

# CASE REPORT

## MUCOSAL MALIGNANT MELANOMA OF NASOPHARYNX: A CASE REPORT

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**ABSTRACT:** Primary mucosal malignant melanomas of sinonasal tract are uncommon tumors comprising 0.3-2% of all malignant melanomas and 4% of all head and Neck melanomas. We are reporting a rare case of mucosal malignant melanoma in a 45 year old female arising from nasopharynx which was excised completely by trans palatal approach followed by irradiation. This case is being reported because of its isolated involvement of nasopharynx, and early age of presentation.

**KEYWORDS:** Malignant Melanoma, Sino nasal tract.

**INTRODUCTION:** Primary mucosal malignant melanoma of the sinonasal tract is an uncommon tumor comprising 0.3-2% of all malignant melanomas and about 4% of head and neck melanomas.<sup>1</sup> In the head and neck region sinonasal tract is the common site and forms about 4% of all sinonasal tract neoplasms.<sup>2</sup> These originate from melanocytes present in the mucosal lining of the respiratory tract. These are known to behave more aggressive and have poor prognosis due to occurrence in occult sites together with lack of early and specific signs contributing to late diagnosis, cervical lymphadenopathy and distant metastasis. Because of rare occurrence and ill-defined pathogenesis, staging and treatment protocols of mucosal melanomas are not well established.

**CASE REPORT:** A 45yrs old female presented to E.N.T OPD, S. V. R. R. Hospital, Tirupati with nasal bleed, head ache and pigmentation of left eye for the past 3 months. Nasal bleed was spontaneous, intermittent and profuse not associated with nasal obstruction. She noticed pigmentation of left eye incidentally, without any visual disturbances. On examination pigmentation of left lower palpebral and bulbar conjunctiva was noted (Fig. 1). Nasal examination revealed bilateral inferior turbinate hypertrophy without any evidence of mass and nasal endoscopy showed an irregular pigmented mass arising from roof and left lateral wall of nasopharynx. Clinically there were no palpable cervical lymph nodes.

C.T scan of paranasal sinuses revealed irregular homogenous mass lesion involving nasopharynx and bilateral inferior turbinate hypertrophy. (Fig. 2, 3) She was subjected to Diagnostic nasal endoscopy and biopsy was taken. Histopathology of the section showed mucosal malignant melanoma. (Fig. 4) Ultrasound neck and C.T abdomen was done to rule out regional and distant metastasis. Under general anaesthesia the mass was excised by Wilson's transpalatal approach and later subjected to irradiation. We followed the patient for 10months, during this period there was no recurrence.

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**DISCUSSION:** Primary mucosal malignant melanoma of the sinonasal tract is an uncommon tumor comprising 0.3-2% of all malignant melanomas and about 4% of head and neck melanomas.<sup>1</sup> In the head and neck region sinonasal tract is the common site and form about 4% of all sinonasal tract neoplasms.<sup>2</sup> These tumors occur in the seventh decade of life and predominantly affects Caucasians.<sup>3</sup> Unlike age and race, it would appear that sex is less significant with regard to the development of mucosal malignant melanoma.<sup>3</sup> These tumors originate from the neural crest derived melanocytes present in the mucosa and stroma of the respiratory tract. Risk factors for the development of mucosal melanomas have not been identified, exposure to formaldehyde was a suggested risk factor.<sup>4</sup> Recently genetic studies revealed that different melanoma subtypes carry different genetic mutations. Curtin et al showed that 39% of mucosal melanomas carry mutations and or increased copy number of KIT.<sup>5</sup> However BRAF mutations, frequently found in cutaneous melanomas are rare in mucosal melanomas.<sup>6</sup>

