



CORPUS PUBLISHERS

Corpus Journal of Case Reports (CJCR)

Volume 3 Issue 1, 2022

Article Information

Received date : March 03, 2022

Published date: March 17, 2022

*Corresponding author

Garima Rawat, M.D.S. Oral & Maxillofacial Pathology, Senior Resident, Department of Pathology, Dharamshila Narayana Superspeciality Hospital, +91-9873776634, New Delhi

Keywords

Mucosal melanoma; Paranasal sinus; Melanocytes; HMB-45

Distributed under Creative Commons CC-BY 4.0

Case Report

Sinonasal Mucosal Melanoma- An Intriguing Case Report

Garima Rawat^{*}, Hema Malini Aiyer¹ and Anshuman kumar²

¹Department of Pathology, Dharamshila Narayana Hospital, India

²Department of Surgical Oncology, Dharamshila Narayana Superspeciality Hospital, India

Abstract

Introduction

Primary Mucosal melanomas of the head and neck region are a rare kind of malignancy arising from the melanocytes. The primary melanomas of the paranasal sinuses are extremely unusual and associated with a poor prognosis due to delayed diagnosis. These lesions tend to metastasize and locally invade tissues more than any other malignant tumour of the head and neck region.

Case report

Here, we present a case of a fifty-seven-year-old male who presented with epistaxis and nasal obstruction. Imaging revealed a soft tissue mass lesion involving the maxillary sinus and nasal cavity with erosion of the floor of right orbit.

Discussion

The mainstay of treatment for these sinonasal mucosal melanomas is surgical resection followed by adjuvant radiotherapy or

Introduction

Sinonasal mucosal melanomas were first described by Lucke in 1869, they comprise rare aggressive neoplasms accounting for 0.5-2% of all melanomas. These lesions are believed to arise from melanocytes, which are derived from the neural crest cells. These melanocytes contain the melanin pigment in the basal and suprabasal layers of the epithelium. Most of the melanomas arise in the skin, although sometimes they may also arise from mucosal surfaces. [1,2] Mucosal malignant melanoma has been described in virtually all sites and organ systems to where neural crest cells migrate. The World Health Organization (WHO) has defined mucosal melanoma as a malignant neoplasm of melanocytes or of melanocyte precursors [3]. It is characterized by a proliferation of atypical melanocytes at the epithelial-connective tissue interface associated with upward migration into the epithelium and invasion of the underlying connective tissue. These melanomas tend to present at an advanced stage, are more aggressive, and usually present in the vertical (nodular) growth phase of the disease [2,4].

We present a rare case of Sinonasal mucosal melanoma involving the right maxillary sinus and nasal cavity in a male in sixth decade of life.

Case Report

A 57-year old male presented to the outpatient department of a tertiary care cancer hospital with the chief complaint of epistaxis/ nasal bleed and nasal stuffiness. The patient consulted a private practitioner who advised NCCT (PNS) which revealed polypoidal soft tissue filling right maxillary sinus causing erosion of floor of right orbit. The patient was referred to our centre for further workup and management. A provisional diagnosis of Lymphoma was considered. The patient underwent Functional Endoscopic Sinus Surgery (FESS) on to obtain a sample for histopathological examination. The histopathology report was suggestive of a Mucosal Melanoma (Figure 2A-C). Immunohistochemistry (IHC) was performed and the tumor cells showed strong cytoplasmic immunoreactivity for HMB-45 and Melan-A, and nuclear staining for SOX-10 (Figure 3A-C). The tumor cells were non-immunoreactive for CD 3, CD 20, CD30 and CK (Figure 4A-C). Based on the Morphologic & IHC features, a final diagnosis of Sinonasal Mucosal Melanoma with epithelioid and spindle cells was made. The patient thereafter underwent a CEMRI face & brain which revealed a soft tissue lesion in the right maxillary sinus causing focal erosion of the postero-lateral & superior wall of right maxillary sinus, the lesion was extending to the extracranial compartment of right orbit. Bilateral level Ib, level II & level III lymph nodes were enlarged. The patient also underwent whole-body PET CT which revealed a FDG avid soft tissue density lesion involving the right maxillary sinus, right nasal cavity with subtle erosion of superior and medial walls of maxillary sinus and floor of right orbit. No obvious extension was seen into the right orbit. Few mildly metabolically active cervical lymph nodes were reported on the right side at levels I and II. No other metastatic lesion was noted anywhere in the body. (Figure 1) The patient was taken up for surgery in the department of surgical oncology and was planned for Right Total Maxillectomy along with Wide Local Excision (WLE) of palatal mucosa along with Modified Neck Dissection (MND) Type III of the ipsilateral side. The surgically resected specimen was sent to the pathology department for intraoperative margin status assessment, which were found to be sufficient and negative on histopathological examination. The specimen received in the pathology department measured 7.0x6.0x4.0 cm and included total maxillectomy, WLE palatal mucosa, maxillary sinus, nasal cavity and part of the zygoma and separate right MND type III. The tumor was a polypoidal lesion attached to the posterior wall of the maxillary sinus. Histopathological findings were consistent with the incisional biopsy findings with more areas showing epithelioid and spindle cell differentiation. All surgical resection margins were uninvolved by tumour and the closest margin (medial margin) was 1.2 cm from the tumor. Twenty-eight lymph nodes dissected were negative for any metastatic malignancy (0/28). The final pathologic TNM staging was done according to AJCC 8th edition staging guidelines for oral mucosal melanoma as under:- Primary Tumour (pT) – pT3: Tumour limited to the mucosa and immediately underlying soft tissue, regardless of thickness or greatest dimension. Regional Lymph Nodes (pN) – pN0: No regional lymph node metastasis. The patient was discharged and had an uneventful recovery. The patient is on follow up since then.

