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Case Report

Primary mucosal malignant melanoma of the nasal cavity

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ABSTRACT

Primary mucosal melanoma of the nasal cavity and paranasal sinuses is a very aggressive and rare disease with only about 0.5 % of malignant melanoma arising from the nasal cavity. There are only few reports from India. We report a rare case of sino-nasal mucosal malignant melanoma in a 58 years old female who presented with blackish coloured sino-nasal mass involving right nasal cavity, spontaneous and recurrent epistaxis and obliteration of the right naso-labial fold with occasional pain in the past 10 months. Contrast enhanced computed tomography scan showed a heterogenous mass involving right nasal cavity, right maxillary antrum and right ethmoidal area. A positron emission tomography computed tomography was also done which showed increased uptake in the region mentioned above. Initial biopsy, the mass was diagnosed as malignant melanoma. Total maxillectomy was performed with plan of post-operative radiotherapy.

Keywords: Malignant melanoma, Nasal cavity, Nose

INTRODUCTION

Sino nasal malignant melanoma, first described by Lucke in 1869, is a rare, aggressive tumour with poor prognosis and frequently delayed diagnosis.[1] The first case of malignant melanoma was reported in India by Kutty and Shreedharan in 1965.[2] It accounts for 0.5-2% of all melanomas.[3] The risk factors are not well known. However, the symptoms develop slowly thus causing a delay in diagnosis ultimately leading to a very poor prognosis. The risk of local recurrence and distant metastasis is very high.[4] The treatment of choice is surgical resection, while radiotherapy and chemotherapy remain a tool to control local and metastatic disease.

CASE REPORT

A 58 years old female presented with a swelling on the nose blocking the right nasal cavity and obliterating the right naso-labial groove. She complained that 9 months back, it started with symptoms of nasal obstruction and intermittent spontaneous epistaxis which was controlled with medications. However, during its course there was a noticeable swelling in the right nasal cavity with blackish discolouration with intermittent episodes of throbbing pain.

On examination there was a blackish ulcerated mass in the right nasal cavity obliterating the right nasolabial groove and pushing the nasal septum completely to the opposite side touching the left lateral nasal wall. However, there was no ulceration of the local skin nor there were any lymph nodes palpable. Neither the Occupational history, environmental exposure history was significant nor there any history of major illness in the past.

Routine haematological and biochemical investigations were within normal limits. Contrast enhanced computed

tomography (CECT) scan of nose and paranasal sinuses revealed a heterogenous mass involving the whole of the right nasal cavity, right maxillary antrum and right anterior ethmoids with few areas of destruction in the medial maxillary wall. Further positron emission tomography (PET) computed tomography scan was done to rule out any lymph node or organ metastasis and it revealed increased uptake in the right nasal cavity and right anterior ethmoids only. Initial biopsy was already done by the patient from outside when patient presented to us and it revealed malignant melanoma.

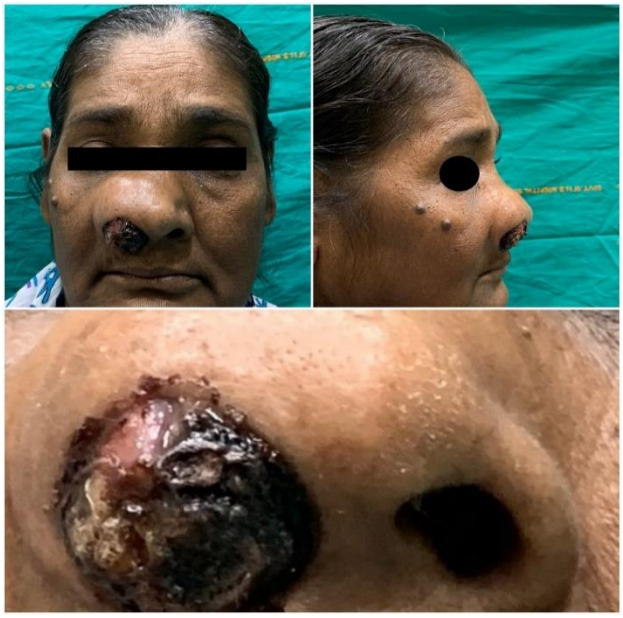

Figure 1: Pre-operative picture of the mass.

