vital signs were notable for a blood pressure of 110/78 mmHg, heart rate of 108 beats per minute, respiratory rate of 24 breaths per minute, and a body temperature of 36.7°C. Cardiovascular examination revealed a prominent second heart sound (P2), a right ventricular heave, and a grade III/VI systolic murmur best heard at the left upper sternal border. Anterior rhinoscopy showed active bleeding from the right nasal cavity, where a highly vascularized mass was visibly protruding. There was no cervical lymphadenopathy or evidence of facial swelling.

A CTscan of the paranasal sinuses revealed a lobulated soft tissue mass measuring approximately 3.4×3.7 cm, occupying the anterior aspect of the right nasal cavity, causing deviation of the nasal septum to the left, and compression of the right nasal ala. There was no evidence of bony erosion, although mild mucosal thickening was noted in the left maxillary and sphenoidal sinuses. The mass exhibited homogeneous soft tissue density without calcification, extending into the right nasal cavity, and partially into the right maxillary sinus. No signs of adjacent bone destruction or orbital involvement were identified.

The radiologic appearance was suggestive of a benign, expansile lesion, and although initially considered a hypervascular tumor, the absence of overt vascular channels and bone remodeling favored a diagnosis more consistent with a schwannoma rather than a vascular neoplasm (Figure 2).

Figure 1. Clinical manifestations of chronic hypoxemia. Left: perioral cyanosis and nasal crusting in an intubated patient. Right: digital clubbing with bulbous distal phalanges and increased nail curvature, commonly associated with cyanotic congenital heart disease.

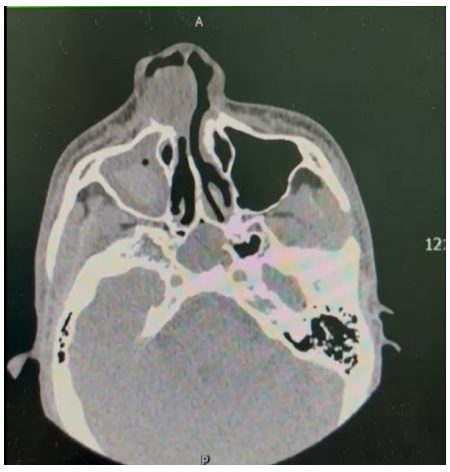

Figure 2. Axial non-contrast CT of the paranasal sinuses showing a well-defined lobulated soft tissue mass in the right nasal cavity with leftward septal displacement, without calcification or bone destruction, suggestive of a benign sinonasal schwannoma.

The preferred operative technique in this case was a midfacial degloving approach, which offered excellent exposure of the sinonasal tract through a combination of intraoral and intranasal incisions—without creating any external facial scars. This method was particularly advantageous for benign but expansile tumors such as schwannoma, especially when located centrally or extending into multiple paranasal compartments. By elevating the midfacial soft tissues en bloc, the surgeon could obtain a wide, panoramic view of the nasal cavity, paranasal sinuses, and anterior skull base, facilitating precise tumor dissection with preservation of surrounding structures.

Although schwannomas are typically non-vascular, their mass effect and proximity to critical anatomical landmarks demand a meticulous and controlled surgical approach. The midfacial degloving technique allows for complete tumor excision with favorable aesthetic outcomes, avoiding the morbidity associated with external incisions used in traditional open approaches such as lateral rhinotomy. In this patient, the tumor was successfully accessed and resected using this method, preserving both function and facial integrity (Figure 3).

Histopathological examination of the nasal biopsy revealed a moderately cellular neoplasm composed of uniform spindle-shaped cells arranged in intersecting fascicles. The tumor exhibited areas with dense cellularity (Antoni A pattern) showing nuclear palisading and Verocay body formation, alongside more loosely arranged hypocellular myxoid regions (Antoni B pattern). There was no evidence of necrosis, nuclear pleomorphism, or increased mitotic activity. Immunohistochemical staining (not shown) would typically demonstrate strong diffuse S100 positivity, supporting the diagnosis of schwannoma, a benign peripheral nerve sheath tumor. In light of the patient's elevated anesthetic risk due to significant cardiopulmonary comorbidity, a more extensive biopsy under general anesthesia was deferred (Figure 4).

