Scientific Research Publishing

International Journal of Otolaryngology and Head & Neck Surgery, 2023, 12, 61-68
https://www.scirp.org/journal/ijohns
ISSN Online: 2168-5460
ISSN Print: 2168-5452

Sino-Nasal Malignant Melanoma: A Case Report

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How to cite this paper: Yee, G.W.Y., Gill, R.S., Kumarasamy, G., Rajesvaran, C. and Rajendran, S. (2023) Sino-Nasal Malignant Melanoma: A Case Report. *International Journal of Otolaryngology and Head & Neck Surgery*, **12**, 61-68.
https://doi.org/10.4236/ijohns.2023.122007

Received: December 21, 2022
Accepted: March 13, 2023
Published: March 16, 2023

Abstract

Melanoma is a malignant tumor and is commonly found second least in the head and neck region, with approximately 10% - 25% of melanomas followed by ear (8% - 11%). We reported a case of sino-nasal malignant melanoma in a 46-year-old Asian lady who presented with epistaxis and dysphagia. By nasoendoscopy and CT scan imaging, the diagnosis of sino-nasal malignant melanoma was later confirmed by histopathology examination along with immunohistochemistry by using Melan A and S100. Malignant melanoma in the head and neck is rare and has a poor prognosis. There has been an increase in the incidence of 5% per year and mortality at 2% per year. This dictates malignant melanoma has rapidly increased compared to any other cancer except lung cancer in women and has contributed to the third highest number of deaths among all cancers. The mortality rate has also increased, and that is attributed to the negligence of symptoms during the initial period. This usually leads to patients seeking consultation during the advanced stage of the disease. In conclusion, this paper is to report that malignant melanoma may be one of the differentials of sino-nasal masses, and it may mimic nasopharyngeal carcinoma in its presentation.

Keywords

Malignant Melanoma, Sino-Nasal, Immunohistochemistry

1. Introduction

Sino-nasal melanoma is a malignant neoplasm of the melanocyte. Melanocytes are derived from the neural crest; melanomas usually occur on the skin. They can arise in other locations where neural crest cells migrate, such as the gastrointestinal tract and brain [1].

The five-year relative survival rate for patients with stage 1 melanoma of the head and neck is more than 90%, compared with less than 20% for those with metastatic disease [2].

The discrepancy in mortality rate is high when compared between primary melanoma (approximately 11%) and metastatic melanoma [3]. Metastatic melanoma patients typically have a low survival rate due to the delay in seeking medical aid including poor efficacies of current cancer therapies. The best treatment options are surgical intervention along with radiation and/or chemotherapy to control the disease [4].

Patients with sino-nasal malignant melanoma present with complaints of epistaxis, nasal block, and persistent rhinorrhoea. Histologically, melanoma's appearance can vary. The cells may appear as stratified squamous epithelium and stroma can be infiltrated by malignant cells. These malignant cells are round to ovoid, exhibit moderate pleomorphic hyperchromatic nuclei and some have prominent nucleoli [2]. Immunoperoxidase studies are very useful, including the use of HMB-45, S100, and Melan A [2].

According to studies, sino-nasal malignant melanoma is highly treatable when discovered in the early stages. However, in the cases of advanced stages as disease metastasizes, the treatment options become limited, and the survival rate decreases to months. The site of the tumor origin is occasionally difficult to identify due to the extensive local destruction it causes and the large size of the tumor. There is a delay in diagnosis due to its nonspecific clinical features.

2. Case Presentation

A 46-year-old Asian lady presented with epistaxis for one year of duration and dysphagia for 6 months. She also experienced the loss of appetite and significant weight loss and was unable to lie flat for the last 3 months. She sought medical attention due to sudden shortness of breath.

At rest, the patient presented with audible stridor and hypernasal speech. Upon neck examination, single left cervical lymph node palpable at level II measuring about 4 × 5 cm, firm, fixed and non-tender. Tuning fork test revealed a negative Rinne's test for both right & left ears, the Weber test was not lateralized. The findings were confirmed with Tympanometry showing a type B for both ears. Pure tone audiometry showed mild to moderate hearing loss over left ear and mild to severe hearing loss over right ear indicating bilateral conductive hearing loss due to mass effect on bilateral eustachian tube. In the oral cavity, the nasal mass was seen to extend inferiorly until the level of oropharynx.

Nasoendoscopy showed a mass occupying both right and left nasal cavities obliterating the post nasal space with thick mucus secretion.

In view of impending air-way compromise, patient underwent tracheostomy under local anaesthesia.

