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Nasopharyngeal amelanotic melanoma (NAM): A case report

Bandana Mehrotra[(1)], Ashok Kumar Kapoor[(2)], Shubhi Pandey Sharma[(3)], Mithlesh Bhargava [(4)], Supriya Mehrotra[(5),] Aditi Dixit[(6)]

(1) Pathologist, Department of Pathology, RML Mehrotra Pathology Pvt. Ltd,, Nirala Nagar, Lucknow, Uttar Pradesh, India.
(2) Pathologist, Department of Pathology, RML Mehrotra Pathology Pvt. Ltd,, Nirala Nagar, Lucknow, Uttar Pradesh, India.
(3) Pathologist, Department of Pathology, RML Mehrotra Pathology Pvt. Ltd,, Nirala Nagar, Lucknow, Uttar Pradesh, India.
(4) Pathologist, Department of Pathology, RML Mehrotra Pathology Pvt. Ltd,, Nirala Nagar, Lucknow, Uttar Pradesh, India.
(5) Pathologist, Department of Pathology, RML Mehrotra Pathology Pvt. Ltd,, Nirala Nagar, Lucknow, Uttar Pradesh, India.
(6) Pathologist, Department of Pathology, RML Mehrotra Pathology Pvt. Ltd,, Nirala Nagar, Lucknow, Uttar Pradesh, India.

Abstract
Introduction: *Nasopharyngeal amelanotic melanomas (NAM) are extremely rare malignant neoplasms. Several prognostic factors, e.g. aggressive treatment, a few mitoses, lymphocytic tumour infiltration; treatment with IL-2 activated killer T-cells and genetically-altered tumour cells may improve the outcome of the patients. Herewith, we report a case of nasopharyngeal amelanotic melanoma.*
Case report : *A male, aged 32 years complained of left nasal obstruction and occasional bleeding from left nostril. He was operated and tumour was excised. Grossly, tumour comprised of multiple small reddish brown soft tissue pieces together measuring 2×2×1cm. All the pieces were sectioned. Microscopically, tumour cells were arranged in sheets or disposed diffusely or at places surrounding ill defined alveolar spaces and showed moderate anisocytosis. Tumour cells had round to oval eccentrically situated nuclei and fair amount of eosinophilic cytoplasm. No identifiable melanin pigment was seen in tumour cells. Immunohistochemistry (IHC) showed strong positivity with anti-S 100, anti-HMB 45 and anti-vimentin antibodies. Weak positive reaction was seen with anti-pan cytokeratin antibody. Negative reaction was obtained with anti-MelanA antibody.*
Negative reaction was also obtained with anti-EMA antibody which ruled out epithelial origin of the tumour. In addition, negative reaction with anti-synaptophysin ruled out neuroendocrine origin of the present neoplasm. Negative reaction was also obtained with anti-MyoD1 which ruled out muscular origin of current neoplasm. Additionally, negative reaction was obtained with anti-CD45 LCA which ruled out lymphomas. Further, negative reaction with anti-CD138 ruled out plasmacytoma.
Conclusion: *Current patient had a polypoidal mass in left nostril. He complained of nasal obstruction and occasional bleeding from left nostril. Patient was finally diagnosed as nasopharyngeal amelanotic melanoma.*
Keywords: *Mucosal malignant neoplasm, melanocyte, neural crest*

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I. Introduction

Nasopharyngeal amelanotic melanoma (NAM) is an extremely rare neoplasm. Its incidence is ~ 1.8% to 8.1% of all melanomas[1]. First patient with NAM was reported by Lucke in the year 1869 [2]. Later, Kutty and Shreedhar reported another case of NAM from India [3]. Most of the patients belong to older age-group; age ranging between 50 to 70 years. However, current patient belonged to younger age-group; he was aged 32 years. He complained of nasal obstruction and occasional bleeding from left nostril. He was operated. Excised tumour was sectioned and examined. Microscopy revealed tumour cells arranged in sheets, disposed diffusely and at places surrounding ill-defined alveolar spaces. IHC examination showed strong positive reaction with anti-S100, anti-HMB 45 and anti-vimentin antibodies. Weak positive reaction was obtained with anti-pan cytokeratin antibody. Negative reaction was obtained with anti-MelanA, anti-EMA, anti-Synaptophysin, anti-MyoD1, anti-CD45 LCA and anti-CD138 antibody. Strong positive reactivity with anti-S100 and anti-HMB 45

