Research Article

Malignant Mucosal Melanoma of Nasal Cavity (melanic variant)

Laxmi Narayan Garg1, Nasib chand2, Bhavna Gulati2, Palak Mahajan2,
Hinna Bansal2 and Subash Goel3

1Department of ENT, 2Department of Pathology, 3Department of Surgery
MMIMS and R Mullan, Ambala (HR), India
Corresponding Author; nasib_chand2004@yahoo.com

[Received-16/03/2015, Accepted-24/03/2015]

ABSTRACT

Malignant melanoma of nasal mucosa and paranasal sinuses is a very rare tumor, which can be melanin producing or
non melanin producing ie amelanotic variant. Rhinosinusal mucosal melanoma constitutes less than 1% of all
melanic tumors and 2-8% of overall cancers developing in nasal fossae and paranasal sinuses. Patient gets medical
attention very late in the course of disease due to ignorance of early mild/nonspecific symptoms in the way of high
degree of local and distant invasion by the disease, and this finally leads to poor prognosis. Studies show average 5
year survival as 20-30% for cutaneous and 10-15% for mucosal melanomas. Here, we present a case of 60 yrs old
male presenting with right nasal mass associated with off and on nasal bleeding. On suspicion of malignancy he was
investigated, confirmed by Biomarkers study and operated after final diagnosis, Later, patient was referred to
Radiation oncology for appropriate treatment.

Key words; malignant melanoma, Rhinosinusal, mucosal, Immunohistochemistry.

INTRODUCTION

Mucosal malignant melanoma of nose is extremely rare and unpredictable presentation in
Otorhinolaryngeal practice. First case of nasal melanoma was reported in1869 by Lucke and
since then, less than 1% of melanic tumors has been reported and only 2-8% of all neoplasms
developed in the nasal fossae and sinuses.1 Nasal/paranasal melanoma is a rare tumor and
has a poor prognosis.2 Primary melanoma of head and neck comprises 25-30% of all melanomas.3

Aerodigestive mucosal melanoma (0.4-4%) mostly occur in the nasal cavity or paranasal
sinuses (maxillary >ethmoid sinuses).4,5 Peak incidence is between 5th and 8th decade of life.
Males are more commonly involved than females. Age and sex in particular, do not affect
prognosis.6,7 Tumor shows aggressive behavior and average five year survival is only 20-30% in
case of cutaneous and 10-15% for mucosal melanomas.
CLINICAL HISTORY
A 60 years old patient presented with six month history of unilateral right sided nasal reddish brown mass (Figure 1) causing right nasal obstruction and recurrent off and on bleeding, with H/o smoking. On anterior Rhinoscopy, a mass was visualized in the right nasal cavity, excised surgically and sent for Histopathological examination (HPE).

DISCUSSION
Melanomas are malignant tumors, that arise from melanocytic proliferation, which in turn derive from neuroectodermal cells, located in the basal layer of skin and mucosa. 80% of melanomas arise from skin and remaining 20% in the head & neck region and other sun exposed areas of the body respectively. Smoking is also thought to be activating factor for melanocytic changes like melanogenic metaplasia. Mucosal melanoma of nasal cavity is rarely encountered lesion and constitutes 1.3% of all malignant melanomas. The tumor carries very poor prognosis, because of local recurrence and early distant metastasis via both lymphatic (5-15%) and other pathways. Commonest site of metastasis is submandibular lymph nodes. Although, there is no accepted staging system (TNM) for nasal/sinusal melanomas, but clinically, we have three stage classification as;

- Stage1 primary tumor without metastases;
- Stage2 primary tumor with loco-regional node metastasis;
- Stage3 primary tumor with distant metastases.

American armed forces institute of pathology, proposed a staging system for nasosinusal and nasopharyngeal melanoma in 2003, stated as;

T1; primary tumor at one anatomical site.

T2; primary tumor at two or more sites.

N1; any lymph node metastasis and M1- any distant metastasis.

