

Primary Sinonasal Malignant Melanoma: An Unusual Clinical Case and Review

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ABSTRACT

Background: Sinonasal mucosal melanoma is a rare and aggressive malignancy, accounting for less than 1% of all melanomas. Due to its nonspecific symptoms and deep anatomical location, diagnosis is often delayed. Management is challenging and typically relies on surgical resection, with or without adjuvant radiotherapy.

Case Presentation: We report the case of a 62-year-old woman with a history of right orbital exenteration who presented with a gradually enlarging left endonasal mass. Initial histopathological examination revealed an undifferentiated tumor. One year later, the patient developed left-sided nasal obstruction and intermittent epistaxis. A second biopsy confirmed the diagnosis of malignant melanoma. Imaging revealed a polylobulated, enhancing mass occupying the left nasal cavity, with extension to the ethmoid cells, olfactory fossa, nasal vestibule, and close contact with adjacent sinus structures. Surgical management included a combined endonasal and Rouge-Denker approach with septectomy, resection of the medial maxillary wall, and a Draf IIa frontal sinusotomy. Complete tumor resection was achieved, and postoperative recovery was uneventful.

Conclusion: Sinonasal malignant melanoma remains a diagnostic and therapeutic challenge due to its rarity and aggressive behavior. This case highlights the importance of a multidisciplinary approach, combining detailed imaging, immunohistochemistry, and tailored surgical strategies to optimize outcomes in this uncommon and often misdiagnosed entity.

KEYWORDS: Sinonasal melanoma, mucosal melanoma, nasal cavity tumor, endoscopic surgery, Rouge-Denker approach, malignant melanoma, case report.

INTRODUCTION

Malignant melanoma is a neoplastic proliferation of melanocytes, neuroectodermal in origin, which may or may not contain melanin pigment. While cutaneous melanomas are the most frequent and well-documented forms, mucosal melanomas represent a rare and particularly aggressive subtype, accounting for less than 1% of all melanomas and approximately 4% of all sinonasal malignancies. Sinonasal mucosal melanoma is a challenging disease to diagnose due to its non-specific clinical presentation. Symptoms such as unilateral nasal obstruction, epistaxis, or rhinorrhea are common but often misattributed to benign inflammatory or infectious conditions. As a result, diagnosis is frequently delayed, and up to 30% of cases may initially be misclassified, especially when the tumor lacks pigmentation.

The exact etiology of mucosal melanomas remains unclear. No definitive environmental or genetic risk factors have been identified, although chronic irritation, trauma, or viral oncogenesis have been proposed in isolated reports. A complete pretherapeutic workup—including nasal endoscopy, cross-sectional imaging (CT, MRI), and histopathological evaluation with immunohistochemistry—is essential to establish a reliable diagnosis and determine the extent of local invasion [1].

The mainstay of treatment is surgical excision with the goal of achieving negative margins, but this is often difficult to obtain due to the anatomical complexity of the sinonasal region. In many cases, adjuvant radiotherapy is considered to improve local control, although its impact on overall survival remains uncertain and varies across published studies. New systemic therapies, such as immune checkpoint inhibitors, are emerging as promising options, but their effectiveness in mucosal subtypes is still under investigation due to limited trial data.

The first reported case of nasal cavity malignant melanoma was described by Lucke in 1869 [2], and fewer than a thousand cases have since been reported in the literature—highlighting the rarity of this disease and the lack of standardized treatment guidelines. A multidisciplinary approach is therefore essential to ensure optimal patient outcomes.

CASE REPORT

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We report the case of 62 years old woman who was referred to our ENT department for left endonasal mass. In 2006, this patient underwent post-traumatic exenteration for the right orbit.

The history of illness started in January 2024 with the appearance of a left endonasal mass increasing gradually of volume. This mass underwent excision with pathological examination, which revealed tumour proliferation of undetermined type.

However, 1 year ago, the patient reported the emergence of left unilateral and intermittent epistaxis with homolateral nasal obstruction.

Clinical examination showed a huge mass offsetting left nasal cavity and preventing her access.



Figure 1: anterior rhinoscopy: mass offsetting left nasal cavity and preventing her access

In December 2024, a second biopsy was performed and revealed indifferent, infiltrating, ulcerated proliferation, primarily suggestive of malignant melanoma.

A nasosinusual CT-Scan showed a left ethmoid-nasal polypoid tissue filling.

