SINONASAL MELANOMA: A RARE CAUSE OF SEVERE NASAL BLEEDING

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% of all melanomas and 4% of all sinonasal tumours. We present a 48year old with a stage II nasal melanocytic melanoma who was treated with primary surgery of the mass.

Keywowords: melanoma, nasal mucosa, head and neck, tumour.

Running theme: Sinonasal melanoma

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% of all melanomas and 4% of all sinonasal tumours. [1, 2] Melanomas are malignat tumours that arise from melanocytes, a neuroectodermal derived cell that are found within the basal layers of the skin, skin adnexa, and mucosal membrane. [3] Melanomas are usually common around the head, neck, and lower extremities. These areas are exposed to sunlight, which is one of the predisposing factors. Areas that are least affected by melanoma are oral and genital mucosa, nail bed, oesophagus, conjunctiva, leptomeninges, vagina, nasopharyngeal and nasal mucosa.[3]

Head and neck mucosal melanoma has a wide range of varying occurrence between 20years to more than 90years with median age of patients being approximate 60years.[4]

There are variable histologic appearance of head and neck mucosal melanoma. The cells range from plasmacytoid, sarcomatoid (spindle cells) to epitheloid.[4,5] It may also vary in melanin content from pigmented tumour to those that are amelanocytic. Desmoplastic melanoma has also been described with features comprising of amelanocytic, poorly circumscribed fascicles and bundles of spindle cells with hyperchromatic nuclei which are set

within a fibrous stroma. These features make it difficult to distinguish it from other neoplasms like including fibrosarcoma, peripheral nerve sheath tumours and spindle cell carcinoma .[4-7]

Mucosal melanoma can be distinguished from other malignancies using immunohistochemistry. They usually stain positive for S-100, HMB-45, vimentin and negative for epithelial membrane antigen and cytokeratins.[4, 5]

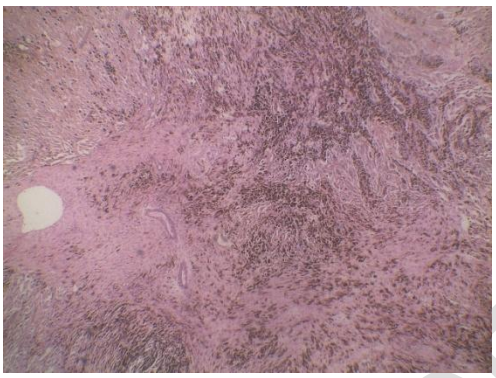
CASE REPORT

A 48year old female who presented with a five month history of a mass in the right nostril which had 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. A solitary right submandibular lymph node was palpated it was firm, mobile and not attached to the underlying structures or overlying skin and measured about 3×3cm. After the initial blood work up and CT scan, the patient was schedule for nasal mass excision with the right submandibular lymph node excision. The sample was sent for histopathology assessment.

Histopathology Findings

A dark brown mass in fragments was received with the largest fragment 5x5x2cm and the rest of the fragments measured 5x5cm in aggregate. Accompanying the nasal excision was an encapsulated submandibular lymph node measuring 3.5x2.5x1.5cm. Cut surfaces of the mass and the lymph node was homogenously dark brown.

Microscopically was an ulcerated tumour with large areas of dark brown pigmentation. The tumour was composed of spindle cells set within a desmoplatic stroma. The nuclei of the tumour cells were markedly pleomorphic with areas of tumour giant cells. Mitoses were frequent. Sections of the submandibular lymph node show 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 II), positive for both S100 and HMB- 45. Patient was lost to follow up during the post-operative period.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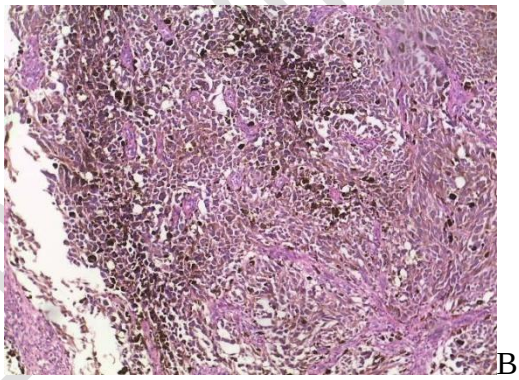
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FIG 1 A micrograph of the tumour with H&E stain showing pigmentation. A is X4 magnification and B is X10 magnification.

