

CASE REPORT

Primary Sino Nasal Mucosal Malignant Melanoma :Our Experience

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ABSTRACT

Malignant melanoma of mucosa of the nose and paranasal sinuses is an extremely rare occurrence and due to this, a universal definitive management protocol is not determined for maximum survival with minimum morbidity. Presentation of such cases is usually delayed due to its symptomatology resembling benign conditions. The disease is subject to definitive diagnosis only after confirmation by immunohistochemistry. Above factors contribute to poor survival of the patient. Early diagnosis and appropriate management as per the extent of the disease is required.

Keywords – sinonasal melanoma, mucosal melanoma, epistaxis

INTRODUCTION

Incidence of malignant melanoma of paranasal sinus ranges from 1/500,000 to 1/1,000,000 in general population⁽¹⁾ and due to such a low incidence rate, the definitive management of various stages of the disease have not been determined. Due to presentation of the patient in advanced stages and with the current modalities of treatment being not tailored for this particular pathology, the 5 year survival is below 30 percent. The current accepted treatment plan involves resection of the tumour with oncologically free margins on frozen section and further adjuvant radiotherapy to prevent regional recurrence. Role of neck dissection in N0 disease is yet to be proven beneficial due to lack of number of cases. We present our encounter with an early case presenting as epistaxis and managed accordingly.

CASE REPORT

A 42 year old male, labourer by occupation, presented to the emergency department with active nasal bleeding with no history of trauma. Bleeding was profuse but stopped with conservative management. He had similar complaints in the past which had been increasing in frequency and severity over past one year. He also had accompanying nasal blockade since last 6 months, more on the right side. Patient was admitted and investigated further.

A computed tomography revealed a moderately enhancing mass in the anterior and posterior ethmoid sinuses and upper 2/3 of right nasal cavity (figures 1 and 2), thinning of the ethmoidal

sinus bones was seen but no intra-cranial, intra-ocular extension was noted, neck was clear. An endoscopic biopsy of the mass was taken under anesthesia and the histopathology and immunohistochemistry (figures 3 and 4) was confirmatory for mucosal melanoma. Patient underwent an endoscopic excision of the mucosa of both anterior and posterior ethmoid sinuses, sphenoid sinuses, mucosa and bones of inferior and middle turbinate with oncological clearance, neck dissection was not performed. Final histopathology was confirmatory but the margins were declared close, so the patient underwent radiotherapy after a PET scan ruled out distant metastasis. The patient is under regular follow up every six months since last 2 years and undergoes diagnostic nasal endoscopies with no evidence of macroscopic recurrence and radiological investigations not suggestive of any tumour mass.

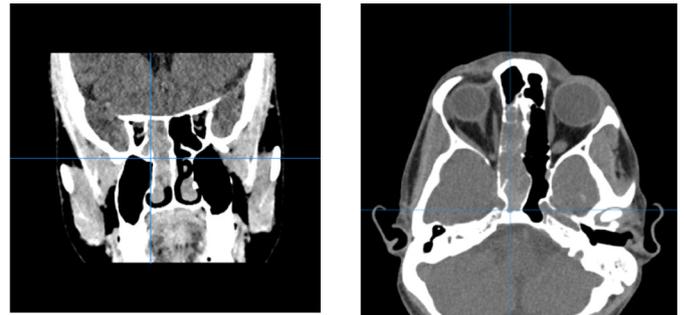


FIG 1 and 2: CT PNS with contrast showing extension of the tumour in the anterior and posterior ethmoid sinuses and sphenoid sinus on the right side.

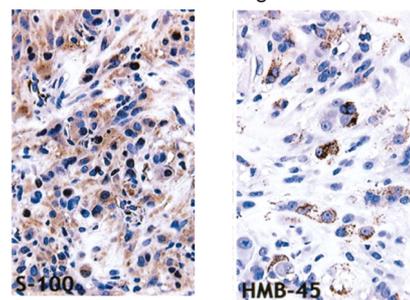


FIG 3 and 4: Immunohistochemistry showing S-100 and HMB-45 positive suggestive of melanoma

DISCUSSION

70-80 percent of sinonasal malignancies are squamous cell carcinomas and only 0.4-4⁽²⁾ percent of head neck malignancies are malignant melanomas. Men are 1.5 times more commonly affected and 80 percent occur in individual's aged 45 and above^(3,4).

