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Case report

An unusual giant isolated mucosal malignant melanoma of nasal cavity – A case report

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ABSTRACT

The mucosal malignant melanoma arises from the melanocytes present in the mucosal membrane. It carries a poor prognosis due to local recurrence, neck node involvement and distant metastasis, making it difficult to identify the possible treatment. The key and critical issue for the management of the malignant melanoma of the nasal cavity is early diagnosis in order to start primary modality of treatment like surgical excision and radiotherapy with adequate follow up. We report a case of malignant melanoma of the nasal cavity in a 65 years old male with presentation of recurrent epistaxis and nasal obstruction. Rarity of this isolated lesion confined to nasal cavity with a giant size warrants its attention for reporting this case.

1. Introduction

Isolated malignant melanoma of the nasal cavity is an extremely rare tumour encountered by Otolaryngologists and has poor prognosis.[1] This tumour usually arises from the melanocytes which are neuroectodermally derived cells found in the basal layer of the skin, skin derived adnexa and some mucosal membranes. Common locations of this lesion are head, neck and lower extremities because they are frequently exposed to sunlight, which is a major predisposing factor. Less frequent sites of involvement are oral cavity, genitalia, conjunctiva, nail beds, orbit, esophagus, nasal cavity, nasopharynx and leptomeninges. The peak age form mucosal melanoma is between 6th to 7th decades with slight male preponderance. The incidence of malignant melanoma is more among whites than the blacks. The age and sex are not related to the prognosis of the disease. Despite the aggressive treatment with all options including surgery, radiotherapy and adjuvant therapy, five year survival rate is 20–46%.[2] We report a case of gaint size mucosal malignant melanoma in the nasal cavity in a 65 years old male without distant metastasis and locoregonal spread.

2. Case report

A 65 year old male attended the out patient department of Otorhinolaryngology with complaints of nasal obstruction and intermittent nasal bleeding since 6 months. On examination, there was a mass of 2 × 2.5 cm irregular, blackish grey in colour and friable mass coming from the left nasal cavity (Fig. 1). The ears, throat and larynx were normal on examination. He had no significant occupational history or environmental exposure to any irritant substance and neither had any past history of major illness. Biopsy confirmed the diagnosis of malignant melanoma. Contrast enhanced computed tomography (CECT) of the nose and sinuses revealed soft tissue mass inside the nasal cavity without any extension into any paranasal sinuses, orbit and cranium (Fig. 2). There was no neck node enlargement or any distant metastasis. The chest X-ray showed no abnormality in the mediastinum and without any evidence of pulmonary metastasis. The patient was underwent complete excision of the mass by endoscopic approach. The mass was attached to the nasal septum anteriorly. Histopathological examination of the excised mass again confirmed the same diagnosis of malignant melanoma of mucosal type where pleomorphic tumour cells revealed melanin pigments (Fig. 3). Immunohistochemical analysis showed positive for S-100, HMB 45 and melan A. Patient was given radiotherapy after surgery. He is now under regular follow up since one year without any evidence of recurrence and distant metastasis.

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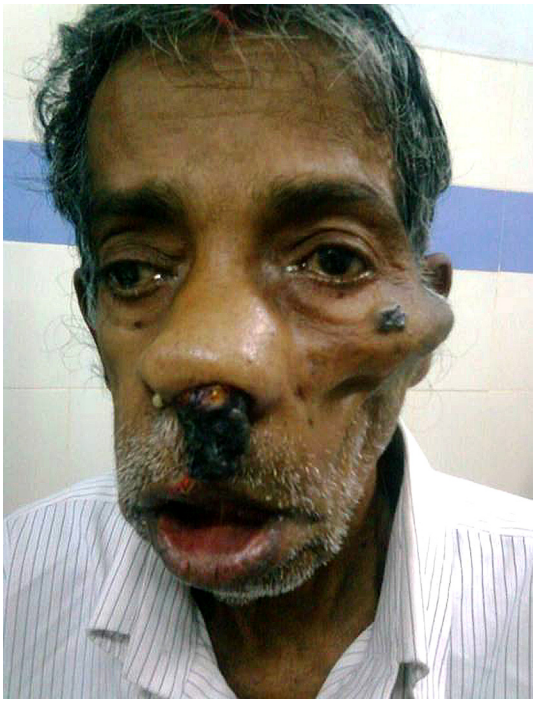

Fig. 1. Patient showing a large blackish grey mass coming from the left nasal cavity.

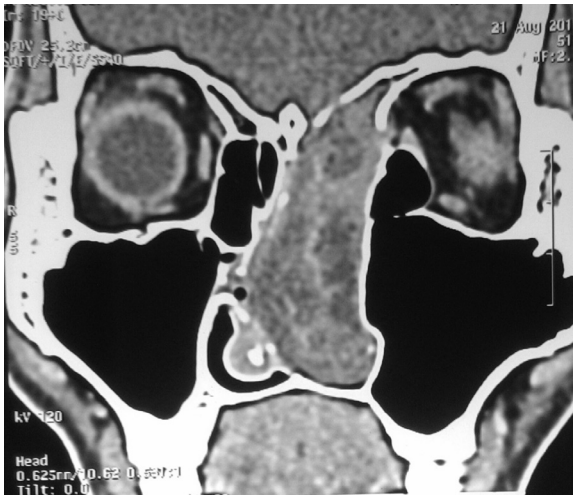

Fig. 2. CT scan of the nose and paranasal sinuses showing a soft tissue mass inside the left nasal cavity without any evidence of bony erosion of surrounding wall.

