PHILIPPINE JOURNAL OF OTOLARYNGOLOGY-HEAD AND NECK SURGERY VOL. 38 NO. 1 JANUARY - JUNE 2023

Sinonasal Mucosal Melanoma: A Rare Intranasal Tumor in an 89-Year-Old Woman

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Keywords: *mucosal melanoma; malignant melanoma; intranasal mass*

Mucosal melanomas are malignant tumors from melanocytes found in epithelium of nasal, oral, reproductive and gastrointestinal mucosa of the body.[1,2] As early as 1869, cases of mucosal melanomas have been described as rare and aggressive but insidious in nature.[3] The mean age of diagnosis in some studies is 60 - 70 years old,[1-7] with early detection proving to be a challenge due to non-specific early stage symptoms.[1,4] They generally have poor prognosis, high tumor recurrence and high prevalence of tumor metastasis in around 23 - 50%.[4,5] Treatment may involve surgical excision, radiotherapy or chemotherapy.[6] However, adequate and appropriate treatment can only be initiated once the diagnosis and staging are established through proper imaging and histopathologic support.[4] We present one such case.

CASE REPORT

An 89-year-old woman consulted our out-patient department (OPD) for right nasal obstruction that started two years prior with progressive hypo-nasal speech. No medications were taken nor any consult done. Ten months prior to consult, there was development of recurrent watery rhinorrhea, progressing right nasal obstruction and occasional hyposmia. A gradually enlarging fleshy mass was noted within the right nasal cavity, as well as intermittent right nasal epistaxis, initially attributed to frequent manipulation of the nares. Increasing nasal mass size and new-onset right facial pain and headache prompted OPD consult. On examination, anterior rhinoscopy showed an obstructing, irregularly shaped, smooth, painless, fleshy-colored right intranasal mass with areas of beefy-red discoloration. Bulging of the right ala was noted as well. *(Figure 1)* Examination of the left nares showed no visible masses or polyps with the nasal septum showing no signs of deviation or breaks in the mucosal surface. Endoscopy of the left nasal cavity revealed the posterior extension of the aforementioned mass from the right posterior choanae. *(Figure 2)* Otoscopic, oral, and neck examination findings were unremarkable. Due to the presence of a unilateral nasal mass with non-specific characteristics and symptoms, initial assessment favored a benign pathology, without totally ruling out malignancy. Patchy tissue discoloration and history of intermittent epistaxis warranted further investigation. Intranasal saline irrigation was initially advised while the patient underwent further work ups.

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Philipp J Otolaryngol Head Neck Surg 2023; 38 (1):61-64

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Contrast-enhanced computed tomography (CECT) imaging of the paranasal sinuses revealed an enhancing, hypodense, soft tissue mass measuring 4.2 x 3.6 x 8.0 cm in size, with areas of necrosis, completely filling up the right nasal antrum, right maxillary, right ethmoid and right frontal sinuses. Extension to the posterior nasal choana (partly obstructing the nasopharynx) and the right middle cranial fossa without osteolytic changes were observed as well. *(Figure 3)* Cranial magnetic resonance imaging (MRI) with contrast showed a heterogeneously enhancing, hyperintense, soft tissue mass on T1 weighted images, occupying the right nasal vault, extending to the posterior choanae bilaterally, partially obliterating the nasopharyngeal lumen, and extending superiorly to fill the ethmoid and the right frontal sinuses as well. A 2.8 x 2.4 x 1.7 cm expansile, peripherally enhancing focus was also seen in the greater wing of the right sphenoid. *(Figure 4)*

Endoscopic excision biopsy of the right intranasal mass was performed for further diagnosis and to provide therapeutic relief of the nasal obstruction. Intraoperatively, the tumor was noted to have polypoid and friable portions, obstructing most areas of the nasal cavity, with attachments observed at the middle third of the posterior nasal septum. Excision of the mass caused moderate bleeding. The patient was discharged home on post-operative oral antibiotics and nasal saline spray with a probable impression of an expansile inverting papilloma.

Histopathological findings revealed multiple anaplastic nuclei with several multinucleated giant cells *(Figure 5A)* and some cells containing brownish intracellular pigments or melanin *(Figure 5B)* with an impression of "pigmented undifferentiated malignancy." Immunohistochemical staining was positive for both MELAN-A and HMB45, confirming the diagnosis of a sinonasal mucosal melanoma. Three-week post-operative endoscopy showed slightly congested turbinates bilaterally, with no intranasal mass in the surgical site. *(Figure 6A)* On the second post-operative month, however, a soft tissue bulge was observed on the right posterior nasal septum. Nonetheless, the patient was relieved of the nasal obstruction, facial pain, headache, and the troublesome intermittent epistaxis.