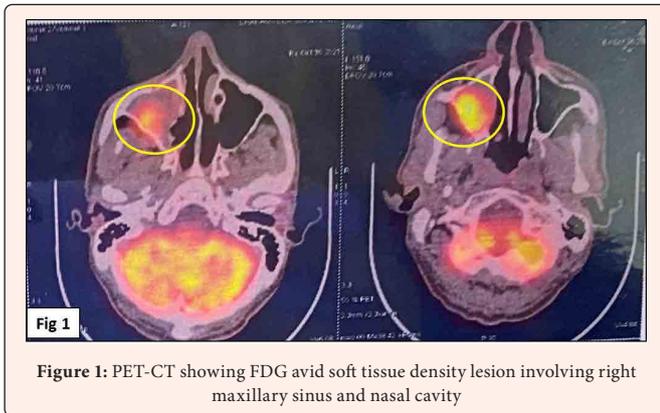


Figure 1: PET-CT showing FDG avid soft tissue density lesion involving right maxillary sinus and nasal cavity

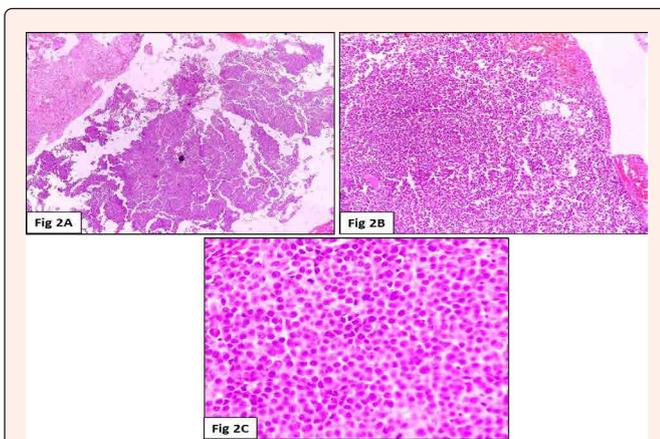


Figure 2:
 A) Photomicrograph showing sheets of round to ovoid shaped cells. (HE-40x)
 B) Photomicrograph showing round to ovoid cells with nuclear pleomorphism. (HE-100x)
 C) Photomicrograph showing tumour cells in sheets with no melanin pigmentation. (HE-400x)

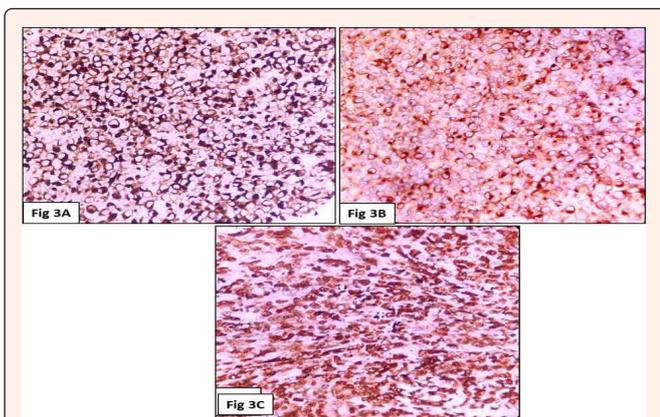


Figure 3:
 A) Photomicrograph showing strong cytoplasmic immunoreactivity for HMB-45. (IHC-400x)
 B) Photomicrograph showing strong cytoplasmic immunoreactivity for Melan-A. (IHC-400x)
 C) Photomicrograph showing strong nuclear immunoreactivity for SOX-10. (IHC-400x)

