

CASE REPORT

A RARE CASE OF PRIMARY MALIGNANT MELANOMA OF NASAL CAVITY

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ABSTRACT: Malignant melanoma of the nose was first described by Lucke in 1869. The incidence of malignant melanoma in the nose and paranasal sinuses ranges between 0.5% and 1%. Mucosal variant is considered more aggressive with distant metastases and poor prognosis. Primary malignant melanoma of nasal cavity is a very rare entity presenting with nasal obstruction and epistaxis. Surgical excision is considered the main stay of treatment,¹ and it has been found to have a high chance of recurrence. In this paper we present a case a primary malignant melanoma of nasal cavity in a mid-aged male patient who presented with nasal obstruction and nasal mass.

KEYWORDS: Malignant melanoma, Nasal cavity.

INTRODUCTION: Malignant melanoma is an aggressive melanocytic neoplasm, usually affecting the skin.² Primary malignant melanoma of the nasal cavity is rarely seen. Clinically, most patients display nonspecific symptoms of unilateral nasal obstruction or epistaxis. The prognosis is generally poor,³ with a mean survival time of 3.5 years. Extensive local invasion and distant metastasis to other organs may occur. The usual treatment of choice is radical excision with wide free margins. Radiotherapy and chemotherapy appear to have little effect.⁴ We report a case of intranasal cavity malignant melanoma in which the patient initially presented with unilateral nasal obstruction and nasal bleed. Pre-operative metastatic lesions were not found.

CASE REPORT: We report a 40 year old male patient who presented with complaint of nasal obstruction and nasal mass in right nasal cavity for 2 months and a single episode of nasal bleeding which was moderate in quantity and stopped spontaneously. Patient is a known case of smoker and alcoholic for past 20 years.

Physical examination revealed fullness in the dorsum of nose more on right side with a blackish mass in the right nasal cavity. Mass felt rubbery in consistency on palpation and was sensitive to touch and non-tender. There was bleeding on probing and was able to probe all around for about a centimeter except the floor and lateral wall. Posterior rhinoscopy showed no discharge or mass extending into the choana. No metastatic lymph nodes were palpable. USG abdomen showed renal cysts and no other mass lesions.

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Fig. 1: Picture of the patient showing the presence of a grayish black mass in the right nasal cavity with bleeding.



Fig. 1

Biopsy was taken and sent for HPE. CT scan of nose and PNS showed a mass in right nasal cavity with no evidence of bony erosion. HPE showed tumour cells predominantly epithelioid variant arranged in nests and sheets. Tumour cells were pleomorphic with increased N:C ratio, vesicular to hyperchromatic nuclei with prominent eosinophilic nucleoli, also binucleated and bizarre form of tumour cells seen. Heavy melanin pigment deposition noted inside and outside the tumour cells. Suggestive of malignant melanoma. Patient was subjected to wide tumour excision via nasal endoscopy because of its localized tumour extent. Specimen sent for HPE also confirmed the diagnosis. The patient had an uneventful post-operative period. He was followed for 6 months post-surgery and showed no signs of recurrence of the tumour.

Fig. 2: Endoscopic image showing the mass arising from the right nasal cavity involving floor and the septum.



Fig. 2

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Fig. 3 & 4: Endoscopic view of the right nasal cavity following a wide local excision with free margins.

