

## Primary Mucosal Malignant Melanoma of Nasal Cavity and Paranasal Sinus

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Primary mucosal melanoma of the nasal cavity and paranasal sinuses is a rare entity. Nasal malignant melanoma develops from the melanocytes residing in mucous membranes that have migrated during embryologic development from the neural crest to the mucosa of nose and sinuses. It is an unusual cause of nasal obstruction and epistaxis seen specially in elderly. This tends to be aggressive tumor with poor prognosis. Here we report a case of primary malignant melanoma of nasal cavity in a 60 year old male presenting with nasal obstruction and epistaxis. Histopathological examination of the resected mass showed a hypercellular tumor composed of fascicles of malignant spindle cells with intracytoplasmic melanin pigment. Immunohistochemistry was not possible in our setup. It was diagnosed as primary mucosal melanoma histomorphologically. The rarity of its occurrence warrants its mention.

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**Key words:** Epistaxis, Nasal obstruction, sinonasal mass, mucosal melanoma, melanin.

### Introduction

**M**alignant melanoma is a malignant transformation of the normal melanocytes, which are derived from neuroectoderm located in the basal layer of skin, skin annexes and more rarely in mucosal membrane.<sup>1</sup> Lucke first described malignant melanomas in 1869. Of all melanomas primary mucosal melanomas (MMM) are rare tumors and only 0.5% malignant melanomas arise in the sinonasal mucosa.<sup>2</sup> Approximately 20% of all melanomas occur in the head and neck, with less than 10% from the mucous membranes of the aerodigestive tract.<sup>3</sup> MMM has more aggressive clinical course.<sup>4</sup> The median age of patients with head and neck mucosal melanoma is approximately 60 years. There is a modest male preponderance.<sup>5</sup> MMM of nose and paranasal sinuses are highly aggressive

tumor.<sup>6</sup> MMM arising in the nasal cavity or paranasal sinuses have a poor prognosis. The nasal cavity is more commonly affected than the paranasal sinuses, and the maxillary antrum is more frequently involved than the ethmoid sinuses.<sup>7</sup> The symptoms of malignant mucosal melanoma are nonspecific and thus, diagnosis is often delayed. The most common clinical presentations of nasal cavity melanomas are epistaxis and nasal obstruction.<sup>8</sup> Malignant melanomas may be misdiagnosed as a polyp or an inverted papilloma, especially when amelanotic. Histopathology report with immunohistochemistry gives accurate diagnosis.<sup>9</sup> Here in we report a case of primary mucosal melanoma of the nasal cavity and paranasal sinus histomorphologically.

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## Case Report

A 60 years old male patient presented with the complaints of swelling in the left nasal cavity causing nasal blockage, obstruction to breathing and epistaxis since 6 months. He also complaints of headache, right eye pain and vision disturbances. He had no other significant medical or surgical history.

On examination, the swelling was a reddish pink colour with brownish discolouration at places, soft in consistency, friable, bleeds on touch; filling the left nostril and protruding through it. There was no lymphadenopathy. Routine investigations of blood, urine, X-ray chest and ECG were normal except mild anaemia. X-ray PNS revealed a homogenous opacity in the left nasal cavity and left maxillary sinus with lateral deviation of nasal septum. Computed tomography (CT) scan showed homogenous enhancing soft tissue lesion in the nasal cavity extending posteriorly in nasopharynx, medially

destroying nasal septum and laterally into left maxillary sinus with suggestive of malignant mass. Patient was taken up for surgical removal of mass under general anaesthesia and the mass was sent for histopathology. Gross examination revealed several bits of blackish irregular, soft, friable tissue; total of (6X5) cm in size. The Haematoxylin & Eosin stained section showed diffusely distributed sheets of tumour cells. Tumour cells are spindle with hyperchromatic nuclei, prominent nucleoli and moderate amount of cytoplasm along with brownish-black melanin pigment in their cytoplasm and melanophages. The tumour cells are arranged in fascicles. Histological features are consistent with malignant melanoma. The patient were evaluated for any other sources of melanoma/or nevus. Immunohistochemistry was not possible due to unavailability in our setup. So, the case was diagnosed as a primary mucosal melanoma of nasal cavity histomorphologically.