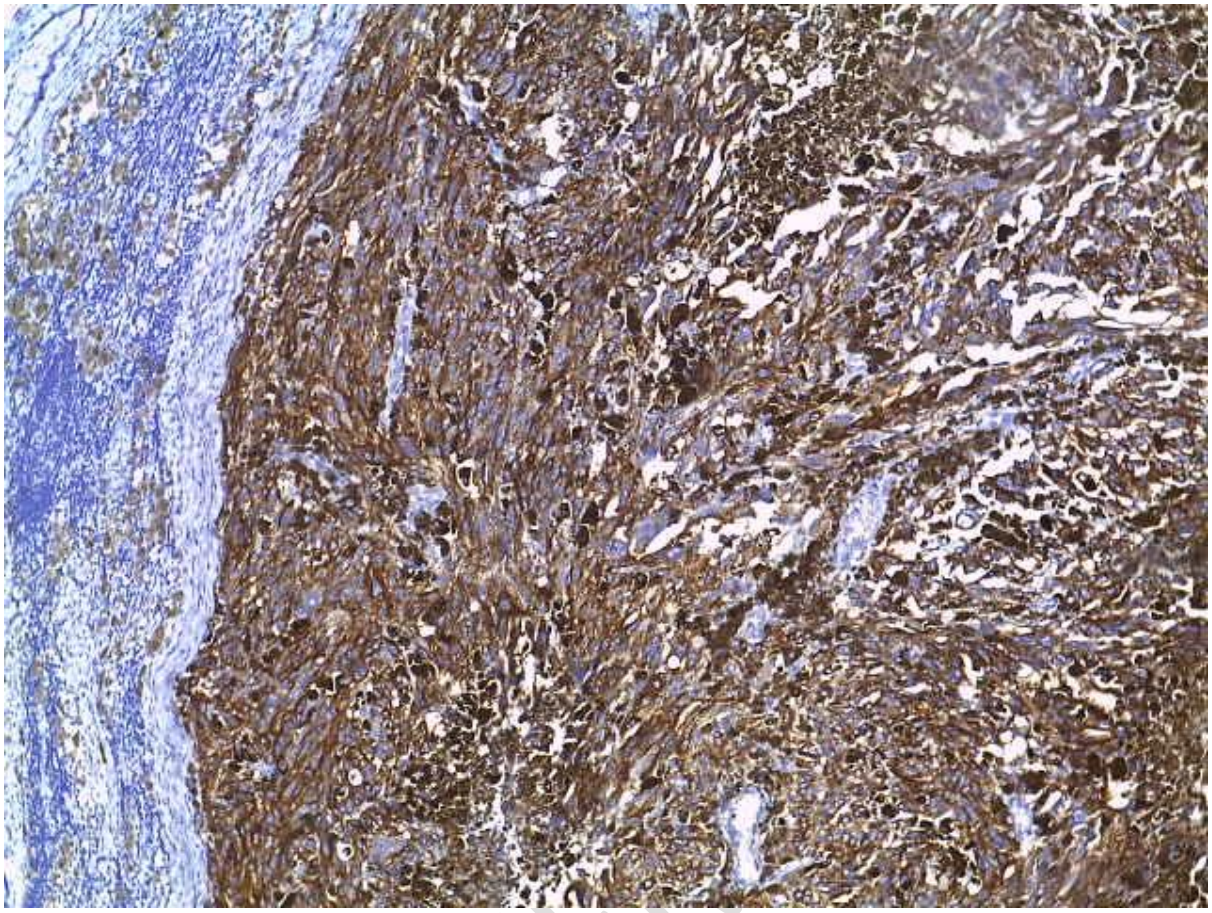

FIG 2A. A micrograph showing HMB positive tumour cells at X10 magnification.

