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Original Article

A Clinico-Radiologic Study of Primary Sinonasal Malignant Melanoma

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ABSTRACT

Aims: To study the clinical and radiological profile in cases of primary sinonasal mucosal melanomas.

Setting and Design: Retrospective, Hospital based.

Subjects and Methods: The study was conducted in Department of Otolaryngology and Head Neck Surgery, SMGS Hospital, Government Medical College Jammu from January 2013 to December 2015 and included a series of six patients. These patients were studied with respect to their clinical and radiological parameters. Subsequently, surgery was carried out depending on the extent of disease.

Results: Six patients with histologically proved primary malignant melanoma of the sinonasal cavity were included in the study. The patients were five males and one female aged 48 - 70 yrs. They presented most commonly with nasal fullness, congestion and epistaxis. Four patients were in stage T3 and two patients were in stage T4. All the patients underwent surgery and regular follow up. One patient showed local failure at sinonasal site for which radiotherapy was given. One patient showed cervical lymph node metastasis for which radical neck dissection was done.

Conclusion: Primary sinonasal tract mucosal malignant melanomas are uncommon tumors that are frequently misclassified, resulting in inappropriate clinical management. These need to be considered in the differential diagnosis of most sinonasal malignancies, particularly carcinoma, lymphoma, sarcoma, and olfactory neuroblastoma.These tumors have poor prognosis owing to higher rates of loco regional recurrence and distant metastasis.

KEYWORDS: Malignancy**,** Melanoma, Sinonasal.

INTRODUCTION

Malignant melanoma is a highly aggressive melanocytic neoplasm, usually affecting the skin. Mucosal melanoma is an exceedingly uncommon condition[1]. National Cancer Database statistics report that melanomas of mucosal origin comprise only 1.3% of all melanomas[2]. However, more than half of all mucosal melanomas arise in the head and neck region. The most common sites for the development of mucosal melanoma are the nasal cavity and paranasal sinuses.

Sinonasal mucosal melanoma accounts for less than 1% of all the malignant melanomas and approximately 3.5% of all sinonasal malignancies[1].

The etiologic and pathologic basis of the disease is not yet fully understood. The epithelium of the sinonasal cavity is ectodermally derived, which could explain the origin of primary (extracutaneous) malignant melanoma in this location. Melanocytes migrating from the neural crest may account for the presence of melanoma in the sinonasal cavity. Zak and Lawson[3] reported the presence of dendritic melanocytes in the epithelium of the sinonasal region. Cove[4] presented a case of a malignant primary multifocal intranasal melanoma arising from a preexisting nasal and maxillary sinus melanosis. Nevertheless, little is known about premalignant melanocytic lesions in the nose. The role of smoking or sun exposure as an etiology for this tumor remains controversial[5]. Their nonspecific clinical features and hidden location cause delays in diagnosis of sinonasal melanomas. Prognosis is always poor due to local recurrence, regional nodal involvement, and distant organ metastasis occurring months or years after the initial diagnosis. Early detection, diagnosis, and treatment of sinonasal melanoma are the key for longer patient survival[6].

Sinonasal malignant melanomas should always be considered in the differential diagnosis of most sinonasal malignancies, particularly carcinoma, lymphoma, sarcoma, and olfactory neuroblastoma.

Radical surgery is rarely feasible due to the intrinsic aggressiveness of the tumor and close relation with important neurovascular structures. Radiology has a paramount role in planning in the surgical resection of sinonasal melanoma since the integrity of the orbital walls, carotid canal, optic canal and skull base must be assessed before the surgery[7]. Chemotherapy and radiotherapy have also shown contradictory results.

The present study reports a series of six cases of primary sinonasal malignant melanomas who presented with different symptoms. Their preoperative evaluation, management and follow-up are discussed in the study.

MATERIALS AND METHODS

The study was conducted in Department of Otolaryngology and Head Neck Surgery, SMGS Hospital, Government Medical College Jammu from January 2013 to December 2015. Six patients were included in the study. Their varied clinical presentation was noted. Radiologic examination (Computed Tomography) was done to know the extent of the disease (for proper staging) and the feasibility of surgery. The present study used a modification of a classification system suggested by Freedman et al[8].

This modified classification system is as follows:

Stage T1: Tumor is limited to one site in the nasal cavity.

Stage T2: Tumor spreads into other nasal structures or the palate.

Stage T3: Tumor extends beyond the ipsilateral nasal cavity into the maxillary or ethmoid sinus, contralateral nasal cavity, or skin.