The patient was compliant with monthly follow-ups for tumor surveillance, as the mass in the posterior septum progressively increased in size by the fourth month. *(Figure 6B)* She was advised to undergo wide excision of the remaining tumor and was referred to the hospital tumor clinic to facilitate possible adjuvant radiotherapy and chemotherapy. The importance of such measures and their effects on the prognosis was explained and emphasized as well. Despite these efforts, the patient repeatedly refused further surgical and non-surgical therapeutic measures, citing old age and the absence of any nasal symptoms as reasons for her refusal.

Figure 1. Right alar bulging with obstructing intranasal smooth, fleshy mass.

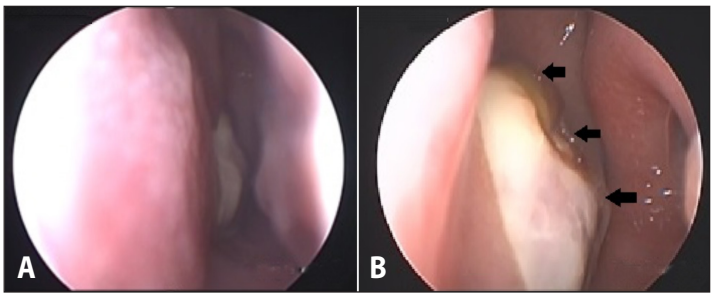

Figure 2. Left nasal endoscopy showing: **A.** left nasal cavity floor; and **B.** posterior extension of right intranasal mass from the right posterior choanae (black arrows)

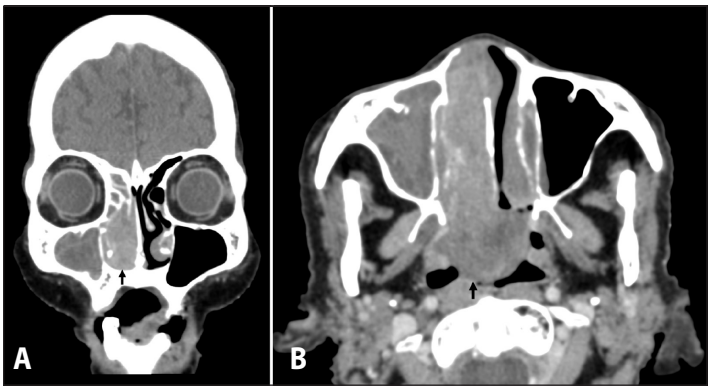

Figure 3. Contrast enhanced PNS CT scans: **A.** Coronal view showing enhancing right soft tissue mass occupying the right nasal vault and ipsilateral sinuses (black arrows); and **B.** Axial view showing right maxillary antral involvement and extension to the posterior nasal choana, partly obstructing the nasopharynx (black arrows).

DISCUSSION

Sinonasal mucosal melanoma is a rare tumor sub-type which makes up <1% of all melanomas and is believed to have an incidence rate of 0.2 – 1 case per million.[8] These intranasal tumors are common in the 7th decade of life.[1-7] At 89 years old, our patient is above average and is considered to have a poorer prognosis due to her more advanced age.[1,5] In general, 1-year survival rates range from 83 – 86% and 5-year survival rates are as low as 16 – 27%.[4] It is more common in males,[2,3] but recent data indicate a shift in tumor predilection

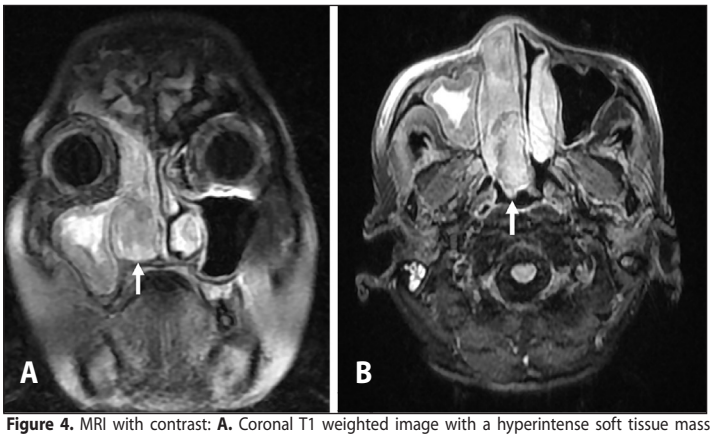

Figure 4. MRI with contrast: **A.** Coronal T1 weighted image with a hyperintense soft tissue mass occupying the right nasal vault (white arrow); and **B.** Axial T1 weighted image showing extension to posterior choanae, partially obliterating the nasopharyngeal lumen (white arrow).

