



## CASE REPORT

# Sinonasal amelanotic malignant melanoma – A diagnostic dilemma



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### KEYWORDS

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**Abstract** Malignant melanoma involving mucosal surface of the head and neck region is rare. Malignant melanoma involving the nasal cavity is even rarer and accounts for less than 1% of cases. Prognosis is generally poor with unpredictable outcome. Regional and distant metastasis is common at presentation. We present a case of a 60 year old female who presented with polypoidal mass in the left nasal cavity and occasional history of nasal bleed. Endoscopic excision of the mass was done with a clinical diagnosis of inverted papilloma. Microscopic examination of the lesion was suggestive of amelanotic malignant melanoma and was confirmed by immunohistochemistry. She subsequently received chemotherapy and radiotherapy.

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## 1. Introduction

Malignant melanoma is a highly lethal neoplasm arising from the neural crest derived melanocytes and primarily affects the skin. 15–20% of all malignant melanoma occur in the head and neck region of which 80% are cutaneous lesions.<sup>1</sup> Malignant melanoma involving the mucosal surfaces in head and neck region is rare. Upper aero digestive tract melanoma accounts for 3–5% of all head and neck tumors.<sup>2</sup> Malignant melanoma involving the sino nasal cavity is even rarer and accounts for less than 1% of all head and neck tumors.<sup>3</sup>

Sinonasal malignant melanoma usually presents with non-specific nasal complaints leading to diagnostic dilemma. They were usually misdiagnosed as carcinomas and inverted papilloma's because they share common age and presentation. Amelanotic melanomas are much more difficult to diagnose because of their appearance. In such a case immunohistochemistry is useful for confirmation of the diagnosis.

Malignant melanoma is aggressive and has regional lymph nodal and distant metastasis at presentation. Wide local excision with radiotherapy is the mainstay of therapy. Prognosis is poor with a mean survival of 3.5 years due to local recurrence, nodal involvement, and distant organ metastasis occurring months or years after the initial diagnosis.

We are presenting a case of amelanotic malignant melanoma of the nasal cavity in a 62 year old female which was excised endoscopically and the patient was given postoperative radiotherapy with chemotherapy after ruling out regional and distant metastasis.

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## 2. Case report

A 60 year old lady presented to the outpatient department with complaints of left side nasal obstruction and nasal discharge for 2 months. Nasal discharge was mucopurulent with occasional blood tinged. She complained of left nasal bleed for twenty days. The nasal bleed was small in amount and used to stop on its own. There was history of forward protrusion of the left eye for 1 month with no complaints of decreased vision or blurred vision. There was no history of headache, sneezing or facial pain.

Anterior rhinoscopy showed a reddish pink friable mass filling the left nasal cavity with minimal discharge on the floor of nasal cavity (Fig. 1). There is mild proptosis of the left eye. Visual acuity and extra ocular movements were normal in the left eye. Posterior rhinoscopy showed the same mass filling the left posterior choana. There was no regional lymphadenopathy, no pigmented lesions elsewhere and no history of regression of the previous pigmented lesion.

Contrast enhanced CT para nasal sinuses of the patient revealed a heterogeneously enhancing lesion filling the left side of the nasal cavity, maxillary sinus, anterior and posterior ethmoids and sphenoid sinuses. There is no evidence of intra orbital or intracranial extension (Fig. 2). Biopsy of the mass was done with clinical suspicion of carcinoma and inverted papilloma of the nasal cavity. It was reported as malignant melanoma. USG abdomen and neck was done to rule out regional and distant metastasis. Chest X ray was normal.

Endoscopic excision of the mass was done under general anesthesia. Pinkish friable mass was seen filling the left nasal cavity, maxillary sinus and anterior and posterior ethmoids and sphenoid sinus. The specimen was sent for histopathology.