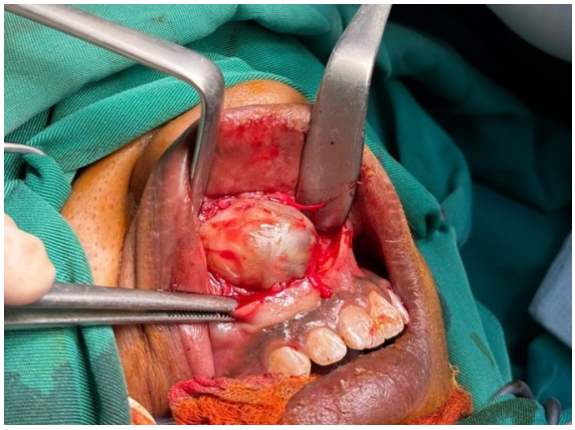

Figure 3. Intraoperative view using the midfacial degloving approach, showing a well-circumscribed vascular mass in the aterior nasal cavity, allowing wide local excision without external facial scarring.

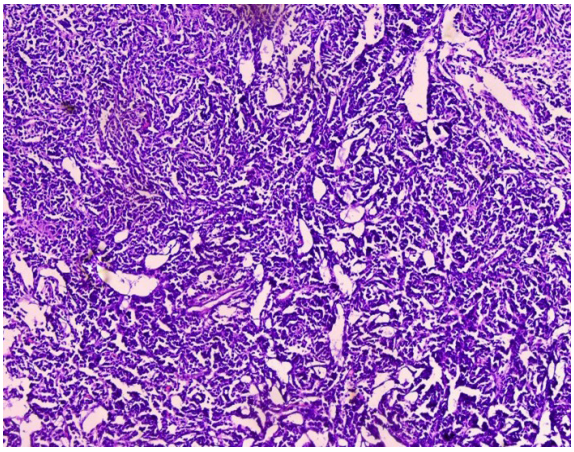

Figure 4. (H&E, 10×): Histopathology of the nasal tumor showing a spindle cell neoplasm with Antoni A and Antoni B areas, nuclear palisading, and Verocay bodies, without necrosis or significant atypia, supporting a benign schwannoma.

A transthoracic echocardiogram confirmed a large secundum ASD measuring 41–60 mm with right-to-left shunting, severe right atrial and right ventricular dilation, interventricular septum flattening (D-shaped LV), and severe pulmonary hypertension with an estimated systolic pulmonary artery pressure (SPAP) of 83 mmHg. Biventricular function was reduced, with a left ventricular ejection fraction of 51%. No thrombus or pericardial effusion was detected.

The patient was diagnosed with a sinonasal schwannoma, in the setting of reversed shunt-associated pulmonary hypertension and decompensated right heart failure (also known as Eisenmenger Syndrome). He was managed conservatively with intravenous furosemide, spironolactone, and adjusted subcutaneous insulin due to steroid-induced hyperglycemia. Tumor biopsy and potential surgical excision were deferred in light of the high anesthetic risk posed by his fragile cardiopulmonary status. He remained hemodynamically stable under close monitoring, with plans for reevaluation of surgical feasibility following optimization of right ventricular function and oxygenation.

Laboratory investigations revealed severe anemia with a hemoglobin level of 7.6 g/dL, likely attributable to recurrent tumor-related epistaxis. This critical decline in hemoglobin highlighted the clinical dilemma: balancing the urgency of surgical intervention against the significant perioperative risk associated with right-to-left intracardiac shunting, and compromised cardiopulmonary reserve.

Literature Review (PICO-based)

Sinonasal schwannoma is a rare benign peripheral nerve sheath tumor of the sinonasal tract, characterized by indolent growth and generally favorable prognosis. While malignant transformation is exceedingly rare, complete surgical excision remains the mainstay of treatment to prevent local recurrence, which can occur if resection is incomplete. Histologic grading and immunohistochemical findings—such as diffuse S100 positivity—are useful for diagnosis but do not reliably predict clinical behavior in all cases.[1–3,7]

In patients with underlying cardiopulmonary compromise, particularly those with reversed shunt-associated pulmonary hypertension due to uncorrected

congenital heart disease, the perioperative risk is significantly heightened. These individuals face elevated risks of right heart decompensation, paradoxical embolism, and intraoperative hemodynamic instability, making surgical planning especially complex, and requiring careful multidisciplinary coordination.[8]

Surgical stress, general anesthesia, and the risk of massive intraoperative bleeding due to tumor vascularity, pose life-threatening consequences in Eisenmenger physiology, including paradoxical embolism, acute right heart failure, and refractory hypoxemia. Current evidence emphasizes the need for multidisciplinary preoperative optimization, including pulmonary vasodilators and invasive hemodynamic monitoring, prior to any elective surgery.[8,9]

Several case reports and small series on sinonasal schwannoma indicate that, while conservative management is generally not the standard approach in otherwise healthy individuals, it may be temporarily justified in patients with non-aggressive, well-demarcated tumors and severe systemic comorbidities that preclude surgery. Given the benign nature and slow growth of schwannomas, observation with symptomatic control may be an acceptable interim strategy in carefully selected cases.[1–4,7]

