Sebaceous Carcinoma Of The Nasal Vestibule: A Case Report
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Abstract
Sebaceous carcinomas are cutaneous appendageal tumors that are very rare in the nasal vestibule. Here we describe a case of a 90-year-old man who presented with an ulcerated mass in the right nasal vestibule existing for 18 months, also with a previous history of "basal cell carcinoma" of the same area. The physical examination revealed a destructive, irregularly shaped mass measuring 1.5 cm in maximum diameter. At follow-up, 12 months later, a new lesion of 0.5 cm in diameter had appeared in the same area. Histologic examination of the mass on scanning magnification was shown, asymmetric, poorly circumscribed, solid aggregates of variable size and shapes, composed of small undifferentiated cells. Sebaceous differentiation was evident in some tumor islands. The undifferentiated cells exhibited strong reactivity with 34bE12 and sebaceous cells in tumor islands expressed CEA and EMA. Mucicarmine and periodic acid Schiff stains with and without diastase were negative, confirming that the vacuolated clear cells were neither mucinous nor glycogen-rich squamous cells. This combination of features is that of a poorly differentiated sebaceous carcinoma.

INTRODUCTION
Sebaceous carcinoma (SC) is an uncommon skin tumor, which usually occurs on the eyelid (1, 2, 3, 4). To the best of our knowledge, vestibulum nasi presentation of SC is extremely rare; there are only two published case in the literature (5, 6). Consequently, in primary tumors of this localization, SC is not ordinarily included at the differential diagnosis. Although the largest sebaceous glands are found in the nose, a common site of extraocular head and neck SC is the parotid gland, where ectopic sebaceous glands are frequently localized in (7). There were a few isolated case reports of nasal SC (5, 6, 8, 9, 10).

CASE REPORT
We describe a case of a 90-year-old man who presented with an ulcerated mass in the right nasal vestibule existing for 18 months, with a history of "basal cell carcinoma" of the same area. On physical examination there was a destructive, irregularly shaped, extensively ulcerated mass measuring 1.5 cm in maximum diameter. An excisional biopsy was performed and the tissue was submitted for microscopic evaluation.

Histologic examination revealed an ulcerated epithelium and an infiltrating malignant neoplasm composed of irregularly shaped lobular formations (Figure 1).

Figure 1: Scanning magnification of the irregularly shaped lobular formation

Many cells showed foamy-bubbly cytoplasm and sebaceous duct-like structures in the center of some lobules (Figure 2). There were numerous mitotic figures in regions with undifferentiated, basaloid cells (Figure 3).
The sebaceous cells were variably sized and contained minimally pleomorphic, centrally located, scalloped or starry nuclei with foamy-bubbly cytoplasm. There were also numerous small, darkly staining, undifferentiated cells with oval shaped nuclei, prominent nucleoli and eosinophilic, occasionally vacuolated cytoplasm. Areas of necrosis were also seen (Figure 4). The undifferentiated cells exhibited reactivity with 34 E12, sebaceous cells in tumoral islands stained positive with EMA (Figure 5), some sebaceous cells and sebaceous duct-like structure stained positive CEA.

No labeling was observed for vimentin and actin. Mucicarmine and periodic acid Schiff stains with and without diastase were negative, confirming that the vacuolated clear cells were neither mucus cells nor glycogen-rich squamous cells. This combination of features was interpreted as those of poorly differentiated SC. Twelve months later, a new lesion of 0.5 cm in diameter had appeared in the same area with the similar histologic appearance. After 6 months of follow up, there has been no evidence of local recurrence and metastasis.

DISCUSSION

SC can be confused with benign and malignant lesions, often resulting in delayed diagnosis, which can then lead to higher morbidity and mortality (1, 2, 8). While a number of reports
have documented SC arising from nose, SC of the vestibulum nasi has extremely rarely been reported in the literature (5, 8). The current report is the third documented case of an SC arising in a vestibulum nasi. To our knowledge, 19 examples of nasal SC have been reported in the literature (5, 8).

