CASE REPORT

Chinese Journal of Medical Research
Chinese J Med Res 2022; 5(2): 5-7

ISSN(e): 2618-091X
ISSN(p): 2663-8053

Recurrent primary Sino-Nasal Mucosal Melanoma- a rare case presentation in Pakistan

Syeda Tehmina Junaid[1], Bushra Syed[2], Asmat Zehra[3], Hina Altaf[4]

[1] *Bab-e-Urdu Road, MA Jinnah Road, Department of E.N.T, Head and Neck Surgery, Dow Medical College, Dow University of Health Sciences and Civil Hospital Karachi, Pakistan*

[2] *Bab-e-Urdu Road, MA Jinnah Road, Dow Medical College, Dow University of Health Sciences Karachi, Pakistan*

[3] *Bab-e-Urdu Road, MA Jinnah Road, Dow Medical College, Dow University of Health Sciences Karachi, Pakistan*

[4] *Bab-e-Urdu Road, MA Jinnah Road, Dow Medical College, Dow University of Health Sciences Karachi, Pakistan*

*Corresponding Author: Dr. Bushra Syed,Bab-e-Urdu Road, MA Jinnah Road, Dow Medical College, Dow University of Health Sciences Karachi, Pakistan Email: bushra_1712@yahoo.com

Abstract

A tumor like Sino-nasal Mucosal Melanoma (SNMM) is a fascinating medical scenario in parameters of Pakistan due to its ultimate rarity, supported by only two relevant cases reported from Pakistan till now, the first case of Mr. Husain in 1978 and the other seen in the Department of Radiation Oncology, Aga Khan University Hospital in Karachi. The tumor is accompanied with adverse prognosis and increased chances of recurrence calling for expert diagnosis and necessary treatment measures. The case highlights the medical scenario of a 46-year-old female who presented with recurrence of SNMM hence the report surrounds its causes and risk factors in addition to, suitable treatment options in the setup of a developing country like Pakistan and its outcomes along with added histopathological context regarding SNMM from experience with the disease.

Keywords: Paranasal Sinuses, Melanoma, Oncology, Nasal Mucosa, Neoplasms.

INTRODUCTION

SNMM is an infrequent and aggressive malignancy arising from melanocytes of the mucous membranes. It constitutes about 0.7 – 1% of all melanomas [4]. Epistaxis and nasal obstruction are the most frequent initial symptoms [4]. SNMM has a poor prognosis, with approximate 5-year survival rate of around 0–30 percent [1]. This may be because the diagnosis of SNMM is often missed at earlier stages [4]. Relapses after primary treatment were seen in 31- 85% of the patients [6]. The best present-day treatment modality is wide surgical resection of the tumor [8]. Adjuvant postoperative radiotherapy has also shown good outcomes [6]. The patient gave us an informed consent for the publication of this case report.

CASE REPORT

Dear editor in chief, we share the case of a 46-year-old female with no known co-morbid, resident of Hyderabad, Sindh, who presented to the outpatient otorhinolaryngology clinic of a tertiary care hospital in Karachi in September 2020 with complete left nasal obstruction, anosmia, unilateral epistaxis and periodic left eye lacrimation. She had been diagnosed with Sino-nasal Mucosal Melanoma (SNMM) in 2015 on imaging as well as histopathology of the biopsied specimen. The tumor had been rejected by lateral rhinotomy at that time, after which her symptoms had resolved. Five years later, the symptoms recurred. Plain Computed Tomography (CT) scan of the paranasal sinuses in axial and coronal sections showed enhancing lesion of 5 x 3 x 4.2 cm in AP x TS x LS dimensions, primarily in the left nasal space. The mass extended into the maxillary sinus, anterior ethmoidal sinus, and orbit. Based on a high suspicion of a recurrent tumor, a decision of diagnostic radical surgical resection with wide margins was made. A mid-facial degloving approach was adopted. The mass was removed completely in piecemeal from all the involved sites. Adjuvant radiotherapy was used to avoid a second recurrence. Histopathological examination revealed malignant spindle and epithelioid tumor cells with marked melanin pigmentation and scattered mitoses. The cells were immunoreactive to S-100, Melan-A and HMB-45, compatible with the diagnosis of recurrent SNMM. The tumor was classified as stage IV-A (T4a, N0, M0) as per the 2013 AJCC-TNM (The American Joint Committee on Cancer — Tumor, Node, and Metastasis) classification for mucosal melanoma of the head and neck. Two follow up visits over a period of six months revealed complete resolution of the mass on examination and the CT scan was also clear. The patient also reported improvement in her symptoms.

