Malignant Mucosal Melanoma of the Maxillary Sinus with an Oro-antral Fistula

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ABSTRACT

Introduction

Sinonasal mucosal melanomas are *uncommon, making up merely 0.5-2% of all malignant melanomas and 4% of melanomas in head and neck region. In the sinonasal region, they are very aggressive, commonly presenting at an advanced stage. Diagnosis is made using histopathological examination and immunohistochemistry by S100 and HMB-45 markers.*

Case Report

We describe a case of 62 year-old female with swelling in the maxillary area and an oroantral fistula at the right maxillary alveolus who underwent right endoscopic sinus surgery with septoplasty with partial medial maxillectomy and oroantral fistula closure which was diagnosed as malignant melanoma on histopathology and immunohistochemistry.

Discussion

Malignant mucosal melanoma is a rare but aggressive disease originating from melanocytes located in the mucosa. It is characterised by swift nonspecific symptom progression and early metastasis. When the tumour is detected early and fully excised, malignant melanomas are mostly curable. However, once metastasis occurs, treatment options are limited.

Keywords

Malignant Melanoma; Sinonasal Melanoma; Sinonasal Malignancy; Oro-Antral Fistula

Sinonasal mucosal melanomas are particularly aggressive, frequently appearing at an advanced stage, and have a 5-year survival rate of 20-30%.[3] Nasal obstruction, epistaxis and swelling are the commonly presenting complaints, although they tend to be nonspecific and hence can delay diagnosis.[1,4] Diagnosis is made using histopathological examination and immunohistochemistry by S100 and HMB-45 markers.[2]

When the tumour is discovered early and fully excised, melanoma is highly curable. However, once metastasis occurs, treatment options are limited, and palliative care becomes the primary treatment modality.[3]

Case Report

A 62 yr female visited our outpatient clinic, reporting dull pain and swelling in the right maxillary region that had persisted for one month. She also complained of swelling involving her right 2nd upper molar tooth. We examined her to find an exophytic growth at right maxillary alveolus involving the gingival mucosa and right 2nd upper molar tooth with an oro-antral fistula (Fig. 1). The examination by touch revealed a wound that felt firm and had a smooth surface. The mass was non-tender and did not bleed on touch. Cervical neck nodes were not palpable. The diagnostic nasal endoscopy revealed a deviated nasal septum towards right, with a greyish mass partially occluding the right nasal cavity.

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Fig. 1. Growth seen at right maxillary alveolus with oro-antral fistula on initial visit

Computed tomography of the paranasal sinuses showed heterogeneously enhancing soft tissue lesion with destruction of medial wall of right maxilla, suggestive of right maxillary sinus malignancy or infected papilloma with bony erosions with extension of mass into right nasal cavity (Fig. 2).

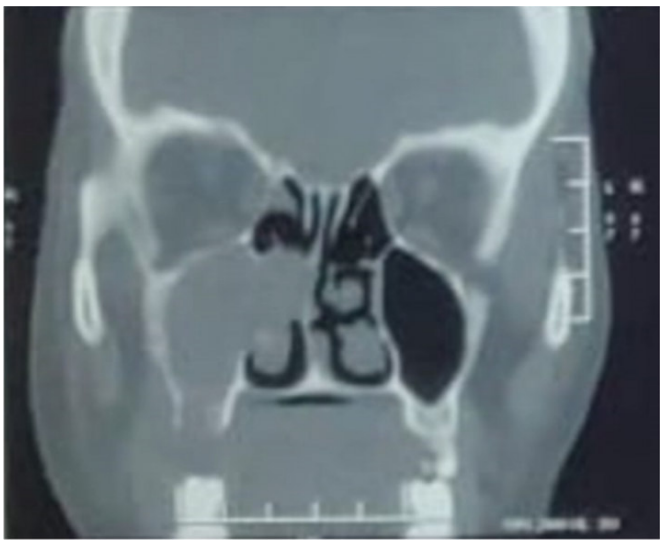

Fig. 2. Coronal computed tomography revealed a soft tissue lesion the right maxillary sinus extending into right nasal cavity with bony erosions.

