International Journal of Enhanced Research in Medicines & Dental Care, ISSN: 2349-1590
Vol. 1 Issue 10, December-2014, pp: (23-26), Available online at: www.erpublications.com

Mucosal Malignant Melanoma of the Nasal Cavity: A Rare Case Report

S. P. Gulati[1], Dr. Raman Wadhera[2], Mamta Rohilla[3], Narender Rohilla[4], Ankit Gulati[5]

[1,2]MS, Senior Professor and Head Professor, Department of Otorhinolaryngology, Pt. B. D. Sharma PGIMS, Rohtak

[3]MS Resident, Department of Otorhinolaryngology, Pt. B. D. Sharma PGIMS, Rohtak

[4]MD, Pathology Resident, Department of Otorhinolaryngology, Pt. B. D. Sharma PGIMS, Rohtak

[5]MS Resident, Department of Otorhinolaryngology, Pt. B. D. Sharma PGIMS, Rohtak

Abstract: Mucosal malignant melanoma (MMM) of the nose is extremely rare. We report a rare case of MMM of the nasal cavity in a 68-year-old male patient presenting with a polypoidal mass in the right nasal cavity. It was increasing gradually and obstructing breathing. A biopsy of the lesion was done with a clinical suspicion of inverted papilloma/carcinoma. Microscopy revealed features suggestive of malignant melanoma with minimal melanin pigmentation. Subsequently wide local excision was done. Diagnosis of malignant melanoma was facilitated by histopathology and immunohistochemistry. Primary mucosal malignant melanoma of the nasal cavity, paranasal sinuses and nasopharynx is an aggressive disease and fortunately rare with current research data being mainly from western population. The disease is known to be more common among elderly population in their 70's and to follow prolonged course due to innocuous symptoms. There are only few reports from the Indian subcontinent. This case is being reported for its rarity in our geographical region and rapid progression of the disease over a span of 3 months.

Key words: Mucosal malignant melanoma, nasal melanoma, sinonasal melanoma.

Introduction

Incidence of malignant melanoma as a whole is 2:100,000. Out of these, mucosal malignant melanomas of head and neck account for less than 1 % of these cases [1]. Mucosal malignant melanoma of the nose and paranasal sinuses is extremely rare. So rare that the majority of ENT surgeons do not see even a single case in their lifetimes. Prognosis is generally poor and unpredictable. Early diagnosis confirmed by immunohistochemistry [2] and radical surgical management appears to offer the best hope for curing the disease. Malignant melanomas may be misdiagnosed as a polyp or an inverted papilloma, especially when amelanotic. Histopathology report with immunohistochemistry gives accurate diagnosis. We report clinical and histopathological features of a case of mucosal malignant melanoma of nasal cavity in a 68-year-old male patient.

Case Report

A 68-year-old male patient came to the outpatient department with swelling in the right nasal cavity causing obstruction to breathing and gradually increasing in size over three months. There were occasional episodes of epistaxis, few drops at a time. He gave a history of tobacco smoking and long periods of exposure to sunlight being a farmer by occupation. On clinical examination, the swelling was reddish pink with brownish discoloration at places, soft in consistency with irregular surface filling up the whole of right nasal cavity.

Fig 1: clinical photograph of the patient with greyish brown mass filling right nasal cavity

On anterior rhinoscopy, turbinates couldnot be visualized. The mass was not bleeding on manipulation. There was no pigmented lesion elsewhere. Clinically, there was no regional lymphadenopathy which was confirmed by ultrasound neck.

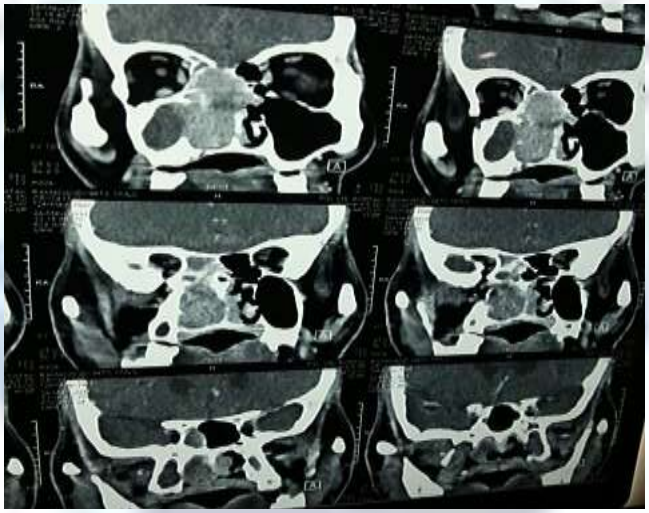

Fig 2: CT scan of nose and paranasal sinus showing extent of soft tissue mass

CT scan of the paranasal sinuses showed a soft tissue mass occupying whole of the right nasal cavity, involving the right anterior and posterior ethmoid sinuses and also blocking the infundibulum of the maxillary antrum. There was no evidence of direct orbital involvement. Superiorly the mass was extending up to the cribriform plate raising the possibility of its erosion at some places; however the patient did not have any signs of meningeal irritation but patient was having complaint of altered smell. There was no involvement of the sphenoid sinuses, no erosion of the nasal septum and the disease was limited to the right side. Septum was deviated to left side. A biopsy of the lesion was done with a clinical suspicion of inverted papilloma/carcinoma.

