A CASE REPORT OF PRIMARY MELANOTIC TUMOUR OF THE NASAL CAVITY

Hanumanth Prasad M[1], Suhas S. S[2], Ravi D[3], Balaji N. K[4], Madhuri M. G[5]

[1]Professor and HOD, Department of ENT, Mandya Institute of Medical Sciences, Mandya.
[2]Senior Resident, Department of ENT, Mandya Institute of Medical Sciences, Mandya.
[3]Associate Professor, Department of ENT, Mandya Institute of Medical Sciences, Mandya.
[4]Assistant Professor, Department of ENT, Mandya Institute of Medical Sciences, Mandya.
[5]Junior Resident, Department of ENT, Mandya Institute of Medical Sciences, Mandya.

ABSTRACT

Primary mucosal melanomas arise from melanocytes located in occult sites and can present along the mucosal membranes lining respiratory, gastrointestinal, and urogenital tract. Sinonasal melanomas are extremely rare. Infrequent tumour incidence has limited the insight about their pathogenesis and associated risk factors along with indefinite protocols for staging and treatment of mucosal melanomas. We report here a case of primary melanotic tumour involving the nasal cavity, which was detected at an early stage and operated. We also describe the histological features, imaging studies, and treatment options for this tumour along with a brief literature review.

KEYWORDS

Mucosal Melanoma, Nasal Tumours, Lateral Rhinotomy.

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INTRODUCTION

Melanoma is a cancer arising from the melanocytes. The primary functions of melanocytes of pigmentation and UV protection in the skin and eye are unambiguous and evident, but their presence in noncutaneous areas is not well understood. Although, the existence of melanocytes has been demonstrated in many mucosal membranes. The function of mucosal melanocytes is still questionable. Evidences supporting other non-pigmentary functions of melanocytes such as antimicrobial and immunological functions are scarce and still debatable.[1,2]

Majority of the melanomas are cutaneous in origin. Cutaneous tumours can develop anywhere on the skin, but are more likely to start on the trunk, chest, and back) in males and on the legs in females.[3] Unlike cutaneous melanomas, primary mucosal melanomas arise from melanocytes located in occult sites and can present along the mucosal membranes lining respiratory, gastrointestinal, and urogenital tract. The sinonasal melanomas are extremely rare and accounts for less than 1% of all melanomas. The rarity of the mucosal melanomas have limited the understanding of possible risk factors and the pathogenesis of this disease resulting in inadequacy of precise treatment options. We report here a rare case of melanoma of nasal cavity, which was diagnosed at an early stage and operated and the biopsy of which affirmed to be malignant mucosal melanoma. We also briefly discuss about cases reported in the past along with a literature review.

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Corresponding Author:
Dr. Suhas S. S,
Senior Resident,
Department of ENT,
Mandya Institute of Medical Sciences,
Mandya.
E-mail: drsssuhas@gmail.com
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CASE REPORT

A 64 year old gentleman presented to us in the outpatient department with the complaints of left-sided headache with left nasal obstruction and occasional blood-tinged purulent nasal discharge since two months and mass protruding out from left nasal cavity since one week. The patient's history revealed progressive left nasal blockage not severe enough to cause total block with recurrent episodes of mild nasal bleed (1-2 episodes/week) for the past one month for which he used to take herbal inhalational powdered medicine for a period of 2 weeks with minimal relief of symptoms and then stopped on his own. He observed a small globular mass protruding from left nasal cavity for which he consulted a local allopathic doctor and was referred here for further evaluation and treatment. There was no history of disturbance in smell, no symptoms, or signs of intracranial involvement.

On examination, a smooth mucosa covered dark-coloured mass was seen in the left nasal cavity completely blocking whole of the vestibule and displacing the left lateral alar cartilage outwards. Diagnostic nasal endoscopy could not be done as mass was blocking the nasal aperture making it impossible to pass the zero degree nasal endoscope. Right-sided nasal endoscopy was normal and there was no extension of tumour into the nasopharyngeal area from the left side.