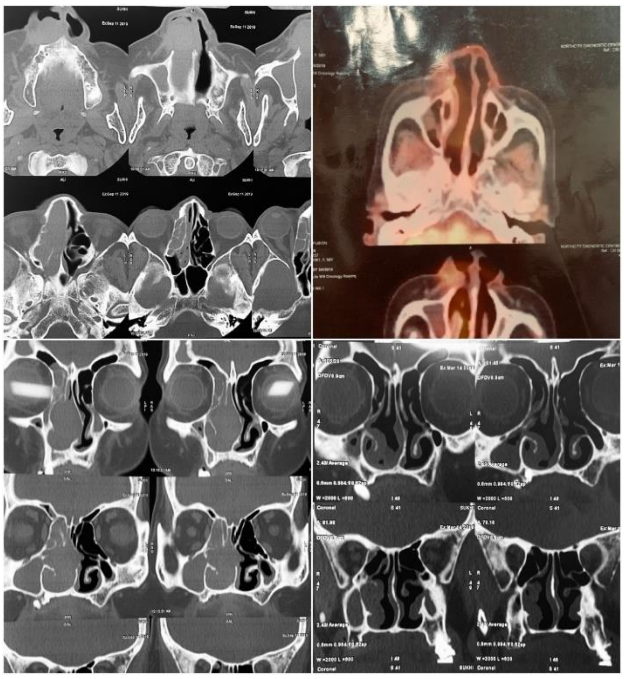

Figure 2: CECT scan and PET scan showing the mass with uptake of FDG.

Total maxillectomy was performed with anterior and posterior ethmoidectomy and the specimen was sent for Histopathological examination which confirmed the diagnosis of malignant melanoma showing tumour cells containing melanin and subsequently concurred by immunohistochemistry by showing positivity in S-100. Patient is currently on follow-up with department of otolaryngology, radiotherapy and medical oncology and is doing well so far.

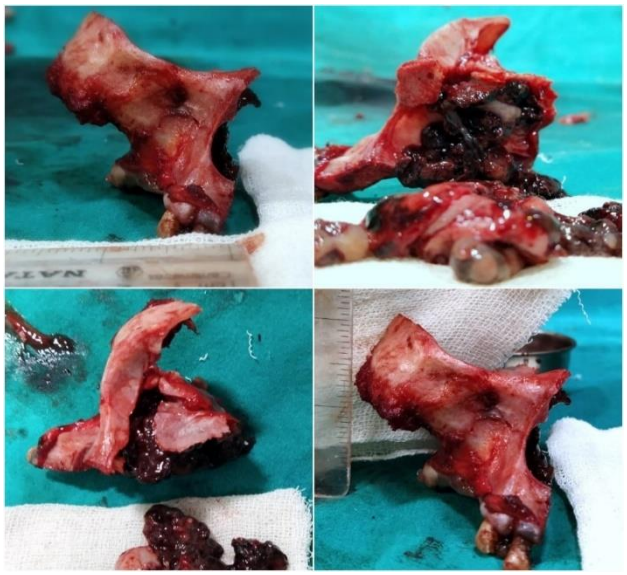

Figure 3: Specimen of right maxilla with blackish mass.

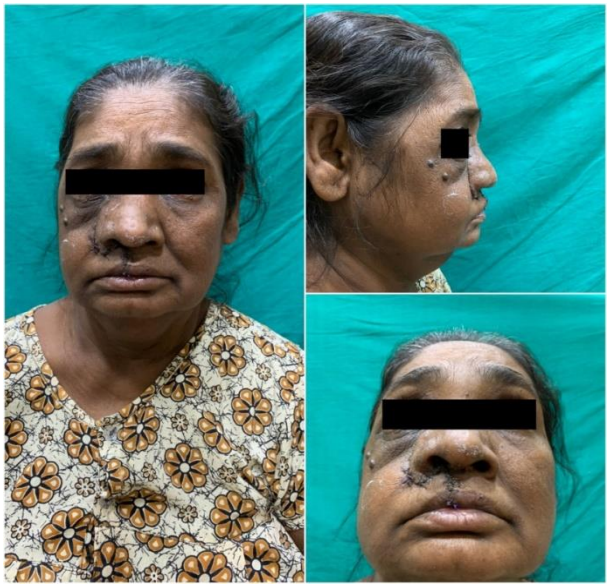

Figure 4: Post-operative image of the patient.