Overall prognosis depends on time at which, patient presents, is also important. It also depends on size and location of tumor, as in our case, patient mainly presented with off and on epistaxis and difficulty in breathing due to progressive nasal obstruction. Unfortunately, the tumor is usually well advanced at time of presentation and also shows recurrences, lymph node involvement and distant metastases respectively. As far as differential diagnosis is concerned, tumor must be differentiated from round cell tumors (benign, intermediate or malignant), Benign tumors like( nasal polyps, osteoma, chondroma, schwannoma, neurofibroma and cementoma ), Intermediate tumors including, inverted papilloma, meningioma, hemangioma, and hemangiopericytoma. The malignant tumors include Squamous cell carcinoma (most common), followed by adenoid cystic carcinoma, adenocarcinoma, neuroblastoma, retiform-cell sarcoma, lymphoma, and undifferentiated small cell carcinoma. HPE is the final tool for correct and definitive diagnosis. Diagnosis of malignant melanoma is challenging particularly, when the tumor shows melanin (high or low) positivity ie (melanic/amelanic variants).

After H&E section examination, the diagnosis is made and further confirmed by Immunohistochemical markers study like S-100 specifically , HMB-45 monoclonal antibody derived (cytoplasmic positivity in 65 – 95% of cases), particularly for primary and metastatic melanomas including amelanotic M, spindle cell M, and acral lentigenous M. It can also be seen in signet ring cell M, myxoid M, small cell M, balloon cell M, and other different anatomical sites melanomas such as Gall bladder, Urinary bladder, anorectal, vulva, sinus region , uterine cervix and other than mucosal sites melanomas such as, bones and Pulmonary blastomas respectively. Other tumor markers are Melan- A
Malignant Mucosal Melanoma of Nasal Cavity (melanic variant)

(very specific for differentiation from sarcoma, plasmacytoma and carcinosarcomas), Vimentin (mesenchymal), Cyclin-D (nuclear), tyrosine and CD 44 (membranous) positivity for tumor cells. In our case also, the excised mass was processed and examined by light microscopy. HPE shows ulcerated surface epithelium, having underlying tumor composed of sheets and clumps of large polygonal cells with abundant cytoplasm, showing little melanin and large hyperchromatic vesicular nuclei along with many macronuclei are seen. Fare number of mitotic figures seen (figure-3) Atypical and typical pseudoglandular pattern was also evident at places. Masson Fontana stain shows red colored nuclei and brownish black cytoplasmic melanin granules (Figure-4). Immunohistochemistry (S-100) shows cytoplasmic pigment (Figure-5)

CONCLUSION
While dealing with a tumor mass arising from nasal mucosa especially, when it is associated with nasal bleeding/or obstruction, is very distressing to patient. A point of suspicion arises regarding diagnosis, treatment and it’s future outcome in terms of prognosis. Clinical diagnosis is sometime evident and is also supported by Endoscopic examination as well as C T Scan. The final diagnosis is based on Histopathological examination (gold standard), which is further confirmed by Immunohistochemical techniques (IHC) including study of Biomarkers of high specificity for malignant melanoma such as S-100, HMB-45 and Melan-A respectively. So early diagnosis and wide Surgical excision followed by postoperative Radiotherapy can only improve survival, Inspite of local recurrence, nodal and distant organ metastasis. Other poor prognostic factors include advanced age, obstructive symptoms of nasopharynx and paranasal sinuses, vascular invasion into liver and skeletal muscles, cellular pleomorphism and high mitotic index respectively.

REFERENCES;
Malignant Mucosal Melanoma of Nasal Cavity (melanic variant)

13 Ciolofan S, Ionita E, Mogoanta CA, Popescu FC, Anghelina F, Chiutu L and Stanciu G.

Figures:

Figure; 1- Clinical photograph showing brownish mass in the Right nasal fossa.

Figure; 2- H&E stained section showing mucosa & sheets of malignant melanocytes having macronuclei (10X).
Malignant Mucosal Melanoma of Nasal Cavity (melanic variant)

**Figure 3** - Section showing sheets of malignant melanocytes (40X).

**Figure 4** - Section showing red nuclei and cytoplasmic brown black pigment (Masson Fontana stain, 40X).

**Figure 5** - Immunohistochemical stain (S-100) showing cytoplasmic positivity (40X).