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CASE REPORT

Papillary cystadenocarcinoma of the tongue

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ABSTRACT
While squamous cell carcinoma is certainly the most common malignant oral cavity neoplasm, one must always be cognizant of a variety of less Common pathologies, including minor salivary gland tumours. Salivary gland tumours of the tongue are rare. The most common type is low-grade mucoepidermoid carcinoma, followed by adenoid cystic carcinoma. Papillary cystadenocarcinoma of the tongue is an extremely rare malignant neoplasm. We report here a case of papillary cystadenocarcinoma in a 60-year-old lady who presented with a large pedunculated mass with localization limited to the base of the tongue. This case was also unusual because the tumour had not metastasized. The patient was treated with surgery and radiotherapy.
Key words: Papillary cystadenocarcinoma, tongue, salivary gland tumour

INTRODUCTION

Papillary cystadenocarcinoma of the salivary glands is an extremely rare and distinct malignant neoplasm that was first classified as a distinctive neoplasm in 1991 by WHO.[1] Until then, it was classified as an atypical type of adenocarcinoma.

It is defined as a low-grade neoplasm by WHO that most commonly arises in the major salivary glands, mainly the parotid gland. In the minor salivary glands, lip, buccal mucosa, palate, tongue, floor of the mouth and the retromolar area are the sites of predilection. However, some aggressive or high-grade variants exhibiting an increased metastatic potential have also been reported.[2,3] It has been suggested that these neoplasms may arise from excretory duct reserve cells. The incidence is rare; however, the adenocarcinomas account for approximately 2–3% of all parotid neoplasms and 15% of all parotid carcinomas. Usually, it is seen in the age range of 20–86 years (mean 58.8 years), with a slight male predominance or no difference with respect to gender.[4]

Clinically, the reported sizes of the neoplasm have ranged from 0.4–6 cm in diameter (mean 2.2–2.4 cm); and the neoplasm presents as firm, solitary, asymptomatic swelling with firm attachment to the surrounding tissues. In approximately 25% of the patients, it manifests with pain or facial paralysis. These tumours are usually unencapsulated and may be markedly invasive.[5]

This neoplasm has revealed more diversity in its histopathological features than had at one time been considered. Histologically, the tumour is characterized by cysts and papillary epithelial projections. As the name implies, the architecture of cystadenocarcinoma is dominated by large cystic structures. The cell linings vary from columnar to cuboidal to simple squamous. Approximately 75% have papillary features. Many of these tumours are of low to moderate grade, but tumours of high grade have been reported. Malignancy is confirmed by the presence of nuclear pleomorphism, mitosis and an infiltrative growth pattern.[2]

Metastasis usually occurs in high-grade adenocarcinoma. About 25% of patients have regional metastasis, while 20% of cases present with systemic metastasis. Approximately 5% of tumours recur locally after excision. The prognosis varies according to the degree of differentiation and extent of the tumour.[6]

Treatment involves local excision. Neck dissection and postoperative radiation therapy are used for clinically positive neck metastases. Postoperatively, patients should be followed closely because these tumours have a propensity for local recurrence; also, a solitary nodal metastasis may arise years after the initial resection.[3]

CASE REPORT

A 60-year-old female patient reported with a mass on the tongue of one and half months' duration. She had a habit of tobacco chewing since 20 years. On intraoral examination, a pedunculated mass which was firm in consistency and measuring about 6 × 4 cm was seen on the ventral surface of the tongue, encroaching towards the dorsal surface. Mass was adherent to the tongue musculature. Patient had right

submandibular lymphadenopathy. Lymph nodes were mobile and nontender. TNM status was $T_2N_1M_0$.

An incisional biopsy was done and the tissue specimen was sent for histopathological analysis. Histologically, hematoxylin- and eosin-stained tissue sections showed stratified squamous epithelium overlying tumour mass in the connective tissue stroma [Figure 1]. Tumour mass was characterized by various-sized cystic cavities in which papillary epithelial projections with thin fibrovascular cores were observed [Figure 2]. Cystic spaces contained mucous [Figure 3]. The papillary projections consisted of one to several layers of columnar epithelial cells with eosinophilic or occasionally clear cytoplasm. Some low columnar-cuboidal epithelial cells were also intermingled among the tumour cells. Moderately atypical epithelial cells with nuclear pleomorphism, including mutinucleation, and evident nucleoli were also seen among the tumour cells [Figure 4]. Mitotic figures were occasionally observed. Tumour cells were positive for PAS and alcian blue staining on the luminal surface, thus indicating presence of mucin in the carcinoma cells. Intracytoplasmic mucin was also demonstrated in several cells. Based on the clinical and pathological findings, the tumour was diagnosed as a papillary cystadenocarcinoma. The excised tissue specimen of the pedunculated mass measured 5.5 × 4.5 cm in size, and the cut surface was white in color with a solid appearance [Figure 5]. It showed the same histopathological features as those of the incisional biopsy specimen.

