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CASE REPORT

Malignant Melanoma of Maxillary Sinus: A Rare Entity

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ABSTRACT

Malignant melanoma of the nose and paranasal sinuses is an aggressive disease, typically presenting at an advanced stage, with a 5-year survival rate ranging between 20 and 30%. Melanomas are tumours arising from melanocytes which are neuroectodermally derived cells located in the basal layers of the skin, adnexa and some of the mucosal membranes. Malignant melanomas developing from maxillary sinuses are extremely rare. Their diagnosis can be confirmed by immunohistochemistry by using anti-S100 and HMB-45 antibodies.

Key Words: Malignant melanoma, Maxillary sinus, HMB-45

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Introduction

Malignant melanoma is a relatively rare malignancy and constitutes approximately about 1-2 % of all malignancies arising in the body. 90% of these occur in the skin. Primary malignant melanoma arising in nasal and paranasal sinuses are rare[1], accounting for less than 1% of all melanomas and have a poor prognosis. In 1965, Kully and Shreedharan[2] reported the first case of melanoma in India. Ravid and Esteeves[3] reported that Lucke in 1869, operated on a 52 year old man suffering from melanotic sarcoma of the nasal mucosa. The first case in American literature was reported by Lincoln in 1885. One third of these melanomas are usually amelanotic lesions. Malignant melanoma of the nose and paranasal sinuses is rare. It accounts for less than 1% of all malignant melanomas and less than 3% of all nasal malignancies. It is also rare for melanoma to metastasize to this anatomical site .The tumour mostly occurs in the 4th decade of life. Clinically, the patients present with complaints of nasal mass, bloody discharge, obstruction and rhinorrhoea. Grossly, melanoma can present as a firm, gray-white or pink-to-black, ulcerated mass. Black colouration is a rarity, and its absence does not rule out melanoma. Histologically, melanoma is variable in appearance and it has been said that it may look like anything; therefore, it is in the differential diagnosis of almost everything. It is identified because of its junctional activity, prominent melanin pigmentation, marked cytological atypia, nuclear grooves, folds and pseudoinclusions, large eosinophilic nucleoli and abundant mitotic figures. The cells can be epithelioid, spindle shaped or extremely bizarre. Their size can range from small (lymphocyte like) to that of giant multinucleated forms. The cytoplasm can be eosinophilic, basophilic, foamy, of the signet ring type, oncocytic, or completely clear (balloon cell melanoma). Melanin can be abundant, scanty, or absent (amelanotic melanoma). Immunoperoxidase studies are extremely useful and include the use of HMB-45, S100, Melan A, and one of the newest diagnostic markers, pigment epithelium–derived factor (PEDF). Immunohistochemically, sinonasal

malignant melanomas are positive for S100 and markers for melanomas including HMB45, Mart-1, and tyrosinase. Most of them do not produce significant amounts of melanin. P16 is expressed in a significant number of these tumours and is mainly related to the deletion of the 9p21 region.

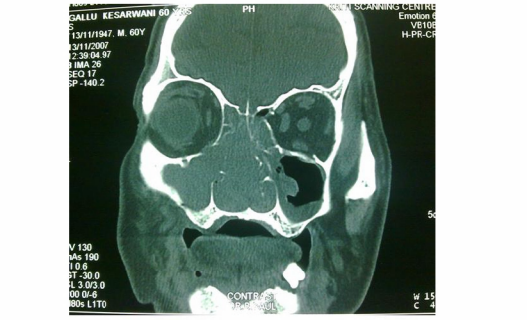
A case of malignant melanoma of the maxillary sinus is documented here, due to its rarity.

Case Report

A 60-year-old male presented with complaints of a mass in the nose, nasal blockage, nasal discharge, epistaxis and fullness over the left cheek. The symptoms increased rapidly over a period of two months. On clinical examination, a fleshy, pinkish white, firm and tender mass was present in the left nasal cavity, which was completely blocking the nasal passage and bled on touch. The left ala was stretched out. The septum was markedly deviated to the right side by the mass. On posterior rhinoscopy, a whitish growth was seen in the nasopharynx. Examination of the throat, ears and larynx were normal.