3. Discussion

Malignant melanoma is an extremely rare and aggressive tumour of the nasal cavity with high incidence of recurrence and distant metastasis. Mucosal malignant melanoma in the head and neck region are extremely rare accounting 0.4–1.8% of all malignant melanoma.[3] Most of the mucosal melanomas arises from anorectal or are seen on the female genitalias. Commonest sites of origin of head and neck region are sinonasal area and oral cavity. Commonest sites of origin in nose are from lateral wall, particularly from middle and inferior turbinates followed by septum of the nasal cavity. Isolated mucosal malignant melanoma of the nasal cavity, nasopharynx, paranasal sinuses is aggressive disease and fortunately rare in Indian subcontinents. The majority of this lesion presents with nasal obstruction and recurrent epistaxis. In

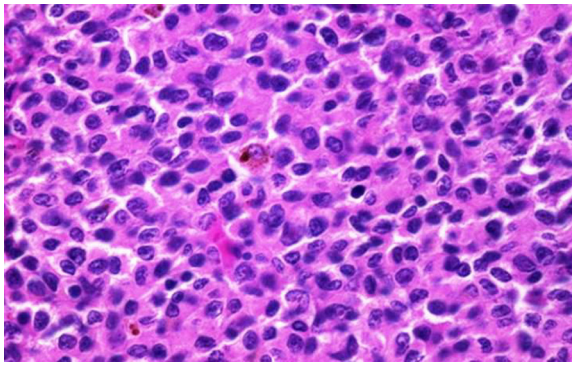

Fig. 3. Microphotograph showing features consistent with mucosal malignant melanoma (Haematoxylin & Eosin ×40).

advanced form, it presents with proptosis, diplopia, pain and facial deformity which are less common. There is ethnic difference with 8.8% afflicting black and Hispanics. The 5 year survival rate in malignant mucosal melanoma range from 10% to 38%.[4] The malignant melanoma is characterized by early local recurrences, frequent metastasis to lymph nodes and viscera, making it one of the most fatal form of sinonasal tumour. The submandibular nodes are most commonly affected. The nasal mucosal malignant melanoma is considered as one of the differential diagnosis of nasal malignancies like carcinoma, lymphoma, sarcoma and olfactory neuroblastoma. The only definitive prognostic factor in case of malignant melanoma is the presence or absence of distant metastasis at the time of diagnosis. Imaging should be done before taking biopsy. CT scan and MRI are utilized to characterize this lesion. CT scan delineates the surrounding bony erosions whereas MRI is thought to gives accurate tumour margin in relation to the surrounding structures. Ultimately tissue biopsy is the only confirmatory for diagnosis. The cytological picture shows cellular smears with single cell population with large nuclear cytoplasmic ratios and hyperchromatic nuclei. There is often multinucleated cells are present. The pigment is seen within cytoplasm and also in the background. Histologically, it is variable appearance and it may look like anything.[5] Grossly the size of this tumour varies from the size of pea to giant one. Immunohistochemical study is not usually necessary for diagnosis but useful for facilitating and completeness of the confirmatory diagnosis. S-100 and homatropine methylbromide (HMB45) were positive for melanoma. S-100 shows high sensitive and low specificity whereas HMB45 reveals highly specific and moderately sensitive in melanoma. If above two stains give unclear results, another marker, a melanoma specific marker called as melan A is classically used which is highly specific for differentiating melanoma from other malignant growth.[6] Historically, malignant melanoma was characterized as radioresistant lesion, but recent observations are in favour that radiotherapy has a major role in this lesion. The overall response rate of 50–75%, if radiotherapy alone is used in localized form of mucosal melanoma. These lesions are more radiosensitive than their cutaneous counter parts. Surgery with radiotherapy should be used for patients with large bulky primary lesions or with regional metastasis. Patients, who don't agree for surgical excision or unresectable local tumour, should be advised for radiotherapy alone as a definitive treatment. Chemotherapy should be reserved for patients with systemic involvement. There are several new treatment modality like biologic and immunomodulatory therapeutic options are currently are being in the trial phase for the patients of mucosal melanoma and the result of such treatment are still eagerly awaited for successful outcome.OK-432, Interleukin 2, lymphokine activated killer cells and BCG vaccine have

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met some success and need further research with large cohort for successful outcome. New modality of radiotherapy like intensity modulated radiotherapy (IMRT) and three dimensional conformal radiotherapy (3-D-CRT) are in investigating phase and show some promise for treating this lesion. Unfortunately the prognosis of sinonasal mucosal melanoma is poor with a mean five year overall survival rate is only15.7%.[7] The poor prognosis of this lesion in sinonasal area may be due to close proximity of vital structures such as skull base, carotid artery, brain and orbit.[8] The poor prognostic factors associated with tumours are tumour size (>3 cm), obstructive symptoms, advanced age, location at sinuses and nasopharynx, vascular invasion into the skeletal muscles, high mitotic count, cellular pleomorphism and distant metastasis.[9] Other factors leading to poor prognosis are late detection and incorrect histopathological diagnosis because of its rarity and low index of suspicion.[10]

4. Conclusion

Primary mucosal malignant is an extremely rare and highly aggressive disease of the sinonasal cavity. It typically presents with recurrent epistaxis and nasal obstruction. Mucosal variety of malignant melanoma as in nasal cavity has poorer prognosis than dermal counterpart. Treatment is surgery and postoperative radiotherapy. Close follow up is needed as local recurrence may happen and resection of the recurrent lesion results in longer survival of the patient.

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