Malignant mucosal melanoma of the nasal cavity: A case report

Devika Gupta, Niti Goyal, Vandana Rana, Rajat Jagani, Davendra Swarup

ABSTRACT

Introduction: Mucosal melanoma of the nasal cavity is a rare tumor. It is seen more commonly in the elderly and is known to have a male preponderance. Patients often present with non-specific symptoms of nasal obstruction or epistaxis. These tumors carry a poor prognosis because of higher rates of locoregional recurrence and distant metastasis.

Case Report: We report a 54-year-old male who presented with submandibular swelling and history of episodes of occasional epistaxis. Microscopic examination of the excision biopsy of the submandibular lymph node supported with immunohistochemistry (IHC) was suggestive of metastatic deposit of malignant melanoma. Clinical examination and radiological imaging for the primary tumor lead to detection of a mass in the right nasal cavity. Histopathology of wide local excision of nasal mass confirmed the diagnosis.

Conclusion: Malignant melanomas can mimic a large number of malignant diseases and early diagnosis by astute pathologist can help achieve attain long time remission.

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CASE REPORT

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Keywords: Malignant diseases, Melanoma, Nasal cavity, Nasal mass, Sinonasal tract, HMG-45

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INTRODUCTION

Melanomas are tumors arising from melanocytes which are neural crest derived cells present in the basal layer of skin, hair follicles, most squamous-covered mucosal membranes, leptomeninges and several other sites [1].

Mucosal melanomas of the nasal cavity, paranasal sinuses and nasopharynx (collectively called sinonasal tract) are extremely rare. They constitute about 1% of all melanomas. Patients have non-specific symptoms which is often the cause of delay in diagnosis. The cases are usually detected when there is extensive local invasion or distant metastasis. Majority of the tumors are melanin rich, however, some have scarcity of pigment and hence called non-pigmented amelanotic tumors. It is often difficult to diagnose pigment deficient melanomas as these can morphologically resemble variety of other malignant neoplasms. Immunohistochemistry is useful in confirming such cases. Treatment is wide local excision with radiotherapy and/or chemotherapy. Prognosis is generally poor and unpredictable.

We are discussing this aggressive tumor because of its rarity and to emphasize the importance of early detection of this lesion and high index of suspicion required when it presents with epistaxis and on histological examination is found to be amelanotic.

CASE REPORT

A 48-year-old male, tobacco chewer, presented with swelling in right angle of mandible of six months duration. The patient related it to dental caries for which he took dental consultation. The patient, however, noticed a gradual increase in the size of the swelling without overlying skin changes. He also complained of occasional episodes of bleeding from right nostril. He underwent fine-needle aspiration cytology (FNAC) of the right submandibular swelling in a local peripheral hospital which was reported as suggestive of non-Hodgkin lymphoma. He was referred to our tertiary care centre for further treatment and management. On general physical examination a single submandibular lymph node, measuring approximately 2x3 cm, was palpated on the right side. Swelling was firm, non-tender and non-mobile with no overlying skin changes. The patient underwent excision biopsy of the submandibular swelling and histopathological examination showed effacement of entire lymph node architecture by a tumor composed of sheets of polygonal cells having high N:C ratio, variable amount of cytoplasm, coarse vesicular chromatin and prominent eosinophilic nucleoli. A few cells focally were found to contain intracytoplasmic pigment. Immunohistochemistry confirmed the diagnosis of metastatic deposit from malignant melanoma. Meanwhile the patient was evaluated in ENT outpatient department for epistaxis. Nasal examination revealed a small, friable, fleshy growth in the right nasal cavity. Nasal endoscopy of the left side was normal. Computed tomography (CT) scan of the head and neck area was inconclusive. Computed tomography scan of chest and abdomen was within normal limits.

