

Intranasal Malignant melanoma: Pathogenesis Epidemiology and Clinical Insight

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Abstract: Malignant melanoma in nasal cavity and paranasal sinuses is a rare and aggressive tumor, comprising a small percentage of all melanomas. This article explores the pathogenesis and epidemiology of this condition, shedding light on its distinct characteristics from cutaneous melanoma. Despite understanding, the 5 year survival rate remains low emphasizing the need for effective diagnosis and treatment strategies. The case report of a 65 year old male with nasal cavity melanoma highlights the clinical presentation and diagnostic challenge associated with this malignancy. The article emphasizes the importance of thorough evaluation including fibroscopic examination and imaging techniques in achieving a conclusive diagnosis. Surgical intervention remains the primary treatment modality with chemotherapy and radiotherapy as adjuncts.

Keywords: Primary mucosal melanoma, Nasal cavity, Pathogenesis, Epidemiology, Clinical presentation, Treatment modalities

1. Introduction

Malignant melanoma is a rare tumour of the nasal cavity and paranasal sinuses accounting for 3.6% of all tumours in these sites and 0.5 - 2.0% of all melanomas.¹ Positive diagnosis of this tumour is made difficult by the non-specific presenting complaints.² Malignant Melanoma of nasal cavity is more aggressive than its cutaneous counterpart. Primary malignant melanoma of nasal cavity arise from melanocytes located in the mucous membrane. Only 0.5% of malignant melanoma arises in nasal cavity.³

The histogenesis of these tumours was in dispute until Zak & Lawson demonstrated melanocytes within the nasal mucosa. Prior to this it was thought that squamous metaplasia was a prerequisite for developing a malignant melanoma. Despite this there are still few reports of an *insitu* element present in association with invasive malignant melanoma.^{4,5}

It mainly occurs in the elderly and the presence of comorbidities can limit the extent of treatment⁶. Treatment options essentially consist of radical surgery and radiotherapy, while chemotherapy is reserved for advanced forms. Despite a better knowledge of this tumour, the 5-year overall survival remains poor and does not exceed 40% in any of the published studies.⁷

Pathogenesis

Melanocytes are dendritic cells that arise in the neural tube. They are located at the dermo-epidermal junction of all mucous membranes. Melanocytes are detected under normal conditions in about 21% of individuals.⁸ Mucosal melanoma is a neuroectodermal tumour arising from these melanocytes.⁹ No risk factor has yet been clearly identified to explain the development of these tumours. In contrast with melanoma of the skin, in which sun exposure is known to be the major risk factor, the risk factors for mucosal melanomas have not been identified. Exposure to formaldehyde has been suspected in several studies.¹⁰ This has not yet been confirmed. It has been hypothesized that smoking may constitute a predisposing factor essentially for mucosal melanoma of the oral cavity.² Few

genetic studies have witnessed gene mutations affecting the tyrosine kinase receptor.¹¹ Some authors have suspected the role of heredity and environment in the pathogenesis of mucosal melanoma in order to explain the different prevalence rates of these tumours between Caucasian (1% of melanomas) and Asian populations (7.5% of melanomas).⁷

Epidemiology

Primary mucosal melanoma of the nasal cavity and paranasal sinuses is a rare tumour, representing between 0.7 and 1% of all melanomas in Caucasian populations and between 4 and 8% of malignant tumours of the nasal cavity and paranasal sinuses.⁸ The prevalence is equal in both sexes.⁶ The patient's age at the time of diagnosis is generally between 60 and 80 years with a mean age between 65 and 70 years. Primary mucosal melanoma can arise in various anatomical sites, but it predominantly (55% of cases) involves the head and neck, in which the nasal cavity and paranasal sinuses is the most frequent site, representing 70% of cases (50% in the nasal cavity, 20% in the paranasal sinuses) followed by the oral cavity in about 17% of cases.²

American Joint Committee on cancer staging mucosal melanoma of the head and neck, 7th edition.