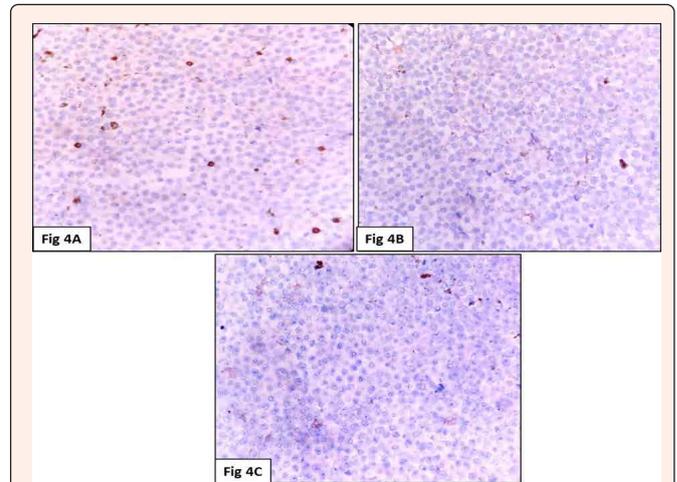


Figure 4:
 A) Photomicrograph showing non immunoreactivity for CD 3. (IHC- 400x)
 B) Photomicrograph showing non immunoreactivity for CD 20. (IHC- 400x)
 C) Photomicrograph showing non immunoreactivity for CK. (IHC- 400x)

Discussion

Mucosal malignant melanoma was first described by Weber in 1859 and after that, it was recognized as a distinct clinical entity and named as “melanotic sarcoma” by Lucke in 1869. This entity represents 0.5-2.0 % of all malignant melanomas and approximately 4% of melanomas of the head and neck region. These lesions most commonly affect adults above fifth decade of life; children are rarely involved. There is slight male predominance [1-4]. the most commonly involved sites are nasal cavity, middle nasal conchae; paranasal sinuses are rarely involved with maxillary sinus being the most commonly affected amongst them. The first symptoms of a sinonasal melanoma are usually delayed and when the symptoms appear they include unilateral nasal obstruction and epistaxis. Fibre-optic nasolaryngoscopy reveal blackish, grey or translucent polypoid masses in case of amelanotic melanomas. Because most melanomas are asymptomatic in their early stages, the diagnosis is unfortunately often delayed until symptoms. Neck nodal metastases to cervical lymph nodes, occurs in 10-50% cases [5-8]. In the case being discussed here, the patient was a 57-year-old male who complained of nasal bleeding and nasal obstruction since 6-7 months. This tumour is characterized by aggressive and invasive behaviour which manifests as both local and distant metastases to sites such as lungs, liver, brain, and bones. Lymphatic metastasis at the time of diagnosis seems to be a crucial prognostic factor for oral melanomas. Histopathologically, mucosal melanoma is composed of sheets or islands of epithelioid melanocytes, which may be arranged in an organoid, or alveolar pattern. The cells have pale cytoplasm and large open nuclei with prominent nucleoli and occasionally they may be plasmacytoid. Sheets and fascicles of spindle cells may also be seen, but are usually a minor part of the lesion. In most instances, the cells of melanoma contain melanin granules, but they may demonstrate no melanin production (amelanotic melanoma). Lack of production may cause diagnostic confusion at the light microscopic level because melanoma can mimic a variety of undifferentiated tumours. Therefore, immunohistochemical studies are a guide to diagnosis. The immunoprofile of melanoma shows S100 positivity and negativity for cytokeratins in over 95% of cases. Although sensitive, S100 is not specific. More specific markers include HMB45, Melan-A or anti-tyrosinase, which stain about 75% of lesions [1-5]. The morphological and immunohistochemical picture was on the same lines in our case also. Surgery remains the mainstay of treatment along with chemotherapy, radiotherapy, and immunotherapy. In the cases where metastasis has already occurred, the disease is considered classically incurable, surgery is considered only for palliative care and other treatment modalities like radiotherapy and chemotherapy take precedence. Melanoma is notorious for its unpredictable and widespread metastasis. Metastatic spread to the bone, usually the vertebrae, is a frequent finding in terminal disease and may be accompanied by multiple metastases to the lymph nodes, central nervous system, lungs and liver. Metastasis to the oral regions is uncommon and usually involves soft tissues, notably the tongue.

