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Sinonasal Melanoma: A Rare Cause of Severe Nasal Bleeding

Leonard Derkyi-Kwarteng[1*], Kafui P. Akakpo[1], Ato A. Brown[2], Abigail N. Derkyi-Kwarteng[3] and Emmanuel Gustav Imbeah[1]

[1]Department of Pathology, UCC-SMS, Ghana.
[2]Department of Anatomy and Cell Biology, UCC-SMS, Ghana.
[3]Ewim Polyclinic, Ghana Health Service, Cape Coast, Ghana.

Authors' contributions

This work was carried out in collaboration among all authors. Author LDK coordinated the work. Authors LDK and KPA were the pathologists who independently reported the case. Authors AAB and ANDK were the clinicians who saw the case. Author EGI were the scientist who did the laboratory test. All authors wrote, read and approved the final manuscript.

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Case Reports

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ABSTRACT

Mucosal melanoma is an aggressive but very rare tumour that can occur within the nasal and paranasal sinuses. It accounts for less than 1.0% of all melanomas and 4.0% of all sinonasal tumours. We present a 48-year-old with a stage IVA nasal melanocytic melanoma who had surgery as primary treatment option.

Keywords: Melanoma; nasal mucosa; head and neck; tumour.

1. INTRODUCTION

Mucosal melanoma is an aggressive but very rare tumour that can occur within the nasal and paranasal sinuses. It accounts for less than 1.0% of all melanomas and 4.0% of all sinonasal tumours. [1]. Melanomas are malignant tumours that arise from melanocytes, a neuroectodermal

**Corresponding author: E-mail: l.derkyi-kwarteng@uccsms.edu.gh;*

derived cell that are found within the skin adnexa, basal layers of the skin and mucosal membrane [2,3]. Melanomas are usually common in sun exposed areas of the body which is a predisposing factor; with lower extremities, head and neck being the areas affected most [3-5]. Areas that are least affected by melanoma are leptomeninges, oral mucosa, nail bed, oesophagus, conjunctiva, vagina, genital mucosa, nasopharyngeal and nasal mucosa [2-4].

The median age of head and neck mucosal melanoma is 60 years with a wide range of occurrence varying from 20 years to over 90 years [2-5].

There are variable histologic appearance of head and neck mucosal melanoma ranging from epitheloid type, sarcomatoid (spindle cells), to plasmacytoid [2-5]. It may also vary in melanin content; from pigmented tumour to those that are amelanocytic [4,5]. Desmoplastic melanoma has also been described with features comprising of amelanocytic, poorly circumscribed fascicles and bundles of spindle cells with hyperchromatic nuclei [2] which are set within a fibrous stroma. These features make it difficult to distinguish it from other neoplasms like fibrosarcoma, peripheral nerve sheath tumours and spindle cell carcinoma [3-6].

Mucosal melanoma can be distinguished from other malignancies using immune histo chemistry. They usually stain positive for S-100, HMB-45, vimentin and negative for epithelial membrane antigen and cytokeratins [3,4].

We report the first sinonasal melanoma in West African population based on our literature search.