The role of adjuvant therapy, such as radiotherapy, remains controversial and is typically reserved for unresectable lesions, residual disease, or rare recurrences. There are no large-scale studies or consensus guidelines regarding the optimal management of sinonasal schwannoma in patients with severe cardiopulmonary compromise, such as those with reversed shunt physiology secondary to uncorrected congenital heart disease. As such, clinical decisions must be highly individualized, balancing tumor-related morbidity with the significant anesthetic and perioperative risks posed by the patient's underlying condition.[1,10]

In patients with sinonasal schwannoma complicated by Eisenmenger syndrome, surgical excision should be considered only after comprehensive cardiopulmonary evaluation and stabilization, as the risks of perioperative decompensation may outweigh the benefits of tumor removal in the short term. Conservative management, including close monitoring, symptomatic control, and delay of surgery until hemodynamic parameters are optimized, may be the safer approach in selected cases with non-invasive tumor features and stable bleeding control. Although complete resection offers the best chance for disease control, the presence of severe pulmonary hypertension and right-to-left shunting necessitates individualized decision-making in a multidisciplinary setting, where preserving cardiopulmonary stability is prioritized over immediate oncologic intervention.[11,12]

METHOD

A literature search was conducted using the keywords "sinonasal schwannoma", "schwannoma", "Eisenmenger syndrome", "vascular tumor management", "surgical risk", and "conservative treatment" on PubMed, Medline, and Google Scholar databases. The search was limited to articles published in the last 10 years (2014–2024), available in full text, and written in English.

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1. Articles published within the last decade;
2. Case reports, case series, or reviews involving sinonasal schwannoma or schwannoma;

3. Reports involving patients with comorbid Eisenmenger syndrome, pulmonary hypertension, or cyanotic congenital heart disease;
4. Articles discussing treatment strategies, either surgical excision or conservative management (observation, medical therapy).

Exclusion criteria included:

1. Articles not available in full text;
2. Reports unrelated to therapeutic considerations or without clinical outcome descriptions.

Relevant information from selected articles was analyzed in terms of treatment decisions, perioperative outcomes, recurrence rates, and risk-benefit considerations for high-risk cardiac patients. The aim was to synthesize applicable evidence to guide the decision-making process in managing sinonasal tumors in patients with Eisenmenger physiology.

RESULT

From a total of 17 articles identified through database search, five were excluded after title and abstract screening due to irrelevance or duplication. The remaining 12 full-text articles were assessed for eligibility, and six met the final inclusion criteria. Of these, two were case reports involving sinonasal schwannoma in patients with major cardiopulmonary comorbidities, including one with uncorrected congenital heart disease. Two narrative reviews discussed perioperative considerations in pulmonary hypertension, and the other two articles addressed recurrence and surgical strategies in schwannoma. However, none specifically explored the management of sinonasal schwannoma in patients with Eisenmenger syndrome. This highlighted the absence of targeted clinical evidence, and underscored the rarity and complexity of managing this combined pathology.

DISCUSSION

Sinonasal schwannoma is a rare benign mesenchymal neoplasm arising from Schwann cells of peripheral nerve sheaths. It accounts for a small fraction of all sinonasal tumors—estimated at less than 4% of head and neck schwannomas—and typically presents as a slow-growing, non-infiltrative mass within the nasal cavity or paranasal sinuses. Although schwannomas are generally indolent and exhibit extremely low metastatic potential, local recurrence may occur, particularly in cases of incomplete surgical excision, or involvement of anatomically complex regions.[1–4,7,10]

The clinical presentation of sinonasal schwannoma can vary, but it most commonly includes nasal obstruction, facial fullness or pressure, headache, and, less frequently, epistaxis. In our patient, recurrent nasal bleeding might have resulted from mucosal erosion due to tumor expansion rather than inherent tumor vascularity, as schwannomas are typically less vascular than hemangiopericytomas.