Gross examination showed multiple irregular grayish brown tissue pieces largest measuring  $2.5 \times 2 \times 2$  cm and the cut surface showed areas of necrosis. No pigment was identified on overall gross examination of the tissue specimen.

Microscopic examination of tumor showed tumor tissue to be arranged in the form of sheets. Tumor cells were moderately pleomorphic with eosinophilic cytoplasm and nuclei were oval to round with coarse stippled chromatin, prominent nucleoli. Mitotic figures are 3 to 4/10 HPF. No melanin pigment was identified. Immunohistochemistry showed strong positivity for S 100. Few of them showed weak HMB 45. Cytokeratin and SMA were negative. Overall features are those of amelanotic malignant melanoma (see Fig. 3).



**Figure 1** Anterior rhinoscopy showing a reddish polypoidal mass in the left nasal cavity.

She has received postoperative radiotherapy along with chemotherapy. She is in regular follow up for the last 6 months and has no recurrence.

## 3. Discussion

Malignant melanomas are neural crest-derived neoplasms originating from melanocytes and demonstrating melanocytic differentiation. 91% of all malignant melanomas were cutaneous, 5% ocular and only 1.3% are mucosal in origin.<sup>1</sup> The most common site for the mucosal melanomas in head and neck region is the upper aero digestive tract. Upper aero digestive tract melanoma accounts for 1–3% of all malignant melanoma.<sup>2</sup> Sinonasal malignant melanomas are extremely rare. In 1967, Lewis and Martin reported an incidence of 0.67% for nasal mucosal melanomas among Caucasians with melanoma.<sup>3</sup>

Our patient is a sixty year old lady and has a history of prolonged exposure to sunlight. There was no history of addictions. Although exposure to UV radiation is a risk factor for cutaneous forms of melanoma it is not for mucosal melanomas.<sup>4</sup> Mucosal melanomas tend to occur in an older age group than their cutaneous counterpart (from the fifth to eighth decade) and men are more commonly affected than women.<sup>5</sup>

In the nasal cavity the most common site for malignant melanomas is the nasal septum followed by the lateral nasal wall but in our case, the origin was most probably from the lateral nasal wall.<sup>6,7</sup> Most common presenting complaint is nasal obstruction as in any other nasal mass. Any tumor with a variable degree of blackish discoloration is a suspect for malignant melanoma but amelanotic forms appear as a friable reddish pink angry looking mass and hence can cause diagnostic confusion with carcinoma and inverted papilloma. Mucosal malignant melanomas usually invade the underlying bone by the time of their presentation.<sup>8</sup> But, in our case, high resolution bone cuts of CT scan do not reveal any bone erosion.

Microscopically in malignant melanoma, most tumor cells appear pigmented and hence diagnosis of melanin rich melanoma is not difficult. Melanin pigment deposition can be demonstrated by Fontanna Mason staining.<sup>9</sup> But diagnostic confusion occurs in amelanotic melanoma. In malignant melanoma, tumor cells are arranged in sheets with oval to round nuclei and prominent nucleoli. Similar histological picture can be found in many other small round cell tumors and sino nasal undifferentiated cancers.<sup>9</sup> Immunocytochemistry remains the gold standard to differentiate these tumors. Immunohistochemistry using S-100 and HMB-45 is more sensitive in differentiating malignant melanomas from other neoplasms.<sup>10</sup> In our case both S 100 and HMB 45 were used and both these markers were positive. Though there was no identifiable melanin pigments in the specimen examined, IHC markers helped in diagnosing amelanotic malignant melanoma.

Malignant melanomas are highly aggressive tumors and have an unpredictable course with poor prognosis. Radical excision of the primary tumor is the main stay of treatment. Malignant melanoma is relatively chemo resistant and radio resistant tumor. Postoperative radiotherapy/chemotherapy is given as adjuvant therapy to control the disease.<sup>11</sup> Endoscopic excision of the tumor was done in our case as there was no evidence of bony destruction on the CT scan. There

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