Positron emission tomography (PET) scan was done which showed an FDG-avid soft tissue density lesion in the right nasal cavity measuring 36x12 mm (Figure 1). Wide excision biopsy of nasal mass was done and sent for histopathological confirmation. Hematoxylin & Eosin (H&E) section of the nasal mass revealed round to polygonal tumor cells disposed in sheets. The cells had high N:C ratio, hyperchromatic pleomorphic nuclei with prominent eosinophilic nucleoli. These cells had variable amount of cytoplasm with only a few cells having intracytoplasmic melanin pigment focally. On IHC the tumor cells were positive for HMG-45 and S-100 and were negative for LCA, Pan-CK and EMA. Based on HPE and IHC the diagnosis of malignant melanoma was offered (Figure 2A–C). This tumor was considered as primary melanoma of nasal cavity due to the absence of previous or concurrent pigmented lesions elsewhere.

The patient was subsequently referred to the radiotherapy department for further management. Presently, one year after the diagnosis the patient is disease free and is on regular follow-up at our hospital.

DISCUSSION

Malignant melanomas originate from neural crest derived melanocytes present in the basal layer of skin, hair follicles and most squamous covered mucosal membranes, leptomeninges and several other sites. Most of the melanomas arise in the sun exposed areas, i.e., head and neck area and on lower extremities. Twenty to twenty-five percent of melanoma cases occur in head and neck region out of which approximately 6–8% originates in mucous membrane of upper aerodigestive tract [2]. The most common site for mucosal melanomas in the head and neck region is oral cavity followed by sinonasal region and lastly pharynx. Sinonasal mucosal melanomas are uncommon and comprise less than 1% of all melanomas and less than 5% of all sinonasal neoplasms [3]. The most common site of origin for melanomas within the nose is nasal septum followed by inferior and middle turbinate [4]. The exact site of origin of the larger lesions often cannot be determined. These tumors are aggressive with high incidence of locoregional recurrence and distant metastasis to lymph nodes and viscera. The incidence of regional lymph node metastasis is 5–15% [5]. The submandibular lymph nodes are most commonly involved. Involvement of regional lymph nodes strongly suggests distant spread as seen in our case [6].

The diagnosis of mucosal melanomas is based on histological finding and IHC because their microscopic features overlap with high grade lymphoma, poorly

Figure 1: Positron emission tomography scan showing FDG-avid soft tissue density lesion within the right nasal cavity.

Figure 2: (A) Section from mass right nasal cavity showing an attenuated respiratory epithelial lining. The subepithelial region is infiltrated by sheets of tumor cells (H&E stain, x100). (B) Section showing round to polygonal tumor cells having high nuclear-to-cytoplasmic ratio, coarse vesicular nuclei with conspicuous, eosinophilic nucleoli (H&E stain, x400). (C) Positive staining of tumor cells for (HMB-45, x200).

differentiated carcinomas, rhabdomyosarcomas, plasmacytomas [7]. They are positive for S-100 protein and specific melanocytic markers such as melan A and HMB-45 antigens [8]. The diagnosis is further complicated by the absence or presence of scanty melanin pigment as seen in our case.

The management of nasal mucosal melanomas has not been uniform. Standard treatment is surgical resection associated with adjuvant radiotherapy and chemotherapy. Surgery along with adjuvant chemotherapy and radiotherapy should be used for patients with either regional metastasis or large bulky primary disease [9]. In our patient wide local excision was performed followed by postoperative radiotherapy. The purpose of presenting the case is rarity of the lesion along with high index of suspicion which will help in early diagnosis and treatment.

CONCLUSION

Malignant melanomas are the greatest mimickers in pathology. They can be mistaken for a variety of tumors especially in the nasal cavity where these tumors usually do not show any junctional activity and also are amelanotic. Hence a high degree of suspicion on part of the pathologist is required to clinch the diagnosis in early stages. Treatment of choice in mucosal melanomas is combination of surgery with radiotherapy. Genesis of targeted immunotherapy and chemotherapy against melanomas based on clear understanding of biology of these tumors will help achieve a higher response rate.

Author Contributions

Devika Gupta – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Niti Goyal – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Vandana Rana – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Rajat Jagani – Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Davendra Swarup – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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