SC generally is a tumor of older adults. Most patients under age 30 have had prior irradiation in the area (4). Extraocular SC appears to arise almost exclusively in middle aged or elderly patients with a peak in the seventh decade of life (4). Most patients have no related systemic associations, and in contrast with sebaceous adenoma, SC is less frequently associated with Muir-Torre syndrome (5, 10-12, 13, 14). The incidence of ocular lesions is slightly higher in women and in the Asian population. In contrary to the eye localization, males are affected twice as frequently as females in nasal localization. The majority of the lesions arise on the head and neck where they usually appear as an ulcerated nodule (4).

In general, the patients with SC have a 5-year survival rate of about 80% (4). Local recurrence is common, and up to a third of affected patients will develop regional lymph node metastases. Regional dissemination to draining lymph nodes seems to be a harbinger of systemic spread (4). Distant metastases are not unusual, occasionally to the lungs, central nervous system, and viscera (5).

A clinical diagnosis of sebaceous carcinoma is made initially in only one third of cases. Approximately 50% of the cases are not recognized in the initial biopsy, and 18% are misdiagnosed as squamous cell carcinoma (4).

The histological differential diagnosis of SC includes sebaceous adenoma, basal cell carcinoma with sebaceous differentiation; clear cell squamous cell carcinoma and metastatic clear cell tumor. The morphologic hallmark of sebaceous differentiation is the detection of sebaceous cells, i.e., epithelial cells with multiple, clear vacuoles containing fat, that impinge the nucleus imparting a scalloped appearance. Special stains such as oil red O may be helpful in confirming the presence of fat, but require frozen sections. Since the specimen in this case report was already fixed in formalin, we were not able to use such stain.

In the current case, clear cell areas were scant and the tumor was composed of mainly basaloid cells. On scanning magnification, the asymmetric pattern, lobulations with varied size and shape and infiltrative border of the tumor supported a diagnosis of carcinoma. Also supporting such diagnosis, there were necrosis and abundant mitotic figures. Therefore, only malignant neoplasms were considered in the differential diagnosis.

Basal cell carcinoma with sebaceous differentiation should be considered in the differential diagnosis, particularly due to the presence of basaloid cells and focal palisade alignment. Misago et al (Table 1) have stated some criteria to differentiate SC from basal cell carcinoma with sebaceous differentiation (4, 5). Basal cell carcinomas usually have cleft formation from surrounding stroma with palisading of follicular germinative cells with scant cytoplasm. On the other hand, SC may occasionally simulate the alignment of palisade with pink cytoplasm cells. Although we did not identify pagetoid spread of neoplastic cells in the overlying epidermis, this may be associated with the localization of the tumor (pagetoid spread is usually seen in eye lesions) (4, 5, 7). Basal cell carcinoma cells are composed of oval monomorphic nucleus with scant cytoplasm and no nucleoli. In contrast, SC cells commonly show severe nuclear atypia with frequent mitosis. Also more often observed in SC are well developed lobular architecture of the sebaceous cells and sebaceous duct like structures (4, 9-13).

Immunohistochemical studies were performed to try to confirm the sebaceous nature of the neoplasm. In normal skin sebaceous glands exhibited positive staining for EMA, the polyclonal anti-CEA antibody reacted with the sebaceous ducts. Sebaceous glands and inner and outer layer of the ductal portion were labeled by the antibody against 34 E12 (CK 1/5/10/14) (4). In the current case, using standard techniques, sebaceous cells demonstrated cytoplasmic / membranous reactivity for EMA and some sebaceous cells and sebaceous duct-like structures showed cytoplasmic reactivity for CEA; undifferentiated cells demonstrated reactivity with 34E12. SC cells express various epithelial markers, including cytokeratins, EMA (4-7, 17-19, 13). Although CEA is a well-established marker for sweat gland differentiation (16, 20, 23), some authors have reported focal labeling with the polyclonal anti-CEA in SC, squamous cell carcinoma, and basal cell carcinoma. (4, 16, 22, 23).

In summary, SC should be included in the differential diagnosis of epithelial tumors of the nose.

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