Nasal Vestibule Melanoma: Case report
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Abstract

Background: Melanomas of the nasal cavity and paranasal sinuses are quite rare, especially in the nasal vestibule.

Case presentation: We report a case of 47 years old male who presented with a 6 months history of progressively worsening left sided nasal blockage and a very rapid swelling of the left lateral nasal wall. Patient underwent an intra nasal excision biopsy and the histopathologic examination confirmed diagnosis of melanoma. Patient died within 6 months after diagnosis regardless of surgical excision and chemotherapy treatment.

Conclusion: Aerodigestive melanoma is a very rare disease. Its diagnosis and treatment is a complex issue. It has a very poor prognosis.

Keywords: Nasal neoplasm; Mucocutanous melanoma; Malignant; Epistaxis; Sinonasal cancer

Introduction

Tumors arising in the nasal vestibule classically are squamous cell carcinomas that have a natural history analogous to that of squamous cell skin cancer. Other types of skin cancers such as basal cell carcinoma and melanoma can also arise in the nasal vestibule as well.

Generally speaking malignant melanoma is less common but much more deadly than basal or squamous cell carcinoma in head and neck area. It is a disease of the skin as well as the mucosa.

In nose primary malignant melanoma is rare, accounting for less than 1% of malignant melanoma [1-9]. Malignant melanoma was first described in the nose by Lucke in 1869. The first case was reported in India in 1965 by Kutty and Sredharan [2]. Mucosal melanoma of head and neck is very rare, representing only 15% of all malignant melanomas of head and neck region [6]. About 70% of head and neck mucosal melanoma occurs in the nasal cavity or paranasal sinus region, while 25% of mucosal melanoma develops in the oral cavity [3]. Ethnicity considerably affects the prevalence of head and neck mucosal melanomas. It occurs in a greater extent in Asians and less frequently in the Western population [3,7]. Furthermore, mucosal melanoma has poorer prognosis than cutaneous malignant melanoma.

Patient information

A 47 years old man presented with a 6 months history of progressive nasal blockage and difficulty of breathing. He developed a left sided nasal swelling for 3 weeks that increased in size rapidly. There were nasal discharge and epistaxis on an off.

Clinical Findings

Examination revealed tender swelling over upper left lateral nasal wall with normal overlying skin. Rigid nasoendoscopy showed mobile polyp like mass occluding the left nasal vestibule and extended to left lateral nasal wall (Figure 1). The rest of the examination showed multiple nevi in head and neck area, but no cervical lymphadenopathy.
Diagnostic Assessment

None contrast MRI of the nasal fossa and the paranasal sinus showed well defined lobulated mass lesion. It was at the center of the anterior aspect of the left nasal fossa, measuring 50x36x31 mm in its largest AP. CC&TR dimensions respectively.

It displays iso to hypointense signal in T1 W1 and iso to hypertense signal in T2 & FLAIR W1s. Associated rarefaction and erosion of the left nasal bone is noted with extension of the mass anteriorly assuming subcutaneous location, yet no definite intraorbital extension. Another thin lesion of similar signal intensity on all pulse sequence is also noted at the right nasal fossa anteriorly measuring 35x28x7 mm in its largest CC. AP &TR dimensions respectively. No associated bone erosion (Figure 2).

Excision of the nasal mass and left nasal necrotic mass originated from vestibule to lateral nasal wall and anterior septum was done within 3days.

Histology revealed invasive nests and masses of variable size with delicate fibrovascular septae , the invasion is formed of pleomorphic cells with large nuclei and prominent nucleoli. Mitotic figures and apoptotic bodies are detected (Figure 3). Diffuse necrosed areas and melanin pigments are observed (Figure 4).

Immunohistochemical staining revealed that the tumor cells are positive to MelanA (Figure 5) and HMB45 (Figure 6), while markers of: cytokeratin, LCA, GFAP and CD99 were negative.

Patient diagnosed as a case of highly malignant melanoma, and advised for further investigation for staging and start management, but he decided to travel abroad for treatment.