In sinonasal tract the commonest site of origin is lateral nasal wall and in particular from the middle and inferior turbinates followed by nasal septum. Disease may also arise from the maxillary and ethmoid sinuses. Isolated involvement of nasopharynx is rare<sup>7</sup>. Although the frequency of the sites of origin is said to correlate with areas of pigmentation, it is of interest that tumors do not arise from the olfactory region where pigmentation is greater. Malignant melanoma in the nose is almost always a primary lesion. It is an unusual site for secondary deposits but patients with malignant melanoma have an increased liability to develop a second primary tumor in another site. Between 10 and 18% of patients with nasal malignant melanoma will present with cervical lymphadenopathy and 4% with lung metastasis.<sup>8</sup> Macroscopically, brownish black coloration is apparent in 70% of cases but up to 10% of tumors may be completely amelanotic. Microscopically these tumors may show an angiocentric pattern due to extensive necrosis and preservation of the tumor cells and blood vessels. Immuno histochemically these tumors stain positive for S-100, HMB-45 and Tyrosinase.

Symptoms are mostly nonspecific, failing to prompt patients to seek immediate medical attention. Usually symptoms are of shorter duration for patients who had nasopharyngeal primary when compared to other locations of head and neck.<sup>9</sup> When symptomatic majority of patients present with epistaxis, unilateral nasal obstruction or a combination of two. The patients may also present with nasal mass, proptosis and Pain which is a rare feature. But our patient had severe headache and there is stigmata of mucosal pigmentation (conjunctival pigmentation).

Mucosal melanoma is always a malignant condition and no benign variants have been described so far. The decision of appropriate treatment should be considered individually and depends on size of tumor, depth of invasion, anatomic location and presence of cervical lymphadenopathy and distant metastasis.<sup>10</sup> Surgery followed by radiotherapy seems to be the optimal treatment. Because of low rate of cervical metastasis at initial presentation prophylactic neck dissection is not recommended.<sup>7</sup> Radiotherapy reduces the likelihood of local failure but probably does not enhance survival.<sup>8</sup> However radiotherapy can be considered alone as definitive therapy in patients with unresectable local disease, elderly patients who are poor surgical candidates, or patients who refuse surgery. The addition of chemotherapy has no impact on survival and Specific immunologic therapy was documented showing promising results but remains investigational.<sup>11</sup>

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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, failure to remove nodes containing melanoma and local implantation during surgery.<sup>12</sup> Regardless of staging system over all prognosis of sinonasal melanoma is poor, with a 5-year survival rate of 24.2%, with 50% of patients dying within 3 years.<sup>3</sup> Majority of patients die with distant metastasis to lung, liver, brain and skin.

**CONCLUSION:** Malignant mucosal melanoma of sinonasal tract is a rare neoplasm with aggressive behaviour and poor prognosis with nonspecific presentation. It should be considered in the differential diagnosis among the patients presenting with nasal bleed and growth in the nasopharynx, even in early age group to ensure its early diagnosis and treatment.