Primary tumor (T)

T3 Mucosal disease

T4a Moderately advanced disease; tumor involving deep soft tissue, cartilage, bone, or overlying skin

T4b Very advanced disease; tumor involving brain, dura, skull base, lower cranial nerves (IX, X, XI, XII), masticator space, carotid artery, prevertebral space, or mediastinal structures

Regional lymph nodes (N)

NX Regional lymph nodes cannot be assessed

N0 No regional lymph node metastases

N1 Regional lymph node metastases present

(M)

M0 No distant metastasis

M1 Distant metastasis present

Staging

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Stage III T3, N0, M0
 Stage IVA T4a, N0, M0
 T3–T4a, N1, M0
 Stage IVB T4B, any N, M0
 Stage IVC Any T, any N, M1

2. Case Report

A 65 - year - old male presented with nasal swelling, blockage and with occasional nasal bleeding from last 3 months. He noticed that a small swelling, approximately 1.5 cm in diameter, on the left side of nose about a month back which had rapidly increased in size. He had a smoking history from last 40 years. On examination, a mass was noticed in left nostril which was bluish - red friable and was completely blocking the nasal passage. Routine Biochemical and haematological investigations were carried out which all were in normal limits.

X - ray PNS

(Water's view) showed soft - tissue shadow over the nasal region.

Chest X - ray showed no abnormality.

CT scan findings suggested provisional diagnosis of inverted papilloma.

Intranasal biopsy and tumour debulking was performed.

3. Pathological Findings

Multiple grey brown soft tissue pieces were received in department of pathology that measured together 5.5x3.0x1.2cm. Largest soft tissue piece measured 3.5x2.0x0.5cm. On cut section surface appeared brownish.

Tissue sections were processed and subsequently stained with hematoxylin and eosin and studied under Magnus Mx21i LED microscope.

Microscopic findings: Sections were lined by pseudostratified ciliated columnar epithelium. Subepithelium showed tumor cells that were arranged in nests sheets and cords (Figure 2). Tumor cells were round to spindle in shape, pleomorphic with high N: C ratio, opened up chromatin, prominent eosinophilic nucleoli and moderate amount of cytoplasm (Figure 3, 4). Prominent melanin pigmentation was seen throughout the section (as is seen in Figure 1, 2) Stroma showed infiltration with inflammatory exudate.

Immunohistochemistry: S100 marker showed positivity for melanoma cells (Figure 5).

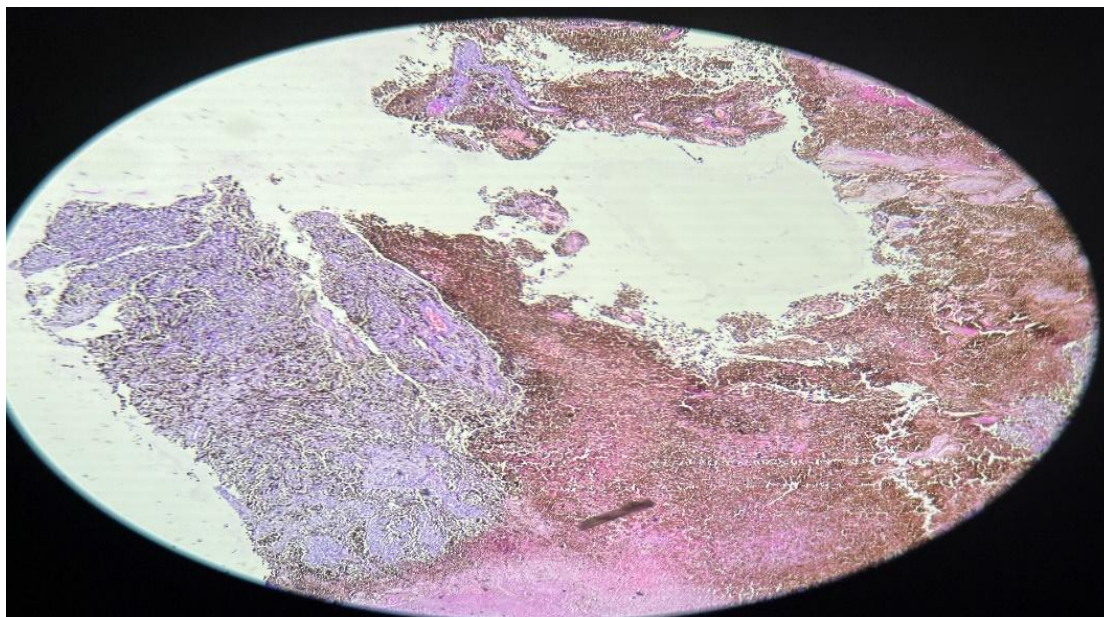


Figure 1: Scanner view showing prominent melanin pigmentation throughout the section