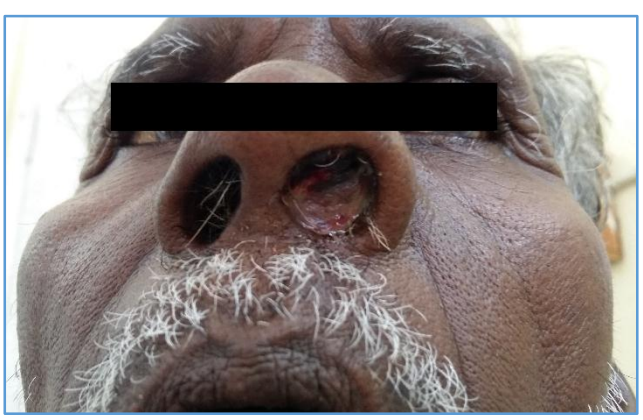

Fig. 1: Clinical Picture showing Dark Coloured Mass in the Left Nasal Cavity

A contrast-enhanced CT scan of nose and paranasal sinuses with neck was done. It revealed a small (33 x 20 mm) mildly-enhancing soft tissue density mass lesion in the anterior aspect of the left nasal cavity without obvious destruction of bony walls. Nasal septum showed undulations. There was no evidence of orbital, nasopharyngeal, or intracranial involvement. Rest of the paranasal sinuses appeared normal. Scan confirmed no regional metastasis.

Biopsy from the left nasal mass showed fragments of a neoplasm comprising of sheets of cells having moderate foamy eosinophilic cytoplasm round to ovoid pleomorphic nucleus with conspicuous nucleoli. Few multinucleated tumour giant cells. Few of these cells have cytoplasm laden with brown pigment. Mitotic activity was present. Features suggestive of poorly differentiated malignancy - Melanoma.

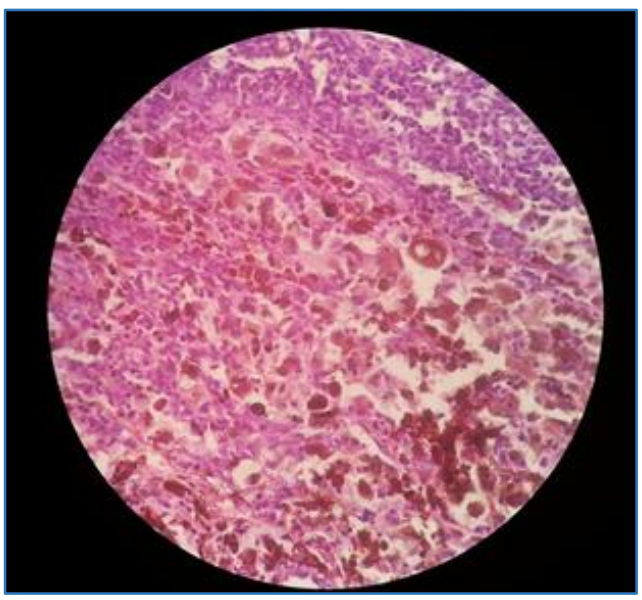

Fig. 2: High-Power Field showing Brown Pigment Laden Cells

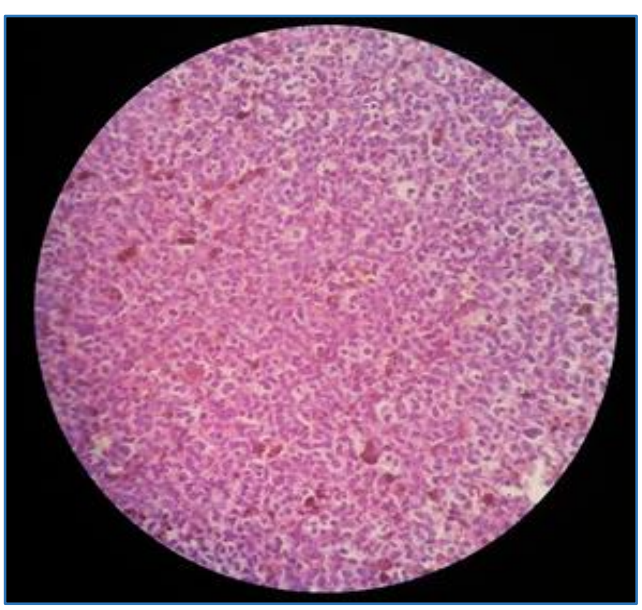

Fig. 3: Low-Power Field showing Malignant Cells

Patient underwent wide excision by lateral rhinotomy approach as the tumour was localised. Mass was found to be arising from left septal mucosa. Mass was excised along with a part of septal cartilage. Histopathology confirmed it to be malignant melanoma.

