CASE REPORT

Nasal and paranasal sinus mucosal melanoma

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ABSTRACT

Background: Nasosinusal melanoma is a rare and highly aggressive tumor with a high potential for metastasis. It originates from melanocytes in the nasal mucosa and primarily affects the lateral nasal wall, nasal septum, and inferior nasal concha. It is less common in the paranasal sinuses. This type of cancer is known for its high recurrence rate and is most commonly seen in individuals between the ages of 70 and 80, with a slightly higher incidence in females. Case report: This case study discusses a 73-year-old female patient with nasal mucosa melanoma. The tumor was first identified 1.5 years before the initial surgery, and the patient received adjuvant treatments, including immunotherapy. She also underwent a second surgery in an attempt to completely remove the tumor. However, almost a year after the second surgery, a new mass was discovered in the nasal cavity. Due to the development of distant metastases, further treatment was discontinued, and the patient was referred to palliative care services. Conclusion: Early diagnosis and appropriate treatment are crucial, although they may not always result in a cure due to the aggressive nature of this type of tumor.

Keywords: Melanoma, Nasal mucosa, Neoplasm recurrence local, Nose neoplasms.

INTRODUCTION

Nasosinusal melanoma is a rare and highly aggressive tumor that originates from melanocytes found in the nasal mucosa[1]. It accounts for 4% of malignant illnesses in the region and has a high potential for metastasis. The most commonly affected areas are the lateral nasal wall, nasal septum, and inferior nasal concha, with involvement of the paranasal sinuses region being less common[2].

Unlike other types of melanoma, nasosinusal melanoma is not related to the exposure to ultraviolet (UV) radiation because of its location, but rather to the exposure to toxic volatile agents, such as cigarettes and formaldehyde. Common symptoms include unilateral nasal obstruction, epistaxis, and, less frequently, rhinorrhea, hyposmia, and frontal headache[3]. However, the non-specific symptoms and location of nasosinusal melanomas often lead to delayed diagnosis and confusion with other pathologies[4].

Factors such as older age, male gender, and amelanotic lesions, as well as tumors originating in the paranasal sinuses, have been linked to a poorer prognosis, with a 5-year survival rate of only 30.69%[2]. Nasosinusal melanomas have a local recurrence rate of 20% and a potential to create distant metastases of 80%[5]. This type of tumor is most commonly seen in individuals between the ages of 70 and 80, with a slight predominance in females[6].

The initial diagnostic test for paranasal sinus tumors is typically computed tomography (CT) scans, often associated with videonasolaryngoscopy. Magnetic resonance

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imaging (MRI) can also provide valuable information about the tumor's characteristics, aiding in staging and determining the best approach for removal[3]. While melanomas are generally resistant to radiotherapy, especially nasosinusal melanomas, studies have shown that radiotherapy can be effective as an adjuvant treatment after surgical removal, particularly in reducing the risk of local recurrence[5]. However, radiotherapy alone has not been found to be an effective treatment for nasosinusal melanoma[7].

The choice of a surgical method for the removal of nasosinusal melanoma depends on the location and size of the lesion[8]. Radical open surgical techniques have been less widely used due to the potential loss of function and the greater aesthetic damage associated with the procedure[2]. In the last two decades, endoscopic nasal surgery has become the preferred method because of its superior results, shorter surgery time, faster recovery, less bleeding, and better visualization of the affected area[2,9].

This article aims to discuss the clinicopathological characteristics of nasosinusal melanoma, a rare tumor that accounts for less than 5% of all head and neck tumors in the United States[10] and is not among the ten most common in Brazil, according to the National Cancer Institute[11]. Due to its low occurrence, knowledge about its origin and predisposing factors is limited. Additionally, there are not any clearly defined guidelines for the evaluation and therapeutic approach of mucosal melanomas[12]. Therefore, this report is relevant in contributing to the development of a solid theoretical framework and to fostering discussions about this type of tumor.