The TNM staging system for mucosal melanomas⁽⁶⁾ is given in tables 1 and 2. Most clinicians also use a simplified 3 stage system⁽¹⁾ as depicted in table 3.

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Table 1: TNM staging for mucosal melanoma

Primary tumor (T)	
T3	Tumors limited to the mucosa and immediately underlying soft tissue, regardless of thickness or greatest dimension; for example, polypoid nasal disease, pigmented or nonpigmented lesions of the oral cavity, pharynx, or larynx
T4	Moderately advanced or very advanced disease
T4a	Moderately advanced disease Tumor involving deep soft tissue, cartilage, bone, or overlying skin
T4b	Very advanced disease Tumor 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 metastases
N1	Regional lymph node metastases present
Distant metastasis (M)	
cM0	No distant metastasis
cM1	Distant metastasis
pM1	Distant metastasis, microscopically confirmed

Table 2 : Anatomic stage/prognostic group

Stage	T	N	M
III	T3	N0	M0
IVA	T4a	N0	M0
	T3-T4a	N1	M0
IVB	T4b	Any N	M0
IVC	Any T	Any N	M1

Table 3 : 3 stage system of grading

STAGE	DISCRIPTION
I	Local disease
II	Lymphnode metastasis
III	Distant metastasis

Cases detected relatively early without extensive sinus spread and not extending into the orbit and cranium and no cervical lymphadenopathy like the one in our case, seem to have a good prognosis but long term assessment is yet to be made. Majority of cases though, are diagnosed late, and thus prognosis is poor and morbidity and mortality is high. Treatment regimens are customized to each case but requires expertise due to close proximity of functionally important structures like the orbit, optic nerve, anterior and middle cranial fossae in case of disease that has spread beyond the sinuses⁽⁷⁾. Treatment regimen for local disease is wide local excision with oncologically free margins^(8,9,10). Presence of lymph node metastasis worsens the prognosis and neck dissection and clearance is warranted^(11,12) though elective neck dissection in an N0 neck is still controversial⁽¹³⁾.

Post operative radiotherapy is indicated even though melanomas have been considered relatively radio resistant^(8,14,15,16) especially due to inadequate resection in

sinonasal melanomas. Most authors are in favour of higher doses per fraction for macroscopic disease and standard fractionation in case of microscopic remnant⁽¹⁷⁾, but the area in the vicinity of a sinonasal melanoma houses some very important structures like the optic nerve, pituitary, and its close proximity to brain parenchyma increases the risk of complications like loss of function, radionecrosis and radiation induced secondary malignancies. A combination of chemotherapy in addition to radiotherapy shows some clinical evidence of better local control but is yet to be thoroughly evaluated and put into routine use. Locoregional recurrences are common and some evidence in favour of irradiation of neck in case of N0 neck after appropriate neck dissection has also emerged^(18,19) For lesions with intracranial extension, carotid artery involvement and optic nerve invasion which are unresectable palliation is the only mode of management^(20,21). Despite the above measures the current overall 5 year survival is 15.7%^(6,22).

CONCLUSION

The rare nature of this particular malignancy makes early detection difficult, hence thorough history and clinical evaluation of patients presenting with complaints of epistaxis, nasal obstruction, sinusitis and exophthalmos is necessary. High degree of suspicion and early use of CT scan, MRI and histopathological modalities will aid in the definitive diagnosis of sinonasal mucosal melanomas. Multi-modality treatment is the key to achieving locoregional control of the disease. Newer avenues of neoadjuvant chemotherapy need to be explored and a well structured and universal treatment protocol has to be setup for mucosal malignant melanomas of the nose and paranasal sinuses.

