Malignant Sinonasal Mucosal Melanoma
A Case Report and Review of the Literature
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Abstract
Sinonasal mucosal melanoma represents an aggressive and rare neoplasm that presents similarly to other common ENT complaints of nasal obstruction and epistaxis but overall portends a poor prognosis. Roughly 5% of malignant melanoma arises in nasal cavity and is much more aggressive than its cutaneous counterpart. This submission provides an overview of malignant mucosal melanoma in the sinonasal cavity, preferred diagnostic methods and modern treatment. Additionally, we report a case of malignant melanoma of the sinonasal cavity in an 88-year-old male who presented with nasal obstruction, epistaxis, proptosis, and maxillary sinus tenderness. CT and MRI imaging along with immunohistochemical staining of S100 and HMB 45 confirmed the diagnosis. The overall rarity of this presentation necessitates a comprehensive examination of the current methods to diagnose and treat this condition.

Case Presentation
- 88-year-old male presented in office with left sided epistaxis.
- The patient had been experiencing epistaxis for several months from the left nostril intermittently that generally resolved with conservative management. He denied any history of nasal packing/cautery or nasal obstructions.
- CT and MRI imaging revealed a large left sinonasal soft tissue mass with involvement of the left maxillary sinus, nasal cavity, and left palate with evidence of bone destruction.
- The differential included benign etiologies such as fibrous dysplasia, myxoid chondrosarcoma, nasal septum hemangioma, sebaceous or dermatoid nevus, and sinonasal undifferentiated carcinoma.
- Immunohistochemical staining of S100 and HMB 45 confirmed the diagnosis. The overall presentation of this patient is consistent with sinonasal mucosal melanoma.

Discussion
- Melanocytes are cells whose primary function is to produce melanin, which is a tan-brown pigmentation that protects against UV radiation.
- Most melanomas are therefore cutaneous; however, melanoma can also arise from the mucosal surfaces of the eyes, reproductive mucosal surfaces, and anywhere in the respiratory tract, including the paranasal sinus and nasal cavity mucosa.
- By definition, large, mucosal tissues are generally not exposed to sunlight and do not require UV protection. It is not entirely clear why melanocytes are found in the mucosal epithelium, but some sources suggest a possible immunologic function of these cells.
- When comparing the 5-year overall survival rate for CM (80%), the rate for MM (only 25%) portrays a much worse clinical outcome.
- When MM arises from the head and neck (MMHN), the 5-year overall survival rate reported is approximately 32% compared to other subsites.
- When comparing these forms of melanoma, locoregional nodal metastasis is far more frequent in MM (21%) compared to CM (9%) and carries a higher mortality than the N3 neck.
- The sinonasal cavity is the most common subsite of MMHN, nearly 70-80% arise in this area, representing roughly 4% of all sinonasal tumors.
- The median age of diagnosis in sinonasal MM is 60-69 years old; however, when examined in the oral cavity, the typical age group is individuals less than 40 years old.
- The typical carcinogens responsible for other sinonasal malignancies are also implicated in MMHN: tobacco, alcohol, formaldehyde, wood dust, and 1,3-butadiene. While these agents have been identified as risk factors for developing MMHN.
- In conjunction, both CT and MRI are useful in assessing overall extent of disease and allow accurate staging and surgical/therapeutic evaluation. PET/CT may be helpful to screen for distant metastasis in patients with local advanced disease.
- As in the cutaneous counterpart, surgery remains the most important therapy for sinonasal melanoma, although it can be difficult to achieve negative margins secondary to the typical lentiginous spreading behavior of MM.
- Even with aggressive surgical resections, the local recurrence rates can be particularly high in ranges from 50%-90%.
- When localized to the sinonasal region, MM may require extensive craniofacial reconstruction when there is involvement of the cribiform plate, orbital exenteration for orbit involvement, and radical nasal resection for diffuse mucosal disease.
- Endoscopic versus open resections yield similar locoregional control, with an overall decreased morbidity with endoscopic approaches.
- Therapeutic neck dissection followed by radiation therapy has been shown to improve regional control in non-advanced MMHN.
- Few systemic treatments exist for advanced mucosal melanoma. Daclizumab and high-dose interleukin (IL-2) in conjunction with MEK, PD, and CTLA-4 inhibitors
- Venetoclax, BRAF mutational inhibitors (V600E) and immunotherapy (anti-PD1, anti-PD-L1, and anti-VEGFR2) may also be considered.
- Conclusions
Although the pathogenesis between mucosal and cutaneous melanoma shares a similar molecular course, patients diagnosed with MMHN have a poorer overall outcome than the cutaneous counterpart. MMHN represents 10% of sinonasal malignancies, with the majority arising in the nasal cavity. When compared to the nasal form of MM, sinonasal MM has a much worse prognosis. Surgery remains the mainstay of treatment for these patients. While complete surgical excision of melanoma offers the only chance of cure, this is often not possible due to the location of the tumor. Even with aggressive surgery, the local recurrence rates can be high, ranging from 50% to 90%. Adjacent radiation therapy does not improve overall survival. Malignant mucosal melanoma should be considered in any patient presenting with persistent nasal or sinus symptoms, especially if there is evidence of local extension or involvement of regional lymph nodes. The key to improving outcomes lies in early detection and aggressive surgical resection, followed by adjuvant radiotherapy. While there is no cure for MMHN, early intervention can significantly improve survival rates.