The clinical presentation looked like a benign inflammatory mass, so the decision was taken to go ahead for surgery rather than taking nasal or oral biopsy. The patient underwent a right-sided full house functional endoscopic sinus surgery (with opening up of all natural ostia of the sinuses) with septoplasty (to get a better approach to the right nasal cavity). Complete clearance of nasal and maxillary mass was done endoscopically along with partial medial maxillectomy to remove the eroded bony parts. All the diseased tissue in the maxilla and oral lesion including the bony erosions was excised with the help of microdebrider in certain areas, so margins could not be obtained and the patient was referred to radiation oncology for adjuvant radiotherapy. In the oral cavity, the oral maxillofacial surgeon performed complete excision of the overhanging mass with clearance of the disease along with 2nd upper molar tooth, gingival mucosa and bony erosions, and primary closure of the oroantral fistula was done using a buccal fat pad and buccal mucosa (Fig. 3). Hard palate was not involved in our patient.

Fig. 3. Excising the growth at right maxillary alveolus and repairing the oro-antral fistula

In histopathological examination a tumour composed of round to oval cells, arranged in sheets and pseudo alveolar pattern with intracytoplasmic and extracellular brownish pigment – melanin - was seen. Thus, a diagnosis of malignant melanoma of right maxillary sinus extending orally as a gingival lesion was made which was later confirmed on immunohistochemistry by the presence of S100 (Fig. 4) and HMB45 (Fig. 5) markers.

Further, the patient was referred to radiation oncology, where she underwent adjuvant radiotherapy with 30 cycles of 60Gy over 6 weeks. She has been disease-free for the last 6 months and is under close follow up.

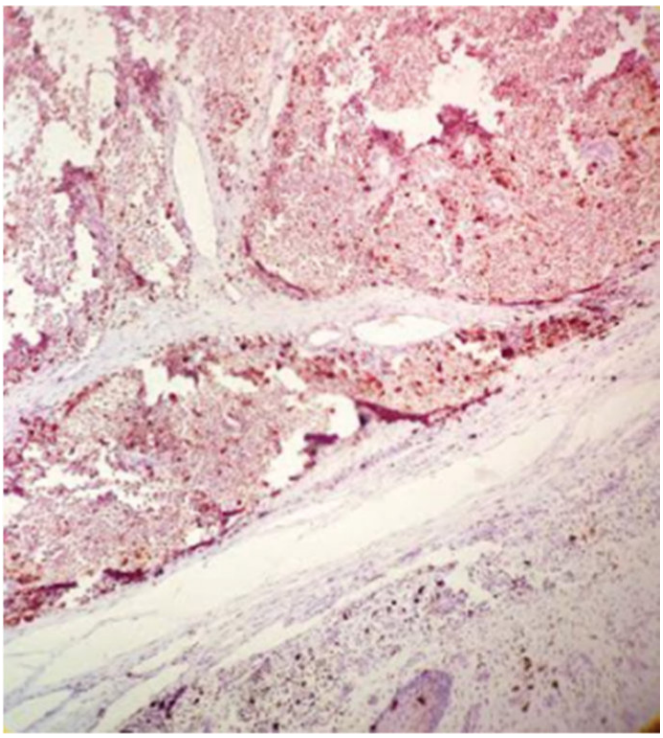

Fig. 4. On Immunohistochemical staining, S100 positivity seen under 4 x 10x magnification

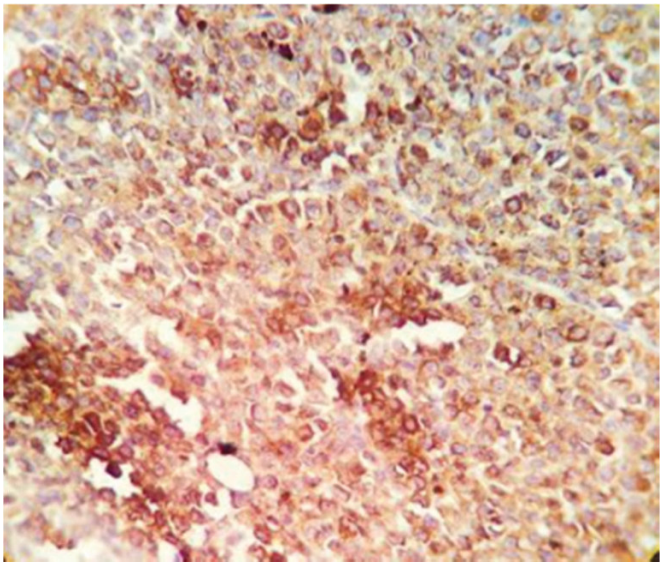

Fig. 5. On Immunohistochemical staining, HMB45 positivity seen under 4 x 40x magnification