CT of head and neck showed large enhancing mass measuring approximately 5.8 × 7. 6cm (AP × W) completely obliterating the nasopharynx, oropharynx, and part of hypopharynx with local infiltration and cervical lymphadenopathies (**Figure 1** and **Figure 2**). The mass is seen extending superiorly to involve both posterior nasal spaces bilaterally and causing effacement of both parapharyngeal

spaces (Figure 3). There is a heterogenous lesion in the left infratemporal fossa with association of erosion of the adjacent wall of left maxillary sinus. The right infratemporal space is preserved.

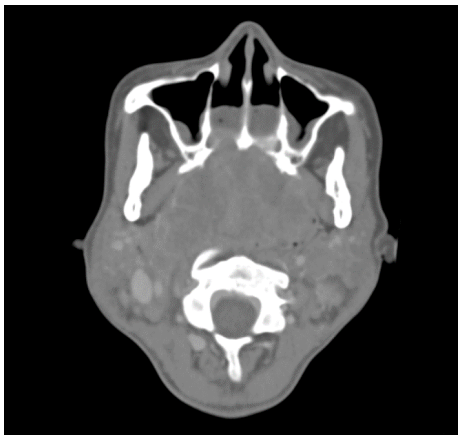

Figure 1. Axial plane. Computed tomography of the head. A large mass blocking the air flow at the bilateral nasal cavity is clearly visible.

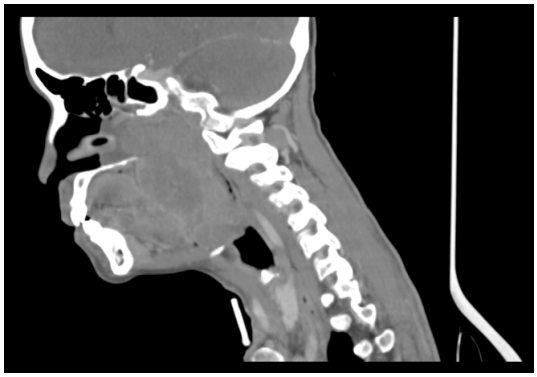

Figure 2. Sagittal plane. Computed tomography of the head. A large mass obliterating nasopharynx, oropharynx, and part of hypopharynx.

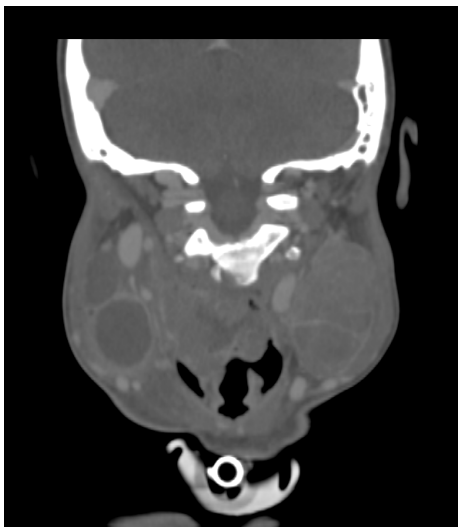

Figure 3. Coronal plane. Large mass causing effacement of both parapharyngeal spaces

CT of thorax and abdomen revealed left lung lesion, multiple hypodense liver lesions, bilateral breast lesions, enlarged mesenteric and pelvic lymphadenopathies, subcutaneous lesions in the left shoulder, bulky left adrenal gland, ill-defined hypodense pancreatic tail lesion, which extends into the splenic vein which may represent metastasis.

FNAC of enlarged cervical lymph node showed hypercellular composed of sheets of malignant looking cells arranged singly and few in clusters (Figure 4 and Figure 5). The cells are highly pleomorphic with high NC ration, irregular coarse chromatin nuclei having prominent nucleoli and multinucleoli. Some of the cells exhibit atypia with very large distorted hyperchromatic nuclei and tumor giant cells. Mitoses and cannibalism are evident. Scattered neutrophils are present in background.