Stage T4: Lesion extends to the orbit, pterygopalatinc fossa, brain, or sphenoid sinus.

The diagnosis of sinonasal malignant melanoma was made on histopathologic examination of the nasal mass. Their management and follow-up is being discussed.

Table 1 – Clinical profile and management data of the patients of Sinonasal Malignant Melanoma

S.No	Age/sex	Clinical presentation	Tumour cell type	Stage	Treatment	Recurrence
1	62/M	Epistaxis, Nasal Obstruction	Epitheliod	T3	surgery	No
2	70/M	Epistaxis, Nasal Obstruction, Hyposmia.	Epitheliod	T4	surgery	Yes (at local site and Radiotherapy was given)
3	65/F	Blurred Vision, Epiphora, Headache	Spindle Shaped	T4	surgery	Yes (Cervical LN metastasis and RND was done)
4	58/M	Epistaxsis	Epitheliod	T3	surgery	No
5	60/M	Pain root of nose, Nasal Obstruction	Epitheliod	T3	surgery	No
6	48/M	Anosmia, Nasal Obstruction	Epitheliod	T3	surgery	No

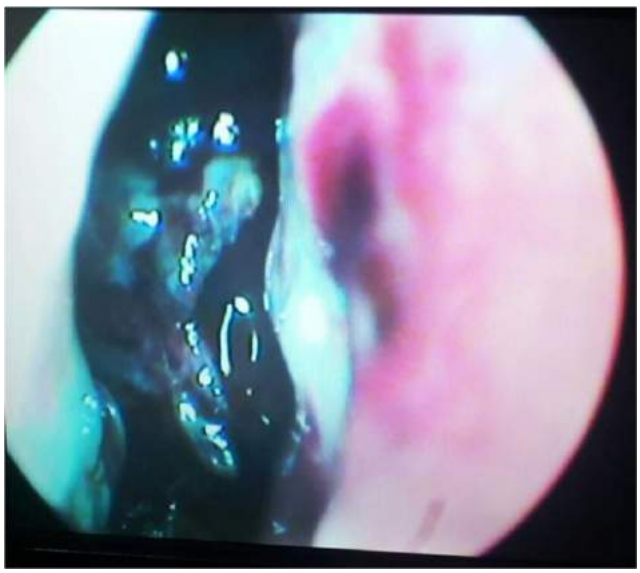

Fig 1: Nasal endoscopy showing blackish brown mass filling the nasal cavity.

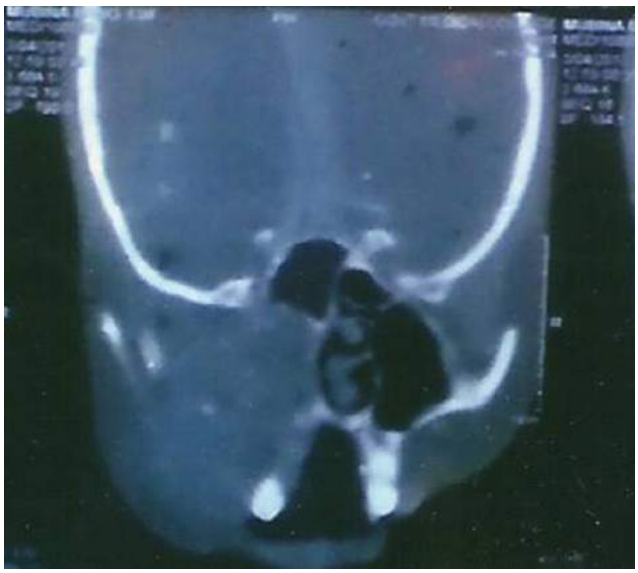

Fig 2A: CT Scan of paranasal sinuses showing extent of soft tissue mass: Coronal view