The routine haematological and biochemical investigations were within normal limits. CT scan revealed opacification of the left maxillary sinus by a nonspecific hypodense, mildly enhancing soft tissue mass, extending into the middle meatus and bulging into the nasopharynx. The mass displaced the nasal septum to the right side, thus thinning it. The left ethmoid, frontal and bilateral sphenoid sinuses were opacified by hypodense nonenhancing soft tissue [Table/Fig 1].

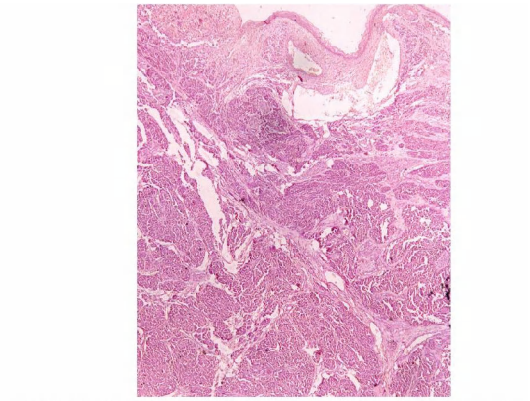
Left maxillectomy was performed and the tumour was removed piece-meal and sent for histopathological examination.



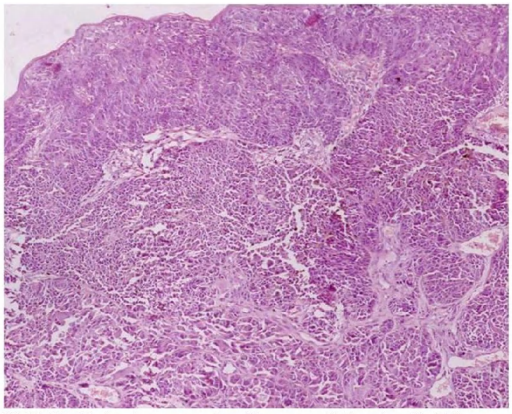
[Table/Fig 1]CT scan showing opacification of left maxillary sinus by nonspecific hypodense soft tissue mass.

Histopathological Examination

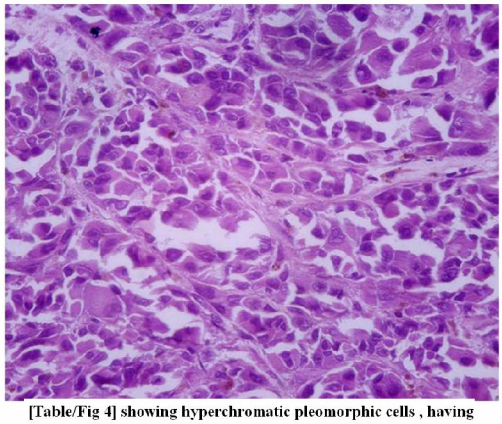
On gross examination, multiple, irregular, friable, dark brown pieces of tissue admixed with few bony pieces ranging from 6x4 to1x1 cm in size were obtained. The cut surface was brown. Histologically, the biopsy material showed stratified squamous epithelium, fibrovascular tissue and tiny pieces of bone and mucous glands [Table/Fig 2],[Table/Fig 3]. Aggregates of cells arranged in alveolar and diffuse patterns were present. The cells were moderately pleomorphic, showing anisokaryosis, round to oval nuclei, vesicular chromatin, prominent nucleoli and abundant cytoplasm. Some of these cells had eccentric nuclei (plasmacytoid) [Table/fig 4].There was evidence of an intracellular and extracellular dark brown pigment, which was later confirmed to be melanin, by S100 and HMB45 antibodies [Table/Fig 5], [Table/Fig 6].



