Malignant Melanoma Of Nasal Cavity

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Citation

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Abstract

Malignant melanoma is a rare tumor in otorhinolaryngology. We are reporting a case of 40 years old male who presented with a mass in left nasal cavity. Biopsy suggested the diagnosis of malignant melanoma. The tumor was excised by medial maxillectomy and surgery was followed by radiotherapy. During 6 months follow up no recurrence was noticed.

INTRODUCTION

Invasive tumors containing abnormal melanocytes are termed malignant melanomas. Malignant melanomas of nasal cavity and sinuses are extremely rare tumors.

Melanomas are tumors arising from melanocytes which are neuroectodermally derived cells located in the basal layers of skin, skin adnexas and some of the mucosal membrane.

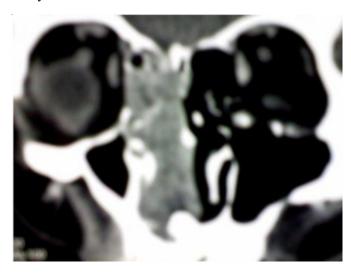
Common sites for melanomas are head, neck and the lower extremities as they are exposed to sunlight, which is one of the predisposing factor. Less commonly they occur in the oral and genital mucosa, nail beds, conjunctiva, orbit, esophagus, nasal mucosa or nasopharynx, vagina and leptomeninges. The cause of melanoma in solar-hidden mucosa is unclear, although smoking may have a role in activation of pre-existing melanocytes leading to melanogenic metaplasia.

CASE REPORT

A 40 years old male presented with history of left sided nasal obstruction for 3 months. Anterior rhinoscopy revealed a fleshy, black, non tender mass in left nostril which was completely blocking the nasal passage. There was no palpable regional lymphnode. Preoperative biopsy of mass suggested the diagnosis of malignant melanoma. CT showed a space occupying lesion filling the left nasal cavity and mass was totally obstructing the maxillary ostium (Figure.1). Medial maxillectomy was done under general anesthesia and tumor was removed en mass with tumor free margins of 2cm (Figure. 2 & 3). The tumor was arising from inferior turbinate and was involving nasal septum. Histopathology study revealed melanocytes in the sub mucosa. The stroma contained large number of spindle shaped cells with hyperchromatic nuclei. The histopathological findings led to

the diagnosis of malignant melanoma.

Figure 1: Coronal CT Showing melanoma filling left nasal cavity



The postoperative course was uneventful. Radiotherapy was given after surgery and 6 months follow up revealed no recurrence or metastasis.

DISCUSSION

Primary mucosal melanoma of the head and neck is an uncommon neoplasm and accounts for 20% of all malignant melanomas. Within the head and neck region, the most common site of origin is the nasal cavity, where the anterior septum is most often involved, followed by inferior and middle turbinate. However it is difficult particularly in large lesions to determine the exact site of origin. The second most common site is the oral cavity, where a predilection for the hard palate and maxillary alveolar gingivae has been found. The peak age for mucosal melanoma is during sixth

to seventh decade. A slight male predominance has also been reported. The presenting sign and symptoms of mucosal melanoma directly relate to the anatomic site of origin. The majority of patients with nasal cavity primaries present with nasal obstruction and epistaxis. Proptosis, diplopia, pain and facial deformity are less common and are indicative of advanced disease. ² Malignant melanomas of the nasal cavity and sinuses are characterized by early and repeated recurrences. ³ Nasal and sinus melanomas usually are advanced at the time of discovery. Metastases are often found at initial presentation for both cutaneous and mucosal melanomas.

Various methods of therapy, including surgery, irradiation alone, irradiation with surgery and chemotherapy have been used in treating malignant melanoma of the nose. The preferred treatment for Sino nasal mucosal melanoma is wide local excision with tumor free margin. Different surgical procedures like lateral rhinotomy, craniofacial resection, maxillectomy and total rhinectomy are done depending on the extent of the lesion. 4 Chen et al stated that surgery is the first choice and post operative radiotherapy gives a better result. 5 There is high incidence of local recurrences even with fresh frozen section to ensure complete removal. More radical surgery, including removal of the eye, palate or external portion of the nose, may not significantly lesson the incidence of local recurrence. 6

Gilligan et al reported absolute local control by radiotherapy alone achieved in 61% of cases. Radical radiotherapy for melanoma of this site can be justified because of local control achieved, low treatment morbidity in patients who are typically elderly and propensity to disseminated disease.

Melanoma is relatively chemo resistant tumor. The main role of chemotherapy remains as palliative treatment in the setting of disseminated stage IV disease. Dacarbazine is currently the only chemotherapeutic agent approved for the treatment of advanced stage IV disease. 8

Figure 2

Figure 2: Per-operative view showing medial maxillectomy approach for tumor excision



Among the recent techniques of immunotherapy, use of genetically altered tumor cells to elicit a T cell response has been used in the treatment of malignant melanomas, by stimulating response to certain melanoma antigens. These include melanoma antigen (MAGEs) -1, -2 and -3 and tyrosinase. These antigens are processed in to peptides and presented to T cells via HLA-A antigens on tumor. In addition, a melanoma antigen called MART is recognized in the context of class II MHC antigens. 9

The overall prognosis and survival rate of these tumors is very poor. The single most powerful predictor of survival is the status of regional lymph nodes. Some of the contributing reasons for their poor prognosis are the delay in detection and inaccurate histological diagnosis, the frequent injudicious therapy and the difficulties in precise removal of tumor. The 5- years survival varies between 10-40% and the median survival is 21-24 months. About one half of patients die within 3 years; of those alive at 5 years, 50% may have residual disease. Virtually all patients eventually die because of persistent local recurrence and spread of the disease. The course of the disease is variable depending upon the host tumor relationship. Some patients succumb to the disease within a few weeks or months of presentation despite aggressive surgical removal, other have a long disease free period followed by dramatic recurrence. 3

CONCLUSION

Malignant melanoma is the least common but most dangerous form of nasal cancer. Early diagnosis is crucial for optimum management since patients often present quite late. When the diagnosis is finally made, the tumor should be excised with wide local excision with tumor free margins.

Figure 3

Figure 3: Specimen of excised tumor



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