

Primary Sinonasal Malignant Melanoma: Diagnosis to Prognosis

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Received: July 23, 2021

Published: October 01, 2021

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Malignant melanoma is comparatively a rare malignancy whose prevalence is nearly 1 - 2% of whole-body malignancies. Out of these, 90% are skin malignancies. Melanomas are malignant neoplasms, they are derived from melanocytes, a neuroectodermal derivatives located in the basal layer of the skin. Malignant melanomas in the head and neck mucosal surfaces are quite rare, which account for < 1% of all melanomas. Malignant melanoma of sinonasal region accounts for 1% of all mucosal melanomas and 3 - 4% of malignant neoplasms of the sinonasal tract. They originate from melanocytes present in mucous membranes, in glands, superficial and deep stroma of septum and turbinates, bones like maxilla and ethmoids. Due to late presentation at an advanced stage, these have a significantly low 5-year survival.

The most common sites in the upper airway in order of frequency are: oral cavity followed by nasal cavity and the paranasal sinuses. It can also spread to orbit, palate and buccal tissues but generally have less nodal or distant metastasis. However, it has got a unique, multi-focal characteristic due to which it is difficult to remove it en bloc while obtaining a clear margin during its surgical excision. For this reason, malignant melanoma of the nose and paranasal sinuses a high recurrence rate and a significantly poor prognosis. Although 0.6 to 9.3% of patients with skin melanoma have metastasis to upper aero-digestive tract mucosal membrane, and the most frequent sites being base and nasal cavity. At various occasions lesions are seen 2 years to 7 years after occurrence of skin lesion. But by the time they appear in the head and neck mucosal surfaces, often the patient already has distant or even disseminated metastasis. First case of melanoma in India was reported in 1965, by Shreedharan and Kully.

Mostly patients present with complaints of unilateral nasal blockage, epistaxis, face swelling, also at times nasal mass or proptosis. It is a malignant condition and no benign variants are reported.

On direct visualization brown-black discoloration is seen in 70 - 75% of cases but nearly 10% of tumors may be amelanotic. On microscopic examination, due to extensive necrosis it appears as angiocentric. Immunohistochemically, these tumors stain positive for S100 and HMB-45 while contrast enhanced CT scan and MR imaging helps in assessing the extent of disease. Talking about differential diagnosis, it includes cutaneous melanoma (metastatic), poorly differentiated carcinoma, and angiocentric NK cell/T-cell lymphoma. Immunohistochemical stains can further narrow the diagnosis.

As the melanoma metastasizes to mucosa in approximately 2.5 - 10% of cases, metastatic lesions should be excluded always. The absence of a cutaneous primary and the presence of mucosal involvement makes a metastasis unlikely.

Distant metastases are seen in the lung, skin, liver, and brain. Which is also the cause for death in a large number of patients. Local recurrence after resection is around 50%. Radiotherapy on the one hand fails to enhance survival rate as it occurs as a result of advance stage and distant metastasis, but it is known to reduce the likelihood of local failure.

Immunotherapy could have been a potential treatment but researches show that it has not been useful. The most frequent treat-

ment failures have been local recurrences; nasal cavity (40%), pharynx (32%) and oral cavity (25%). It has a very low survival rate, 5-year survival rate varies from 5 to 30%. So, in order to overcome such high rate of recurrence, even after properly carried out early and initial surgery, more radical resection is required and even in initial and localized cases. Out of all the presently available treatment modalities, most preferred is surgical intervention further followed by post-op radiotherapy which offers the best possible outcome. Although survival rates are still very poor even after aggressive management.

On reviewing the literature, it was found that in most of the studies 5-years survival rate was 6.5 - 34% with above 50% of patients not surviving even for 3 years. The prognostic factors include the tumor thickness, tumor depth of invasion, and involvement of nodes. Although site is also important, in comparison to sino-nasal melanomas oral cavity have a higher frequency nodal metastasis. Its important to note that at the time of presentation almost 10 - 18% of patients have cervical lymphadenopathy and nearly 4 - 5% already have lung metastasis. Recurrences, cervical lymphadenopathy, and distant metastasis can occur at any, some authors suggested that in 55% of patients it happens within 1 year of diagnosis. Mucosal melanomas behave differently in comparison to skin melanomas as already discussed, mucosal melanomas are considered more malignant and have poorer prognosis. Radical surgery with adequately broad safety margins makes up the pillar of treatment, together with radiotherapy, although in many cases treatment option left is only palliative and prognosis is very poor. Even additional use of chemotherapy has failed to improve prognosis and has no impact on survival. Close and regular follow-up is recommended as recurrence as well as systemic metastasis is common and prognosis is also poor but early detection and aggressive resection of recurrent tumor with clear safety margins may improve survival in few patients.

So, take home message is that, the primary sinonasal malignant melanoma is extremely rare which must be differentiated from other sinonasal tumors. Early appropriate diagnosis can be achieved with tissue examination and immunohistochemistry studies. Surgical treatment with broad margins of resection followed by adjuvant chemotherapy and radiation therapy offer

the only possible hope of survival. As the prognosis is very poor, even minor nasal symptoms specially any discoloration in mucosa should be addressed at the earliest.

Volume 3 Issue 11 November 2021

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