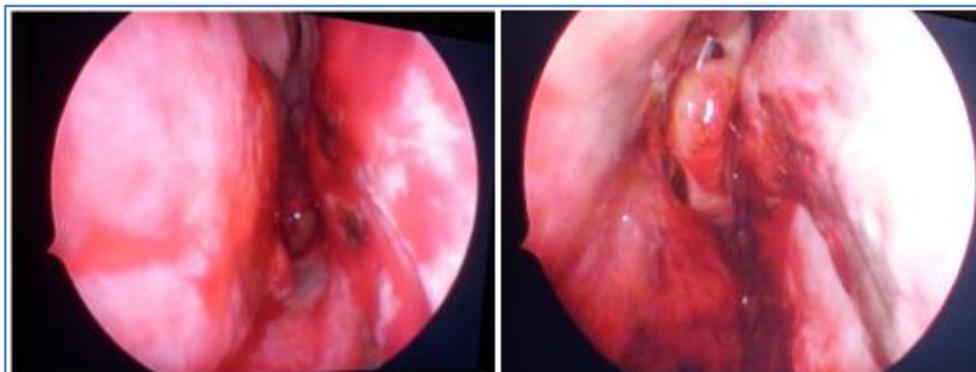


Fig. 3 & 4

DISCUSSION: A primary intranasal cavity melanoma is a rare and usually lethal disease. The etiologic and pathologic basis of the disease is not yet fully understood. In 1974, Zak and Lawson reported the presence of dendritic melanocytes in the epithelium of the sinonasal region. In 1979, Cove presented a case of a malignant primary multifocal intranasal melanoma arising from a preexisting nasal and maxillary sinus melanosis. Nevertheless, little is known about premalignant melanocytic lesions in the nose. The role of smoking or sun exposure as an etiology for this tumor remains controversial.

An intranasal melanoma usually manifests as a solitary lesion rather than multiple foci. The anatomic distribution includes the nasal cavity, turbinates, the nasal wall, antrum, ethmoid sinus, vestibules, frontal sinus, and maxillary sinus. Among these locations, the most commonly reported site is the nasal cavity, followed by the maxillary sinus. In the nasal cavity, the most common sites are the nasal septum (41%), middle turbinate (29%), inferior turbinate (23%), lateral nasal wall (7%), and nasal floor (1%). Most nasal melanomas appear as pigmented, polypoid, fleshy and friable masses, and thus might be initially diagnosed as benign lesions. The precise site of tumor origin is occasionally difficult to identify due to the large size of the tumor and the extensive local destruction it causes.

Clinically, most patients present with symptoms of epistaxis, unilateral nasal obstruction and congestion, and pain and swelling of the cheek and nose.

Diagnosis of mucosal melanomas requires high index of suspicion. Complete history with head and neck examination including diagnostic nasal endoscopy should be done. Histologic findings and immunostain findings are essential in making the diagnosis due to its rare nature and microscopic similarities with certain other conditions like lymphoma, rhabdomyosarcoma, plasmacytoma, olfactory neuroblastoma, and poorly differentiated carcinoma that can lead to misdiagnosis. Mucosal melanomas show immunohistochemistry positive for S100 protein, vimentin, and specific melanocytic markers like Melan-A and HMB45 antigens. On magnetic resonance imaging the tumour shows abundance of melanin which detected by, hyperintensity on T1-weighted imaging and hypointensity on T2-weighted imaging. However, sometimes, the melanin in malignant mucosal melanoma may be scanty or not detectable.⁷

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Mucosal melanomas mostly show early vascular and lymphatic invasion, multiple metastasis and satellite formation, which eventually lead to recurrence locally. If there is local recurrence it is associated with poor prognosis as there is increased risk of distant metastasis. Poor prognostic factors include advanced age, local and distant metastasis, tumor size >3cm, vascular invasion, high mitotic count, cells showed marked pleomorphism and delay in detection, inaccurate diagnosis.^{5,6,7}

The most fundamental treatment is wide resection of the primary tumor whenever possible. Surgery provides the best chance of controlling the disease. Radiotherapy combined with surgery is recommended in cases of local recurrence or incomplete lesion removal. Optimal radiation doses remain uncertain usually high doses of 50-55 Gy in 15 or 16 daily fractions over 21 days. DTIC, whose efficacy is still controversial.

Distant widespread metastases to the liver, lungs, and brain, and regional metastasis to subcutaneous tissues are the major causes of death in most cases. The 5-year survival rate is estimated to be less than 40%.

CONCLUSION: Mucosal malignant melanoma (MMM) of the upper aero digestive tract (UADT) has an incidence of 0.5-3% amongst malignant melanomas of all sites. They are more common in men as compared to women.⁸ Mucosal melanomas are more aggressive than cutaneous melanomas and have a poorer outcome. Primary malignant melanoma of nasal cavity is one rare disease which requires complete evaluation because of its aggressive nature and high chances of local and distant metastases and poor detection rates. Once diagnosis is confirmed with HPE and confirmed about no distant metastasis is noted, as seen in the case that we have presented, endoscopic excision with wide free margins is found to be adequate with less chance of recurrence. Primary mucosal malignant melanoma of the nasal cavity has poor prognosis. So, early diagnosis with high index of suspicion, adequate treatment and long term regular follow up is of utmost importance for the management of the disease.

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