DISCUSSION

Primary mucosal melanoma corresponds to 0.5 to 2% of all malignant melanoma. It has no known risk factors unlike its cutaneous counterpart where sun exposure is

considered to be a risk factor.[5] The mean age of presentation of mucosal melanoma is generally in the 6th decade and it is thought to happen more in white people with a male: female ratio of 2:1.[6,7]

Etiopathogenesis is unclear but it is believed melanomas are believed to arise from melanocytes in the mucosal tissue which originate from cell migration from neural crest during embryonic period.[8]

The primary prevalent symptom of sino nasal mucosal melanoma is unilateral nasal obstruction, nasal congestion and bleeding. Other associated symptoms are nasal discharge, swelling and deformity of nose with headache. Duration of symptoms is variable between 3-24 months.[9] The Ballantyne's classification does not take tumour size, histology or local extension into account.

Table 1: Classification of malignant melanoma.

Ballantyne's classification	
Stage I	Tumour confined to original site
Stage II	Tumour with regional lymph node metastasis
Stage III	Tumour with distant metastasis

Table 2: 7th edition of American joint committee on cancer (AJCC) classification.

AJCC classification	
Primary tumour (T)	
T3	Mucosal disease
T4a	Moderately advanced disease; tumour involving deep soft tissue, cartilage, bone or overlying skin
T4b	Very advanced disease; tumour involving brain, dura, skull base, lower cranial nerves (IX, X, XI, XII), masticator space, carotid artery, prevertebral space or mediastinal structures.
Regional lymph nodes (N)	
Nx	Regional lymph nodes cannot be assessed.
N0	No regional lymph node metastasis.
N1	Regional lymph node metastasis present.
Metastasis (M)	
M0	No distant metastasis
M1	Distant metastasis present
Staging	
Stage III	T3, N0, M0
Stage 1VA	T4a, N0, M0 T3-T4a, N1, M0
Stage 1VB	T4b, any N, M0
Stage IVC	Any T, any N, M1

Diagnosis includes clinical examination, diagnostic nasal endoscopy, CECT scan which will show a heterogeneous mass. 3D reconstruction can be useful if facial reconstruction is planned pre-operatively. MRI is also useful in better delineation of the mass for example, T2 weighted MRI scan can distinguish fluid retention of paranasal sinus from the mass. Also, MRI is better in cases when orbit is involved. And finally, PET-CT scan is essential to detect any local metastasis or distant metastasis. In this case PET-CT was done and the uptake was only seen in the right nasal cavity and right maxillary antrum. No local uptake of any regional lymph nodes was noted.

Due to severe aggressiveness of disease T1 and T2 are not comprised in this classification. The recent 8th edition of AJCC has no changes in the classification of sino nasal melanoma.

The 1st line of treatment is surgical resection along with lymph node dissection but prophylactic neck node dissection at N0 is not recommended since it has been seen that the presence of occult node metastasis is relatively low.[10,11] Local recurrence occurs despite surgery in around 50% cases.[12]

Radiotherapy is recommended to prevent recurrence but doesn't guarantee remission. Chemotherapy is mostly used in advanced stages or when distal metastasis is present.[13] Cisplatin, interferon-α, actinomycin-D are commonly used.

Confirmation of diagnosis is ultimately dependant on Histopathology and Immunohistochemistry. Grossly, the tumour varies from small to large mass. High nuclear cytoplasmic ratio with hyperchromatic nuclei and large nucleoli along with pigment melanin in both cytoplasm and background are seen in cytology. In order to differentiate melanin from other pigments like haemosiderin, Prussian blue and Mason Fontana silver stain are both used to bring out the melanin pigment more clearly. Immunohistochemical markers include S-100, HMB-45, vimentin, cytokeratin etc.

Table 3: Prognosis.

Prognosis	Good	Bad
Age	<50 years[16]	>60 years
Tumour size	<3 cm	>3-4 cms[17]
Area	Isolated septal tumour	Tumours in paranasal sinus
Invasion	Not invading muscle or bone	Invasion into skeletal muscle and bone
Metastasis	No distant metastasis	Presence of distant metastasis

The 5 years overall survival of this disease is 20-40% with mean survival of 17-28 months. Local recurrence occurs in 50% cases.[14] The high rate of recurrence is mainly due to: vascular invasion, submucosal lymphatic spread, multifocal nature, inadequate first line surgical resection, presence of distal metastasis, and common sites of metastasis are lungs, liver, bone. Metastasis are found in 40-76 % of cases.[15]

CONCLUSION

Primary mucosal malignant melanoma is an extremely rare clinical entity especially in the Indian sub-continent. Early diagnosis by biopsy followed by histopathology and immunohistochemistry is essential for better prognosis. Wide surgical excision is preferred with adjuvant radiotherapy to prevent local recurrence however recurrence rate is high and prognosis is poor according to the current data. Gene therapy could be a way to improve the standard of treatment in near future.

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