With a right lateral rhinotomy incision, the mass was exposed. It was soft in consistency. The mass seemed to arise from right lateral nasal wall showing focal black pigmentation at base. Tumor was found attached to ethmoid sinuses, medial wall of maxillary sinus was invaded, maxillary sinus was filled with pus. The turbinates appeared to be normal. Wide local excision of the mass was done. Tumor was extending up to cribriform plate, removed with endoscope. Nasal septum was healthy. The specimen was sent for histopathology. Postoperative period was uneventful. The wound healed well and suture removal was done on seventh day. After receiving histopathology of malignant melanoma, he was subjected to ultrasound abdomen to rule out distant metastasis in the liver.

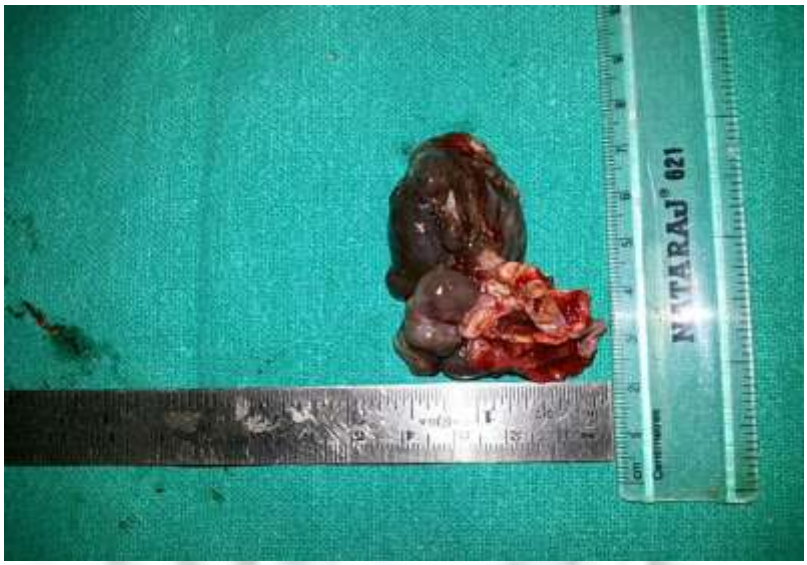

Fig 3: The mass removed from right nasal cavity

On histopathology, the tumor tissue showed different growth patterns. Tumor cells were oval to spindle shaped growing in solid fascicles, whorls, ribbons and reticulated pattern. There was mucosal infiltration seen. Cells showed brownish pigment most probably melanin.

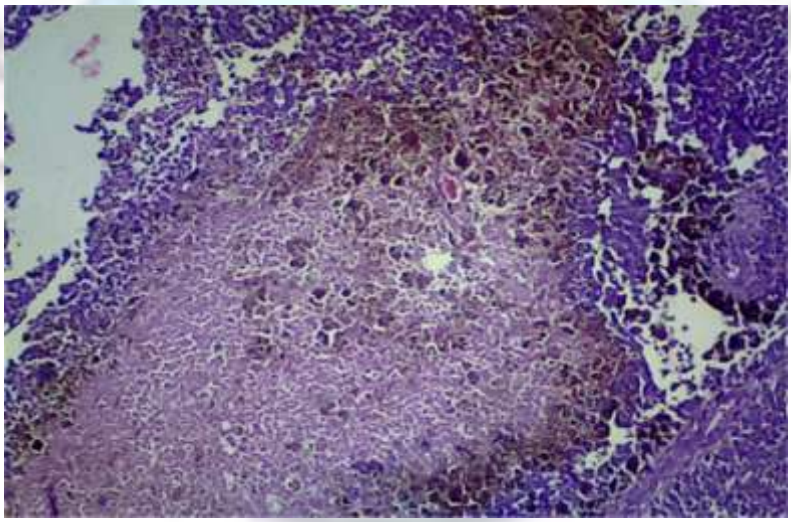

Fig 4: Malignant Melanoma 400X

Immunohistochemistry revealed tumor cells to be HMB 45 positive. This gave the definitive diagnosis of Mucosal Malignant Melanoma.

Discussion

Malignant melanomas are neural crest-derived neoplasms originating from malignant transformation of melanocytes in the basal layer of mucosa. First case of sinonasal malignant melanoma was reported by Lucke in 1869 and has been reported in about 2-8% of sinonasal tumors. Of the head and neck malignant melanomas, over 90% are cutaneous, 5% ocular, 2.2% peripheral unknown primaries and only 1.3% are mucosal [1].Mucosal malignant melanoma (MMM) of the upper aerodigestive tract (UADT) represents 0.5-3% of malignant melanomas of all sites. UADT MMMs are more common in men than in women [2]. Mucosal melanomas are far more aggressive than cutaneous ones and have a much poorer outcome.

