Sebaceous Carcinoma Ex-Pleomorphic Adenoma: A Rare Phenotypic Occurrence

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Primary sebaceous carcinoma of salivary glands is a rare entity with approximately 22 de novo documented cases. Similar tumor arising in a benign mixed tumor has only been reported once. We report a second case of sebaceous carcinoma in a pleomorphic adenoma and discuss the clinicopathologic features, histogenesis, and the differential diagnosis of this unusual tumor.

Index Words: Sebaceous carcinoma, carcinoma ex-pleomorphic adenoma, salivary gland, pleomorphic adenoma

Sebaceous differentiation can be seen as a limited feature in many known benign and malignant salivary gland tumors and may contribute to diagnostic difficulties. Primary sebaceous neoplasms of salivary gland origin are rare and comprise of various adenomas and carcinoma. Sebaceous carcinoma arising in pleomorphic adenoma is a rare occurrence with only one documented case in the English literature. The rarity in this setting and the considerations of primary dermal origin are central to the proper diagnosis of these cases.

We report a new case of sebaceous carcinoma in a patient with a long-standing pleomorphic adenoma and discuss the histogenesis, differential diagnosis, and the clinicopathologic features of this tumor and the literature.

Case Report

A 58-year-old man with a history of a left parotid mass for 15 years presented with a recent rapid growth of the mass. Subsequent fine-needle aspiration biopsy of the mass rendered a "suspicious for malignancy" diagnosis. A computerized tomography scan showed a 2.5 cm mass in the left superficial lobe of the parotid gland with focal peripheral calcifications. There was no evidence of metastatic disease either by computerized tomography scan or magnetic resonance imaging. The patient underwent total parotidectomy and regional lymph node dissection.

Pathologic Findings

The parotid specimen contained an ill-circumscribed mass that measured 3 cm in maximum dimension. Grossly, the tumor was light-tan to brown with areas of fibrosis and was firm to soft in consistency. The tumor manifested solid and partially cystic areas with focal calcification. Histopathologically, a malignant component dominated the lesion (Fig 1). Residual areas of a benign mixed tumor with hyalinized and myxoid stroma were present. The benign mixed tumor comprised less than 20% of the tumor volume and was interspersed between the malignant epithelial areas. Nests of large polygonal cells with centrally located nuclei and clear cytoplasmic vacuoles characteristic of sebaceous differentiation surrounded by basal cells with scant cytoplasm characterized the carcinoma. There were focal areas of squamoid cells and keratinization. The mitotic rate was from 12 to 15 per 10 high power field (magnification × 200). The cytoplasmic vacuoles in the sebaceous cells were negative for periodic acid-Schiff and Oil red O. Immunohistochemical stains for androgen receptor manifested focal nuclear positivity in the malignant cells, but were negative in the benign component of the tumor. All lymph nodes dissected were negative for metastatic disease.
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Discussion

Sebaceous differentiation in classifiable benign and malignant salivary gland tumors has not uncommonly been reported.\(^1\) However, primary sebaceous adenomas and carcinomas\(^2,5-7\) are rare and may lead to differential diagnostic difficulties. Similar tumors developing in pleomorphic adenoma are rare, with only a single case report in the English literature.\(^2\) The rarity and unfamiliarity of this occurrence may lead to differential diagnostic difficulties with primary dermal origin.

Our case showed invasive sebaceous carcinoma in a background of pleomorphic adenoma and resembles, in presentation and phenotypic manifestations, the earlier reported case by Tsukada et al.\(^2\) Both tumors occurred in the left parotid, were nearly the same size, and were treated by superficial parotidectomy and lymph node dissection. In both cases, no evidence of lymph node metastasis was found at the time of presentation. Histologically, the malignant sebaceous features dominated our tumor and were composed of nests and clusters of neoplastic cells with sebaceous features within areas of hyalinized stroma of a benign mixed tumor. The previously reported case, however, recurred twice in 2 years.

Patients with primary salivary sebaceous carcinomas have ranged in age from 17 to 92 years, with a mean of 60 years, without sex predilection. The tumors have ranged in size from 0.6 to 8.5 cm, and almost all were located in the parotid gland. The primary treatment consisted of superficial or total parotidectomy with or without neck lymph node dissection. Sixteen patients are alive and well and six have had local recurrence and metastasis with a follow-up ranging from 10 to 30 years.\(^2,5-9\) In the

Figure 1. (A, B and C) Areas of pleomorphic adenoma with cartilaginous and fibrotic stroma and sebaceous carcinoma. (D) High-power view of sebaceous carcinoma showing malignant cellular nests with cytoplasmic vaculizations with central and peripheral large nuclei.
only documented sebaceous carcinoma ex-pleomorphic adenoma, the patient experienced two recurrences after his initial parotidectomy. Because of the extreme infrequency of primary salivary origin efforts to exclude local extension or metastasis from a nearby primary dermal sebaceous carcinoma or adnexal carcinoma must be made.

Carcinoma ex-pleomorphic adenoma may present within pleomorphic adenoma without evidence of infiltration of the surrounding tissue (noninvasive) or invasive.10,11 Patients with only intratumoral carcinomas usually run a benign course. Olsen and Lewis,12 in a study of 73 cases, found that the histologic subtype was not a significant prognostic factor. However, Tortoledo et al13 found that the histologic subtype has a significant impact on survival; others have also shown that tumor grade is a significant prognostic factor.14

The histogenesis of salivary sebaceous carcinoma remains a subject of debate. Some investigators have suggested that it may represent an aggressive form of mucoepidermoid carcinoma based on the presence of mucinous cells and glandular components in some of these tumors. An origin from displaced epidermal derived sebaceous tissue to the parotid during embryogenesis has also been proposed.15 More plausible is an evolution from an undifferentiated intercalated or striated ductal cell capable of sebaceous, ductal, and squamous differentiation.9 Typical sebaceous glands have been noted in normal parotid salivary glands in all age groups and have been attributed to differentiation from ductal cells.16,17

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References