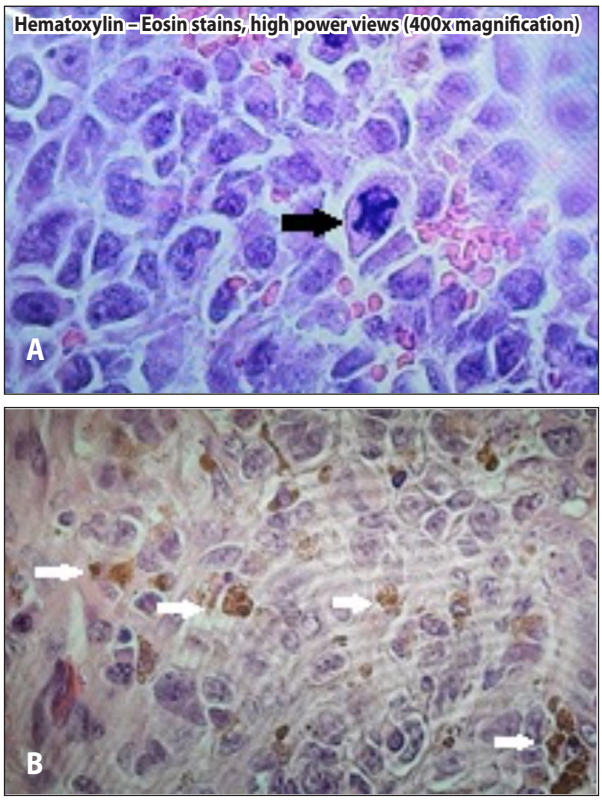

Figure 5. Histopathologic slide, Hematoxylin – Eosin stains, high power views (400x magnification): **A.** Anaplastic nuclei (black arrow); and **B.** Intracytoplasmic melanin (white arrows).

towards females.[1] On top of its rare incidence globally, this tumor is less common in Asians as more cases are observed in Caucasians.[1]

Disease identification based on history and tumor presentation alone tend to be difficult. Non-specific or absent symptoms and atypical tumor presentation often lead to delays in diagnosis.[5,6] The most common presenting symptoms for patients with benign and

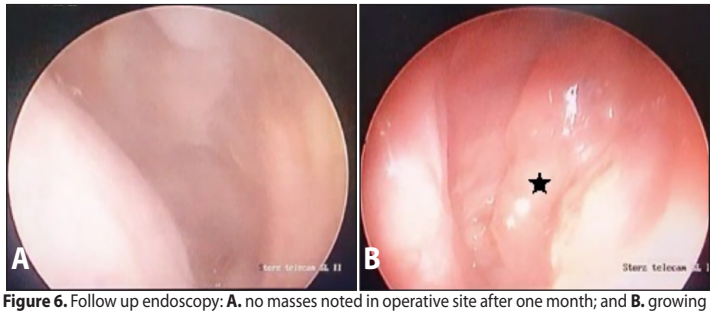

Figure 6. Follow up endoscopy: **A.** no masses noted in operative site after one month; and **B.** growing residual tumor on the right posterior nasal septum (black star) at four months.

malignant intranasal tumors combined were nasal obstruction (97%), rhinorrhea (49%), hyposmia (32%), and intermittent epistaxis (17%)[9] – most of which are non-specific. For cases of sinonasal mucosal melanomas the most observed were unilateral nasal obstruction and epistaxis, similar to our patient's experience.[2,4] Other symptoms include rhinorrhea, hyposmia, frontal headache, facial pain, proptosis and diplopia.[6,9] Epistaxis, in our case, was initially attributed to the patient's frequent nasal manipulation downplaying the potential diagnosis of a malignancy. This should have been given greater weight in the initial assessment of the tumor, as it is a common symptom in malignant intranasal masses.[10]