suggested the origin of tumour cells from the neural crest[4]. Tumour was finally diagnosed as nasopharyngeal amelanotic malignant melanoma.

II. Case Report :

A 32-year-old male complained of left nasal obstruction and occasional bleeding from left nostril. He was operated and tumour was excised. Grossly, tumour comprised of multiple small reddish brown soft tissue pieces, together measuring 2×2×1cm. All the pieces were sectioned. Histopathologically, sections showed tumour cells arranged in sheets or disposed diffusely or at places surrounding ill-defined alveolar spaces. Tumour cells showed moderate anisocytosis and had round to oval eccentrically placed nuclei and fair amounts of eosinophilic cytoplasm. Areas of necrosis were seen. The underlying stroma showed infiltration by chronic inflammatory cells. No identifiable melanin pigment was seen in tumour cells. Based on the morphological features, the following differentials were considered: poorly differentiated neoplasm, alveolar rhabdomyosarcoma, plasmacytoma, lymphoma and melanoma. Subsequently, IHC was performed.

In view of the following differentials, IHC was performed first with Pancytokeratin(PCK), CD45LCA, CD138, EMA, Vimentin, out of which vimentin showed strong and diffuse immune-reactivity, PCK showed weak positivity which was an aberrant presentation while the rest of the markers were negative ruling out poorly differentiated carcinoma, lymphoma and plasmacytoma. Further IHC was performed with synaptophysin, S-100, MyoD1 out of which S-100 showed strong diffuse nuclear and cytoplasmic positivity while the others were negative ruling out neuro-endocrine origin of the tumor as well as the possibility of alveolar rhabdomyosarcoma. Finally, IHC was performed with HMB45 and MelanA, out of which HMB45 showed diffuse strong cytoplasmic positivity and MelanA was negative in tumor cells, confirming the diagnosis of amelanotic melanoma.

III. Discussion:

Mucosal melanomas are very rare neoplasms [1]. About 2% of melanomas may be amelanotic[5]. Nasal mucosal melanomas constitute <1% of all melanomas[6]. Mucosal amelanotic melanomas are known to have a poor prognosis as compared with cutaneous amelanotic melanomas[7], However, prognostic features of cutaneous and mucosal amelanotic malignant melanomas (AMM) appear similar. Mucosal AMM reach to dangerous vertical growth phase in terms of tumour thickness and depth of invasion at the time of diagnosis. In addition, mucosal melanomas appear to be refractory to treatment than cutaneous melanomas. Other poor prognostic features may be distant metastasis and vascular invasion. In a separate report, a patient with amelanotic melanoma gave history of recurrent tumours. However, the previous patient was successfully treated with Pembrolizumab[8]. Another important feature of current neoplasm may be local recurrence of the tumour. Recurrence may occur after 6 months or after several years following surgery[1,8,9]. Another patient gave history of chronic *Biri* smoking, tobacco chewing and inhalation of formaldehyde [10].

Present tumour had immunological features of melanoma without pigment production. Melanogenesis may require 3 important tyrosine-related proteins (TRP), e. g. tyrosinase, TRP-1 and TRP-2. Failure to produce melanin may suggest lack of one of these proteins[11]. Moreover, melanin may either protect the melanocytes from cellular toxins or it may augment tumour growth and tumour –progression[11]. Another patient gave history of prolonged exposure to sunlight. However, exposure to UV rays does not appear to be a risk factor[7]. Genetically altered tumour cells have been used as immunotherapy. Further, tumour-spread may occur to regional lymph nodes through lymphatics. Moreover, vascular invasion and spread may also occur in liver, lungs, bone and brain [10]. Additionally, one of the patients with NAM also developed Parkinson's disease [9].