Therapeutic intervention

Patient did not receive any therapeutic intervention in our facility.

Follow up and Outcomes

Few months later contact with his family member confirmed patient expired regardless of treatment.
Discussion

Worldwide numbers of cases diagnosed with melanoma are rising. There is 600% upswing in melanoma incidence over the past 50 years [4]. Most likely this is as a result of increased public awareness of the earliest signs of skin melanomas, or due to higher detection rates from vigorous surveillance.

It was believed that melanoma is a disease of old age, but now there is increase in incidence in younger age groups. Most probably this is due to increased sun exposure due to the perceived benefit of sunscreen, or inadequate usage of the sunscreen [5].

There are several risks factors that have been related to malignant melanoma. Most of all is sun exposure. Other risk factors include skin Fitzpatrick types II and I, blonde/red hair, outdoor summer jobs/hobbies (5,9). Meta-analysis, Gandini et al found possible positive correlation between an individual’s number of nevi and the overall risk of melanoma [4,9]. In the case presented above some of the risk factors are there. He has an outdoor job, which makes sun exposure, in a region like Middle East, one risk factor. In addition he has multiple nevi scattered over his face and neck. None of them looks suspicious. Possibilities of any were in the nasal vestibule is difficult to be ruled out. Besides early notice and detection of such kind of tumor in nasal vestibule is almost impossible most of the time. This makes nasal melanoma, both mucosal and vestibular, usually diagnosed in late stages.

Nasal melanoma often present with unilateral nasal obstruction, swelling and discharge. Unfortunately apparently benign nature of these signs and symptoms can lead to a delayed presentation. Epistaxis is present in almost all cases of nasal melanoma [8]. This was observed in our case. The patient was having nasal blockage for 8 months, where he related that to an attack of upper respiratory tract infections and sinusitis. For that he did not seek medical care. Only when he noticed swelling at one side of his nose that was increasing dramatically over a period of two weeks he started to look for the cause. By the time of presentation already the mass involved the nasal bone and septum. There was macroscopic ulceration of the lesion as well. All of that indicates bad prognostic features in our case.

The most valuable prognostic issues are number of metastatic nodes, tumor burden and ulceration [5,9]. The prognostic factors in nasal cutaneous melanoma depend on the tumor thickness and ulceration. These follow the Clark qualitative systems and the Breslow quantitative system [5]. While for cutaneous melanoma TNM staging system is followed, for mucosal melanoma the AJCC staging begins with stage III disease as the most limited form of disease [3].

Furthermore, it is noticed that mucosal melanoma have lower prevalence of regional lymph node metastases compared to cutaneous melanoma, both at presentation and at recurrence. Generally primary site recurrence ranges from 55% to 66% and 16% to 35% for nodal recurrence. Most recurrences occur within the first 3 years [6]. Although in the case presented up there is no appreciably palpable neck lymph node, the risk of subclinical disease is high. Micrometastases are diagnosed after sentinel or elective lymphadenectomy [5,9] for the purpose of staging. Plus PET scanning is another work up to look up for lymph nodes metastasis and disseminated disease.

Moh’s micrographic surgery is adopted for initial excision in melanoma especially at the nose, to minimize disfigurement that result from wide local excision with adequate margins [3,5]. The role of postoperative radiotherapy is not conclusive in treatment of cutaneous malignant melanoma. For that reason radiotherapy is usually applied as an adjuvant modality used for positive surgical margins, local recurrence, or palliation. Likewise, chemotherapy/immunotherapy is used with an adjuvant or palliative intention. Immunotherapy with interleukin (IL-2) and interferon Alfa (IFN-a) are used with cisplatin. This is currently effective in a small percentage of patients [6].

In summary nasal melanoma, cutaneous and mucosal, is rare tumor. It is relatively radioresistant as well as chemoresistant with high rate of recurrence. The prognosis is extremely poor due to late presentation as well as early hematogenous metastasis.

References
