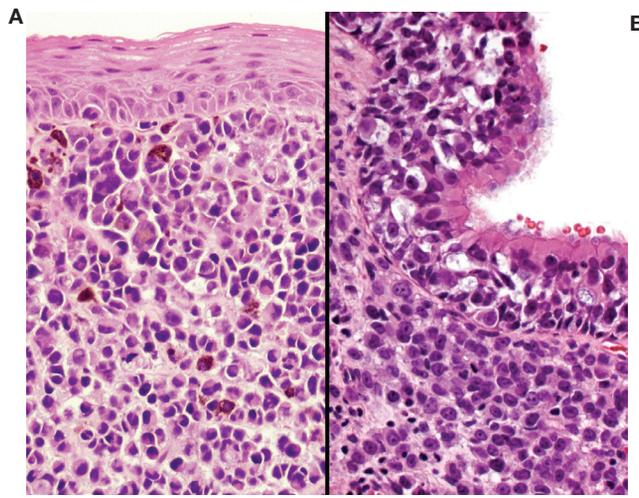


## Sinonasal tract mucosal melanoma

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*Figure 1. Mucosal melanoma can be confirmed as a primary tumor when there is surface origin, junctional activity, or Pagetoid spread. A: Neoplastic melanocytes are noted within the junctional area at the base of the squamous epithelium. The tumor cells are rhabdoid to plasmacytoid, with several cells containing melanin pigment. B: Pagetoid spread of melanoma cells is seen within a respiratory epithelium. The neoplastic cells are undifferentiated in appearance and without pigmentation.*

Mucosal melanoma is a neural-crest–derived neoplasm that originates in melanocytes and demonstrates melanocytic differentiation. Exposure to formalin and ultraviolet light are known etiologic factors; another possible factor is exposure to radiation. Approximately 15 to 20% of all skin melanomas develop on the head and neck, but less than 1% of all melanomas develop in ocular or mucosal sites, including the sinonasal tract.

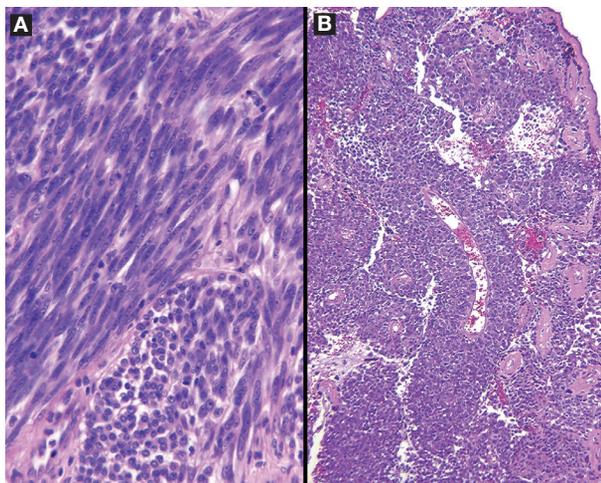
Most patients with mucosal melanoma present during the fifth to eighth decades of life; there is no predisposition to either sex. Japanese people have the highest incidence. Patients present with nasal obstruction, epistaxis or nasal discharge, polyps, or melanorrhea (black-flecked, melanin discharge). Breslow thickness and Clark level are not used to evaluate mucosal melanomas in the sinonasal tract.

Wide local excision in combination with radiation

provides the best outcome, although overall survival is poor and recurrences are common. Patients with obstructive symptoms, multiple sites of involvement, tumors of 3 cm or larger, an undifferentiated tumor on histology, a high mitotic count, and an elevated stage are more likely to have a poor prognosis.

Mucosal melanomas are frequently polypoid masses, growing as large as 6 cm, and they demonstrate surface erosion or ulceration. The color of melanin-producing tumors is generally black to dark brown. Histologically, these tumors have a protean architecture and cytology, and they frequently mimic other types of primary tumor. The presence of junctional activity and epidermal migration (Pagetoid spread) indicates that a patient has a primary tumor (figure 1). Destructive growth is common, and bone invasion, lymphovascular invasion, and perineural invasion are easily identified.

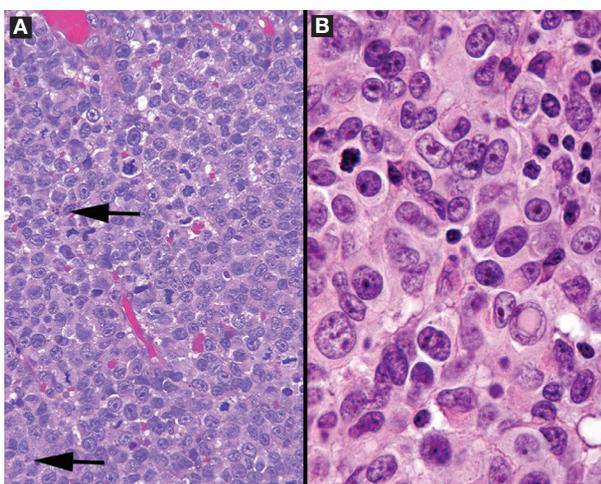
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**Figure 2.** **A:** This melanoma has a spindled appearance. Nucleoli are present, along with isolated mitoses. **B:** Several vessels are present in this tumor, and the neoplastic cells are arranged immediately around them (peritheliomatous pattern).

Many different patterns of growth can be seen, including sheet-like, fascicular, organoid, storiform, and peritheliomatous; the peritheliomatous pattern is quite distinctive and unique to mucosal melanoma (figure 2). The cells may be polygonal, epithelioid, plasmacytoid, rhabdoid, or undifferentiated (figure 3). The nuclear chromatin is usually vesicular, with prominent, enlarged, irregular, and brightly hypereosinophilic nucleoli. Intranuclear cytoplasmic inclusions are also quite characteristic (figure 3).

The presence of melanin helps confirm the diagno-



**Figure 3.** **A:** This image shows a sheet of undifferentiated to polygonal neoplastic cells. Note the prominent nucleoli and easily identifiable mitoses. Cytoplasmic melanin pigment (arrows) is sparse. **B:** The neoplastic cells have a high nucleus-to-cytoplasm ratio and prominent nucleoli. There is a single large intranuclear cytoplasmic inclusion.

sis, but many tumors do not produce melanin. Tumor necrosis and increased mitoses, including atypical forms, are common. A melanin bleach (Fontana-Masson stain) can help confirm cytoplasmic melanin.

Several immunohistochemical studies can be used to confirm melanoma, including S-100 protein, Sox10, HMB-45, melan-A, and tyrosinase. Because of the remarkable plasticity of the neoplastic cells, the histologic differential diagnosis includes olfactory neuroblastoma, sinonasal undifferentiated carcinoma, rhabdomyosarcoma, leiomyosarcoma, plasmacytoma, lymphoma, malignant peripheral nerve sheath tumor, and mesenchymal chondrosarcoma (the latter when the chondroid matrix is not seen).

### Suggested reading

- Dauer EH, Lewis JE, Rohlinger AL, et al. Sinonasal melanoma: A clinicopathologic review of 61 cases. *Otolaryngol Head Neck Surg* 2008;138(3):347-52.
- Prasad ML, Busam KJ, Patel SG, et al. Clinicopathologic differences in malignant melanoma arising in oral squamous and sinonasal respiratory mucosa of the upper aerodigestive tract. *Arch Pathol Lab Med* 2003;127(8):997-1002.
- Thompson LD, Wieneke JA, Miettinen M. Sinonasal tract and nasopharyngeal melanomas: A clinicopathologic study of 115 cases with a proposed staging system. *Am J Surg Pathol* 2003;27(5):594-611.