Melanomas are believed to arise from cells derived from neuroectodermal origin. In the orbit, these precursors are normally found in the uvea and the conjunctiva, but other orbital tissues may harbor neuroectodermal cells capable of transformation into melanoma. Blue nevi and cellular blue nevi are also believed to be derived from neural crest cells migrating with developing other orbital tissues may harbor neuromelanocytic tumor of the orbit and central nervous system. It is difficult to definitively classify the nevus identified in this case because of its small size, but the histologic features are different from blue nevus or nevus of Ota. The histologic features of the nevus cells resemble those of type A nevomelanocytes of dermal common acquired or congenital nevi. Therefore, it may represent an epithelial analog of dermal common acquired or congenital nevus. The nevus was intimately associated with the melanoma, and it is reasonable to speculate that the melanoma arose from this nevus. However, definitive proof that the nevus is indeed a precursor lesion is lacking. To our knowledge, of the approximately 50 cases of primary orbital melanoma reported in the literature, this is the first case suggested to be associated with an occult nevus of non–blue nevus type.

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Undifferentiated Sinonasal Carcinoma With Nasolacrimal Duct Obstruction

Sinonasal undifferentiated carcinoma (SNUC) is an uncommon malignancy of the nasal and paranasal sinuses. First recognized by Frier et al in 1986 as a distinct clinical and pathologic entity, it was recently classified as a large cell subtype of poorly differentiated neuroendocrine carcinoma by Mills. Common initial symptoms include nasal obstruction, epistaxis, and facial pain. Late-stage SNUC may exhibit an orbital mass, causing proptosis, diplopia, and visual loss. Orbital invasion occurs in 59% of nasal and paranasal malignant tumors, occasionally demonstrating dental symptoms. We report a case of SNUC in a patient with recurrent dacryocystitis and epiphora.

Report of a Case. A 71-year-old Chinese woman was first seen in April 1999. Her chief complaints were sudden onset of swelling and pain over the right medial canthal area for 1 week. Her medical history was remarkable for recurrent dacryocystitis with tearing and discharge. The problem had been waxing and waning for 1 year. There was no nasal bleeding or congestion. Her symptoms were resolved with oral antibiotics each time. An examination revealed a tender swelling at the right medial canthal region. There was no regurgitation from the punctum on massaging. The clinical diagnosis was dacryocystitis secondary to lacrimal drainage obstruction. Incision and drainage were offered for infection control and specimen culture, but she preferred treatment with antibiotics. Within 1 week, the inflammation subsided. Plain x-ray films of the skull and sinuses were unremarkable. A distal block was confirmed during syringing and probing of the lower canalicus.

An endoscopic examination revealed no intranasal mass. Endonasal dacryocystorhinostomy was performed, and the osteotomy was created by bone punch. No lesions were found in the lacrimal sac. To minimize postoperative fibrosis, 0.4 mg/mL of mitomycin C was applied around the osteotomy for 5 minutes. A silicone tube was inserted, linking the sac to the nasal cavity.

Two weeks after the operation, extensive granulation tissue was found in the right nasal cavity and immediately removed. The patient was instructed to clean the nasal cavity daily with isotonic solution. Within 1 week, the infection control and specimen culture were unremarkable. A distal block was confirmed during syringing and probing of the lower canalicus.

Computed tomography revealed a large mass in the lacrimal sac, extending from the medial orbit and the nasal cavity through the osteotomy (Figure 2). An incisional biopsy from the right nasal mass was performed, and a 3 × 3 × 2-mm specimen of firm, reddish tissue was submitted for histopathologic evaluation. Microscopic examination revealed 2 large, well-defined, irregular round lobules surrounded by fibroinflammatory connective tissue. The tumor was composed of solid sheets of large round and polygonal cells with hyperchromatic nuclei and prominent nucleoli (Figure 3). Many cells had vacuolated cytoplasm and periodic acid-Schiff–positive cytoplasmic granules. A central area of necrosis was present in one of the lobules (Figure 4). One mitotic figure was present in 40 high-power fields. There was moderate polymorphonuclear infiltration. No vascular invasion was observed. Special stains for mucin were negative. Immunohistochemical staining showed a positive epithelial marker for cytokeratin (AE1/AE3) (Figure 5), but negative markers for leukocyte common antigen (lymphoid), S100 protein (melanocytic), desmin (muscle), and neuron-specific enolase (neuroendocrine). The section stained for HMB45 had no tumor present. In situ hybridization for Epstein-Barr virus–encoded RNAs was negative. The pathologic diagnosis of SNUC was confirmed through consultation with William R. Green, Eye Pathology Laboratory, Johns Hopkins Hospital (Baltimore, Md).