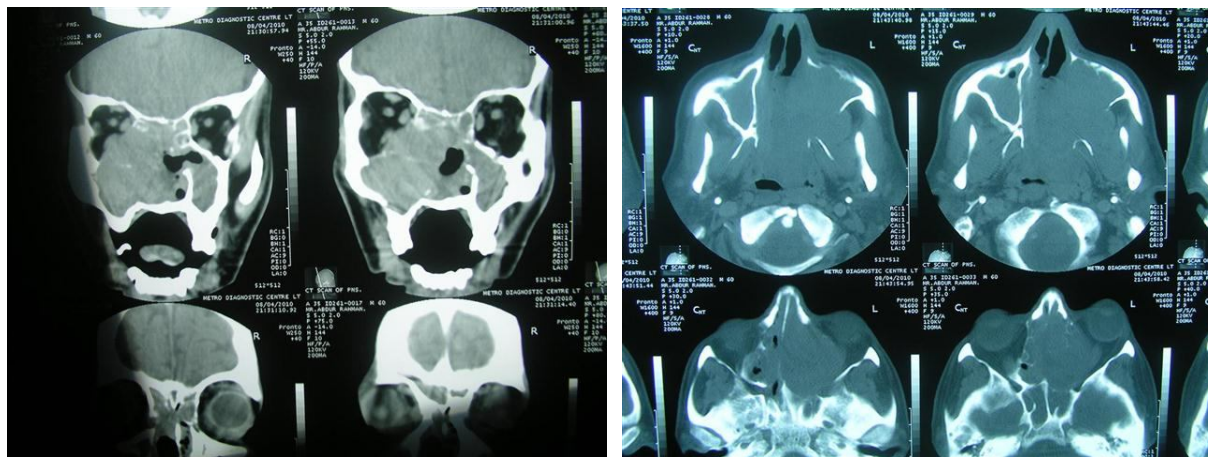


Fig 1&2: CT image of nasal melanoma involving left nasal cavity and paranasal sinus

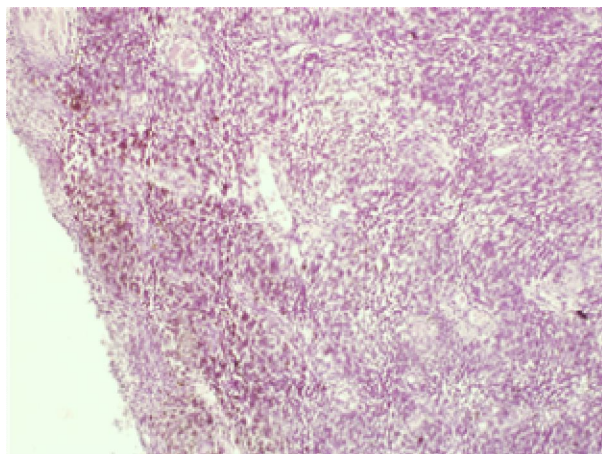


Fig 3: Polypoid melanoma (40X)

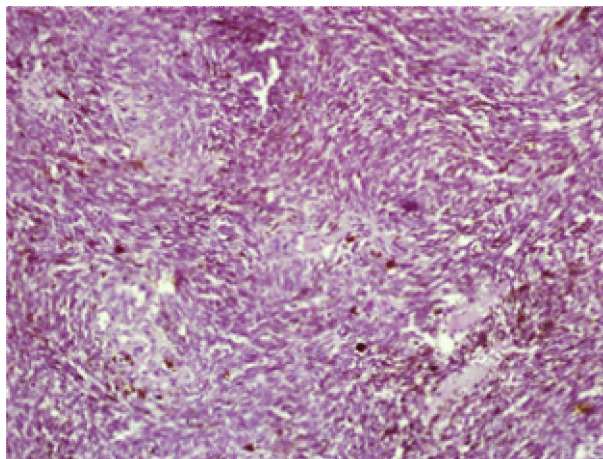


Fig 4: Melanoma cells in fascicles (100X)