CASE REPORT – ENDONASAL MELANOMA

A 73-year-old female patient, M.T. C.S., presented to the otorhinolaryngology department reporting that she had undergone resection of a lesion in the left nasal cavity, located in the tail of the left inferior turbinate, 1.5 years previously. She reported having undergone pharmacological therapy after the initial surgery, but was unsure of the specific medications she had taken - she believed they were immunotherapy. However, the patient did not have any medical reports documenting this treatment, only biopsy results of the lesion showed a diagnosis of melanoma. She also reported experiencing nasal obstruction for approximately 30 days and had noticed a lesion in the region of her uvula. CT scans (figure 1) and video nasolaryngoscopy revealed a brown lesion occupying the entire left nasal cavity, cavum, and pharynx. The patient was advised to undergo surgical treatment via nasal endoscopy to remove the lesion, which was performed by a team of otorhinolaryngology and head and neck surgery specialists. The procedure involved resecting the lesion, along with the medial wall of the maxillary sinus, the inferior concha, and the posterior two-thirds of the nasal septum. Due to the extension of the lesion, the uvula and part of the soft palate also had to be resected (figure 2). Additionally, part of the posterior wall of the cavum was also removed (figure 3). The anatomopathological examination revealed epithelioid malignant neoplasm and ulcerated fusocellular, consistent with malignant melanoma, affecting the nasopharynx and posterior oropharynx. However, the anterior margin of the turbinate, septum and medial wall of the maxilla were clear of lesions. The margins of the lesion on the uvula and palate were also

clear, indicating that no further resection was necessary. The patient was then referred to the oncology department, where she was recommended to undergo treatment with radiotherapy as part of her treatment plan.

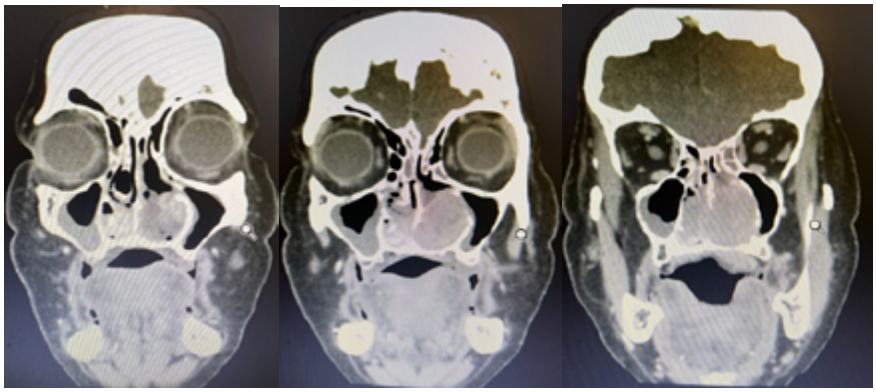

Figure 1: A mass present in the left nasal cavity, with a noticeable lateral shift of the medial wall of the right maxillary sinus.

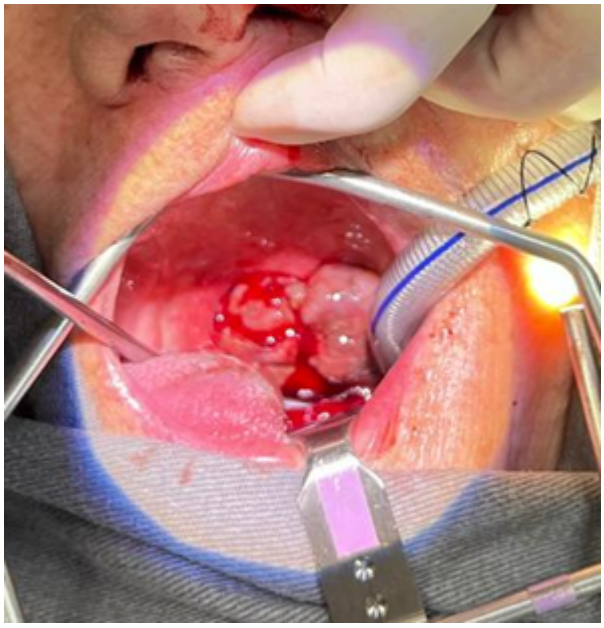

Figure 2: Aspect of the oral lesion - uvula.