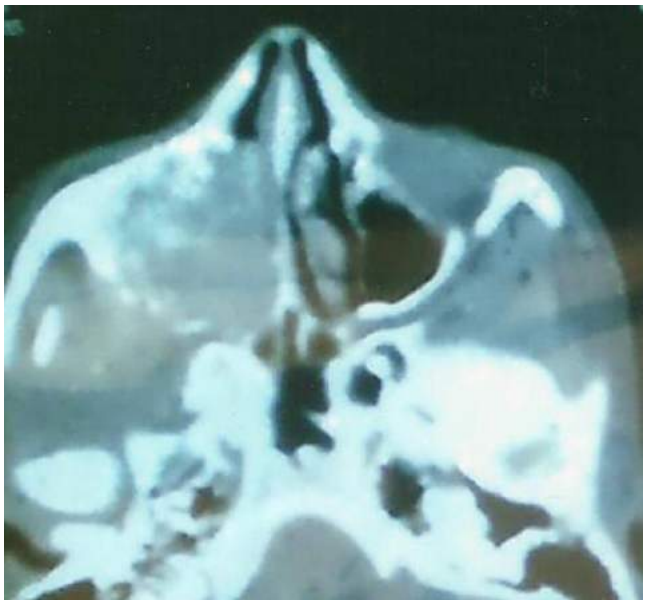

Fig 2B: CT Scan of paranasal sinuses showing soft tissue mass: Axial view

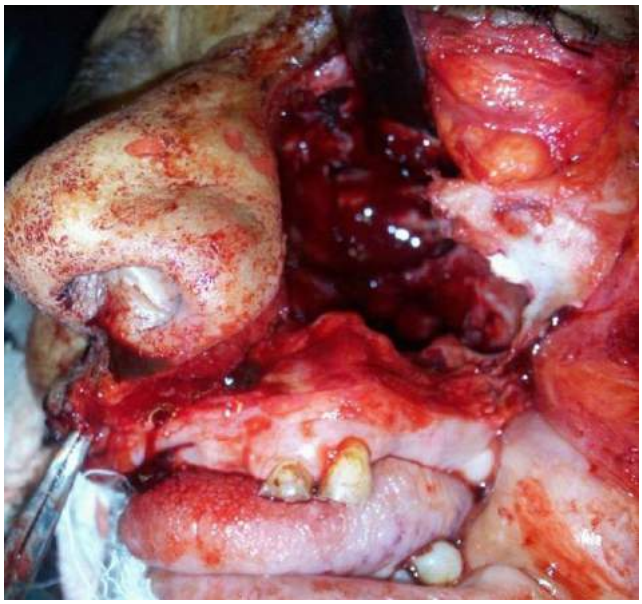

Fig 3: Intraoperative picture of partial maxillectomy being performed

RESULTS

Six patients with histologically proved primary malignant melanoma of the sinonasal cavity were included in the study. The patients were five males and one female aged 48-70 yrs (mean age, 60 years). They presented most commonly with nasal fullness and congestion (n=4) and epistaxis (n=2) as depicted in Fig 1. Imaging (Fig 2A, 2B) was done in all the patients for proper staging. Four patients were in stage T3 and two patients were in stage T4 (Table 1).

Depending upon the extent of the lesion, the patients underwent surgery which included partial maxillectomy (Fig 3), total maxillectomy or total maxillectomy with orbital exentration. The median follow-up period was 8 months (range 6-18 months).

One patient showed local failure at sinonasal site for which radiotherapy was given. One patient showed cervical lymph node metastasis for which radical neck dissection was done. None of the patients had distant metastasis.

DISCUSSION

Melanomas are tumors arising from melanocytes which are neuroectodermally derived cells located in the basal layers of skin, skin adnexa and some of the mucosal membrane. Mucosal melanomas of the head and neck are very rare, representing only 0.4% to 1.8% of all malignant melanomas[9]. These tumors originate from the melanocytes present in the mucous membranes; however, the risk factors for cutaneous melanomas (eg, sun exposure) do not apply for mucosal tumors[9,10]. Melanocytes are widely distributed throughout cutaneous and mucosal surfaces. They are present in nasal mucosa, in the glands, superficial and deep stroma of the septum and turbinates, being particularly common in the supporting cells of the olfactory epithelium[3].

In the head and neck region, nasal and oral cavities are the most commonly affected sites. Malignant melanoma comprises 3.5% of all nasal tumors and is second commonest malignant neoplasm (23%) in that region[1]. The commonest site of origin in nose is from the lateral nasal wall and in particular from the middle and inferior turbinates, followed by the nasal septum. Disease may also arise from the maxilla and ethmoids. The precise site of tumor origin is occasionally difficult to identify due to the large size of the tumor at presentation and the extensive local destruction it causes[11].

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[12]. The peak age incidence is between 5th and 8th decade and is seen slightly more common in males[13]. In the present study, the mean age of presentation was 60 years with 5 males and 1 female.

An intranasal melanoma usually manifests as a solitary lesion rather than multiple foci[5]. The majority of patients present with unilateral nasal obstruction or epistaxis or both as seen in our cases. Occasionally patients may notice proptosis (2%) in addition to other symptoms and sometimes swelling of the nose or an actual mass is visible at the vestibule (6%). Pain is rarely a feature. But one of our patient had severe pain at root of nose.Duration and symptoms before medical attention is sought average four to nine months, with a range of one to two years[14] due to innocuous symptoms.

