

Unusual Presentation of Malignant Melanoma in the Nasal Cavity

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Abstract: ***Background:** Nasal cavity melanoma is an extremely rare tumor and more aggressive than its cutaneous counterpart. They occur in elderly patients of both genders and are commonly associated with a poor prognosis. **Aim:** To present a case of primary malignant melanoma of the nasal cavity in a 55-year old female and increase the awareness of the general populace of its presentation and management. **Case Presentation:** A 55-year old female house wife presented with six months' history of gradual, progressive left nasal obstruction and intermittent epistaxis. Three months prior to presentation she noticed dark coloured nasal growth which completely blocked the left nasal cavity with associated persistent mucopurulent nasal discharge. There was no known co-morbidity. Per nasal biopsy was done under local anaesthesia. Microscopy showed large epithelioid and few spindle cells showing hyperchromatic nuclei, macronucleoli and both intra- and extra-cellular melanin pigments that led to a provisional diagnosis of malignant melanoma. She subsequently had lateral Rhinotomy and was managed accordingly. The patient is doing well 8 months post-operatively. **Conclusion:** Primary mucosal malignant melanoma of the nasal cavity is a sporadic, aggressive tumour with poor prognosis and late detection. The clinical and radiographic features of these tumours are nonspecific; therefore, an accurate diagnosis requires histologic assessment. So, early diagnosis with high index of suspicion is essential for the management of the condition.*

Keywords: malignant melanoma, nasal cavity, mucosal

1. Introduction

Malignant Melanomas are tumours arising from melanocytes, which are derived from neuroectoderm located in the basal layers of skin, skin annexes, and more rarely in the mucosal membrane. Common sites for melanomas are head, neck and the lower extremities, less common sites being the oral mucosa, nail beds, conjunctiva, orbit, esophagus, nasal mucosa or nasopharynx, vagina and leptomeninges [1].

Sinonasal malignant melanoma is an extremely rare tumor and more aggressive than its cutaneous counterpart. Lucke recognized it as a distinct clinical entity in 1869 on resecting a "melanotic sarcoma which arose from the nasal mucous membrane" from a man [2]. sinonasal tract mucosal malignant melanomas is rare, accounting for between 0.3% and 2% of all malignant melanomas and about 4% of head and neck melanomas [3-5]. Interestingly, the head and neck represent the most common site of mucosal malignant melanoma [6-8].

Epidemiological studies by Chiu and Weinstock [7] calculated the incidence of oronasal melanoma to be 41/10⁸ population per year. Nasal mucosal melanoma has a poor prognosis. The 5-year survival varies between 25 and 31% [7-9] and the median survival is 21– 24 months [7, 9].

Traditionally patients are treated by radical surgical procedures like lateral rhinotomy, total rhinectomy, maxillectomy and cranio-facial resection [9]. Stammberger *et al.* [10] and Vermeersch and Moerman [11] have pioneered endoscopic management of sinonasal malignancies but it has still not achieved the popularity it deserves [12]. Here we present a rare case of a 55-year old female.

2. Presentation of Case

A 55-year old female house wife presented with six months' history of gradual, progressive left nasal obstruction and intermittent epistaxis. Three months prior to presentation she noticed dark coloured nasal growth which completely blocked the left nasal cavity with associated persistent mucopurulent nasal discharge. No associated history of headache, vomiting, blurring of vision or visual loss, toothache, loosening or loss of teeth. She had no history of cough, yellowness of the eyes, abdominal swelling or bone pains. No known co-morbidity.

Examination revealed a middle-aged woman not in obvious painful or respiratory distress, afebrile not pale anicteric acyanosed. There was fullness in the left cheek with healthy skin. There was flattened dorsum of the nose with a blackish mass occupying the entire left nasal cavity, it bleeds on contact. Septum was slightly deviated to the right and the right nasal cavity appeared normal (Figure 1). Oral cavity/oropharynx showed good oral hygiene and no hard or soft palatal bulge. Other examinations were also normal.

CT scan showed a heterogenous and peripherally enhancing mass in the left nasal cavity and non-enhancing mass in the left maxillary sinus. There was no bony destruction. Per nasal biopsy was done under local anaesthesia.

Macroscopic evaluation of the resection revealed multiple tumour fragments all measuring 4x3x1cm appearing brown-black in colour with soft consistency.

On microscopic examination of the sections for both frozen and permanent sections, diffuse sheets of malignant epithelial tumour cells having hyperchromatic to vesicular nuclei with prominent eosinophilic nucleoli, scanty cytoplasm with abundant melanin pigment were seen. A diagnosis of malignant melanoma of the nasal cavity was established.

Immunohistochemical examination confirmed melanocytic origin of the proliferating tumour cells. Thus, the main marker of melanocytes namely HMB-45, showed diffuse positive cytoplasmic immunostaining was different in various parts of the tumour; emphasizing moderate intensity areas alternating with intensely stained parts. Also, Vimentin Mesenchymal markers and S-100 showed diffuse positive cytoplasmic staining in the tumour. Unlike the other markers, cytoplasmic immunostaining for Melan-A was focally positive in the tumour.

She subsequently had lateral Rhinotomy. Operative findings were black nasal mass arising from the lateral wall of the nose at the anterior portion of the inferior turbinate, inferior and middle meatuses. Clearance biopsy was done. Left antral proof puncture yielded mucopurulent effluent. The microscopy, culture and sensitivity yielded no growth. Patient had intravenous Augmentin, metronidazole and 5% dextrose saline for 24 hours, then antibiotics converted to oral for another six days. She remains symptom free eight months post-operatively.