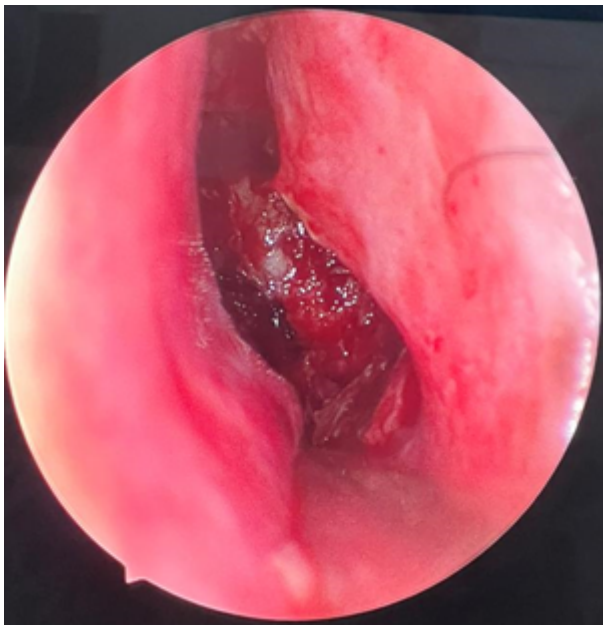

Figure 3: View of the lesion affecting the cavum.

Figure 4: Pigmented coloration is present in the endonasal lesion.

Eight months after the surgery performed in our department, the patient returned with complaints of nosebleeds and a visible mass in her left nasal cavity. A chest CT scan revealed the presence of pulmonary nodules and osteolytic lesions in the costal arches, indicating the likelihood of metastases. Further examination through a CT scan of the sinuses showed a lesion occupying the entire left nasal cavity, with erosion extending into the left frontal sinus. As a result, the patient was referred for chemotherapy and palliative care.

DISCUSSION

Nasosinusal melanoma is a rare type of tumor known for its high metastatic potential and poor prognosis[13]. The average age of diagnosis is in the seventh decade of life[14], which is consistent with the case presented, in which the disease was diagnosed at the age of 73.

Malignant sinonasal pathologies typically present with non-specific initial symptoms, such as nasal obstruction, facial pain, rhinorrhea, and epistaxis. These symptoms can also be present in benign pathologies, such as allergic processes[15]. However, an unsatisfactory response to medical treatments for inflammatory diseases, a negative investigation for allergies, and unilateral symptoms may raise suspicion of a nasosinusal tumor[16].

The most common symptoms of nasal melanoma include unilateral nasal obstruction, a visible mass, and epistaxis[2]. Other differential diagnoses for nasal melanoma include nasosinusal inverted papillomas[4]. Inverted papillomas are typically benign tumors with a polylobular appearance and firm consistency, and they can cause symptoms such as epistaxis, nasal obstruction, rhinorrhea, and facial pressure[16]. Another potential diagnosis is squamous cell carcinoma (SCC), which accounts for 61% of nasosinusal malignancies[15]. Nasosinusal SCCs are malignant epithelial neoplasms that originate in the epithelium lining of the paranasal sinuses and nasal cavity[17]. Patients with SCC may be asymptomatic in the early stages, but as the disease progresses, they may experience nasal obstruction, epistaxis, sinusitis, proptosis, facial pain, and diplopia[18]. Nasosinusal adenocarcinoma, the second most common type of nasosinusal malignancy after SCC[15], can cause symptoms such as nasal discharge, nasal obstruction, and epistaxis[19].