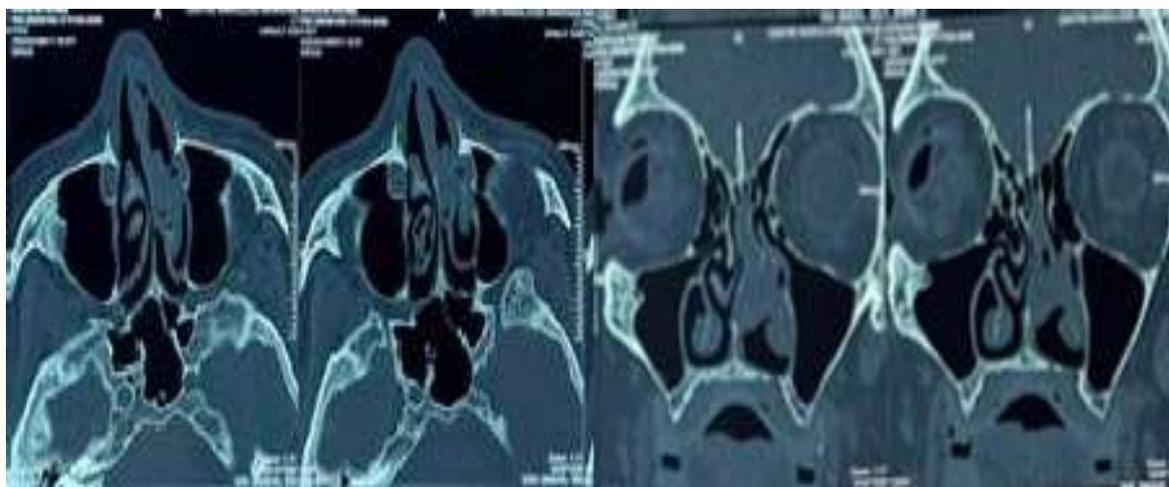


Figure 2: naso-sinusual CT-Scan: left ethmoid-nasal polypoid tissue

A cervical-facial MRI was performed and revealed the presence of a tissue mass with irregular contours in the left nasal cavity, with T1 iso-signal, diffusion hypersignal, enhanced heterogeneously after gadolinium injection, measuring 34.14.5 mm and extending over 38

mm. This mass encompasses the left middle turbinate, pushes back the ipsilateral upper and middle turbinates, and internally comes into contact with the nasal septum. Externally, it comes into close contact with the middle and lower nasal passages and the inner wall of the left maxillary sinus, pushing back the ipsilateral nasolacrimal duct. Superiorly, the mass fills the ipsilateral olfactory fossa and comes into contact with the ethmoid cribriform plate. Anteriorly, it fills the upper part of the nasal vestibule and comes into contact with the ipsilateral nasal wing.