On imaging, sinonasal schwannomas typically appear as well-defined, non-infiltrative soft-tissue masses with homogeneous or heterogeneous contrast enhancement. Bony remodeling may be seen due to mass effect, but frank bony destruction is uncommon.[1,4]

Histologically, schwannomas are characterized by a biphasic pattern of Antoni A (hypercellular, spindle cells in palisades or Verocay bodies) and Antoni B (hypocellular, myxoid areas). Cellular atypia and mitotic activity are typically minimal.[4]

Immunohistochemically, schwannomas demonstrate strong, diffuse positivity for S100 protein, and may also express SOX10. Other markers such as CD34, STAT6, or bcl-2, which are more characteristic of solitary fibrous tumors or hemangiopericytomas, are typically negative in schwannomas.[1,4]

The patient was diagnosed with a sinonasal schwannoma based on characteristic radiological and histopathological features. However, clinical management was significantly complicated by the presence of severe pulmonary arterial hypertension with reversed cardiac shunt, secondary to an uncorrected secundum atrial septal defect (ASD).[1–4]

This condition represents an advanced and irreversible stage of congenital heart disease physiology, marked by right-to-left shunting, chronic hypoxemia, and elevated pulmonary vascular resistance. The coexistence of a space-occupying sinonasal tumor and a cyanotic cardiac defect posed substantial perioperative risk, making surgical planning particularly challenging. Even though schwannomas are typically benign and slow-growing, their anatomical location and symptomatic burden necessitated careful multidisciplinary decision-making in the context of high anesthetic and cardiovascular risk.[5,6]

The pathophysiological hallmark of Eisenmenger syndrome involves increased pulmonary vascular resistance that exceeds systemic pressures, reversing the shunt direction. This leads to chronic hypoxemia, secondary erythrocytosis, paradoxical embolism risk, and impaired cardiopulmonary reserve. Any form of general anesthesia or surgical stress may precipitate acute decompensation, arrhythmia, right heart failure, or sudden death in these patients. Thus, the management of any non-cardiac surgical condition in Eisenmenger physiology must be approached with extreme caution.[9,13]

Literature on the management of sinonasal schwannoma in patients with reversed shunt-associated pulmonary hypertension remains virtually nonexistent, likely owing to the extreme rarity of this clinical combination. While sinonasal schwannoma is generally regarded as a benign and indolent tumor, complete surgical excision remains the definitive treatment to prevent local recurrence.[12]

Endoscopic surgical approaches have increasingly replaced traditional open techniques due to their superior visualization, lower morbidity, and improved cosmetic outcomes. In contrast to vascular tumors such as hemangiopericytomas, schwannomas are typically less prone to intraoperative bleeding, although surgical difficulty may still arise from their proximity to critical anatomical structures in the sinonasal tract.[14]

In high-risk patients with significant cardiopulmonary compromise, including those with pulmonary hypertension and right-to-left shunting, careful preoperative planning, anesthesia risk stratification, and potential staged interventions are essential. However, no standardized perioperative protocols exist for managing sinonasal schwannoma in this rare subset of patients, and clinical decisions must be individualized based on tumor behavior and cardiopulmonary stability.[8,15]

For patients without significant comorbidity, surgery is typically curative, and recurrence rates drop considerably with clear margins. In contrast, for our patient, the combination of high anesthetic risk, severe pulmonary hypertension (SPAP >80 mmHg), and right ventricular dysfunction led to a consensus decision to defer surgical intervention. This highlights the critical importance of individualized risk-benefit analysis when treating otherwise surgically curable tumors in high-risk cardiopulmonary patients.[12,16]

In cases where surgical resection is considered, several approaches are available depending on tumor size, location, and extent. In our case, the selected surgical technique was the midfacial degloving approach, which allows wide exposure of the nasal cavity and paranasal sinuses without external facial incisions. This method involves intraoral and intranasal incisions followed by elevation of soft tissue structures to gain

access to the tumor. It is particularly suited for large, vascular sinonasal tumors, such as hemangiopericytomas, as it facilitates complete tumor excision while maintaining satisfactory cosmetic outcomes. However, in patients with compromised cardiopulmonary function, such as those with Eisenmenger syndrome, the surgical and anesthetic risks remain substantial.[16,17]

Conservative management of sinonasal schwannoma, while not the standard approach, may be cautiously adopted in select high-risk scenarios. Observation and symptomatic treatment - such as nasal bleeding control, analgesia, and airway maintenance - may serve as interim measures in patients deemed temporarily unfit for surgery due to comorbid conditions such as severe pulmonary hypertension and reversed cardiac shunt.[18]

Unlike highly vascular tumors, schwannomas typically exhibit a slow-growing, non-aggressive a course, allowing clinicians a window for stabilization and multidisciplinary planning. In our case, surgical excision was deferred while initiating supportive therapy aimed at optimizing cardiac function, improving oxygenation, and closely monitoring tumor progression both clinically and radiologically.[9]

There is no documented role for chemotherapy in benign schwannoma, and radiotherapy is generally not indicated unless there is a malignant transformation or recurrent, or symptomatic growth not amenable to surgery. Thus, the chosen conservative strategy, while non-curative, reflects a risk-adjusted management pathway tailored to the patient's unique cardiopulmonary vulnerability.[13]