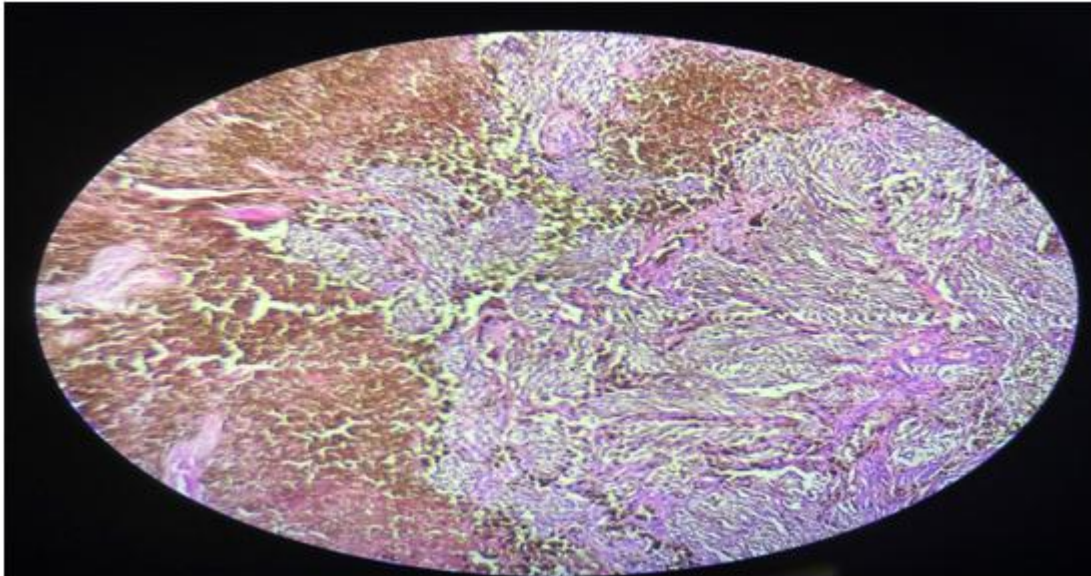


Figure 2: Low power view showing architectural arrangement of tumor cells

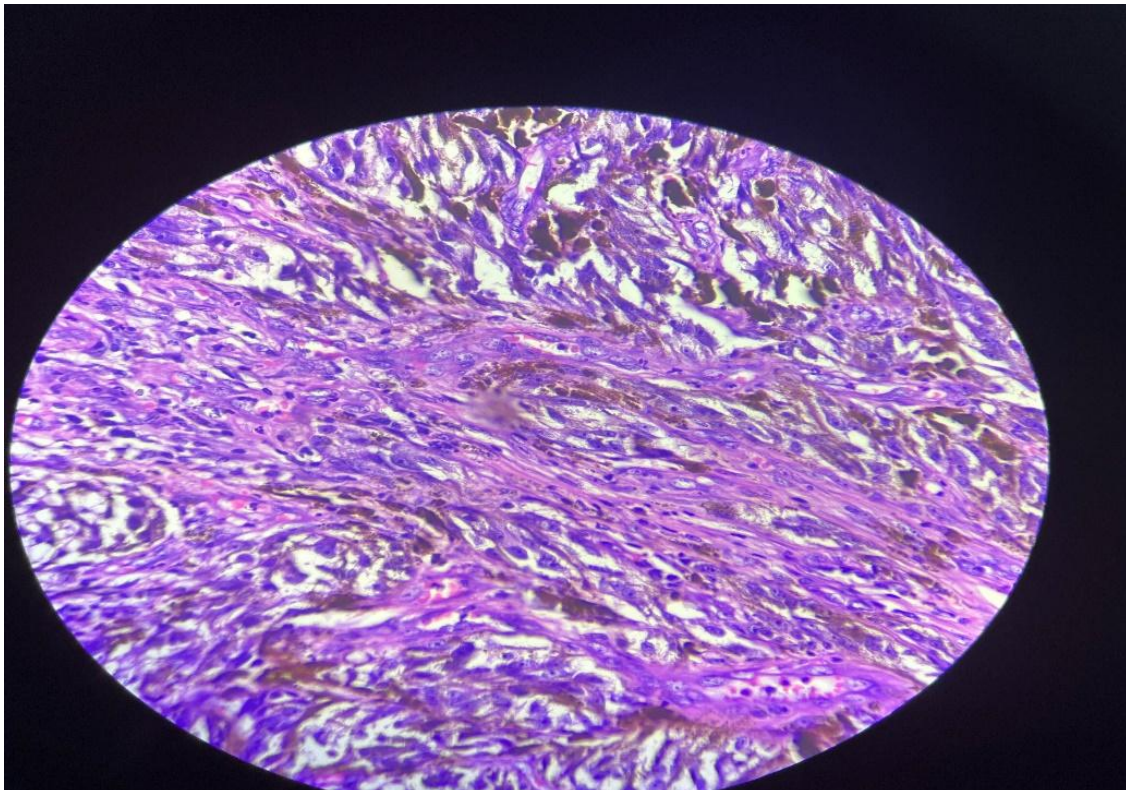


Figure 3: High power view showing cytological