Fig. 4: Postoperative Picture

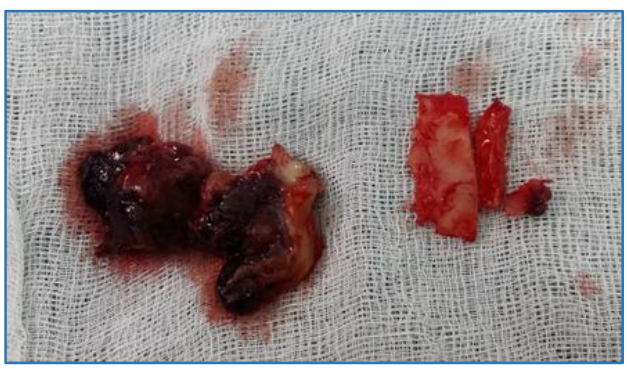

Fig. 5: Excised Biopsy Specimen

DISCUSSION

Melanocytes are the neuroectodermal-derived cells present in the basal layers of the skin/mucosal membranes, which are responsible for production of brown pigment called melanin, which gives the skin its tan or brown colour. This melanin protects the deeper layers of the skin from some of the harmful effects of the sun. The presence of melanocytes is optimum at sunlight exposed skin areas like the face, neck, and extremities. The uncontrolled, abnormal proliferation of these melanocytes results in a tumour known as cutaneous melanoma, malignant melanoma, or mucosal melanomas.

In contrast to cutaneous melanomas, mucosal melanoma is a rare tumour representing only about 1.4% of all melanomas[4]. Primary mucosal melanomas can arise from any part of mucous membrane present in the body, but predominantly arises from melanocytes located in mucosal membranes lining respiratory, gastrointestinal, and urogenital tract. Mucosal melanoma of the nasal cavity, paranasal sinuses, and nasopharynx is still a rarer entity and accounts for about 4% of all sinonasal malignancies[5]. Incidence of nasal cavity melanoma is 0.3 per million and for paranasal sinuses 0.2 per million.[4]

Mucosal melanomas are aggressive tumours with poor prognosis compared to other variants of melanoma. Majority of these mucosal tumours arise from occult sites in congruous with lack of early and specific signs contributing to late diagnosis and unfavourable prognosis. Infrequent tumour incidence has limited the insight about their pathogenesis and associated risk factors along with indefinite protocols for staging and treatment of mucosal melanomas.

Malignant mucosal melanomas are predominantly seen in elderly age group with the peak incidence being in the 7th

decade of life. Aetiopathogenesis of this disease is still unclear. Exposure to formaldehyde in case of sinonasal type and cigarette smoking in case of oral melanomas has been suggested as the possible risk factors.[5,6,7] Curtin et al[8] in his research study showed that genetic mutations and/or increased copy number of KIT (Receptor tyrosine kinase) in 39% of mucosal melanomas.

Even though few case reports have showed a male dominance, there is no significant sexual preponderance. Nasal septum is considered as the most common site of the tumour (41%) followed by middle turbinate (29%), inferior turbinate (23%), lateral nasal wall (7%), and nasal floor (1%).[9] Progressive unilateral nasal obstruction, epistaxis, mass in the nasal cavity are the usual complaints and there is a tendency to misdiagnose as benign lesion as they appear pigmented, polypoid, fleshy, and friable mass. These nasal tumours presents at an advanced stage in initial presentation due to the wide space available to accommodate their growth and the obscure anatomy of the sinuses.

Because of their overlapping and indeterminate microscopic features, it could be easily misdiagnosed as lymphoma, rhabdomyosarcoma, plasmacytoma, olfactory neuroblastoma, and poorly-differentiated carcinoma. Hence, diagnosis of mucosal melanomas is mainly based on proper histologic findings and immunostain.

As a rule, patients with localised disease should undergo surgery followed by postoperative radiotherapy for better local control of the disease.

The majority of the patients present with epistaxis and progressively increasing nasal obstruction.[10] At initial presentation, these tumours are fairly advanced due to the ample space available to accommodate their growth and the obscure anatomy of the sinuses. The natural course of malignant melanomas is marked by early local recurrences, extensions, and frequent metastasis to lymph nodes and viscera making it one of the most dangerous forms of nasal and paranasal sinus tumours. The incidence of regional lymph node metastasis on admission is approximately 5.15%. The submandibular lymph nodes are most commonly involved. A high index of suspicion is required to make an early diagnosis. Surgical treatment remains the treatment of choice for sinonasal melanoma, although complete surgical removal is often limited by surrounding structures, so negative margins are not easy to achieve. There was not found statistically significant difference in overall survival between patients treated by surgery alone or patients treated with surgery and radiotherapy or chemotherapy or combination of all three treatment modalities.

CONCLUSION

Despite the rarity of this tumour, melanoma of nasal cavity can be a differential for mass in the nasal cavity. The clinical features, histology, imaging studies, and the indefinite treatment protocol pose a challenge in the diagnosis and treatment. Therefore, accurate knowledge, experience, and a multidisciplinary team approach is required for early diagnosis and management.

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