DECLARATION

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REFERENCES

1. Patel SG, Prasad ML, Escrig M, Singh B, Shaha AR, Kraus DH, Boyle JO, Huvos AG, Busam K, Shah JP. Primary mucosal malignant melanoma of the head and neck. *Head Neck* 2002; 24: 247-257
2. Berthelsen A, Andersen AP, Jensen TS, Hansen HS. Melanomas of the mucosa in the oral cavity and the upper respiratory passages. *Cancer* 1984; 54: 907-912
3. Batsakis JG, Regezi JA, Solomon AR, Rice DH. The pathology of head and neck tumors: mucosal melanomas, part 13. *Head Neck Surg* 1982; 4: 404-418
4. Snow GB, van der Waal I. Mucosal melanomas of the head and neck. *Otolaryngol Clin North Am* 1986; 19: 537-547
5. Popoviæ D, Milisavljeviæ D. Malignant tumors of the maxillary sinus: A ten year experience. *Med Biol* 2004; 11: 31-34
6. Lydiatt WM, Patel SG, O’Sullivan B, Brandwein MS, Ridge JA, Migliacci JC, Loomis AM, Shah JP. Head and neck cancers—major changes in the American Joint Committee

- on cancer eighth edition cancer staging manual. CA: a cancer journal for clinicians. 2017 Mar;67(2):122-37.
7. Townsend C. Sabiston textbook of Surgery. 17th ed. In: Townsend C, Beachamp D, Mattox K. Townsend Textbook of Surgery. Elsevier Health Sciences, 2004: 833-867
 8. Mucke T, Holzle F, Kesting MR, et al. Tumor size and depth in primary malignant melanoma in the oral cavity influence survival. *Int J Oral Maxillofac Surg* 2009;67:1409–2115.
 9. Rapini RP, Goliz LE, Greer Jr RO, et al. Primary malignant melanoma of the oral cavity. A review of 177 cases. *Cancer* 1985;55:1543.
 10. Meleti M, Leemans CR, deBree R, et al. Head and neck head mucosal melanoma: experience with 42 patients, with emphasis on the role of postoperative radiotherapy. *Head Neck* 2008;30:1543–51
 11. Tanaka N, Mimura M, Ogi K, et al. Primary malignant melanoma of the oral cavity: assessment of outcome from the clinical records of 35 patients. *Int J Oral Maxillofac Surg* 2004;33:761.
 12. Tanaka N, Amagasa T, Iwaki H, et al. Oral malignant melanoma in Japan. *Oral Surg Oral Med Oral Pathol* 1994;78:81.
 13. Larson DD, Larson JD. Head and neck melanoma. *Clin Plast Surg* 2010;37:73–7
 14. Penel N, Mallet Y, Mirabel X, et al. Head and neck mucosal melanoma. *Am J Clin Oncol* 2005;26:626–30.
 15. Ballo T, Garden AS, Myers JN, et al. Melanoma metastatic to cervical lymph node: can radiotherapy replace formal dissection after local excision of nodal disease? *Head Neck* 2005;27:718–21.
 16. Owens JM, Roberts DB, Myers JN. The role of postoperative radiotherapy in the treatment of mucosal melanomas of the head and neck region. *Arch Otolaryngol – Head Neck Surg* 2003;129:864–8
 17. Wada H, Nemoto K, Ogawa Y, et al. A multi-institutional retrospective analysis of external radiotherapy for mucosal melanoma of the head and neck in northern Japan. *Int J Radiat Oncol Biol Phys* 2004;59:495–550
 18. Krengli M, Masini L, Kaanders JHAM, et al. Radiotherapy in the treatment of mucosal melanomas of the upper aero-digestive tract. A rare cancer network study. *Int J Radiat Oncol Biol Phys* 2006;65:751–9.
 19. Lee SP, Shimizu KT, Tran LM, et al. Mucosal melanoma of head and neck. The impact of local control on survival. *Laryngoscope* 1994;104:121.
 20. Townsend C. Sabiston textbook of Surgery. 17th ed. In: Townsend C, Beachamp D, Mattox K. Townsend Textbook of Surgery. Elsevier Health Sciences, 2004: 833-867
 21. Grewal DS, Lele SY, Mallya SV, Baser B, Bahal NK, Rege JD. Malignant melanoma of nasopharynx extending to the nose with metastasis in the neck. *J Postgrad Med* 1994; 40: 31-3
 22. Wong JH, Cagle LA, Storm FK, Morton DL. Natural history of surgically treated mucosal melanoma. *Am J Surg* 1987; 154: 54-57.