details along with melanin pigmentation

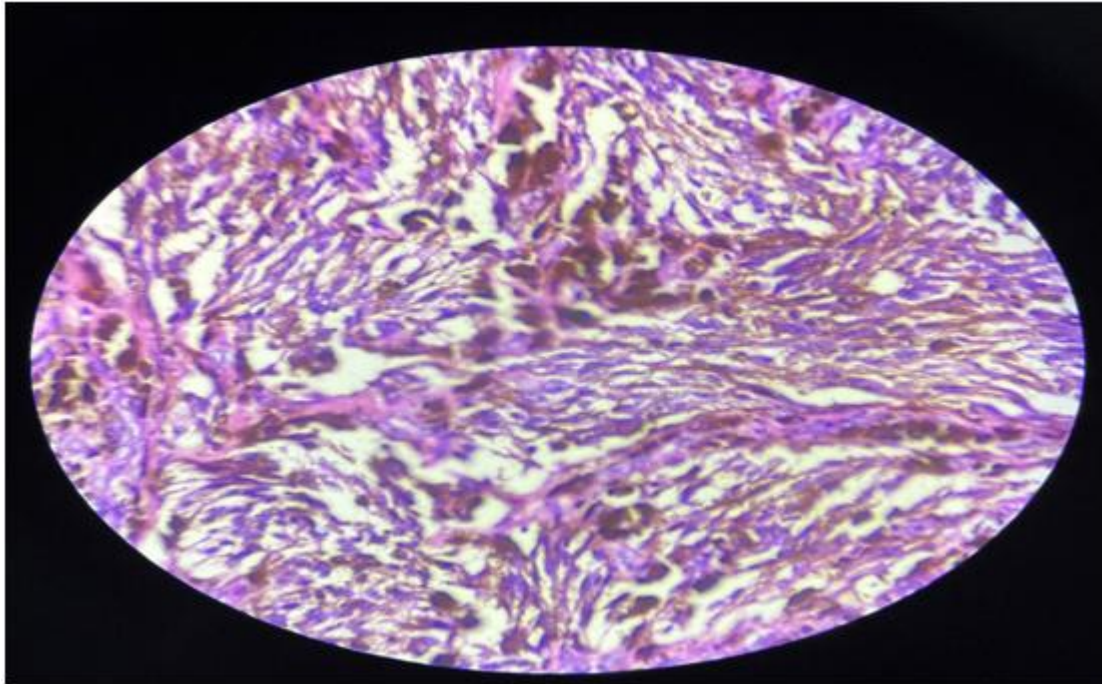


Figure 4: High power view showing cytological atypia in form of hyperchromasia, pleomorphism, high N: C ratio.