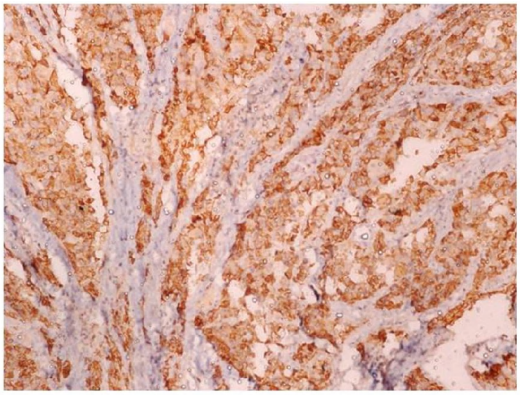
[Table/Fig 2]showing stratified squamous epithelium and tumour cells arranged in alveolar pattern, separated by fibrovascular tissue (HE X 40)



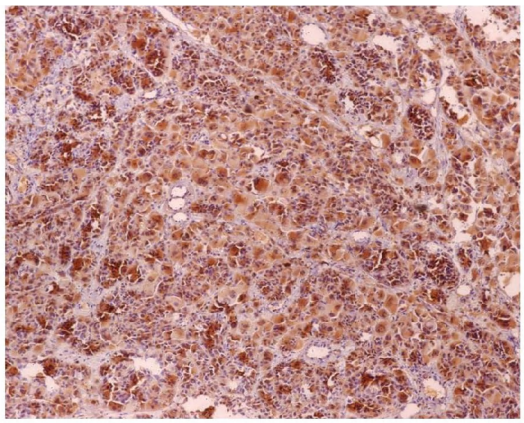
[Table/Fig 3] showing aggregates of tumour cells in sub epithelial zone reaching upto epithelial layer (HE X 40)



[Table/Fig 4] showing hyperchromatic pleomorphic cells , having plasmacytoid appearance (HE X400)



[Table/Fig 5] showing HMB 45 positivity(IHCX100)



[Table/Fig 6] showing S100 positivity (IHCX100)

Discussion

The present case was diagnosed as malignant melanoma on light microscopy, which was later confirmed by immunohistochemistry. Before immunohistochemistry, more commonly occurring neoplasms of the nose and nasopharynx like lymphoma, sinonasal undifferentiated (anaplastic) carcinoma, olfactory neuroblastoma (esthesioblastoma) and small-cell neuroendocrine carcinoma, were also considered. However, the pattern of cell arrangement, morphology and immunohistochemistry helped in ruling out these possibilities.

In the present case, due to the advanced age of the patient, lymphoma was the nearest possible diagnosis but it was ruled out due to lack of the monomorphic appearance of cells in comparison to the present case which showed marked pleomorphism and positivity for HMB-45 and S-100 on immunohistochemistry.

Sinonasal undifferentiated (anaplastic) carcinoma usually has medium sized cells arranged in nests, lobules, trabeculae and sheets having a high mitotic rate, extensive necrosis and prominent vascular invasion, while in the present case, cells were arranged in an alveolar pattern, showing pleomorphism and had moderate cytoplasm. Sinonasal undifferentiated carcinoma is usually positive for cytokeratin and epithelial membrane antigen, but is negative

for S-100, but in our case, S-100 was positive.

Olfactory neuroblastoma (esthesioblastoma) was also considered in the differential diagnosis due to its site of occurrence, but it was ruled out due to the absence of the Homer Wright rosette or Flexner-Wintersteiner rosettes, morphology, pattern of cells, prominent fibrillary or reticular background and absence of positive staining for synaptophysin.

Small-cell neuroendocrine carcinoma was considered in the differential diagnosis due to age, site and clinical presentation (epistaxis, proptosis, and obstructive phenomenon), as in malignant melanoma. Small-cell neuroendocrine carcinoma was ruled out morphologically due to the absence of small hyperchromatic cells.

Conclusion

The present case highlighted the importance of malignant melanoma in differential diagnoses of tumours occurring in the nose and paranasal sinuses, as it has a very aggressive behaviour and a poor prognosis. Immunohistochemistry is also helpful in further confirming the diagnosis, especially in amelanotic melanoma and in ruling out other tumours of the nose and paranasal sinuses.

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