IV. Conclusion:

A rare case of amelanotic melanoma of the nasal mucosa in a 32 year old male has been discussed in this case report. Mucosal melanomas are usually refractory to treatment and have a poorer prognosis than cutaneous melanomas. The diagnosis of this entity relies on clinical examination, radiological findings, H&E and immunohistochemical study of the biopsy of the lesion. Tumour extension assessment also holds significance for staging of the tumour and further treatment and management. Surgery with wide local excision is the treatment of choice for well localized tumours along with adjuvant radiotherapy. Potential of immunotherapy in the management of this tumour in still under research.

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Legends to Figures

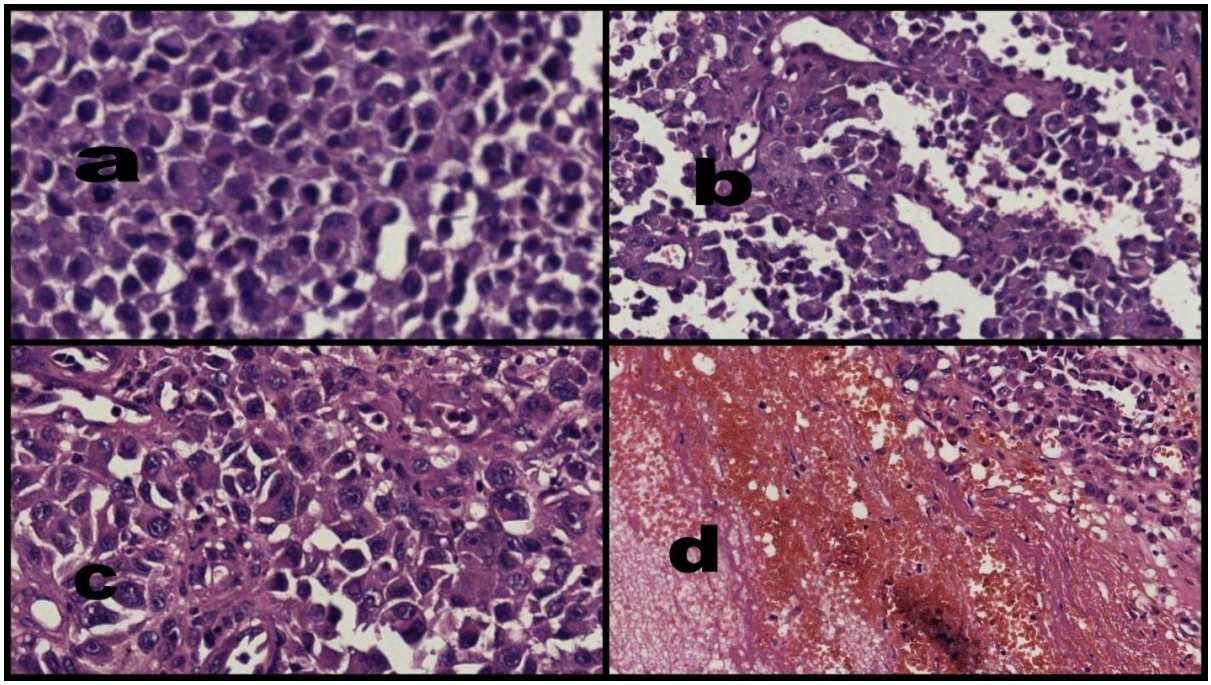

Figure 1 (a) Photomicrograph shows tumour consisting of sheets of proliferated tumour cells (HE×400). **(b)** Photomicrograph shows tumour cells arranged in ill- defined alveolus-like pattern (HE×400). **(c)** Photomicrograph shows cellular tumour. Tumour cells showed moderate anisonucleosis, mild nucleomegaly with round to oval eccentric nuclei and fair amount of eosinophilic cytoplasm (HE×400). **(d)** Photomicrograph shows areas of necrosis, haemorrhage and tumour (HE×100).