AJCC-TNM (The American Joint Committee on Cancer — Tumor, Node, and Metastasis) classification.

DISCUSSION

The medical literature does not contain an extensive volume of publications on SNMM in Pakistan, mainly because of the rarity of this disease in this region. Sino-nasal mucosal melanoma (SNMM) is an infrequent and difficult to treat malignancy, first described by Lucked in 1869 [1]. Mucosal melanomas arise from the neural crest driven melanocytes present in the mucosal membranes of the body [2]. The most common site of primary mucosal melanomas is the head and neck (41%), of which melanomas of the nasal cavity, paranasal sinuses and conjunctiva are the most frequent [3]. SNMM constitutes about 0.7 – 1% of all melanomas [4]. The common sites for SNMM are the septum, inferior and middle turbinate's, the lateral nasal cavity wall, and the paranasal sinuses. Of the paranasal sinuses, the maxillary sinus is the most frequently involved [1]. Our patient's tumor primarily occupied the left nasal space and the maxillary and ethmoidal sinuses. Orbital extension was also observed in our case, which has not been reported much. Due to its rarity, a specific cause of SNMM has not been identified yet. However, some risk factors include increased age, female gender, formaldehyde exposure, cigarette smoking and inhaled carcinogens [3]. We did not find any such risk factors in our patient's history, except female gender and age. No association was found between SNMM and cancer-causing viruses like papilloma viruses, herpes viruses, and polyomavirus [3]. SNMM has a slow clinical development pattern but its early signs and symptoms are similar to those of inflammatory, benign conditions [4]. Therefore, an early definitive diagnosis is usually missed and the tumor is detected at an advanced stage [4]. The symptom spectrum constitutes of epistaxis, ulcers, anosmia and nasal obstruction, out of which epistaxis and nasal obstruction are the most common [4]. The approximate 5-year survival rate of SNMM is around 0–30 percent [1]. To form a diagnosis immunohistochemical and morphological findings are used [1]. For staging the SNMM the AJCC-TNM classification is conventionally used [4]. The tumor, in our case, was also staged using this classification as IV-A (T4a, N0, M0) because it was a moderately advanced local disease with no evidence of metastasis or regional lymph nodes involvement.

SNMM secures second rank after squamous cell carcinoma among all other malignancies of the nasal region [5]. With a noxious tumor like SNMM there remains a high possibility of local recurrence years after the initial treatment [5]. Up to 31-85% of patients may encounter a relapse [6]. The prognosis, in such cases, becomes poor with only about 24% of the patients being able to survive. Locoregional recurrence may point towards metastasis in some patients, demanding unavoidable aggressive treatments [5]. In our patient, despite locoregional recurrence, no metastasis was found on investigation. An aggressive tumor like this one take 14.7 months on average for local recurrence [5]. However, our patient was in remission for more than this expected duration. Factors that contribute to increased risk of local recurrence and metastasis include initial diagnosis at older age, atypical and vague clinical symptoms, paranasal sinus origin, presence of ≥10 mitoses per high power field, increased melanotic pigmentation, pseudopapillary or sarcomatous architecture, perineural or Vaso-lymphatic annexation, and undifferentiated cell morphology [7]. It is also noteworthy to state that a recurrent tumor may at times present with variations both histologically and in pigmentation when compared to the primary cancer [5]. In our case, histopathological examination of the resected tumor revealed malignant spindle and epithelioid tumor cells with marked melanin pigmentation and scattered mitoses. The tumor cells were immunoreactive to S-100 and HMB-45, aiding us in the accurate diagnosis of mucosal malignant melanoma.

Wide surgical resection to achieve clear margins is the preferred treatment modality [8]. However, it is hard to achieve free margins because of the intricate anatomy of the Sino-nasal area and the locally invasive nature of this tumor [6]. To enable better access during surgery for clearing margins, a trans-facial approach is preferred [5]. end bloc resection of low-grade tumor masses is possible; however, piecemeal resections of larger tumors were to have similar oncological efficacy to end bloc resections by external surgical techniques [9]. Adjunct radiotherapy has shown better local control even in patients with advanced disease [6]. In our case, a decision of surgical resection was made, based on high suspicion of a recurrent tumor. A mid-facial degloving approach was adopted, as it has better cosmetic and functional outcome compared to the transracial approach [10]. The mass was resected completely in piecemeal from all the involved sites of the Sino-nasal region. Adjuvant radiation therapy was used to avoid a second recurrence.