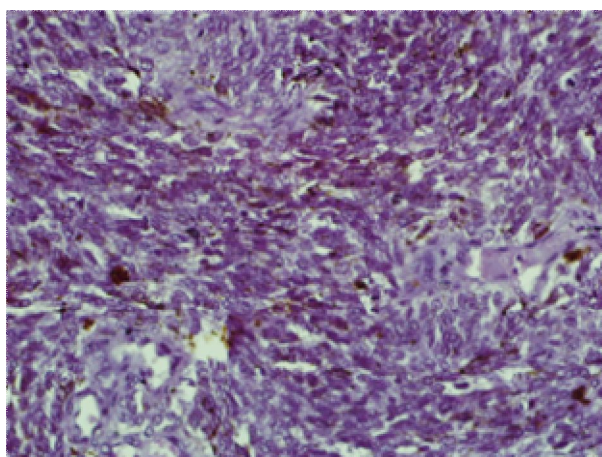


Fig 5: Intracellular melanin, melanophages

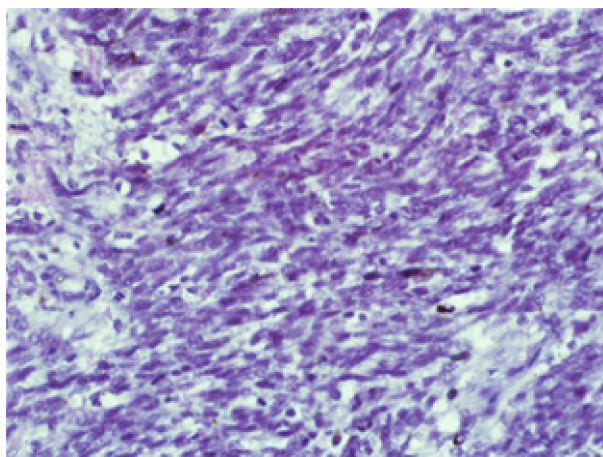


Fig 6: Multiple mitotic figures (400X)

### Discussion

Malignant mucosal melanoma is an uncommon devastating disease. At the time of presentation, the lesions are at an advanced stage which lowers the survival rate of patients. The incidence of malignant melanoma of the head and neck and nasal cavities varies from 0.4%-4%. Sino nasal malignancies are difficult to treat and have a poor prognosis. One reason for the poor prognosis is the close proximity to vital structures such as the skull base; brain, orbits, and carotid artery.<sup>10</sup> There are no specific etiologic factors though formaldehyde exposure and tobacco smoking have been implicated in the pathogenesis. MMM may present as polypoidal or sessile, friable to rubbery lesions with or without ulceration.

The size varies from 10 mm to >50 mm. The colour ranges from brown, black, pink or white, depending upon the amount of melanin production.<sup>4</sup> In the nasal cavity, the most frequent site of occurrence is the nasal septum, lateral wall, inferior turbinate and rarely floor and roof of nose. Its presence in paranasal sinuses is due to its extension.<sup>1</sup> Diagnostic evaluation includes a thorough physical examination of the head and neck including assessment of overall facial asymmetry, extraocular muscle evaluation, pupillary response, and signs of globe displacement. Intranasal exam may include flexible or rigid endoscopy for visualization of the nasal cavity. Imaging studies are usually obtained prior to biopsy. Both CT and MRI may be utilized to further characterize

sinonasal malignancies. Ultimately biopsy is the only accurate mean of obtaining tissue diagnosis.<sup>10</sup> Histologically their microscopic appearance is quite variable, and three general cell types are described; small polygonal, large polygonal, and spindle shaped. The nuclei are often large and vesicular, with prominent nucleoli. Multi nucleated tumour giant cells may be present. They have a higher incidence of pleomorphism and mitotic figures than their cutaneous counterparts.<sup>2</sup> On histopathological examination, melanoma mimics many other tumours including lymphoma, sarcoma, and poorly differentiated carcinoma. So, pathological diagnosis hinges on identification of intracellular melanin. However demonstrable melanin is seen in 50-70% cases.<sup>6</sup> A diagnosis of melanoma depends on identification of melanin pigment that is confirmed by Fontana-Masson silver stain and the appropriate immunohistochemical staining pattern that included HMB-45 antibodies, Melan-A, tyrosinase and antimicrophthalmia transcription factor. S-100 protein is always positive in melanomas.<sup>4</sup> MMM tends to be aggressive tumour and overall prognosis and survival rate ranges between 10-40% with mean survival rate ranges being 21-24 months. Poor prognostic factors include local and distant metastasis, local recurrence, vascular invasion and a second primary. Single most powerful predictor is absence of regional lymph nodes. The mainstay of treatment is wide surgical excision; chemotherapy and radiotherapy being not very effective.<sup>1</sup>

Primary mucosal malignant melanomas of the nasal cavity are a rare, aggressive tumour with poor prognosis and late detection. So, early diagnosis with high index of suspicion is essential for the management of condition.

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