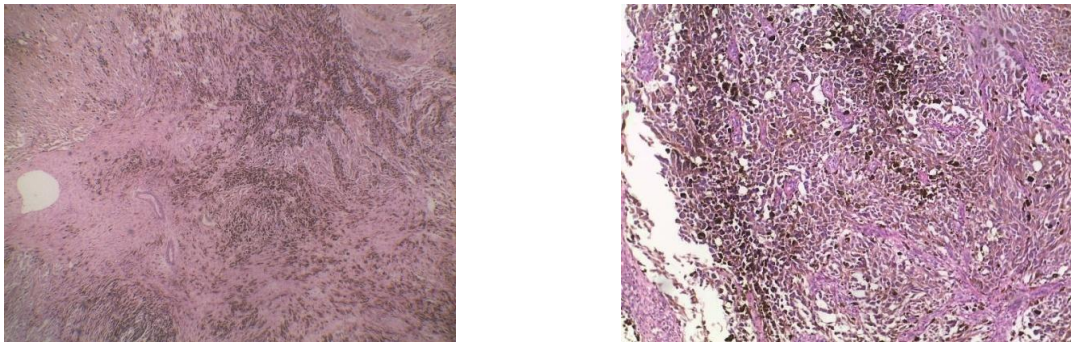
2. CASE REPORT

A 48-year-old female who presented with a five-month history of a mass in the right nostril which has increased in size over the period and was associated with recurrent epistaxis. She had no history of night sweats, weight loss or fever. Examination shows a black friable mass within the right nasal cavity attached to the floor and lateral nasal wall with splaying of the nasal bridge. An enlarged solitary right submandibular lymph node measuring 3x3 cm was palpated. After the initial blood work up and CT scan, the mass was excised with submandibular lymphadenectomy. The sample was sent for histopathology assessment.

2.1 Histopathology Findings

A dark brown mass in fragments was received with the largest fragment 5x5x2 cm. Accompanying the nasal excision was an encapsulated submandibular lymph node measuring 3.5x2.5x1.5 cm. Cut surfaces of the mass and the lymph node was homogenously dark brown.

Microscopy shows an ulcerated tumour with large areas of dark brown pigmentation. The tumour was composed of spindle cells set within a desmoplastic stroma. The nuclei of the tumour cells were markedly pleomorphic with tumour giant cells. There were frequent mitoses both normal and abnormal. The submandibular lymph node also shows a similar tumour. Immunohistochemistry for S 100 and HMB- 45 were all positive. A final diagnosis of right nasal malignant melanoma with right submandibular lymph node involvement (stage IVA), positive for both S100 and HMB- 45. Patient was lost to follow up during the post-operative period.



A B

Fig. 1. A microphotographs showing features consistent with pigmented malignant melanoma. A (H&E, 4x) and B (H&E, 10x)

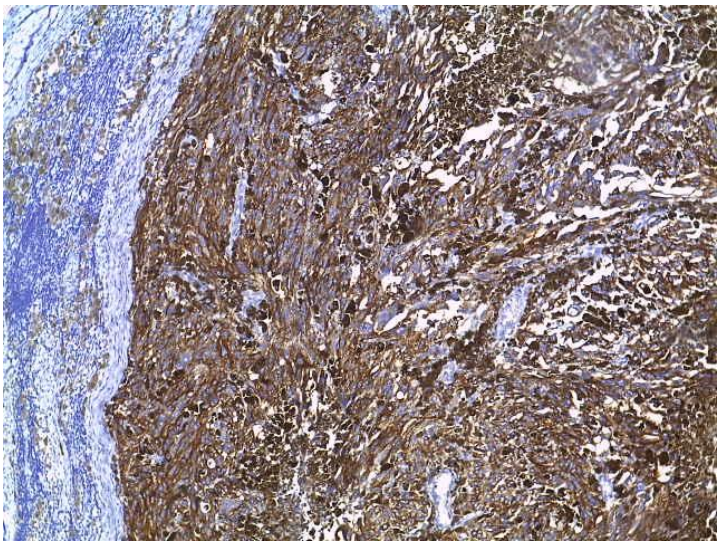


Fig. 2A. A microphotograph showing HMB positive tumour cells of malignant melanoma (40x)