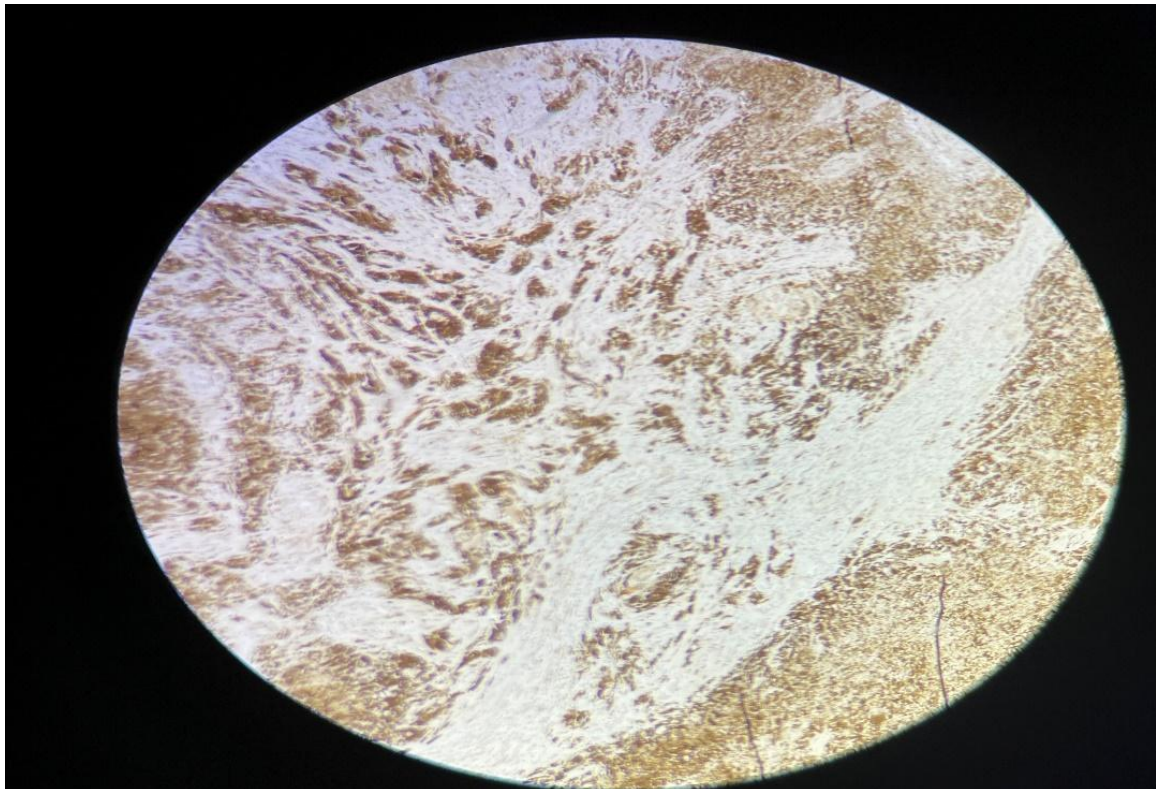


Figure 5: IHC positivity with S100 for melanoma cells

4. Conclusion

Malignant Melanoma of nasal cavity is an extremely rare tumour. Only 0.5% of malignant melanoma arises in nasal cavity.³ The most common presenting complaints are nasal obstruction and epistaxis. Nasal obstruction is unilateral, permanent and progressive, either isolated or associated with other symptoms.² Unilateral symptoms must be considered to be suspicious and justify thorough fibroscopic or endoscopic investigation of the nasal cavity. Intranasal examination defines the appearance of the

tumour (sessile, nodular, polypoid or granulating), its size and implantation. It may be slate - coloured, reddish, crimson, brownish or black, which is highly suggestive of the diagnosis.⁸ The tumour surface can be homogeneous or heterogeneous, with a friable consistency and the tumour may be covered by a greyish exudate. An imaging assessment comprising computed tomography (CT) of the facial bones and magnetic resonance imaging (MRI) is an essential part of the local staging of the tumour. Malignant melanoma is characterized by heterogeneous contrast enhancement. A spontaneous high - intensity

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signal on T1 with a low - intensity signal on T2 would be characteristic of melanoma. This unusual appearance, sometimes observed with other types of tumours (angiosarcoma, cylindroma and aesthesioneuroblastoma) appears to be related to the high melanin content and/or bleeding inside the tumour.² However pathological diagnosis remains confirmatory. Surgery remains the mainstay of treatment followed by chemotherapy and radiotherapy.

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