In the nasal cavity, the most frequent site of occurrence is the nasal septum (anterior portion) and the lateral nasal wall. Clinically most patients have symptoms of nasal obstruction or epistaxis or both. On examination, nasal melanomas tend to be large, bulky, friable mass which bleed with manipulation. Clinical appearance of the tumors may be indistinguishable from benign polyposis, especially in amelanotictumors. The nasal melanomas project into the involved cavity and may have a somewhat polypoidal configuration. The consistency is firm, friable or gelatinous. Incidence of regional lymph node metastasis on presentation is about 5 to 15% with submandibular nodes being most commonly affected. About 10% of the patients may have distant metastasis on presentation [3].

Early diagnosis requires high index of suspicion. Definitive diagnosis is confirmed only by immunohistochemistry of the surgical specimen with tumor being S-100 and HMB 45 positive [2].Because of its rarity, there can be some differences in opinions regarding preferred modality of treatment. Surgery in the form of wide local excision remains the choice of treatment today. Different surgical procedures like lateral rhinotomy, maxillectomy or craniofacial resection are done depending on the extent of the disease.

Historically, mucosal malignant melanoma was characterized as a radioresistant disease. But some studies have shown that post-operative radiotherapy may give better local control. It should be used as adjuvant therapy in cases with regional metastasis or large bulky primary disease [4]. Radiotherapy alone was reported to give absolute local control in 61% cases by Gillian et al. [5].

Mucosal malignant melanoma is relatively chemoresistant tumor. Palliative chemotherapy is used in advanced disease with dacarbazine. Immunotherapy has shown a great promise in the treatment of this disease in recent studies. OK 432, Interleukin 2, lymphokine activated killer cells, BCG (Bacilli Calmette Guerin) vaccines have shown partial success but further trials in this regards are needed [6,7].

MMM tends to aggressive tumor and overall prognosis and survival rate ranges between 10 to 40% with mean survival being 21 -24 months [8]. Poor prognostic factors include local and distant metastasis, local recurrence, vascular invasion and a second primary [9]. Single most powerful predictor is absence of regional lymph nodes. Other contributing factors for poor prognosis are delay in detection and inaccurate histological diagnosis because of its rarity and low index of suspicion [10].

Our patient came with history of nasal obstruction and epistaxis. A nasal mass was seen protruding from the right nasal cavity. After appropriate investigations, he was subjected to surgery and the histopathology report with immunohistochemistry gave the diagnosis of malignant melanoma. He was also given postoperative radiotherapy. Now, he is symptom free and is under regular follow up considering the high rate of recurrence.

References

[1]. Jones Andrew S. Mucosal malignant melanoma. In: Scott Brown's Otorhinolaryngology, Head and Neck Surgery. 7th ed. Vol.2Hodder Arnold: UK; 2008: 2406-16.
[2]. Bothale KA, Maimoon SA, Patrikar AD, Mahore SD. Mucosal malignant melanoma of the nasal cavity. Indian J Cancer. 2009:46; 67-70.
[3]. Dwivedi R, Dwivedi R, Kazi R, Kumar S, Agarwal SP. Mucosal melanoma of nasal cavity and paranasal sinus. J Can Res Ther. 2008:4; 200-202.
[4]. Harwood AR, Cummings BJ. Radiotherapy for mucosal melanomas. Int J Radiat Oncol Biol Phys. 1982:8; 1121-6.
[5]. Gilligan D, Slevin NJ. Radical radiotherapy for 28 cases of mucosal melanoma in nasal cavity and sinuses. Br J Radiol. 1991:64; 1147-50.
[6]. Mundra RK, Sikdar A. Endoscopic removal of malignant melanoma of nasal cavity. Ind J Otolaryngol Head Neck. 2005:57; 341-3.
[7]. Sober AJ, Koh HK, Wittenberg, Washington C V Jr. Melanoma and other skin cancer. In: Braunwald E, Hauser SL, editors. Harrison's Principles of Internal Medicine. 15th ed. Vol. 1: USA : Mcgraw Hill 2001: p.554-9.
[8]. Conley JJ. Melanomas of the mucous membrane of the head and neck. Laryngoscope 1989:12;1248-54..
[9]. Prashant D, Kujur P, Sharma N, Prashant V. Primary Oral Malignant Melanoma. Journal of Case Reports. 2013:3; 272-6.
[10]. Errarhay S, Mamouni N, Mahmoud S, El fatemi H, Saadi H, MesbahiO et al. Primary Malignant Melanoma of the Female Genital Tract: Unusual Localization. Journal of Case Reports. 2013:3; 169-75.