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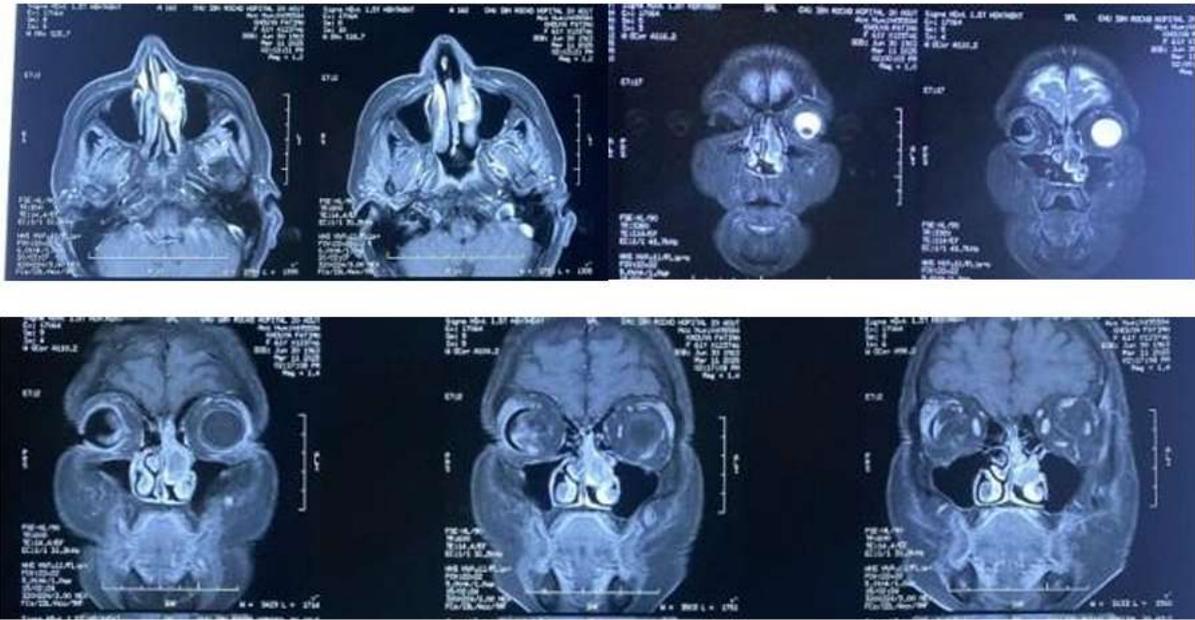
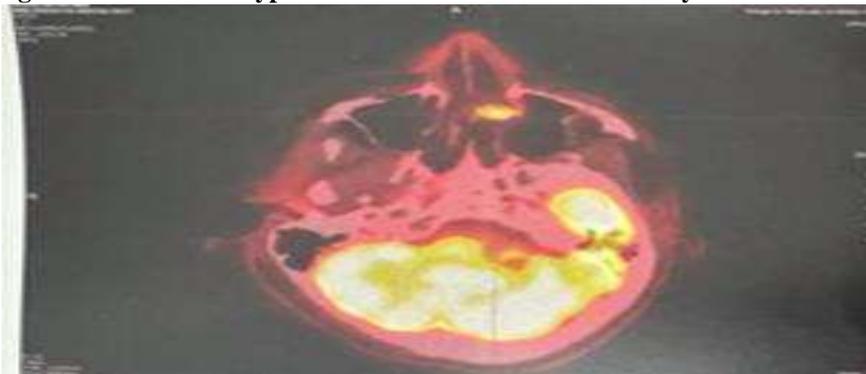


Figure 3: cervico-facial MRI: tissue mass in the left nasal cavity enhanced heterogeneously after gadolinium injection, measuring 34.14.5. 38mm

A PET scan revealed hyper metabolism in the left nasal cavity and left choana, where there was thickening.

Figure 4: PET- Scan: hypermetabolism in the left nasal cavity and left choana



For the therapeutic options, we proposed a surgical management to the patient. Two approaches were undertaken. The initial approach was endonasal with aspiration at the tumour shiver. The mass came into contact with the roof, septum and lateral wall of the maxillary sinus. The second approach was the Roux-denker approach, transantronal. A septectomy was performed with exeresis of the internal wall of the left maxillary sinus with evidention of the sinus, as well as milling of the roof of the left nasal cavity opposite the insertion of the tumour. A Draf II A was performed in the frontal sinus. The remains of the tumour were resected. A packing with 3 nasal swabs was performed.

DISCUSSION

Primary mucosal melanoma of the nasal cavity and paranasal sinuses is a rare and aggressive malignancy that poses significant diagnostic and therapeutic challenges. It represents approximately 0.5% to 1% of all melanomas and 4% of sinonasal malignant tumors, predominantly affecting adults over 60 years of age, with no clear sex predominance [3,4]. The etiology remains unclear, though chronic inflammation, trauma, and exposure to inhaled irritants have been hypothesized as contributing factors [5].

Our patient's history of post-traumatic orbital exenteration and delayed onset of symptoms may have contributed to both diagnostic ambiguity and disease progression. The presentation with unilateral nasal obstruction and intermittent epistaxis is typical, but often nonspecific, leading to misdiagnosis or underdiagnosis in early stages. Up to 30% of mucosal melanomas are initially misinterpreted as benign or undifferentiated neoplasms, especially when pigmentation is absent or minimal [6].

The initial biopsy in our case was inconclusive, reflecting the histopathological complexity of mucosal melanomas, which may exhibit spindle, epithelioid, or mixed cellular patterns.

Immunohistochemical analysis remains essential for accurate diagnosis, typically requiring positivity for melanocytic markers such as HMB-45, Melan-A, and S-100 [7].

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Imaging plays a pivotal role in staging and surgical planning. The CT scan delineates bony erosion and sinus involvement, while MRI provides superior assessment of soft tissue and skull base invasion. In our case, the tumor extended superiorly to the olfactory fossa, laterally to the maxillary sinus wall, and anteriorly to the nasal vestibule, highlighting its aggressive local behavior and complex topography.

