Mucosal Malignant Melanoma of Sino-Nasal Region with Orbital Involvement: Case Report

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Abstract We report a case of melanocarcinoma of sino-nasal region in a 42 year old female who presented with brownish-colored sino-nasal mass involving left orbit, spontaneous epistaxis, facial swelling on left side, and ulceration of overlying skin, cervical and distant metastasis in liver over a short time span of 3 months. On biopsy, the mass was diagnosed as melanocarcinoma of nose and paranasal sinuses. 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 populations. The disease is known to be more common among elderly population in their seventies and to follow a prolonged course due to innocuous symptoms. This case is being reported for its rarity in our geographical region, early age of presentation and rapid progression of the disease over a span of 3 months.

Keywords Malignant · Melanoma · Paranasal sinuses · Melanocyte

Introduction

Sino-nasal tract mucosal malignant melanomas are rare tumors that need to be considered in the differential diagnosis of most sino-nasal malignancies, particularly carcinoma, lymphoma, sarcoma, and olfactory neuroblastoma. These tumors originate from the melanocytes present in the mucous membranes. They carry a poor prognosis due to local recurrence, cervical lymphadenopathy, and distant metastasis, making it difficult to identify the best treatment. Radical surgery is rarely feasible due to the intrinsic aggressiveness of the tumor and close relation with important vascular-nerve structures. Chemotherapy and radiotherapy have also shown contradictory results. There are only few reports from the Indian sub-continent [1–3]. We report a case of melanocarcinoma of sino-nasal region in a 42 year old female who presented with brownish-colored sino-nasal mass involving left orbit, facial swelling on left side and ulceration of overlying skin, cervical and distant metastasis in liver over a short time span of 3 months.

Case Report

A 42 years old female presented in ENT out patient department with swelling over left side of face and nasal blockage on same side since 2 months and externally visible brownish-colored mass in left nasal cavity. She had also history of epistaxis and swelling over left side of neck. The swelling started over lateral part of the nose on left side, below left eye and rapidly increased in size to involve eye and left cheek with ulceration of overlying skin. Swelling was associated with severe throbbing type of pain. No significant occupational history or environmental

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Fig. 1 Swelling over left side of face involving left eye with ulceration of overlying skin

exposure to of any irritant substance was obtained. Neither was any history of any other major illness in past.

On examination, a huge swelling was present over left side of face with ulceration of overlying skin, extending from nose up to left maxillary region and superiorly involved the left orbit covering it almost entirely (Fig. 1). A grayish brown mass was seen filling the left nasal cavity up to the nasal vestibule and also seen from right nostril with destruction of nasal septum (Fig. 2). Left submandibular lymph node was enlarged. Investigations revealed hemoglobin of 9.5 gm%, total lymphocyte count of 10,800 and differential leucocyte count of polymorph 70%, lymphocyte 28%, and eosinophil 2%. The erythrocyte sedimentation rate was 54 mm/hour. Routine biochemical investigations revealed normal glucose, renal and liver function tests. Tests for HIV using enzyme-linked

Fig. 2 Greyish brown mass filling the left nasal cavity up to the nasal vestibule with enlarged submandibular lymph node

Fig. 3 CT Scan of paranasal sinuses (coronal section) showing a moderately enhancing soft tissue mass lesion in the nasal cavity, left maxillary and ethmoid sinuses with destruction of nasal septum and bony medial wall of maxilla

immunosorbent assay were negative for both HIV 1 and 2. X-rays of the chest were normal. Biopsy of the mass showed melanocarcinoma. Fine needle aspiration cytology from left submandibular lymph node revealed metastasis of melanocarcinoma cells. Computerized tomography scan of paranasal sinuses revealed a moderately enhancing soft tissue mass lesion in the nasal cavity and extending into left

Fig. 4 CT Scan of paranasal sinuses (axial section) showing involvement of left orbit

maxillary and ethmoid sinuses and left orbit causing destruction of bony medial wall of maxilla, ethmoid, floor of orbit, and cribriform plate (Figs. 3, 4). The mass was seen to extend into subcutaneous plane of face and displaced eye-ball supero-laterally. No intracranial extension was seen. USG abdomen revealed both hypo and hyper echoic shadow in both lobes of liver likely to be metastasis. Due to the advanced disease at the time of presentation, we subjected the patient to chemo-irradiation.

Discussion

Mucosal melanomas of the head and neck are very rare, representing about 0.4–1.8% of all malignant melanomas [4]. These tumors originate from the melanocytes present in the mucous membranes; however, the risk factors for cutaneous melanomas (e.g., sun exposure) do not apply for mucosal tumors [4, 5]. 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 [6]. Most mucosal melanomas are anorectal or are found on the female genitalia. In the head and neck region, nasal and oral cavities are the most commonly affected sites. 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. Although, the frequency of the sites of origin is said to correlate with areas of pigmentation in the noses of normal Ugandan Africans it is of interest that tumors do not arise from the olfactory region where pigmentation is greatest [7]. 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 [8]. Most of the tumors occur in the seventh decade of life. There are geographic/racial differences in the incidence of head and neck mucosal melanomas. The disease predominately affects Caucasians though it has been reported in Negroes. In Ugandan Africans, oral and nasal primary melanomas form 10% of all melanomas. The proportion of oral melanomas to all melanomas in Japanese people is as high as 7.5%.

Macroscopically, brownish-black coloration is apparent in 75% of cases but up to 10% of tumors may be completely amelanotic. Microscopically, sino-nasal 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 2–9.3% of cases, metastatic lesions should always be excluded [9]. The absence of a cutaneous primary and the presence of mucosal involvement makes a metastasis unlikely. Immunohistochemical stains for epithelial and lymphoid/natural killer markers will rule out a poorly differentiated carcinoma and an angiocentric nasopharyngeal T-cell/natural killer cell lymphoma, respectively.

The majority of patients present with unilateral nasal obstruction, epistaxis or a combination of two. 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%) [10]. The patient in our case first noticed swelling over face which rapidly increased and involved the eye on the left side. Nasal obstruction and epistaxis were symptoms which followed associated with cervical metastasis and mass appearing at the nasal vestibule. There is delay in presentation that varies from 1 month to 2 years, the average being 4 months, due to innocuous symptoms. Pain is rarely a feature. But our patient had severe pain. Mucosal melanoma is always a malignant condition and no benign variants have been described so far. The prognosis of the sino-nasal melanomas is extremely poor, with a 5 years survival of 6.5–34% and with more than 50% of patients dying within 3 years. 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 sino-nasal primaries [11]. Between 10 and 18% of patients with nasal malignant melanoma will present with cervical lymphadenopathy and 4% with lung metastasis [8]. Local recurrence, cervical lymphadenopathy, and metastatic disease can occur at any time though few authors reported that 55% of patients manifested this within 1 year of diagnosis [12]. Systemic metastases are found in the lung, liver, brain, and skin and the vast majority of patients die of or with such secondary disease. Our patient had single cervical metastasis and multiple secondaries in the liver at presentation. These lesions are most commonly treated with wide local resection. The likelihood of local recurrence after resection is approximately 50%. Radiotherapy reduces the likelihood of local failure but probably does not enhance survival, which is primarily impacted by advanced stage and the presence of regional metastases. Definitive radiotherapy 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.

Conclusion

Primary mucosal malignant melanoma of the nasal cavity, paranasal sinuses, and nasopharynx is an aggressive disease with a rare presentation in a middle aged female with aggressive manifestation. Physicians and surgeons should be aware of the different ways in which this disease can present to ensure its early diagnosis and treatment.

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