On physical examination nasal melanomas tend to be large, bulky, friable masses which bleed with manipulation.[9] Clinical appearance of the tumors may be indistinguishable from benign polyposis[15]. Macroscopically, brownish-black coloration is apparent in 75% of cases but upto 10% of tumors may be

completely amelanotic. Microscopically, sinonasal mucosal melanomas may show an angiocentric pattern due to extensive necrosis and preservation of the tumor cells around blood vessels. Immunohistochemically, these tumors also stain positive for S100 and HMB-45. The differential diagnosis includes metastatic cutaneous melanoma, poorly differentiated carcinoma, and angiocentric T-cell/natural killer cell lymphoma. Because cutaneous melanoma metastasizes to mucosa in 2% to 9.3% of cases, metastatic lesions should always be excluded[16].To establish a diagnosis of primary intranasal malignant melanoma, the pathologist should exclude the presence of a malignant melanoma at other sites, most commonly the cutaneous region. In 1989,Going and Kean presented a primary intranasal melanoma with the presence of melanoma-in-situ of the adjacent mucosa next to the invasive lesion[17].The finding was considered an unusually well-documented feature found within the primary intranasal melanoma.

Mucosal melanoma is always a malignant condition and no benign variants have been described so far. The prognosis of the sinonasal melanomas is extremely poor, with a five-year survival of 6.5% to 34% and with more than 50% of patients dying within three years[18]. The tumor thickness, depth of invasion, and nodal involvement are important prognostic factors. The site of the tumor is also important. It appears that the melanomas of the oral cavity have a higher frequency of lymph node metastases than do the sinonasal primaries. Between 10 and 18% of patients with nasal malignant melanoma will present with cervical lymphadenopathy and 4% with lung metastasis [19]. Local recurrence, cervical lymphadenopathy and metastatic disease can occur at any time though Gallagher[20] reported that 55% of patients manifested this within one year of diagnosis. Systemic metastases are found in the lung, liver, brain and skin and the vast majority of patients will die of or with such secondary disease. One of our patient developed cervical metastasis.

The most fundamental treatment is wide local resection of the primary tumor whenever possible. Surgery provides the best chance of controlling the disease[21]. The likelihood of local recurrence after resection is approximately 50%. Radiotherapy (RT) reduces the likelihood of local failure but probably does not enhance survival, which is primarily impacted by advanced T stage and the presence of regional metastases. Definitive RT may occasionally cure patients with unresectable local-regional disease or at least provide long-term palliation. The addition of chemotherapy has no impact on survival.

Radiotherapy combined with surgery is recommended in cases of local recurrence or incomplete lesion removal[11]. Optimal radiation doses remain uncertain. Gilligan and Slevin[22] suggested high doses of 50-55 Gy in 15 or 16 daily fractions over 21 days. In addition, Thompson et al[10] also recommended performing simple excision of the involved cervical nodes except in cases when there were simultaneously more than two enlarged ipsilateral cervical glands, for which a radical neck dissection was recommended .

Seo et al[23] performed chemotherapy in conjunction with administration of the antiestrogen agent, tamoxifen, to treat their patients, and obtained satisfactory responses despite the fact that one of the tumors was unstainable for estrogen receptors. Other chemotherapy regimes, such as Vinca-alkaloids, alkylating agents, and antimetabolites, have been tried, but all yielded disappointing results[24].

In general, such tumors are associated with a poor prognosis and unpredictable course. The prognosis seems unrelated to the size, site, or pigmentation of the tumor.

The 5-year survival rate is estimated to be less than 40%[5]. Loree et al[25] concluded that head and neck mucosal melanomas, including nasal cavity lesions, and early-stage cases at presentation (i.e., stages I and II) showed more-favorable outcomes (32%) compared to those with stage III or IV (0%) according to the TNM system. Therefore, early detection and appropriate treatment should therefore be emphasized.

CONCLUSION

Sinonasal malignant melanoma, although rare should always be kept in the differential diagnosis of sinonasal neoplasms in elderly individuals presenting with epistaxis and nasal congestion. The clinical and radiographic features of these tumors are nonspecific; consequently, an accurate diagnosis requires histologic evaluation. These tumors have an unpredictable course and a very poor prognosis due to local recurrences and distant metastases developing unexpectedly. Early detection followed by surgical excision with clear margins remains the main stay of treatment.

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