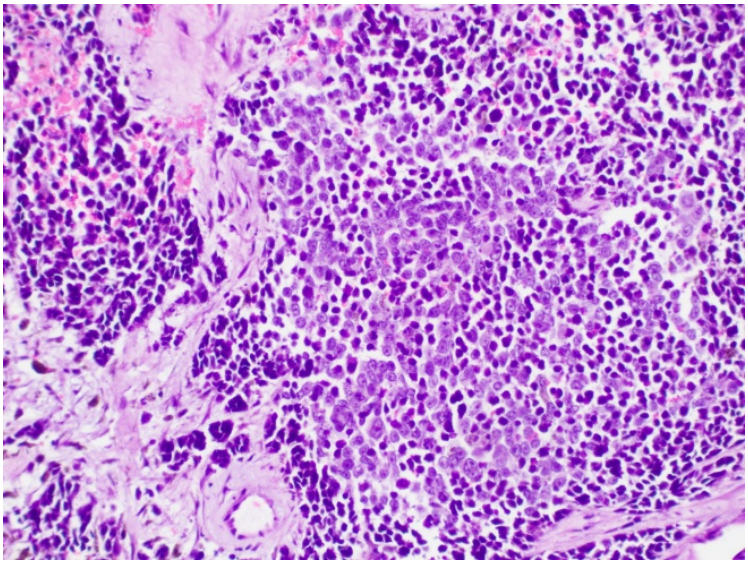

Figure 4. Left nasopharyngeal mass 200× (magnification) haematoxylin and eosin.

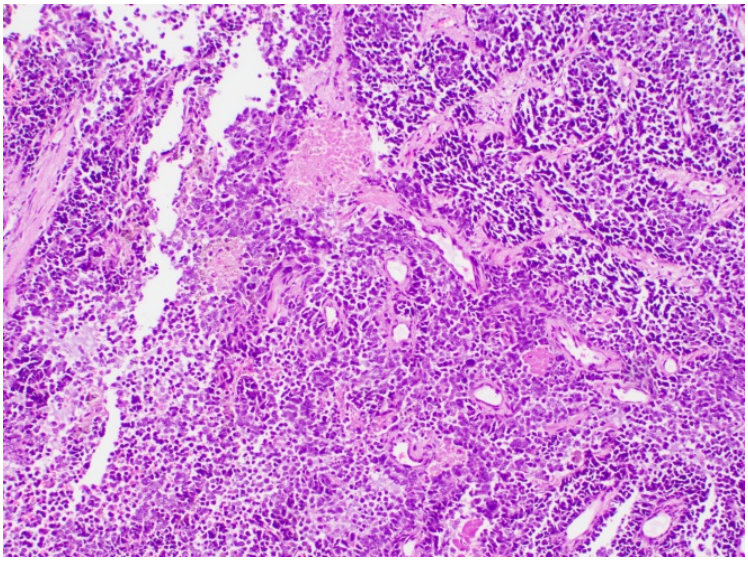

Figure 5. Right nasopharyngeal mass 100× (magnification) haematoxylin and eosin.

In view of the clinical findings, the working diagnosis was towards nasopharyngeal carcinoma as NPC is endemic in our place of practice. Biopsy was taken and histopathological examination revealed malignant melanoma with extensive tumor necrosis. The malignant cells were positive for Melan A (Figure 6) and S100 (Figure 7). The cells were negative for CKAE1/AE3, Synaptophysin, Chromogranin A, NSE, INSM-1 and LCA. CD5 6 is inconclusive, Ki-67 is high (80% - 90%).

The patient was staged as advanced stage of sino-nasal malignant melanoma. However, even before breaking the bad news, patient defaulted follow up and eventually passed away before initiation of therapy.

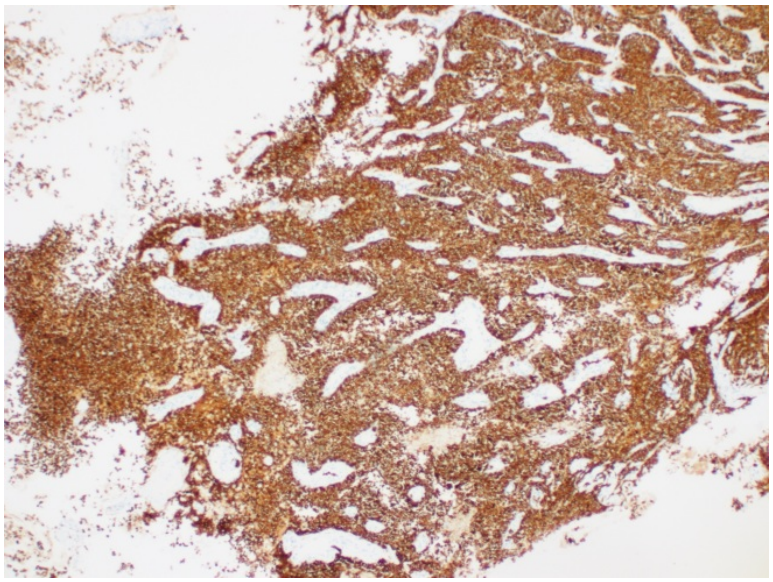

Figure 6. Immunoperoxidase staining positive for Melan A in 40× (magnification).