On physical examination, tumors may vary in appearance and size. Some present as a soft, irregular and polypoid mass,[9] while others manifest as a solid, voluminous, occasionally black or dark in color, dry, obstructing intranasal mass.[6] The presence of slate-colored, reddish, crimson, brownish or black colored tissue is highly indicative of the disease.[5,9] However, 20 – 25% of the time, an amelanotic tumor with an atypical appearance may be present.[6] The location of tumor origin also varies; the nasal septum being most common, but the inferior turbinate, middle turbinate and sinuses are also possible.[3,5,7,9] In our patient, tumor attachments were noted on the posterior nasal septum. At this point, the irregularly shaped, fleshy mass with beefy-red areas, and tumor attachments on the nasal septum already presented some malignant features. However, because benign intranasal tumors share similar characteristics,[10] further diagnostics were needed.

Diagnostic imaging aids in further tumor characterization, tumor staging and surgical planning.[5,6] A CECT scan shows tumor extent and the character of the surrounding bony structures. This can also help in identifying the nature of the tumor. In some cases of mucosal melanoma, findings may include heterogeneously hypodense, expansile and aggressive masses with or without osteolytic changes.[5,8] In our patient, the CT scan showed an expansile mass with non-osteolytic changes. As these features also appear in benign intranasal tumors, committing to a malignancy would have been difficult at that time. The surgeons were

not satisfied with the tumor delineation on CT scan, and additional MRI showed an enhancing, hyperintense, expansile, soft tissue mass on T1 weighted images that was hypointense on T2 weighted images. This radiologic finding is characteristic of melanomas on MRI probably due to high melanin content.[11] It should be noted that other tumors may also present with such features, hence the finding's poor diagnostic specificity.[5,11]

Diagnosis can only be established through histological examination of tissue biopsy.[5,12] Although it can be argued that biopsies may cause further dissemination of tumor cells into the circulation,[13] no alternative diagnostic measures are available to fully assess sinonasal mucosal melanomas. The dual goal of an excision biopsy was to provide an adequate surgical specimen and address the patient's primary complaint of nasal obstruction and bleeding. With the surgeons considering a more benign pathology pre-operatively and considering the patient's age and socio-economic situation, this appeared to be the more efficient diagnostic and therapeutic approach while improving quality of life.

Common histologic findings in melanoma, such as presence of anaplastic nuclei and observed intracytoplasmic melanin were found on our patient's tissue samples. In some cases, findings may include large fusiform or epithelioid cells with abundant eosinophilic cytoplasm or presence of undifferentiated cells, which tend to mislead clinicians towards the diagnosis of other types of tumors.[6] Again, this makes the diagnosis of the disease challenging, even at a cellular level, favoring confirmatory immunohistochemical staining. Markers that are positive include S100 protein, HMB45, Melan-A, tyrosinase, vimentin, cytokeratin and MITF.[5,6] In our patient, HMB45 and Melan-A were positive, confirming our diagnosis of a sinonasal mucosal melanoma.

Optimal management involves removal of as much tumor tissue as possible. Surgical procedures are still seen to provide better outcomes than adjuvant radiotherapy and chemotherapy.[14] Amongst the different surgical options, an open approach is still the definitive management of choice for sinonasal mucosal melanomas.[12] In recent years, however, endoscopic techniques have shown similar survival rates,[14] making them viable alternatives for surgeons in certain situations. Our patient was satisfied post-operatively and opted for a better quality of life rather than an attempt to cure, thus denying further management. This case highlights the value of endoscopic surgery as a viable option in cases presenting with an obstructing intranasal mass, due to its ability to provide biopsy material and immediate symptom relief. In addition, patients with advanced age may benefit from the less invasive procedure with a lower risk of morbidity. The downside to such a conservative surgery is the possibility of residual tumor. In such cases, proper patient education regarding the disease, its outcomes, and the possibility of a more definitive treatment, should always be given emphasis, as it plays a big part in patient care and post-operative management.

In conclusion, the value of having a high index of suspicion for, and adequate knowledge of common and uncommon disease entities cannot be overemphasized. Choosing appropriate and timely approaches play a key role in the diagnosis and treatment of such rare tumors as sinonasal mucosal melanomas. Adequate patient education and counseling should support patient decisions and optimize quality of life.

ACKNOWLEDGEMENTS

The authors would like to acknowledge Dr. Emilaine Balatibat for her invaluable contributions to the development of this paper.

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