Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

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Mélanome of the Nasal Cavity: About A Case

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DOI: https://doi.org/10.36347/sjmcr.2025.v13i07.021 | **Received:** 28.05.2025 | **Accepted:** 03.07.2025 | **Published:** 14.07.2025

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Abstract | **Case Report**

Reporting a rare instance of nasosinusal malignant melanoma is the goal. A 63-year-old patient was seen to have had nasal blockage and recurrent seurosanguinating secretion evolving for 3 months. The diagnosis was proposed by nasosinusa endoscopy, which revealed a malignant process with dark pigmentation. Mucinous melanomas of the nasal cavity and paranasal sinuses are rare and extremely aggressive tumors. In conclusion, mucous malignant nasosinusal melanoma is a rare histological kind that does not exhibit any particular symptoms. Typically, the tumor is gray-blackish and invasive. Histopathological with immunohistochemistry (HMB-45) is the diagnosis. The therapy protocol needs to be examined by a multidisciplinary team and is not codified. The outlook is not good.

Keywords: Immunohistochemistry, radiation, immunotherapy, mucosal malignant melanoma.

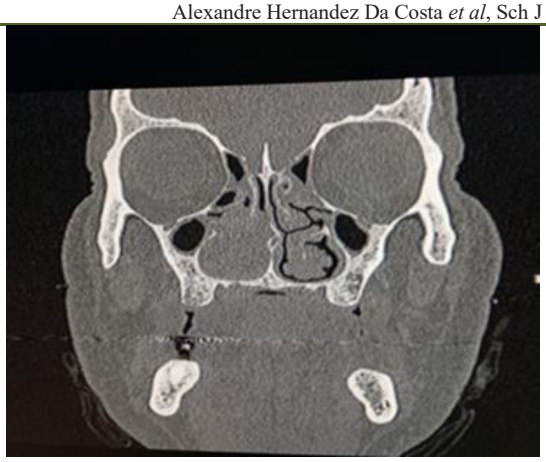
INTRODUCTION

Mucinous melanomas of the nasal cavity and paranasal sinuses are rare and extremely aggressive tumours. Their incidence ranges from 0.2 to 1 per million population [1,2]. The current standard of care is initial surgery for resectable lesions. Overall survival at 5 years varies from 20 to 40% depending on the series [3,4]. These melanomas derive from melanocytes present in the respiratory epithelium of the nasal cavities and sinuses. Their preferred site is the inferior turbinate, rarely the middle turbinate. We present a case of nasosinusal melanoma of the right nasal fossa, involving the 3 turbinates and lysis of the inner wall of the right maxillary sinus, posteriorly towards the choanae and superiorly towards the homolateral ethmoidal cells.

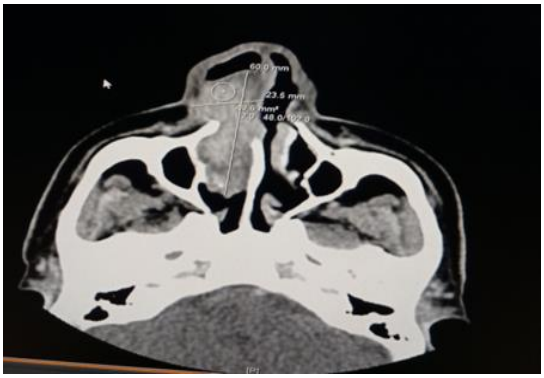
OBSERVATION

63-year-old man, chronic smoker 06p/A, weaned for 5 years, history of hypertension, on calcium channel blocker 5 mg/day, headache, consulted for unilateral right nasal obstruction and serosanguinous secretion for 3 months. Rhinological examination revealed a blackish budding tumour filling the right nasal fossa with serosanguinating secretion. Cervical examination revealed cervical and right submandibular adenopathy ±20mm. Skin examination was unremarkable. Nasal endoscopy was difficult to perform, given the total filling of the mass in the nasal cavity and the risk of bleeding from the slightest contact.