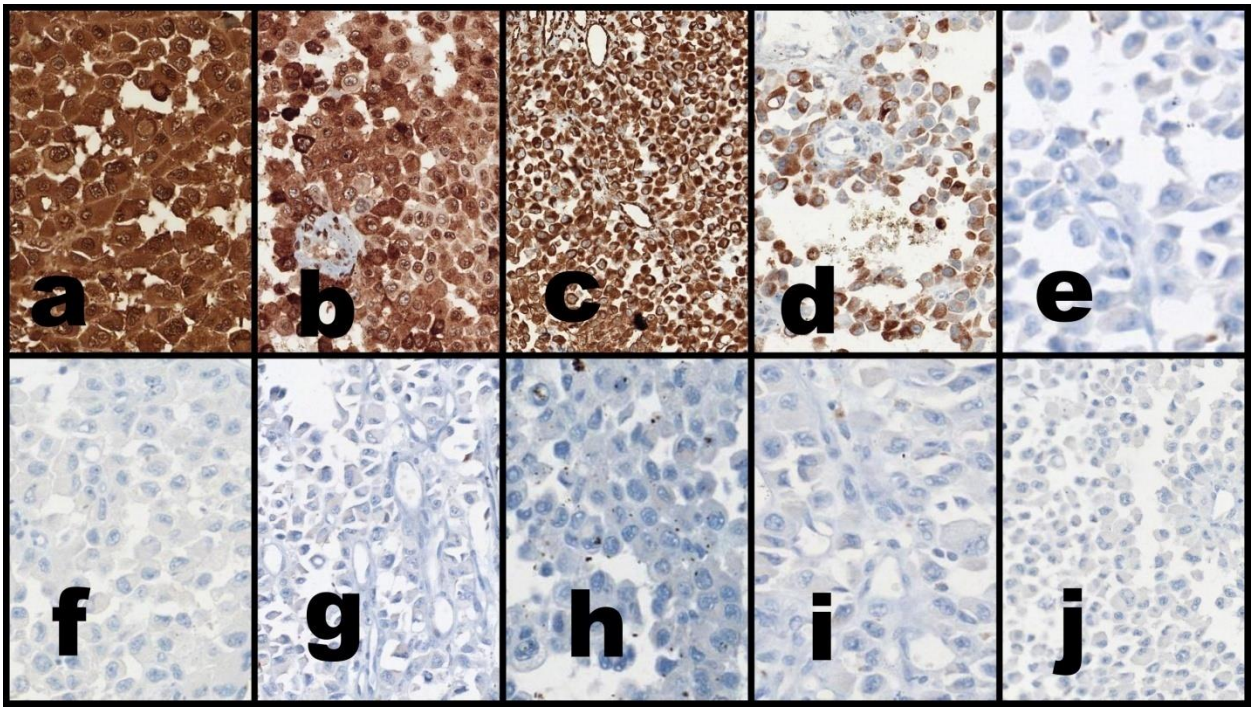

Figure 2 (a) Tumour cells show strong positivity with anti-S100 antibody. **(b)** Strong positivity of tumour cells with anti-HMB 45 antibody is seen. **(c)** Anti-vimentin antibody stained the tumour cells. Strong positive reaction is seen. **(d)** Anti-pan cytokeratin antibody showed mild (1+) reactivity with tumour cells. **(e)** Anti-CD 45 LCA antibody failed to stain the tumour cells. **(f)** Anti CD-138 antibody did not stain the tumour cells. **(g)** Anti-EMA antibody did not stain the tumour cells. **(h)** Negative reaction was obtained with anti-MelanA antibody **(i)** Anti-MyoD1 antibody did not stain the tumour cells. **(j)** Anti-Synaptophysin antibody gave negative reaction with the tumour cells.

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