Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology 30 (2018) 252–256

Contents lists available at ScienceDirect

Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: www.elsevier.com/locate/jomsmp

Case Report

Mucinous adenocarcinoma with signet ring cell features of the sublingual gland: A case report with an immunohistochemical analysis

Masashi Nagata [a,b,*], Kimihide Kusafuka [c], Ryoji Yoshida [a], Kenta Kawahara [a], Yoshihiro Nakagawa [a], Masafumi Nakamoto [a], Masanori Shinohara [a,d], Hideki Nakayama [a]

[a] Department of Oral & Maxillofacial Surgery, Graduate School of Medical Sciences, Kumamoto University, Kumamoto, Japan
[b] Department of Dentistry and Oral Surgery, Aso Iizuka Hospital, Fukuoka, Japan
[c] Pathology Division, Shizuoka Cancer Center, Shizuoka, Japan
[d] Ito Dento-Maxillofacial Hospital, Kumamoto, Japan

ARTICLE INFO

Article history:
Received 16 May 2017
Received in revised form
16 December 2017
Accepted 25 December 2017
Available online 3 February 2018

Keywords:
Mucinous adenocarcinoma
Sublingual gland
Signet ring cell feature
Immunohistochemistry
Poor prognosis

ABSTRACT

Mucinous adenocarcinoma (MAC) is a highly aggressive tumor, and its origin in the salivary glands is extremely rare. We report one case of salivary MAC showing highly invasive features with signet ring cells and immunohistochemical analysis. A 77-year-old Japanese woman who noticed painless swelling of the right floor of the mouth received surgical treatment and was diagnosed with primary MAC of the sublingual gland. The tumor consisted of two components: a glandular pattern and diffuse pattern with large mucinous lakes containing numerous signet ring cells. The tumor cells were immunopositive for CEA, CA125 and CK7 and strongly positive for Her-2 but negative for CK20, androgen receptor, GCDFP-15 and p63. The prognosis was poor, and the patient developed peritoneal metastasis and died. This case suggests that signet ring cell features in MAC may be a phenotype of aggression, and Her-2 may be a therapeutic target for MAC.

1. Introduction

Mucinous adenocarcinoma (MAC) is an elusive tumor of the salivary glands because of its rarity and its diagnostic uncertainty. The most common localization is the large intestine (colon), and MAC has been shown to constitute 10%–20% of colorectal cancers, followed by the pancreas, ovary, lung, prostate and breast [1]. MAC of the head and neck is exceedingly rare and accounts for only about 3% of salivary gland tumors. Salivary gland MACs are characterized by high aggression due to a high rate of local recurrence and nodal metastasis [1,2].

The unifying pathologic description of MAC in the current standard textbooks is, "small clusters and single carcinoma cells floating in large pools of extracellular mucin compartmentalized by fibrous septa". In addition, at least half of the tumor cells should produce mucus [3]. Histologically, MAC may be similar to mucinous eccrine carcinoma of the skin, mucinous carcinoma of the breast and colloid carcinoma of the bowel [4]. An important feature of MAC is that the mucus-producing cells show positive staining on mucicarmine and Alcian blue staining. However, these cells are also characteristic for other salivary gland tumors. A definitive diagnosis of MAC as a primary salivary gland tumor is typically achieved by exclusion of metastatic disease. Due to these complexities associated with discrimination, immunohistochemical staining is helpful in the differential diagnosis.

MAC is usually treated by surgery; however, a standard treatment has not been established due to its rarity. The role of chemotherapy and radiotherapy is controversial. The prognosis depends on the stage of the disease [5]. We herein report the clinicopathologic and immunohistochemical findings in a case of MAC occurring in the sublingual gland.

* Corresponding author at: Department of Oral & Maxillofacial Surgery, Graduate School of Medical Sciences, Kumamoto University, 1-1-1 Honjo, Kumamoto, 860-8556, Japan.
E-mail address: nagatama0213@gmail.com (M. Nagata).

