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Case Report

Familial adenoid cystic carcinoma of sublingual salivary glands

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ABSTRACT

Tumors of the sublingual salivary gland are extremely rare. Most of the sublingual tumors are malignant, adenoid cystic carcinoma (ACC) and mucoepidermoid carcinoma being the most common histological types. Here we report the first case of a familial occurrence of ACC in a family in which the father and daughter were treated for ACC of the sublingual salivary gland. A 75-year-old man was referred with a swelling of the floor of the mouth on his right side and diagnosed as ACC of sublingual salivary gland synchronous with adenocarcinoma of the lung. One year later, his daughter presented, at the age of 46 years, with a swelling of the floor of the mouth on her right side, which was also diagnosed as ACC. This is the first case of familial recurring ACC of sublingual salivary gland worldwide.

1. Introduction

Sublingual salivary gland neoplasms are uncommon representing up to 2.6% of all salivary gland tumors [1]. Most of them are malignant, and ACC and mucoepidermoid carcinoma are the most common types [2,3]. ACC is a malignant tumor originating from salivary glands, and has some characteristic features, including slow growth, perineural invasion, local recurrence, and distant metastasis. ACC represents less than 1% of all head and neck malignancies. Among salivary gland tumors, it is the fifth most common epithelial neoplasm following mucoepidermoid carcinoma, adenocarcinoma not otherwise specified (NOS), acinic cell adenocarcinoma, and polymorphous low-grade adenocarcinoma (PGLA). Greater than half of the ACC occur in the parotid glands and submandibular glands. The most common intraoral site is the palate [4–6].

A familial occurrence of any salivary gland neoplasm is extremely rare, with only few reports found in the literature, including pleomorphic adenoma [7–10], Warthin's tumor [11–14], malignant lymphoepithelial lesion (MLEL) [15,16], carcinoma of the submandibular gland [17], and acinic cell carcinoma [18,19]. With a brief literature review, we report a familial occurrence of ACC involving sublingual salivary gland. To our knowledge, this is the first report described in the literature.

2. Case reports

2.1. Case 1

A 75-year-old man was referred to our hospital with a swelling of the floor of the mouth on his right side and paralysis of the ipsilateral tongue in January 1995. The patient had noticed the painless swelling of the floor of the mouth in the right side for the last 4 months and the mass had been growing gradually ever since. He had no history of smoking and drinking. Intraoral examination showed a mass measuring about 48 mm × 38 mm, which was elastic, movable, and firm, with an ulcer in the floor of the mouth on the right side involving sublingual gland. The tongue was pushed up by the tumor (Fig. 1A). Computed tomography (CT) scan of the head and neck region revealed a mass arising in the right sublingual space, which is over the mylohyoid muscle and under the mucosa line, infiltrating a part of the tongue, and invading the floor of the mouth on the left side (Fig. 2A). There was no evidence of metastatic lymph nodes. The initial histopathological diagnosis by the incisional biopsy was ACC, which showed cribriform growth pattern, with a clinical staging of T3N0M0 (Fig. 3A). However, detailed systemic examination with thoracoabdominal CT scan revealed multiple nodular lesions in the right middle lobar bronchus (Fig. 2B). The lesion was the highly suspected non-small cell lung carcinoma, as confirmed by the cytology of bronco fiber. After all, the lesion was diagnosed as primary lung cancer that was histopathologically confirmed adenocarcinoma by a transbronchial

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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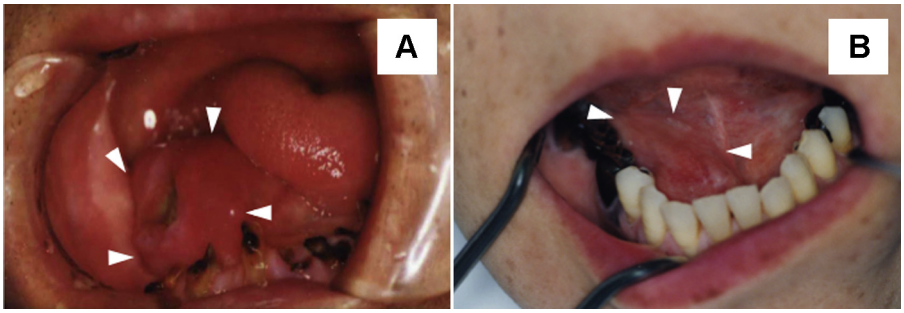

Fig. 1. Intraoral views showing (A) a firm, exophytic mass with an ulcer in the floor of the mouth on the right side (arrow heads); (B) a non-tender mass in the floor of the mouth on the right side (arrow heads).