The diagnosis of nasosinusal melanoma is often delayed, and it is not uncommon for it to occur after the development of distant metastasis. This is because of its non-specific symptoms, painless nature, and location in a region that allows for silent growth[20]. Therefore, endoscopic evaluation is crucial for accurately assessing the extent of the lesion, obtaining biopsy material, and differentiating between different tumors[15]. In the case being studied, metastases were not detected until 2 years and 3 months after the initial diagnosis.

Surgical resection is the primary treatment for nasosinusal melanoma[7]. It is essential to perform a radical resection of the primary tumor with clear margins in order to achieve the best outcome for patients[21]. In this particular case, the initial lesion was surgically removed, but it recurred locally 1.5 years later. A second surgery was performed, which successfully obtained clear margins, but unfortunately, the tumor recurred again eight months later.

A debated point in the management of melanoma is whether open or endoscopic surgery is the better approach. According to studies, the endoscopic approach offers comparable oncological and survival results to open surgery, with lower morbidity and better aesthetic and functional outcomes for patients[5]. However, open surgery may still be necessary in cases of extensive tumor invasion in regions such as the orbit, skull base, hard palate, and soft tissue of the cheek[5]. In the case of the patient presented, endoscopic surgery was chosen because of its benefits and the location of the tumor.

Adjuvant therapies to surgery are still being studied and developed. One of these treatments, used in the patient presented in this case, was immunotherapy. Immunomodulators such as imatinib, nivolumab, ipilimumab, and binimetinib are commonly used to treat melanomas[2]. However, the analysis of these drugs through randomized studies is hindered by the poor prognosis and rarity of the disease, and their efficacy is still being evaluated[2]. Another treatment used in the patient's case was postoperative radiotherapy. While studies have shown that radiotherapy can help control local recurrences, it has not been shown to improve overall survival in patients[7]. Unfortunately, in M.T.C.S.'s case, a new recurrence occurred 8 months after the second surgery, despite the use of postoperative radiotherapy.

Melanomas of the nasosinusal mucosa have a high local recurrence rate, often occurring approximately one to two years after treatment[21]. In the case of the patient studied, the first recurrence occurred a year and a half after the initial intervention (performed at another facility), and the second recurrence happened only 8 months after the second surgery (performed at our facility). This type of tumor is known for its high rate of therapeutic failure and poor prognosis, often leading to distant metastases[7]. The most common sites for these metastases are the liver, lungs, bones, and central nervous system[2]. In the case of M.T.C.S., the metastases were discovered 2 years and 3 months after the initial diagnosis.

This case of nasal melanoma is particularly relevant due to the low incidence of this type of tumor and the limited information available on the subject. Nasal melanoma affects a region that is difficult to see and has non-specific symptoms, making it challenging to diagnose accurately. Continued research on nasal melanoma may lead to advancements in treatment, including new adjuvant therapies, surgeries, or therapeutic approaches that can significantly impact doctors' clinical decisions and improve patient outcomes. Furthermore, publishing articles on rare tumors can raise awareness of the disease, encouraging early screening and diagnosis. Each new article adds valuable information that can be used by other researchers to conduct more in-depth studies.

CONCLUSION

Nasosinusal melanoma is a rare tumor with a poor prognosis. Surgical therapy is currently the most effective approach to treating this disease. However, further research is needed to develop adjuvant therapies that can improve outcomes for patients undergoing surgery. It is crucial for patients to be closely monitored by their medical team because of the high rates of recurrence and the potential for distant metastases.

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ETHICAL APPROVAL

The study in question was submitted and approved by O Comitê de Ética em Pesquisa Envolvendo Seres Humanos do Campus Centro Oeste Dona Lindu da Universidade Federal de São João del – Rei – CEPCO, receiving Certificado de Apreciação ética CAAE number 68636723.0.0000.5545.

Authorship requirements

-Substantial contribution to the study outline or data interpretation: LMNM; JMCM; AVZ

- Participation in writing the preliminary version: DFS, PASS

- Participation in the review and approval of the final version: DFS, PASS

- Compliance with being responsible for the accuracy or integrity of any part of the study: AVZ

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