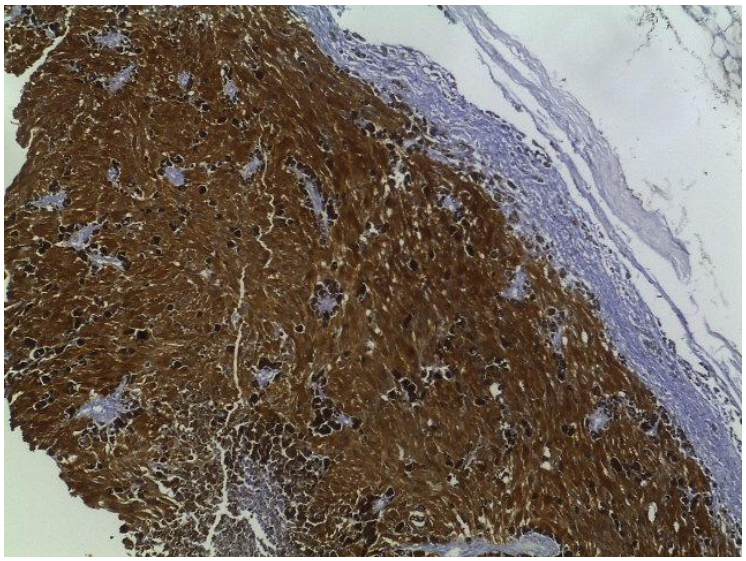


Fig. 2B. A microphotograph showing S100 positive tumour cells of malignant melanoma. (magnification 40x)

3. DISCUSSION

Melanomas are tumours that arise from the melanocytes which are derived from neuroectodermal cells of the basal layer of the epidermis of the skin and skin adnexa and some mucosal membrane [3,4] Although the cause of melanoma is mucosa is unknown, risk factors like formaldehyde exposure and smoking, play a role in activation of pre-existing melanocytes leading to melanogenic metaplasia [7,8]. Our patient presented with a nasal melanoma which is an area of rare occurrence with no history of smoking or formaldehyde exposure.

The occurrence of malignant melanoma of the nose was first described by Lucky in 1869 [2-4,9]. The incidence of malignant melanoma in the nose and paranasal sinuses between 0.5-1.0% which commonly occurs in the 5^{th}-6^{th} decade with equal sex distribution [9]. This patient meets the criteria for age and gender as described by literature.

The patient presented with epistaxis and obstruction of the nasal cavity which are the common presentations of nasal melanoma [2-4]. Other presentations are diplopia, pain proptosis, and facial deformity are less common and are indication of advanced disease [2-4].

The tumours cells were spindly in appearance with heavy pigmentation and were within a desmoplastic stroma background. Though head and neck mucosal melanoma has variable histological appearance, a sarcomatoid (spindle cell) tumour cell is one of them. The other forms of histologic appearance are plasmacytoid and epitheloid [2-4]. It may be pigmented (melanocytic) and non-pigmented (amelanocytic) [6,10-12]. Mucosal melanoma can be distinguished from other tumours using immunohistochemical stains. Immuno histochemically, melanomas stain positively with vimentin, S100, HMB 45 and negatively with cytokeratins and epithelial membrane antigen.

The various investigations for nasal melanoma include MRI, CT scan, chest X-ray, bone scan and/or positive emission tomography. Our patient had a CT scan done with showed a nasal mass [12].

Nasal melanoma can be staged using the AJCC staging system 8th edition [13]. Our patient has a stage IVA disease which corresponds to T4aN1Mx.

The patient underwent excision of the primary tumour which is the main treatment option for patients with head and neck melanoma [2-4]. Usually patients who undergo primary tumour excision receive postsurgical radiotherapy [2-4] but unfortunately our patient was lost to follow up. Patients with locally unresected tumours undergo definitive radiotherapy which may be for palliative or even cure in some cases [2].

4. CONCLUSION

Sinonasal melanoma is a rare mucosal tumour that occurs in the head and neck region presenting with epistaxis and nasal obstruction. Although rare, sinonasal melanoma should be considered as a differential diagnosis in patients presenting with intranasal mass that present with obstruction and epistaxis and mean age of 60 years. This tumour has a varying histological and stain usually positive to S100, HMB 45 and vimentin and negative for cytokeratin and EMA. The mainstay of treatment is surgery and postsurgical radiotherapy. The features are similar in the African population.

CONSENT

Consent for this publication was obtained from the patient.

ETHICAL APPROVAL

As per international standard, written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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