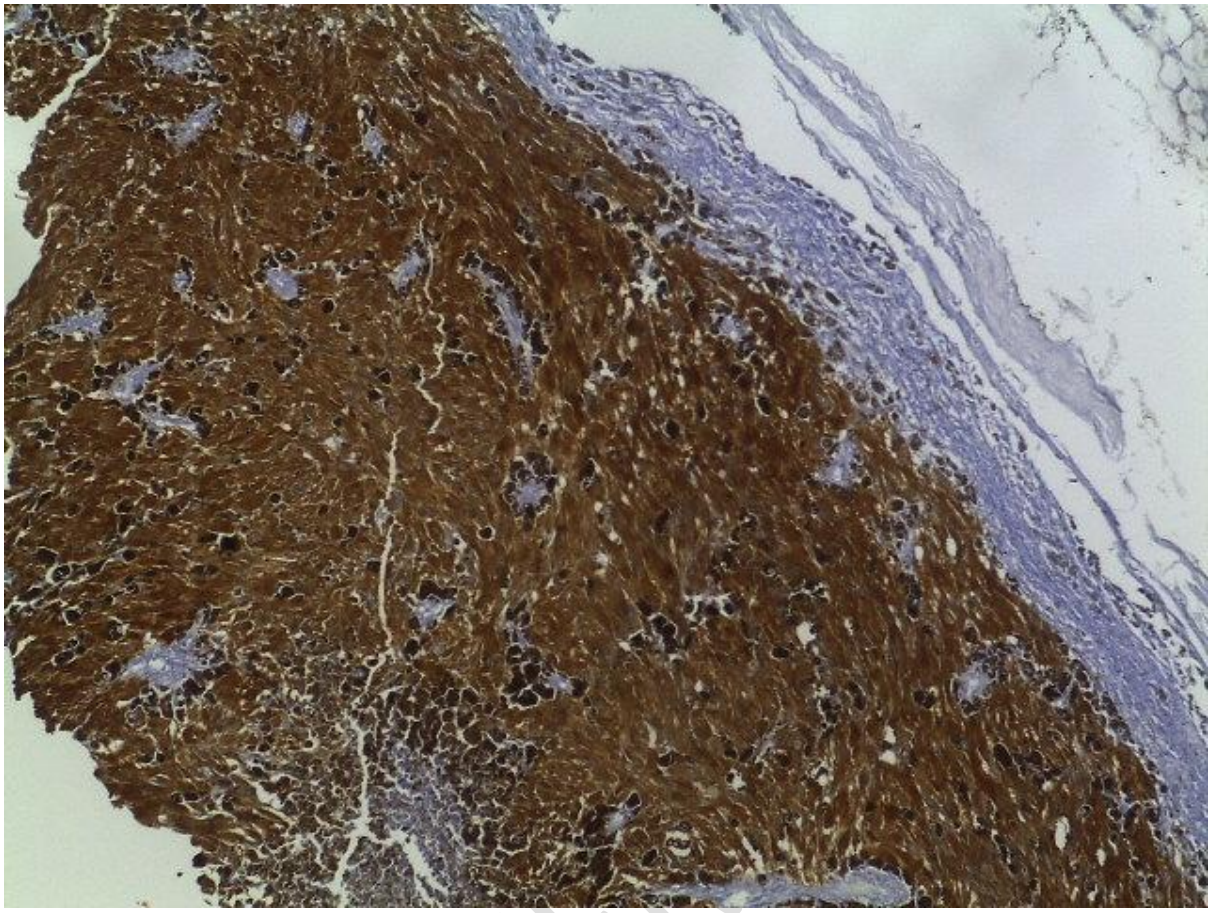

FIG. 2B. A micrograph showing S100 positive tumour cells at X10 magnification.

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. The common sites of melanoma occurrence are areas of sunlight exposure which is a predisposing factors; these areas include head, neck and lower extremities. Less frequently, melanoma occur at the oral and genital mucosa, leptomeninges, conjunctiva, oesophagus, nasal mucosa or nasopharynx and nail bed.[6-9] Although the cause of melanoma in solar-hidden mucosa is unclear, smoking and exposure to formaldehyde may play a role in activation of pre-existing melancytes leading to melanogenic metaplasia.[6-9] 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.[10] The incidence of malignant melanoma in the nose and paranasal sinuses between 0.5-1% which commonly occurs in the 5th-6th decade with no gender predilection.[11] 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 commonest presentation of nasal melanoma. Other presentation are proptosis, diplopia, pain and facial deformity are less common and are indicative of advanced disease.

The tumours cells were spindly in appearance with heavy pigmentation and were within a desmoplastic stromal 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. It may be pigmented (melanocytic) and non-pigmented (amelanocytic).[4, 5, 12, 13] Mucosal melanoma can be distinguished from other tumours using immunohistochemical stains. Immunohistochemically, melanomas stain positively with vimentin, S100, HMB 45 and negatively with cytokeratins and epithelial membrane antigen. In a series of immunohistochemistry reported by Badwein et al, they reported 100% for S100 (14out of 14 cases), 86% HMB 45 (12 out of 14 cases), 90% vimentin (9 out of 10 cases) and 0% cytokeratin (0 out of 9 cases). S100 and HMB 45 were all positive for our patient.[4, 5]

The various investigations for nasal melanoma include CT scan, MRI, chest X-ray, bone scan and/or positive emission tomography. Our patient had a CT scan done with showed a nasal mass.[14, 15]

Nasal melanoma can be staged using the AJCC staging system. In this staging system, stage I disease corresponds with a confined primary tumour, stage II positive cervical lymph node

and stage III distant metstasis.[4, 16-18] Our patient disease has gone beyond the primary site to the submandibular lymph node and therefor had a stage II disease.

The patient underwent excision of the primary tumour which is the main treatment option for patients with head and neck melanoma. Usually patients who undergo primary tumour excision receive postsurgical radiotherapy 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.

Conclusion

Sinonasal melanoma is a rare mucosal tumour that occurs in the head and neck region presenting with epistaxis and nasal obstruction and the tumour is usually positive to S100, HMB 45 and vimentin. The mainstay of treatment is surgery and postsurgical radiotherapy.

CONSENT
Consent for this publication was obtained from the patient.

ETHICAL APPROVAL
It is not applicable.

COMPETING INTERESTS
Authors have declared that no competing interests exist.

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