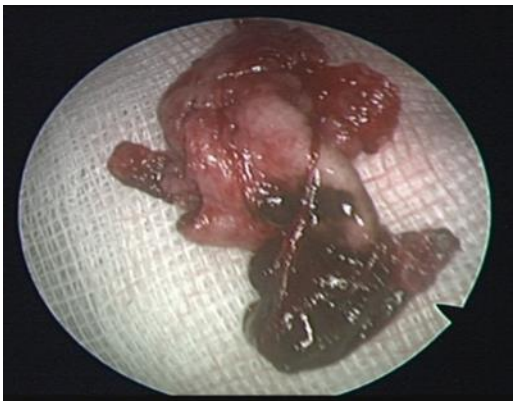
Computed tomography (CT) of the facial fossa with injection of contrast medium revealed a lesional process filling the right nasal fossa as far as the choanae and extending towards the right maxillary sinuses, with bone lysis (figure 1and 2). MRI of the nasal cavity showed the presence of a tumoral process filling the entire right nasal cavity with hypersignal on T1 and T2 sequences, diffusion hypersignal without significant diffusion restriction with moderate enhancement after gadolinium injection, and measuring 61 × 20 × 35 mm with invasion of the three (3) middle and upper inferior turbinates creating a contralateral mass effect and deviation of the nasal septum to the left side. A biopsy was performed to remove the tumour. Anatomopathological examination of the biopsy revealed a malignant melanoma. The distant extension workup was positive, with a secondary pulmonary nodule, and bone scintigraphy was normal. The tumor is classified as stage 1 according to the TNM classification based on the 2010 American Joint Committee on Cancer (AJCC) classification of mucinous melanomas of the nose and paranasal sinuses.



Frontal sinus CT scan reveals a nasal cavity mass filling the entire right nasal cavity



Axial CT scan showing a mass filling the right nasal fossa



Surgical excision of lesions with healthy tissue margin

Surgical treatment consisted of an endonasal endoscopic antero-medial right maxillectomy. Homolateral lymph node dissection of the right cervical chain II, III and IV was performed at the same time. Pathological examination confirmed the diagnosis of melanoma. Adjuvant treatment with external radiotherapy was performed at a dose of 54 Gy. After surgical excision, the disease progressed towards locoregional recurrence in the right axillary lymph nodes, which were resected surgically with a positive pathological result, or towards visceral metastases, as verified in our clinical case with pulmonary expectation.

DISCUSSION

Malignant melanoma is a neoplastic proliferation of melanocyte-like cells of neuroectodermal origin, with or without melanin pigment [4]. Nasosinus melanomas account for 1 to 2% of all melanomas and 3.6% to 12% of nasosinus malignancies [1,4,5,6].

It is a pathology of the elderly, with an average age ranging from 50 to 70 in the literature [4,5,7,8]. Gender distribution varies from one series to another [1,4,8]. In the nasal cavity, the tumor frequently involves the septum, the lateral wall of the nasal cavity and the turbinates, as in our patient's case. However, it is often

difficult to pinpoint the tumour's starting point, given its rapid spread.

The maxillary sinus is the first sinus to be affected [7,9]; the etiopathogenesis is poorly understood, but certain factors have been incriminated, such as smoking and formaldehyde.

Clinical symptoms are dominated by nasal obstruction and epistaxis, often unilateral [6,7]. Nasal endoscopy reveals a hemorrhagic, blackish-gray or slate-colored mass filling the nasal cavity.

Clinical examination should verify the absence of cutaneous melanoma. The frequency of lymph node metastases at the time of diagnosis varies from 0 to 23%. The incidence of distant metastases ranges from 0 to 14% [8,9].

Diagnosis is histological. It is based on the detection of intracellular melanin. In achromic forms, the diagnosis is made by electron microscopy, through the demonstration of premelanosomes, or by immunohistochemistry showing a positive reaction with antivimentin, anti-S 100 and anti-HMB 45[2,4].

Tumor extension is essentially local, and the majority of patients are classified as stage at the time of diagnosis. Magnetic resonance imaging (MRI) reveals tumour extension to the orbit, infra-temporal fossa and cavernous sinus. Bone relationships are studied on CT scans [8,9].

Their management requires a multidisciplinary approach, with cross-sectional imaging (CT and MRI) playing a fundamental role in therapeutic decisions. In the initial work-up, CT and MRI allow precise assessment of location, volume and extension into the sinus cavities, nasopharynx and choanae, as well as orbital and endocranial extension.

Treatment is primarily surgical. Endoscopically-guided endonasal surgery can be proposed for tumours located in the anteroinferior part of the nasal cavity and not exceeding 1cm [4].

Fractionated external radiotherapy is used alone or after surgery, with controversial results.
Chemotherapy is used as adjuvant treatment in advanced forms, with disappointing results [9,11,12].

Evolution and Prognosis

The evolutionary risk is that of the occurrence of metastases, which are sometimes late. The survival rate is between five and ten years. After surgical excision, the disease may progress to locoregional recurrence in the skin or lymph nodes, or to visceral metastases. In our patient's case, this was within the 5-year limit, with axillary adenopathy after 2 years and pulmonary expectation.

CONCLUSION

Mucinous melanomas of the nasosinus cavities are neoplasms with a poor prognosis due to their high propensity for metastasis. The main treatment consists of surgery followed by radiotherapy. At present, there is no general agreement on the benefits of new adjuvant drug therapies for this indication, in contrast to the treatment of primary cutaneous melanoma.

Conflicts of Interest: The authors declare that they have no conflicts of interest in relation to this article.

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