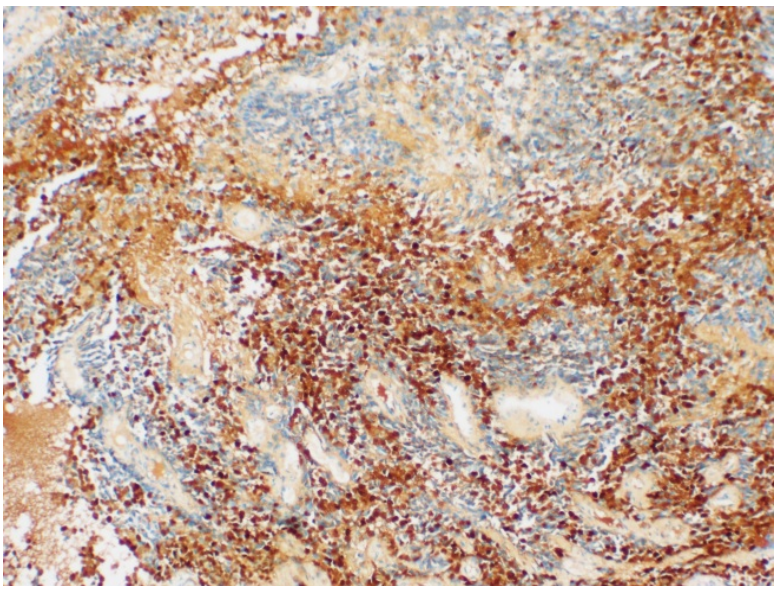

Figure 7. Detected S100 protein in immunoperoxidase studies seen in 100× (magnification).

3. Discussion

Sino-nasal malignant melanoma is uncommon as mucosal melanoma of the head and neck depict approximately less than 1% of all reported melanomas [5]. It is an aggressive, and fast progressing tumour which results in 4% of sino-nasal malignancies [6]. Malignant melanoma in the sino-nasal region is exceptionally rare with mean 5-year survival is 0% - 30% [6]. This tumour arises from melanocytes derived from neural crest tissue, which are the progenitors of the lesion and etiopathogenesis remains unclear. The prevalence of this condition is of mean age ≥ 60 years and more commonly found in males [6]. The disease is found more in white populations compared to blacks, and in individuals with higher exposure to sunlight [7].

Sino-nasal malignant melanoma is commonly arise from nasal cavity, inferior, middle nasal conchae, septum, lateral wall of nasal cavity and facial sinuses [6] which is dissimilar to nasopharyngeal carcinoma, which originates from the fossa of Rosenmuller (**Table 1**).

The most common symptoms reported are unilateral nasal obstruction, nasal congestion, and bleeding with variable duration between 3 - 24 months. In our case, patient presented with the symptom of epistaxis and dysphagia which is similar to the symptoms mentioned above, which has been diagnosed clinically as nasopharyngeal carcinoma as differential diagnosis. Nasoendoscopy usually reveals the large mass obliterating nasal cavity. Some cases described the mass as reddish pink with black pigmentation at base, and firm in consistency with irregular surface [8].

Computed Tomography (CT) scan of the head showed that large enhancing

Table 1. Comparison between sinonasal carcinomas.

	Incidence	Common site	Histopathology	Early presentation
Melanoma	M > F 6^{th}-7^{th} decade	Septum Nasal cavity Facial sinuses	Undifferentiated neoplasm (epithelioid, spindled, plasmacytoid, desmoplastic)	Nasal obstruction Epistaxis
Squamous cell carcinoma	M > F 7^{th} decade	Nasopharynx (fossa of rossenmullers)	Differentiated and ketatinized	Nasal obstruction Epistaxis Aural fullness
Adenocarcinoma	M > F 6^{th}-7^{th} decade	Ethmoid sinus Upper nasal cavity	Cynlindroma Subtype: tubular, cribriform, and solid	Nasal obstruction Nasal congestion Anosmia
Adenoid cystic carcinoma	F > M 5^{th}-6^{th} decade	Oral cavity Soft palate	Glandular cells-cribriform	Facial pain Facial drooping
Olfactory neuroblastoma (esthesioneuroblastoma)	M > F 3^{rd}-7^{th} decade	Cribriform plate Upper nasal vault	Small round blue cell tumours	Nasal obstruction Eye symptoms Endocrine symptoms Facial fullness Headache

Abbreviations: M: Male; F: Female.

mass, and in our case, the mass has completely obliterating the nasopharynx, oropharynx, and part of hypopharynx with local infiltration and cervical lymphadenopathies. This report displayed the classic symptoms and diagnostic findings of nasopharyngeal carcinoma.