Surgical excision with clear margins is the primary curative modality, although this goal is often difficult due to the intricate anatomy of the sinonasal region. A purely endonasal approach may be insufficient for large or posteriorly extending tumors. In this case, a dual approach was adopted—endonasal resection complemented by a Rouge-Denker (transantral) approach, allowing improved exposure of the maxillary sinus and posterior nasal cavity. A septectomy and Draf IIa frontal sinusotomy were also necessary to access all involved compartments and reduce the risk of recurrence.

Despite aggressive surgical efforts, mucosal melanomas have a high rate of local recurrence (up to 60%) and distant metastasis (up to 50%), with overall 5-year survival rates ranging from 20% to 40% [8,9]. The role of adjuvant therapy, particularly radiotherapy, remains debated. Some studies suggest it may reduce local recurrence, although its impact on overall survival is unproven [10]. Immunotherapy (e.g., anti-PD-1, CTLA-4 inhibitors) and targeted therapies (in cases with c-KIT mutations) have shown promise in cutaneous melanomas, but their role in mucosal subtypes remains investigational, partly due to the rarity of these tumors and limited trial data [11].

Our case underscores the importance of a multidisciplinary approach involving ENT surgeons, radiologists, pathologists, and oncologists. Early and accurate histological diagnosis, combined with high-resolution imaging and tailored surgical planning, are essential to optimize outcomes. Given the rarity and poor prognosis associated with this pathology, further multi-institutional studies are needed to develop standardized guidelines and assess the efficacy of novel systemic treatments.

CONCLUSION

Sinonasal mucosal melanoma is a rare and aggressive malignancy that often presents with nonspecific symptoms, leading to delayed diagnosis and therapeutic challenges. Our case highlights the importance of maintaining a high index of suspicion for malignancy in patients with persistent unilateral nasal symptoms, particularly in the elderly. Accurate histological diagnosis, guided by immunohistochemistry, is essential, as is detailed radiological assessment to determine the extent of local invasion.

Surgical resection remains the mainstay of treatment. However, due to the anatomical complexity of the sinonasal region, a combined approach may be required to achieve optimal tumor control, as illustrated by the dual endonasal and Rouge-Denker approach in our patient. The high risk of recurrence and metastasis necessitates close postoperative follow-up and may warrant consideration of adjuvant therapies in selected cases.

This case underlines the critical need for individualized, multidisciplinary management strategies and contributes to the growing clinical experience necessary to improve outcomes in this rare and challenging pathology.

REFERENCES

- 1) Kharoubi S. Mélanome malin des fosses nasales: considérations cliniques et thérapeutiques à propos de trois cas. *Cancer Radiothérapie*. 2005 Mar;9(2):99-103.
- 2) Poissonnet G, Castillo L, Dassonville O, Ettore F, Birtwisle-Peyrottes I, Santini J, et al. Les mélanomes malins nasosinusiens : revue de la littérature à propos de 12 cas. *Rev Laryngol Otol Rhinol* 1997; 118:155–61.
- 3) Thompson LD. Mucosal melanoma of the head and neck. *Head Neck Pathol*. 2010;4(1):53–58.
- 4) López F, Suárez C, et al. Sinonasal mucosal melanoma: diagnosis, treatment, and outcomes. *Ann Surg Oncol*. 2016;23(1):296–302.
- 5) Batsakis JG, Regezi JA. The pathology of head and neck tumors: mucosal melanomas. *Head Neck Surg*. 1979;1(5):387–392.
- 6) Ascierto PA, et al. Melanoma: a multidisciplinary approach. *Crit Rev Oncol Hematol*. 2008;66(1):1–20.
- 7) Prasad ML, Patel SG, et al. Primary mucosal malignant melanoma of the head and neck: a proposal for a staging system. *Head Neck*. 2004;26(1):17–27.
- 8) Amit M, Na'ara S, et al. Patterns of failure and postrecurrence outcomes in sinonasal mucosal melanoma. *Head Neck*. 2017;39(4):773–780.
- 9) Temam S, et al. Postoperative radiotherapy for primary mucosal melanoma of the nasal cavity and paranasal sinuses. *Arch Otolaryngol Head Neck Surg*. 2005;131(10):851–856
- 10) Lund VJ, Howard DJ, Harding L, Wei WI. Management options and survival in malignant melanoma of the sinonasal mucosa. *Laryngoscope*. 1999;109(2 Pt 1):208–211
- 11) D'Angelo SP, et al. Efficacy of anti-PD-1-based immunotherapy in patients with mucosal melanoma. *Cancer*. 2020;126(8):1657–1665.