https://doi.org/10.1016/j.ajoms.2017.12.008

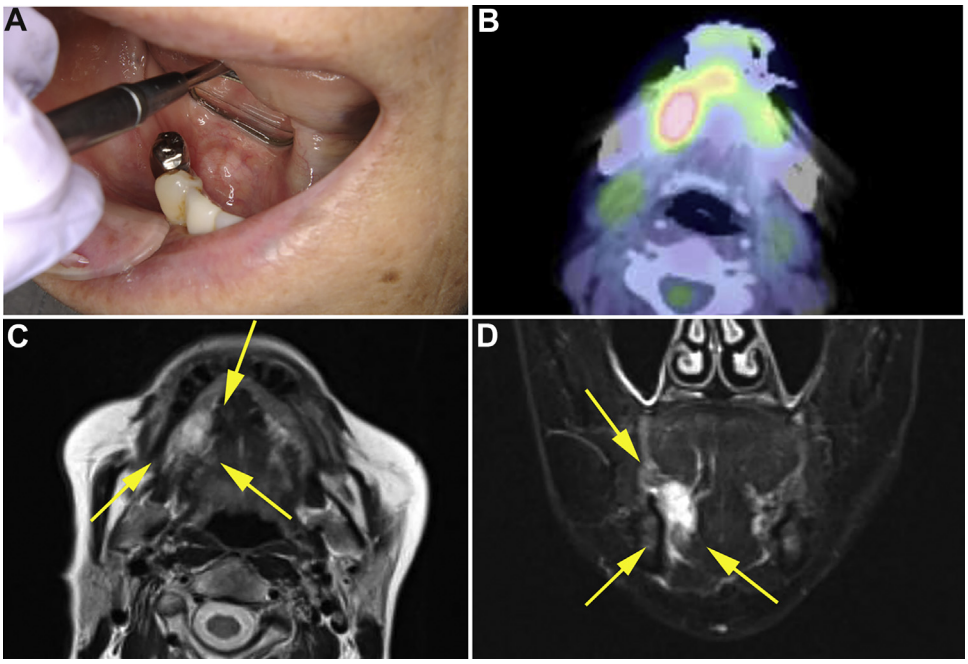

Fig. 1. Intraoral photography and imaging examination findings. (A) The tumorous mass on the right side of the oral floor. (B) Positron emission tomography-computed tomography showing an irregular mass with an SUVmax of 7.6. (C,D) Magnetic resonance imaging showing an indistinct marginal mass with high intensity on T2-weighted imaging (*yellow arrows*).

2. Case report

2.1. Clinical findings

A 77-year-old Japanese woman noted painless swelling of the right floor of the mouth from December 2012. On a physical examination, we noted a non-ulcer diffuse swelling in right oral floor (Fig. 1A). On palpation, a hard, elastic, thumb-sized mass was recognized. No regional lymph node swelling was noted. Computed tomography (CT) showed an irregular mass measuring 30 mm × 20 mm in the right sublingual gland, and positron emission tomography-computed tomography (PET-CT) showed that the SUVmax was 7.6 (Fig. 1B). Magnetic resonance imaging (MRI) showed high intensity in the region on short TI inversion recovery (STIR) imaging, suggesting invasion of the mylohyoid muscle (Fig. 1C and D). There is no distant metastasis and double cancer by image findings. An incisional biopsy was performed, and the histological diagnosis was "mucinous adenocarcinoma". After the diagnosis, right sublingualectomy and selective submandibular neck dissection were performed (Fig. 2A; sublingualectomy specimen). No lymph node metastases were found histologically. However, diffuse carcinoma cell invasion was observed around the submandibular gland. Fifteen months later, malaise and renal impairment was admitted and CT scan recognized peritoneal metastasis. A whole-body examination revealed no other primary malignant disease or local recurrence. Fine-needle aspiration cytology from ascites was performed, and the cytological finding was carcinoma cell, which had signet-ring cell features, and diagnosis was peritoneal carcinomatosis of MAC. After two months, she developed cachexia, and died. Autopsy was not permitted.

2.2. Pathological findings

The gross specimen was a whitish or yellowish-white, circumscribed, 2.2 × 2.1 × 2.4-cm sized submucosal mass that was not encapsulated (Fig. 2B). Variably sized mucin lakes were loculated by fibrous septa and accounted for more than 50% of the total tumor area (Fig. 3A and B). Within the mucin pools, detached tumor cells floated individually and in small clumps, either solid or clustering around microlumina. Some individual cells manifested a signet ring cytomorphology but never showed dominance. The tumor cells were arranged in solid clusters and tended to form secondary

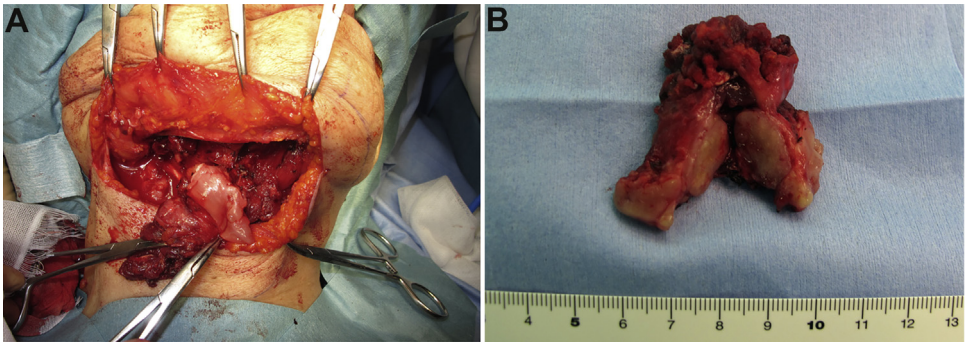

Fig. 2. Photograph of sublingualectomy and surgical specimen. (A) Right sublingualectomy and selective submandibular neck dissection were performed. (B) Macroscopically, the tumor was a whitish or yellowish-white, circumscribed, submucosal mass but not encapsulated.