Discussion

Mucosal melanomas in the head and neck region are infrequent, typically occurring in individuals in their 60s and 70s. Compared to their cutaneous and uveal counterparts, mucosal melanomas often present at a later stage and exhibit more aggressive behaviour, leading to swift symptom progression and early metastasis.

Regardless of pigmentation, intranasal masses that exhibit rapid growth and occult malignancy characteristics should always be looked into in the context of melanoma.[3] Neoplasms confined to the paranasal sinuses have a tendency to remain asymptomatic until they start invading surrounding structures. As a result, the poor prognoses stems from late detection and their proximity to crucial structures like the brain, skull base, orbits, and carotid artery.[5] Tumours extending beyond the roof of the maxillary sinus may penetrate the eye or the posterior wall, accessing nerves and blood vessels at the skull base and the pterygomaxillary space, potentially leading to a direct invasion of the brain.[5] Early detection of these malignancies is crucial to prevent such complications.

The diagnostic evaluation entails a comprehensive physical examination of the head and neck which includes assessing facial asymmetry, evaluating extraocular muscles, observing pupillary response and looking for signs of globe displacement.

The nasal exam includes flexible or rigid diagnostic nasal endoscopy. Both computed tomography and magnetic resonance imaging can be used to further categorize sinonasal malignancies.[5] Diagnosis is based on histopathological examination and Immuno-histochemistry. The detection of anti-S100 antibodies is very sensitive but not specific while anti-HMB45 antibodies are highly specific and somewhat sensitive. When staining the two markers yield uncertain results; Melan A, a melanoma-specific marker, is normally used, as it has proved to be extremely specific in distinguishing melanoma from other cancers.

Malignant mucosal melanomas are classified based on their primary anatomical locations, differentiating between cancers of the maxillary sinus, nasal cavity or

ethmoid sinuses. The TNM framework does not provide a standardised staging system for paranasal sinus melanoma. The majority of clinicians divide it into three stages. Stage I is localized tumour, stage II is local tumour with lymph node metastases, and stage III is distant metastasis.[5] Malignant melanoma in the sinuses is treated on a case-to-case basis depending on location and extent.

When there are no distant metastases, the goal is usually to completely resect the tumour. However, due to the proximity of vital structures like the eyes and brain, it is difficult to secure positive margins free of disease in affected sites. Numerous recent studies indicate that radiation therapy may be beneficial following the surgical removal of mucosal melanoma. So even if these tumours have historically been considered resistant to radiation, complete excision of tumour and radiation therapy postoperatively is the current accepted protocol for resectable lesions.[6,7] In recent years, immunotherapy and targeted therapies have been upcoming treatment modalities that have improved the survival rates and outcomes in patients. At more advanced stages, chemotherapy—often including actinomycin D, interferon, and cisplatin—is generally used, primarily for palliative care or when surgical options are not feasible.[2]

These patients require regular follow-ups including radiological evaluations every year to exclude metastasis. In the cases where tumours have unresectable margins or brain involvement, carotid artery encasement, or bilateral optic nerve involvement, the preferred treatment option is palliative care.[5,6,8]

Conclusion

Malignant melanoma is a rare cancer arising from melanocytes in the later stages of life. These tumors are often missed due to late appearance of nonspecific symptoms. Histopathology and immunohistochemistry provide the definitive diagnosis. These patients require timely follow-ups. Through this case we learn that even in the absence of specific symptoms, timely detection of intranasal and oral lesions can help to improve prognosis of sinonasal malignant mucosal melanomas.

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