Figure 1: Black intranasal mass in the left naris

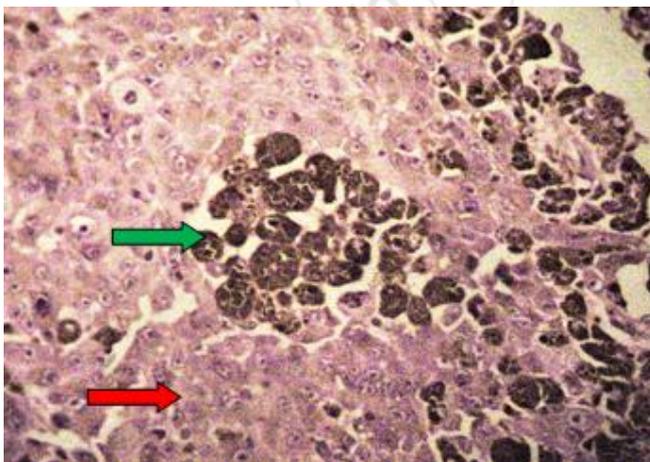


Figure 2: Photomicrograph showing round to oval melanocytes with large nuclei and prominent eosinophilic (cherry red) nucleoli (red arrow) admixed with melanophages (green arrow) (H&E Mag. X 100)

3. Discussion

Mucosal malignant melanoma of the nose and paranasal sinuses is a rare tumour. Holdcraft and Gallagher [13] cited a number of studies documenting the incidence of this lesion. Sinonasal melanoma represented 0.6–0.7% of all

melanomas, 2–9% of melanomas of head and neck and 3.6–4% of all nasal tumours. In the head and neck, the commonest sites for mucosal malignant melanoma are oral cavity (49%), followed by sinonasal involvement (40%) and pharynx (11%) [14]. The commonest site within the nose is the nasal septum, followed by the inferior and middle turbinates [15]; our case was from the inferior turbinate which is a rarer form.

Melanocytes in the mucosa of the upper respiratory tract are the progenitors of the lesion. Formaldehyde exposure and tobacco smoking have been suggested as possible etiological factors [8]. About 5% of nodular melanomas lack pigment (amelanotic melanoma). The tumor occurs between 50- 70 years of age, and both sexes are equally affected with no race predilection [16]. Though, Thompson *et al.* [17] gives an incidence of 11.7% among non-Caucasians.

Malignant melanomas of the nasal cavities and sinuses are characterized by early and repeated recurrences. At presentation, 70-80% of cases are localized, 10-20% have regional lymph node, and <10% have distant metastasis. However, during the course of disease, an additional 20% may develop nodal metastasis and 40-50% may develop distant metastasis to lungs, brain, bone and/or liver [18]. Vascular and neural invasion is seen in 40% cases. However, in our case, the tumor was localized to the left nasal cavity which is similar to the case reported by Medhi *et al.* [16].

Diagnosis of mucosal melanoma is mainly based on histologic findings and immunostain because their microscopic features could be easily misdiagnosed as lymphoma, Rhabdomyosarcoma plasmacytoma, olfactory Neuroblastoma and poorly differentiated carcinoma [19]. They are positive for S-100, vimentin, and specific melanocytes markers such as Melan-A and HMB 45 antigens. These markers were also positive in this index patient. Sometimes the melanin in malignant mucosal melanoma is scanty or absent [20].

Surgery is the cornerstone of therapy [1,21], although wide free margins of resection are difficult to achieve. Local dose escalation with intensity modulated radiotherapy (IMRT) yields good treatment results with respect to local and distant tumor control as well as survival, while treatment-related toxicity can be minimized [22]. Different chemotherapeutic regimens have been tried but all with unsatisfactory results. Local, regional recurrences and distant metastasis still occur despite the implementation of aggressive treatment, including surgery, radiation and adjuvant therapy [23]. Five-year survival rate is between 20 and 46% [24].

Dacarbazine is currently the only chemotherapeutic agent approved for the treatment of advanced disease [25] though other drugs are being researched [26]. Our patient only had surgery because it is a local disease recent studies showed that immunotherapy might have a role in the treatment of malignant melanoma [25]. This is not available in our environment and even if it were to be it will be too expensive for the patient to afford.

Other poor prognostic factors include: advanced age, obstructive symptoms, tumor size >3 cm, location in paranasal sinuses and nasopharynx, vascular invasion into skeletal muscle and bone, high mitotic count, marked cellular pleomorphism and distant metastasis [16].

Local recurrence is a major factor in failure of treatment and is related to several mechanisms, such as incomplete removal (anatomic relationships are complex), multifocal tumor, diffuse submucosal lymphatic spread, transformation of melanocytes at the periphery of the excision into melanoma, failure to remove nodes containing melanoma, and local implantation during surgery [27].

4. Conclusion

Primary mucosal malignant melanoma of the nasal cavity is a sporadic, aggressive tumour with poor prognosis and late detection. They occur in elderly patients of both genders and are commonly associated with a poor prognosis. The natural history is variable, with local recurrence or distant metastasis developing unpredictably. The clinical and radiographic features of these tumours are nonspecific; therefore, an accurate diagnosis requires histologic evaluation. So, early diagnosis with high index of suspicion is essential for the management of the condition.

5. Competing Interests

Authors have declared that no competing interests exist.

Authors' Contributions

All authors contributed accordingly to this work

Consent

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

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