Papillary cystadenocarcinoma is defined as a low-grade carcinoma by WHO. The present case was considered to be low-grade carcinoma because the tumour tissue was encapsulated by dense fibrous connective tissue and showed no regional lymph node metastasis and did not recur within one year of follow-up.

DISCUSSION

The occurrence of papillary cystadenocarcinoma has been noted in the ovary, gallbladder, bile duct, pancreas, mammary gland, thyroid gland and upper respiratory tract. However, papillary cystadenocarcinoma in the salivary gland is still a rare

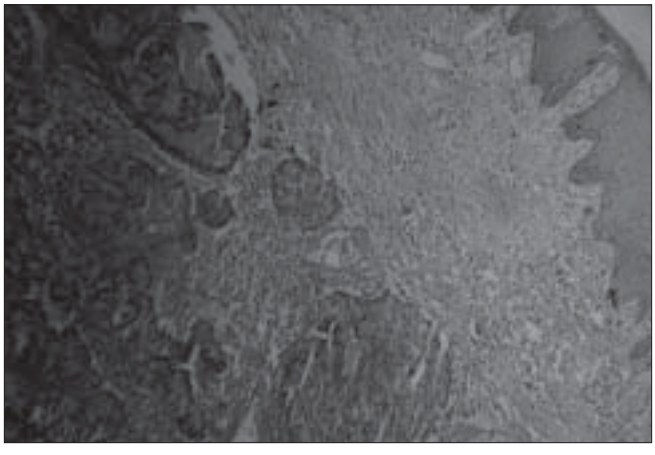

Figure 1: Low magnification of tissue specimen showing tumour mass which is encapsulated

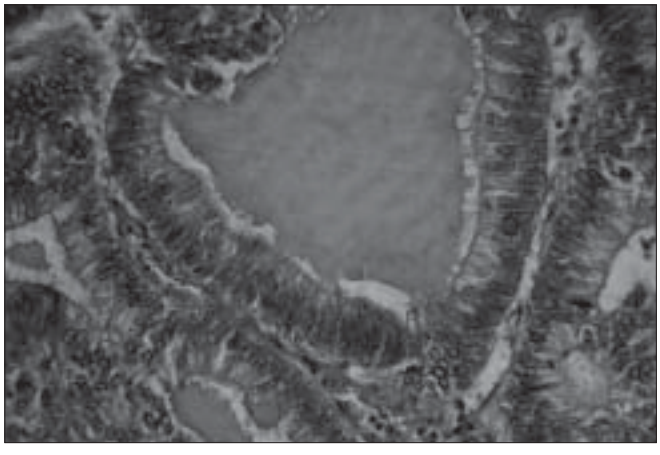

Figure 3: Higher magnification of papillary projections consisting of a single to several layers of columnar epithelial cells with either eosinophilic or occasionally clear, cytoplasm

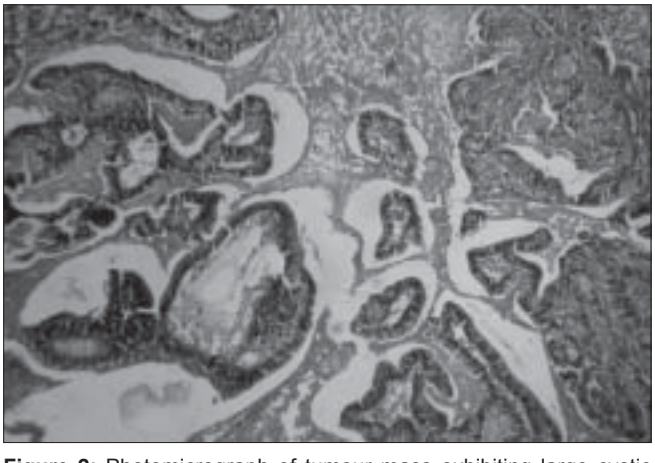

Figure 2: Photomicrograph of tumour mass exhibiting large cystic cavities and intracystic proliferation of the tumour tissue with papillary epithelial projections