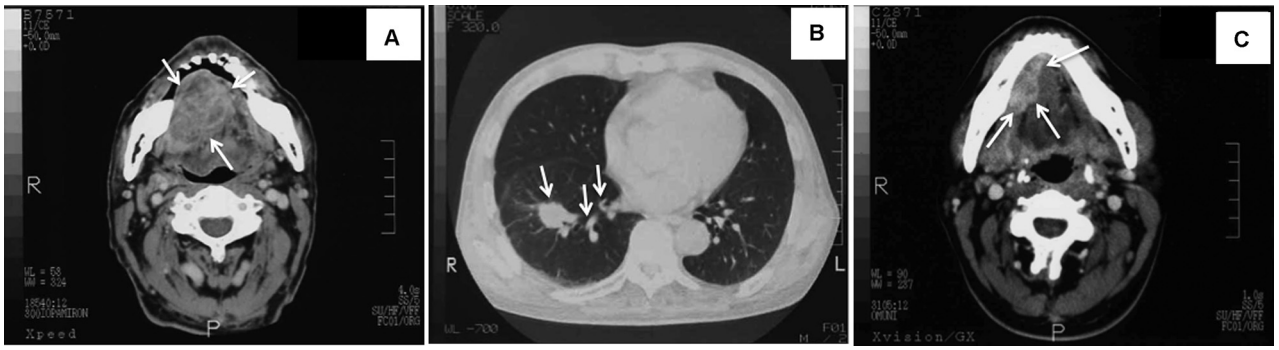

Fig. 2. Computed tomography (CT) images showing (A) a mass infiltrating a part of the tongue and invading the floor of the mouth on the left side (arrows); (B) multiple nodular lesions in the right middle lobar bronchus (arrows); (C) a mass involving the floor of the mouth on the right side and adjacent to the lingual cortex of the mandible (arrows).

biopsy. Because ACC is slow-growing, the patient was admitted to another hospital to treat the lung cancer first. However, in October 1995, he eventually died of vertebral bone metastasis and the progression of lung cancer.

2.2. Case 2

One year later, the patient's daughter at the age of 46 years presented with a swelling of the floor of the mouth on her right side. She had noticed a painless swelling and a slowly growing mass without any associated symptoms since January 1995, when her father had been first referred to the hospital. She also denied a history of drinking and smoking. Intraoral examination showed a firm mass measuring about 12 mm × 28 mm in the floor of the mouth on the right side (Fig. 1B). CT scan identified the lesion arising from the right sublingual salivary gland, extending into the floor of the mouth (Fig. 2C). Histopathological examination by the incisional biopsy revealed a typical ACC with cribriform growth pattern (Fig. 3B). No metastases were found in her regional lymph nodes and other organs with CT scan of both the head and neck region and thoracoabdominal region. Based on these findings with the grading of T2N0M0, she underwent tumor resection with selective neck dissection of the right submandibular triangle by pull-through method and marginal mandibulectomy. The tumor was resected *en bloc* with safety margins. There was a surgical finding that the tumor followed the sublingual gland topography, and occupied the space of part of the gland. Histopathological findings of surgical specimen showed an evidence of a tumor replacing the parenchyma of the sublingual gland. Furthermore, histopathological sections showed evidence of perineural invasion with the formation of cribriform pattern in the primary lesion. A diagnosis of ACC with cribriform growth pattern was made based on the characteristic clinicopathologic findings. None of the lymph nodes showed metastasis in the histopathological sections. Chemotherapy

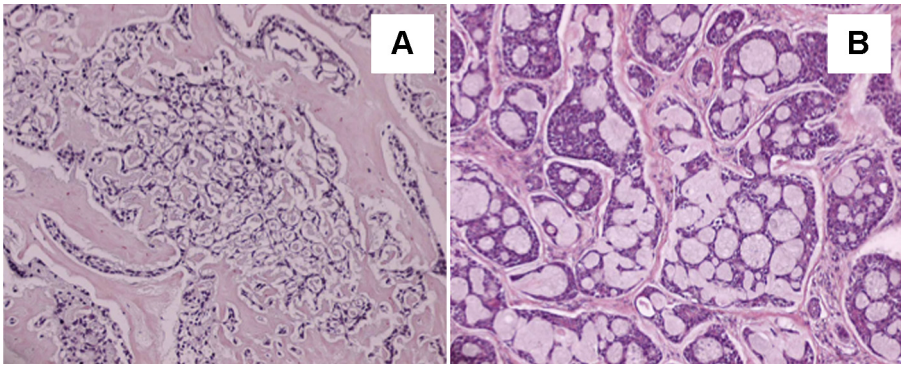

Fig. 3. Histopathological findings revealed (A) tumor cells having large and ovoid nuclei with scanty cytoplasm, form strands are embedded in a prominent, eosinophilic hyaline stroma; (B) tumor cells composed of epithelial and myoepithelial cells proliferate and form nests which was characterized as typical cribriform pattern (H&E staining, original magnification 100×).