The reported 5-year survival rate for mucosal melanoma has ranged from 4.5% to 29% with the median survival rate of 18.5 months after initial diagnosis. The median



survival is affected by whether there is lymph node involvement (18 months) or not (46 months). Cutaneous melanomas can be graded by Clark levels or the Breslow tumour thickness grading system. The former classification assesses the depth of invasion, whereas the Breslow's system measures the thickness of the tumour from the surface of the epidermis to the greatest depth of the tumour. The risk for developing metastatic lesions from primary cutaneous melanomas increases with tumour thickness. The Breslow and Clark grading systems have not been validated as prognostic predictors in oral melanomas, probably owing to the rarity of this lesion. Additionally, in contrast to cutaneous melanomas, most sinonasal melanomas are larger than 4 mm at the time of initial presentation. This factor, together with inadequate resection of margins and higher stage at initial diagnosis, may contribute to the discrepancy in patient's 5-year survival rates between cutaneous melanoma (80%) and sinonasal melanomas (15%). In general, the survival rates are poor and are worse for those with metastasis. Other factors associated with poor prognosis include vascular invasion, necrosis, a polymorphous tumour cell population, and increasing age [2,4,7-8]. Sinonasal melanoma is a neoplasm that often tends to undergo regression. Clinically, colour variation is perhaps the most important hallmark of primary cutaneous melanoma. The change in colour to white, off-white, blue-white and grey-white is a sign of (spontaneous) regression in malignant melanoma. Histopathologically the process starts with a dense lichenoid infiltrate of lymphocytes and ends with fibrosis and/or melanosis within a thickened papillary dermis. The dense infiltrate of lymphocytes permeates the thin melanoma and destroys the atypical melanocytes in the epidermis and the papillary dermis. The aetiology of regression in melanocytic lesions is multifactorial, and the contributing mechanisms are yet to be completely elucidated. A literature search indicates that host-immune-mediated responses, particularly the CD8-positive Cytotoxic T Lymphocytes (CTLs) play a significant role in the development of regression. The role of the immune system in regression is further supported by the high incidence of CD4-positive T lymphocytes and Th1 cytokines in regressed tumours, as well as the high levels of tumour-specific antibodies and CTLs in the peripheral blood of patients with regressed tumours [8-9].

Conclusion

Early diagnosis and treatment are mandatory for a better prognosis concerning malignant melanoma of the oral cavity. Clinicians must carefully examine the oral cavity and any growing pigmented lesion must be biopsied. Amelanotic melanomas can be diagnosed by immunohistochemical examination of tissue from the lesion.

References

1. Rani GS, Kumar TV, Kolasani B, Begum MR, Srinivasan AP (2014) Primary Malignant Melanoma of Maxilla: Report of a Case with Discussion. *Case Rep Dent* 2014: 624306.
2. Chaitanya YC, Chaitanya PV, Reddy MV, Bhavya B (2018) Oral malignant melanoma. *J Indian Acad Oral Med Radiol* 30: 158-160.
3. Barnes L, Eveson JW, Reichart P, Sidransky (2005) World Health Organization classification of tumours. Pathology and genetics of head and neck tumours. IARC Press: Lyon 2005, France.
4. Pour MSH (2008) Malignant melanoma of the oral cavity: A review of literatures. *Indian J Dent Res* 19: 47-51.
5. Devi S, Sinha R, Singh RK (2015) Malignant melanoma maxilla. *Natl J Maxillofac Surg* 6(1): 115-118.
6. Hasan S, Jamdar SF, Jangra J, Al Beajji SM (2016) Oral malignant melanoma: An aggressive clinical entity - Report of a rare case with review of literature. *J Int Soc Prev Community Dent* 6(2): 176-181.
7. Manigandan T, Sagar GV, Amudhan A, Hemalatha VT, Babu NA (2014) Oral malignant melanoma: A case report with review of literature. *Contemp Clin Dent* 5(3): 415-418.
8. Ebenezer J (2006) Malignant melanoma of the oral cavity. *Indian J Dent Res* 17(2): 94-96.
9. Gondivkar SM, Indurkar A, Degwekar S, Bhowate R (2009) Primary oral malignant melanoma--a case report and review of the literature. *Quintessence Int* 40(1): 41-46.