Both systemic workup for disseminated malignancy and the serum immunoglobulin A titer for Epstein-Barr virus viral capsid antigen yielded negative results. The patient refused radical surgery. She was treated with palliative radiotherapy and chemotherapy. At the 9-month follow-up, the size of the tumor appeared to be stable, with no evidence of distant metastasis.

Comment. Sinonasal undifferentiated carcinoma is a distinctive and highly aggressive tumor, arising from the Schneiderian epithelium of the

Figure 1. A firm mass in the right medial canthal area.

Figure 2. Coronal computed tomographic scan shows a large mass in the area of the right lacrimal sac, displacing the globe laterally. The mass extends from the nasal cavity through the osteotomy site.
Figure 3. Two well-defined irregular round tumor lobules contain solid sheets of large round and polygonal cells with hyperchromatic nuclei and prominent nucleoli (hematoxylin-eosin, original magnification ×10).

Figure 4. Central necrosis and tumor cells with vacuolated cytoplasm (hematoxylin-eosin, original magnification ×40).

Figure 5. Positive immunohistochemical stain for cytokeratin AE1/AE3 epithelial marker (original magnification ×40).
nasal and paranasal sinuses. The tumor is often undiagnosed, owing to the limitation of the exact clinical course and histologic features. Sinonasal undifferentiated carcinoma is not well documented. Common symptoms are facial pain, nasal obstruction, and epistaxis. Sharara et al reported an orbital invasion in 3 patients with SNUC; 2 of these patients were initially evaluated for dental symptoms. The correct diagnosis is usually made at a late stage, when the orbital mass begins causing proptosis, diplopia, and visual loss.

Microscopically, the tumor cells are small, with high nuclear-cytoplasmic ratios and numerous mitoses. These cells form nests, trabeculae, or sheets, and show no rosette formation. Vascular invasion and necrosis are commonly found. In our case, the tumor was composed of large round and polygonal cells that appeared to belong in the large cell subtype of the poorly differentiated neuroendocrine carcinoma group I neuroectodermal tumor described by Mills. Both SNUC and sinonasal neuroendocrine carcinoma can exhibit histologic features similar to blue round cell tumors. Immunohistochemical staining differentiates SNUC from sinonasal neuroendocrine carcinoma and esthesioneuroblastoma. Sinonasal undifferentiated carcinoma usually expresses cytokeratins and epithelial membrane antigens. S100 and neuron-specific enolase stains are usually negative.

The positive staining for cytokeratins in our case suggests the epithelial origin.

The treatment requires a combination of radical surgery, irradiation, and chemotherapy. Computed tomography is indicated to rule out tumor invasion into the skull base, orbit, and pterygopalatine fossa. Magnetic resonance imaging is useful to delineate between tumor and normal tissue and to indicate areas of residual or recurrent disease at follow-up.

Malignant tumors that cause nasolacrimal duct obstruction (NLDO) are rare. No malignancies were found in a recent report of 166 patients with primary acquired NLDO. Preoperative computed tomographic imaging was not performed because our case appeared to be a typical NLDO with secondary infection. Postoperative computed tomographic imaging revealed no lesion in the maxillary and paranasal sinuses except for a mass in the lacrimal sac (Figure 2). This case could represent early SNUC arising in the anterior nasal cavity without extensive nasal or sinus extensions. Given the lack of bloody tears and the presence of a distal nasolacrimal block, a lacrimal sac tumor is unlikely. The correct diagnosis was made after dacryocystorhinostomy, when the tumor extended into the lacrimal sac from the nasal cavity through the osteotomy site. This case illustrates that malignancy should always be considered in NLDO.

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