In terms of tumor biology, sinonasal schwannomas are typically benign, encapsulated neoplasms with low proliferative activity and extremely limited metastatic potential. However, despite their indolent nature, long-term surveillance remains essential, particularly due to the potential for local recurrence in cases of incomplete excision or regrowth from residual nerve sheath tissue.[17]

Recurrences have been reported even years after initial treatment, underscoring the importance of structured follow-up. Therefore, serial nasal endoscopy and periodic imaging -such as MRI or CT- are advised to monitor for tumor stability, recurrence, or complications, especially in patients with delayed surgical intervention due to comorbid conditions.[12,17]

Managing the hematological and cardiovascular risks associated with Eisenmenger syndrome in this context was equally complex. Our patient had significant cyanosis (SpO2 66% room air), compensated polycythemia, and right heart strain. He required supplemental oxygen, diuretics, and optimization of insulin therapy due to steroid-related hyperglycemia. Any elevation in systemic vascular resistance or pulmonary vasoconstriction -whether due to hypoventilation, infection, or stress- can precipitate right ventricular failure. Therefore, the perioperative period is particularly dangerous.[8]

There are no randomized trials evaluating surgical timing or safety thresholds in Eisenmenger patients undergoing non-cardiac surgery. However, expert consensus guidelines suggest that only urgent, life-saving surgeries should be performed in this population, unless pulmonary vascular resistance is reduced pharmacologically.[5]

Another layer of complexity in the management of sinonasal schwannoma in high-risk patients is the judicious use of corticosteroids, especially when addressing tumor-associated inflammation or peritumoral edema. While schwannomas are typically not highly edematous or inflammatory, mucosal congestion and secondary obstruction may

warrant short-term anti-inflammatory therapy to improve airway patency and patient comfort.[9]

In our case, a short course of high-dose methylprednisolone was administered to alleviate nasal mucosal inflammation and support respiratory function, particularly in the setting of underlying cyanotic heart disease, and compromised oxygenation. However, this intervention required close metabolic monitoring, as the patient exhibited steroid-induced hyperglycemia. Insulin titration was initiated in collaboration with an endocrinology team to maintain euglycemia during the corticosteroid treatment window.[18]

This case underscored the need for multidisciplinary collaboration involving otolaryngology, cardiology, anesthesiology, and endocrinology. Each specialty contributed to decision-making in a context where guidelines were limited, and clinical judgment was paramount. The evolving nature of the patient's cardiovascular status also necessitated flexibility in treatment planning.[14]

Moreover, psychological and social considerations are also crucial in managing complex patients with dual diagnoses such as this. Chronic hypoxia, limited functional capacity, and the psychosocial burden of congenital heart disease can impair adherence to medical recommendations and follow-up. It is essential to provide adequate counseling, family education, and psychosocial support, especially in young adult patients with long-standing untreated congenital defects.[8]

The rarity of sinonasal schwannoma in itself poses challenges in establishing standardized treatment protocols. When superimposed on Eisenmenger physiology, the situation becomes even more complicated. This dual pathology demands not only medical insight but also ethical considerations about risk tolerance, quality of life, and patient autonomy. Patients should be involved in shared decision-making, with transparent discussions about the risks of intervention versus observation.[1]

Emerging therapeutic options for pulmonary hypertension may offer future opportunities for more aggressive tumor management in Eisenmenger patients. Agents such as endothelin receptor antagonists, phosphodiesterase-5 inhibitors, and prostacyclin analogues have shown efficacy in reducing pulmonary vascular resistance and improving symptoms. In carefully selected patients, preoperative optimization using these agents could theoretically expand surgical candidacy, although this approach must be pursued with caution and individualized assessment.[9]

Additionally, evolving surgical and anesthetic techniques —including awake intubation, regional anesthesia adjuncts, and intraoperative cardiopulmonary support— may further reduce perioperative risk in the future. However, their application remains limited in routine practice due to the need for highly specialized teams and equipment.[13]

In conclusions, this case reflected a rare but important intersection between oncologic and congenital cardiac pathology. It emphasized the limitations of applying standard treatment algorithms to complex patients and the necessity of case-by-case evaluation. Conservative management, though not curative, served as an appropriate bridge strategy in this high-risk setting.

Future multicenter registries or pooled case series, may help illuminate trends and outcomes in patients with sinonasal tumors and severe cardiac comorbidities. Until then, clinicians must rely on interdisciplinary consensus, cautious clinical judgment, and careful longitudinal follow-up in managing such intricate cases.

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