## REFERENCES:

1. Chiu NT, Weinstock MA. Melanoma of oronasal mucosa: population- based analysis of occurrence and mortality. Arch Otolaryngol Head Neck Surg 1996; 122: 985–8.
2. Lentsch EJ, Myers JN. Melanoma of the head and neck: current concepts in diagnosis and management. Laryngoscope 2001; 111: 1209–22.
3. Thomas J. Gal, Natalie Silver, Bin Huang. Demographic and treatment trends in Sinonasal Mucosal Melanoma. Laryngoscope 2011; 121: 2026-2033.
4. Holmstrom M, Lund VJ. Malignant melanomas of the nasal cavity after occupational exposure to formaldehyde. Br J Ind Med. 1991; 48: 9–11.
5. Curtin JA, Busam K, Pinkel D, Bastian BC. Somatic activation of KIT in distinct subtypes of melanoma. J. Clin. Oncol. 2006; 24: 4340–6.
6. Poynter JN, Elder JT, Fullen DR, Nair RP, Soengas MS, Johnson TM, Redman B, Thomas NE, Gruber SB. BRAF and NRAS mutations in melanoma and melanocytic nevi. Melanoma Res. 2006; 16: 267–73.
7. Lester D. R. Thompson, Jacqueline A. Wieneke, Markku Miettinen. Sinonasal Tract and Nasopharyngeal Melanomas. Ame. J. Sur. Path 2003; 27(5): 594-611.
8. Conley J, Pack GT. Melanoma of the mucous membranes of the head and neck. Arch Otolaryngol 1974; 99: 315–9.
9. Berthelsen A, Andersen AP, Jensen TS, et al. Melanomas of the mucosa in the oral cavity and the upper respiratory passages. Cancer 1984; 54: 907–12.
10. Lund VJ. Malignant melanoma of the nasal cavity and paranasal sinuses. Journal of Laryngology and Otology 1982; 96: 347-55.
11. Seo W, Ogasawara H, Sakagami M. Chemohormonal therapy for malignant melanomas of the nasal and paranasal mucosa. Rhinology 1997; 35: 19–21.
12. Ballantyne AJ. Malignant melanoma of the skin of the head and neck: an analysis of 405 cases. Am J Surg 1970; 120: 425–31.

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fig.1 Showing pigmentation of palpebral and bulbar counjunctiva.

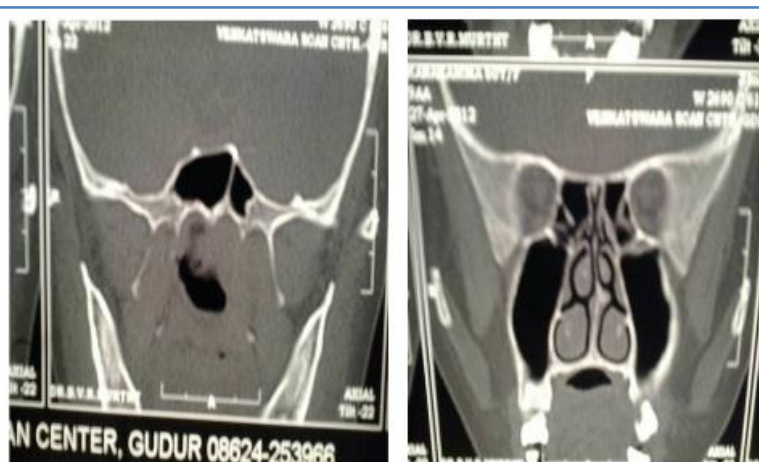


Fig. 2, 3: C.T Paranasal sinuses showing involvement of nasopharynx alone with out involvement of nose and paranasal sinuses

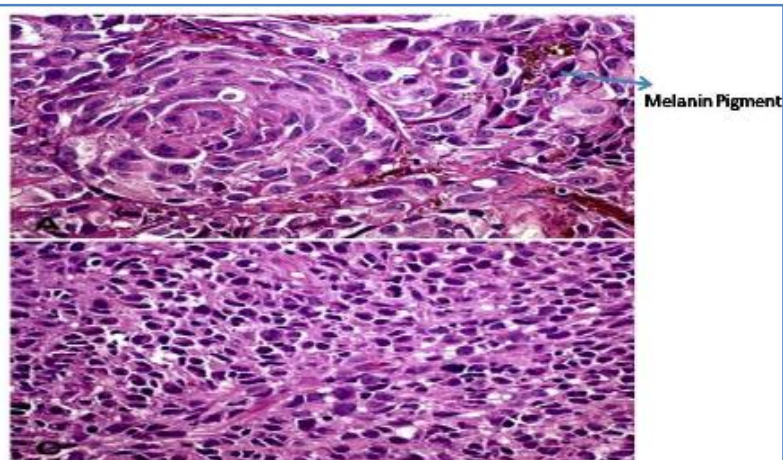


fig. 4 : Histo pathology slide showing intra cellular and extra cellular pigmentation with sheets of cells in vascular stroma.

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