After surgery, the patient was discharged and was reviewed on follow-up after a month. On two subsequent follow-ups over the period of 6 months, her symptoms improved with complete resolution of the lesion on examination. A follow-up CT scan showed no signs of recurrence. The patient remains healthy to date.

CONCLUSION

The scarcity of SNMM in the setting of Pakistan and its clinical comparability with inflammatory conditions is a dominant cause that many practioners do not include SNMM in their differential diagnosis. This tumor eventually ends up being diagnosed as an advanced staged disease and thus has a poor prognosis. SNMM is a locally invasive aggressive tumor that contributes to its high recurrence rate. Surgical aspect of the case emphasizes that careful clearance of the tumor margins during surgery is crucial to prevent recurrence. After the surgery and radiotherapy, a close follow-up is necessary for early detection and prompt intervention that may improve the patient's outcome.

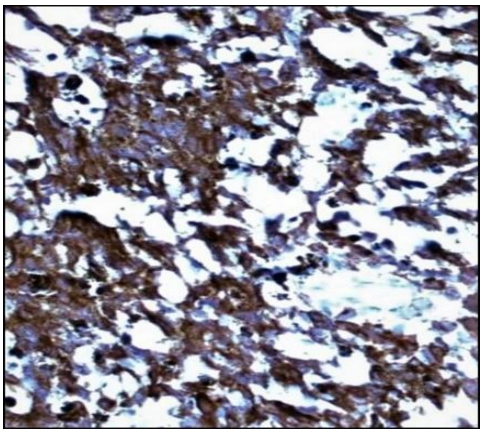

Figure 1: Immunoreactivity of tumor cells to Melan-A

Figure 2: Tumor after surgical resection

Acknowledgement

We acknowledge Dr. Syed Assad Hassan Rizvi for his contribution in this case report.

Ethics Approval/Disclosure

Not applicable.

Patient Consent

An informed consent was taken from the patient prior to writing the manuscript.

Author Contributions

TJ fully contributed to the critical write-up of this case report.
BS fully contributed to the critical write-up of this case report.
AZ fully contributed to the critical write-up of this case report.
HA fully contributed to the critical write-up of this case report.

Conflict of Interest

None declared.

Financial Support

None declared.

REFERENCES

1. Alves IS, Berriel LG, Alves RT, Pinto MB, Oliveira CF, Cazzotto AC, *et al*. Sinonasal melanoma: a case report and literature review. Case reports in oncological medicine. 2017:8201301.
2. Dupin E, Le Douarin NM. Development of melanocyte precursors from the vertebrate neural crest. Oncogene. 2003;22(20):3016-23.
3. Ascierto PA, Accorona R, Botti G, Farina D, Fossati P, Gatta G, *et al*. Mucosal melanoma of the head and neck. Critical reviews in oncology/hematology. 2017;112:136-52.
4. Gilain L, Houette A, Montalban A, Mom T, Saroul N. Mucosal melanoma of the nasal cavity and paranasal sinuses. European annals of otorhinolaryngology, head and neck diseases. 2014;131(6):365-9.
5. Bridger AG, Smee D, Baldwin MA, Kwok B, Bridger GP. Experience with mucosal melanoma of the nose and paranasal sinuses. ANZ journal of surgery. 2005;75(4):192-7.
6. Dauer EH, Lewis JE, Rohlinger AL, Weaver AL, Olsen KD. Sinonasal melanoma: a clinicopathologic review of 61 cases. Otolaryngology—Head and Neck Surgery. 2008;138(3):347-52.
7. Kalogeria EM, Kalyvas D, Tosios KI, Tsiklakis K, Sklavounou A. Recurrence in a patient with a 10-year history of Sinonasal mucosal melanoma manifesting as facial swelling. Journal of clinical and experimental dentistry. 2017;9(12):e1492.
8. Andrianakis A, Kiss P, Pomberger M, Wolf A, Thurnher D, Tomazic PV, *et al*. Sinonasal mucosal melanoma: treatment strategies and survival rates for a rare disease entity. Wiener klinische Wochenschrift. 2021:pp.1-1.
9. Wellman BJ, Traynelis VC, McCulloch TM, Funk GF, Menezes AH, Hoffman HT, *et al*. Midline anterior craniofacial approach for malignancy: results of end bloc versus piecemeal resections. Skull base surgery. 1999;9(01):41-6.
10. Har-El G. Medial maxillectomy via midfacial degloving approach. Operative Techniques in Otolaryngology-Head and Neck Surgery. 1999;10(2):82-6.