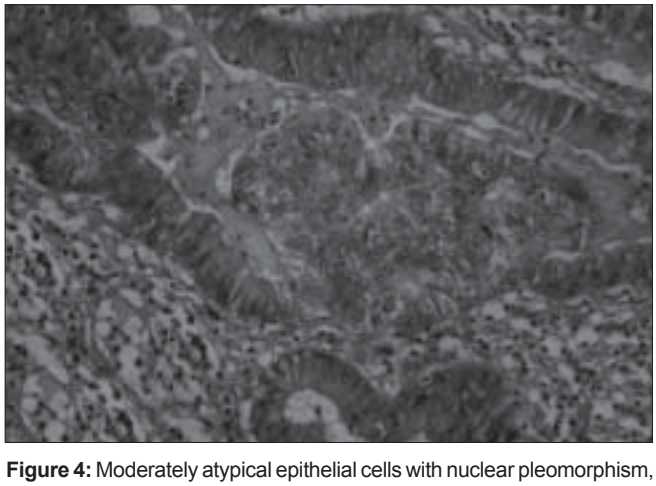

Figure 4: Moderately atypical epithelial cells with nuclear pleomorphism, including mutinucleation and evident nucleoli seen among the tumour cells

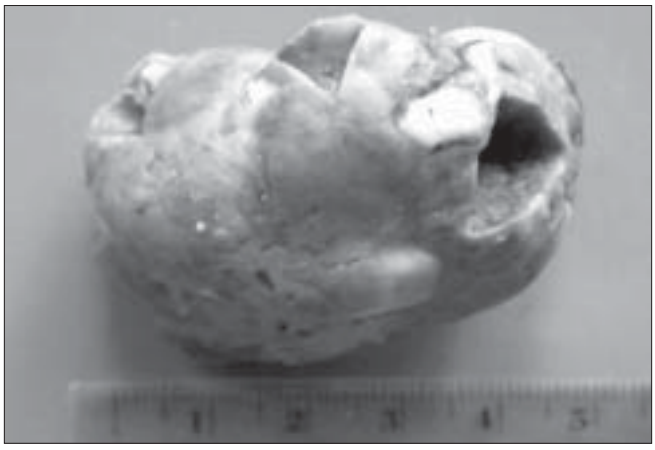

Figure 5: Gross specimen of pedunculated mass measuring 5.5 x 4.5 cm

neoplasm[7,8] and has previously been designated to be either adenocarcinoma, papillary cystadenoma, papillary-cystic carcinoma, papillary adenocarcinoma, cystadenocarcinoma or low-grade papillary adenocarcinoma.[9]

According to the WHO definition,[1] papillary cystadenocarcinoma characteristically exhibits cysts and papillary endocystic projections. However, cystic and/or papillary structures are not peculiar histological features to this neoplasm and may be observed as either main or minor features in a variety of salivary gland neoplasms. It is therefore important to distinguish the following salivary gland tumours from papillary cystadenocarcinoma when papillary cystic growth is evident in a tumour tissue: acinic cell carcinoma (ACC) with a papillary cystic growth pattern, mucoepidermoid carcinoma (MEC), salivary duct carcinoma (SDC), polymorphous low-grade adenocarcinoma (PLGA) and cystadenoma. This lesion must also be differentiated from metastatic papillary carcinomas arising in other organs, including the thyroid, ovary, intrahepatic bile duct, pancreas and gastrointestinal tract. The present case showed no acinar differentiation in the histological findings, while the parenchymal epithelial cells showed a columnar appearance rather than either a tombstone cuboidal or hobnail-like conformation, which are common in adenoid cystic carcinoma with a papillary cystic growth pattern.[10] Hence acinic cell carcinoma with a papillary cystic growth pattern was ruled out.

Other salivary gland tumours such as MEC, SDC and PLGA exhibit the typical histological characteristics in parts of the tumour tissue, in addition to a papillary cystic structure; and therefore, a differential diagnosis of these tumours from the papillary cystadenocarcinoma is not as difficult as in the case of ACC with a papillary cystic growth pattern.

The benign cystadenoma could be ruled out by the presence of cytologic atypia. The possibility of a metastatic carcinoma was ruled out as no positive data was observed in the preoperative evaluation of the patient.[11]

The papillary cystadenocarcinomas have usually been reported to have a good prognosis. However, clarification of the type of adenocarcinoma with a histologic description should be obtained and a close follow-up would be necessary in order to determine an appropriate treatment approach.

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