Abstract

Carcinoma ex pleomorphic adenoma is a rare malignant salivary gland tumor. The carcinomatous component of the carcinoma ex pleomorphic adenoma is mostly one type such as adenocarcinoma NOS, salivary duct carcinoma and undifferentiated carcinoma. We present a case of carcinoma ex pleomorphic adenoma including two carcinomatous components. The tumor occurred in the palate of a 70-year-old man. Histopathologically, the tumor was composed of both benign pleomorphic adenoma and the carcinoma area that showed adenocarcinoma NOS and squamous cell carcinoma. Finally this case was diagnosed as carcinoma ex pleomorphic adenoma including two carcinomatous components.

Key words: Carcinoma ex Pleomorphic Adenoma, Adenocarcinoma NOS, Squamous Cell Carcinoma, Palate, Minor Salivary Gland, Malignant Salivary Gland Tumor

I. Introduction

Carcinoma ex pleomorphic adenoma (CXPA) is defined as a carcinoma observed in pleomorphic adenoma and commonly occurs in the major salivary gland, CXPA is ranked as the sixth most common carcinoma in salivary gland tumors. As the components of the carcinoma originate from ductal epithelium and myoepithelial cells as well as benign pleomorphic adenoma, it is known that the carcinoma presents various morphological patterns and modes of invasion. The carcinoma generally includes only one kind of carcinoma component such as adenocarcinoma NOS, salivary duct carcinoma and undifferentiated carcinoma. However, The malignant component is often difficult to subclassify as one of exactly defined carcinoma categories, and existence of multi-carcinomatous components in one tumor is rare.

We here report a very rare case of CXPA with two kinds of carcinomas: adenocarcinoma NOS and squamous cell carcinoma, arising in the palate.
II. Case Report

A 70-year-old male was admitted to Department of Oral and Maxillofacial Surgery, Tokyo Women’s Medical University, Medical Center East, with bleeding from mass in the right palate. He had become aware of the existence of a tumor mass on the right palate since 2003 and visited an oral surgery office of a certain hospital. He was diagnosed with a benign tumor and followed up, but the follow-up was discontinued at the discretion of the patient. In 2008, he suddenly had bleeding from the mass and visited the emergency department of our hospital. His past medical history included diabetes, high blood pressure, glaucoma and prostatic hypertrophy.

On the oral examination, a painful tumor mass extending from the right hard palate to the soft palate was recognized. The mass was poorly-marginated and 36 × 22 mm in size. There was a grey necrotic part in the center of the tumor mass (Fig. 1). It was soft and no inductions were observed. On the MRI findings by T2 enhanced imaging, the mass shows circular and well-circumscribed form and high intensity area with a low intensity area considered to be ulcer, in both the coronal and the sagittal sections (Fig. 2). Biopsy was carried out and diagnosed with CXPA.

A tumor resection of the right palate was carried out under general anesthesia. Histopathologically, the tumor was circumscribed by a comparatively thick fibrous connective capsule (Fig. 3A) and the inside structure of the tumor was broadly divided by three parts. Most part of the tumor consisted of tubular structures with eosinophilic cytoplasm and oval-shaped nuclei (Fig. 3A). Hyalinization, calcification and chondromatous parts were also recognized in part of the stroma, suggesting benign pleomorphic adenoma (Fig. 3B). Continuously, growth of small luminal structures with atypia such as hyperchromatic nuclei and increase of nuclear to cytoplasm ratios, was recognized (Fig. 3C). PAS-positive element was observed within both the luminal structure and cytoplasm of tumor cells, indicating adenocarcinoma (Fig. 3D). Furthermore, squamous differentiation was also seen in the tumor nest with severe cellular atypia, suggesting well-differentiated squamous cell carcinoma (Fig. 3E). Finally, this case was diagnosed as CXPA with two carcinoma components; adenocarcinoma and squamous cell carcinoma.

About three years has elapsed since the surgery. There are no abnormalities in the part of injury, there are no problems in eating or swallowing, and no recurrences or metastasis has been observed.

III. Discussion

CXPA accounts for 1-5% of all salivary gland tumors, 5-15% of all malignant salivary gland tumors, and approximately 6% of carcinoma in all pleomorphic adenomas1-4). CXPA often occurs in females of the middle and old age. The most frequent site is the parotid gland (67 to 84 %) in major salivary glands, and the palate is the most frequent site in the minor salivary glands5,6,7). On the other hand, Buchner et al.8) researched 380 cases of minor salivary gland tumors, and reported that the frequency of the carcinoma (ex pleomorphic adenoma) was 0.5% in the minor salivary gland tumors. They also reviewed the English literatures and the number of CXPA was only 14 out of 1441 cases in minor salivary gland tumors. Moreover, other reports pointed out that incidence of CXPAs ranged from 0.6 to 3.8%8). Therefore, the case of CXPA arising in the palate is thought to be extremely rare.

Carcinoma components constituting CXPA are mostly adenocarcinoma NOS, salivary duct carcinoma and
undifferentiated carcinoma. However, other malignant tumors such as mucoepidermoid carcinoma, adenoid cystic carcinoma, myoepithelial carcinoma, acinic cell carcinoma had been also reported\(^3\). Carcinoma component of squamous cell carcinoma is rare and a rate is less than \(10\%\)^5. Furthermore, carcinoma component is generally one kind and multi-components are rare. Only one case of CSPA including two carcinoma components;
salivary duct carcinoma and squamous cell carcinoma occurred in the buccal region \(^5\). To the best of knowledge, our case is the first report of CXPA including both adenocarcinoma and squamous cell carcinoma.

CXPA is generally treated by extensive surgical excision, and its prognosis is relatively good. The prognosis is related to factors such as tumor size, the proportion of the necrotic part, infiltration into the capsule or surrounding tissues, finding of cell nucleus division, invasion into blood vessels and nerves\(^1\,4,7,10,11\). However, there had been few reports discussed about relation between kind of carcinoma components in the tumor and prognosis\(^10,11\). Component of high-grade malignancy such as adenocarcinoma NOS, undifferentiated carcinoma or salivary duct carcinoma shows poor prognosis\(^4\). Katabi et al.\(^11\) reported 43 cases of CXPA and component of myoepithelial carcinoma often showed extensive infiltration and metastasis, and it led to an adverse prognosis. Therefore, kind of carcinoma component is considered as one of important factors for predict prognosis. In this case, we observed both squamous cell carcinoma and adenocarcinoma NOS as carcinoma component. Incidence of squamous cell carcinoma is approximately a rate of 10\(^5\),\(^9\), and multi-carcinoma components are unusual in CXPA. Therefore, prognosis has been unknown. Strict and sufficient follow-up is required, although three years have elapsed since the surgery.

**IV. References**