To establish the diagnosis, biopsy of the lesion for histochemistry and immunohistochemistry is the gold standard, although this procedure maybe associated with high risk of dissemination of the tumor cells into circulation [5]. The cytological findings are found to show positive staining with anti-S-100, HMB-45, and anti-vimentin especially in metastatic malignant melanomas. Not all mucosal melanomas have positive staining with anti-S-100 [6] and therefore, another set of markers must be investigated to establish diagnosis.

In our case, diagnosis was established as the malignant cells were positive for Melan A and S100. Another subtypes of malignant melanoma, also known as amelanotic melanoma has high affinity for perineural spread and low rate of lymphatic metastasis [1] which may be misdiagnosed. In such cases, immunohistochemistry is useful in reaching the diagnosis.

For this aggressive disease, treatment can be curative with surgical removal which includes traditional open or endoscopic surgery [9] and followed by radiotherapy or palliation with chemotherapy or radiotherapy. Radiotherapy aids in local control and is indicated in the presence of positive surgical margins, local recurrence, locally advanced tumour, or sometimes for palliative purposes. However, it has also been reported to not improve overall survival [6].

Despite many different regimes, most have shown unsatisfactory results with median Overall Survival (OS) and Disease-Specific Survival (DSS) being 32 and 50 months as reported [9]. Sino-nasal malignant melanoma is known to have a high recurrence and metastasis rate, and therefore patients should maintain lifelong follow-up with PET/CT which is a valuable tool for assessment. The factors for poor prognosis of this condition include advanced age, size of the tumor, location of the tumor (especially paranasal sinuses and nasopharynx), cellular pleomorphism and distant metastasis. It has no significant associations observed for gender, margin status, or complete resection [9].

4. Conclusion

Sino-nasal malignant melanoma with its rare occurrence, it is important to exclude other differential diagnoses with histopathology and immunohistochemistry examination. The significance of establishing the diagnosis enables us to guide patients thoroughly for treatment. It is vital to get patients with the correct management and to the aggressive nature of this disease.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Zito, P.M. and Scharf, R. (2021) Melanoma of the Head and Neck. StatPearls Publishing, Tampa, FL.

[2] Bhartiya1, R. and Prasad, K.M. (2015) Malignant Melanoma of Nasal Cavity—A Case Report. *Journal of Clinical and Diagnostic Research*, **9**, ED21-ED22. https://doi.org/10.7860/JCDR/2015/17009.6995

[3] Liu, Y.X. and Saeed Sheikh, M. (2014) Melanoma: Molecular Pathogenesis and Therapeutic Management. *Molecular and Cellular Pharmacology*, **6**, 228.

[4] Thomson, L.D., Wieneke, J.A. and Miettinen, M. (2003) Sinonasal Tract and Nasopharyngeal Melanoma: A Clinicopathological Study of 115 Cases with a Proposed Staging System. *The American Journal of Surgical Pathology*, **27**, 594-611. https://doi.org/10.1097/00000478-200305000-00004

[5] Kepekci, A.H., Kig, C. and Gundogan, G.I. (2018) The Evaluation of Malignant Mucosal Melanoma of Nasal Cavity with a Rare Occasion. *International Journal of Physiology, Pathophysiology and Pharmacology*, **10**, 139-143.

[6] Alves, I.S.S., Berriel, L.G.S. and Alves, R.T. (2017) Sinonasal Melanoma: A Case Report and Literature Review. *Case Reports in Oncological Medicine*, **2017**, Article ID: 8201301. https://doi.org/10.1155/2017/8201301

[7] Samstein, R.M., Carvajal, R.D., Postow, M.A., Callahan, M.K., Shoushtari, A.N., Patel, S.G., Lee, N.Y. and Barker, C.A. (2016) Localized Sinonasal Mucosal Melanoma: Outcomes and Associations with Stage, Radiotherapy and PET Response. *Journal of the Sciences and Specialties of Head Neck*, **38**, 1310-1317. https://doi.org/10.1002/hed.24435

[8] Bothale, K.A., Maimoon, S.A., Patrikar, A.D. and Mahore, S.D. (2009) Mucosal Malignant Melanoma of the Nasal Cavity. *Indian Journal of Cancer*, **46**, 68.

[9] Na'ara, S., Mukherjee, A., Billan, S. and Gil, Z. (2020) Contemporary Multidisciplinary Management of Sinonasal Mucosal Melanoma. *OncoTargets and Therapy*, **13**, 2289-2298. https://doi.org/10.2147/OTT.S182580