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Case Report
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A Rare Multiple Salivary Gland Neoplasm: Mucoepidermoid Carcinoma of the Hard Palate and Adenoid Cystic Carcinoma of the Submandibular Gland: A Case Report and Literature Review

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Abstract

Introduction: Most tumors of salivary glands appear as a solitary mass in a single gland. It's extremely rare to have a case where tumors with different characteristics develop and coexist.

Case Report: Our patient is a 38-year-old female, a known case of low-grade Mucoepidermoid Carcinoma (MEC) of the hard palate. The tumor was resected in 2019. After three years, she complained of left-sided submandibular swelling for the past 7 years. On January 31st, 2023 excision of the mass was performed, and the final surgical pathology turned out to be Adenoid Cystic Carcinoma (ACC).

Conclusion: Despite the rareness of metachronous occurrence of salivary gland cancer with different anatomic locations and histopathology, it is still encountered in medical practice. Here, we emphasize the importance of follow-up with periodic clinical and radiological examinations.

Keywords: Salivary gland tumors; Salivary gland neoplasms; Multiple malignant salivary gland neoplasms; Head and neck pathology

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Introduction

Salivary Gland Cancer (SGC) is a rare disease, accounting for 3% to 11% of all head and neck cancers and 0.2% of total body malignancies, with an annual incidence of 0.5 to 2 patients/100,000 people [1,2]. Salivary gland cancer has an equal gender distribution, and the majority of cases arise in the 6th decade [3]. The World Health Organization (WHO) recognized 33 different malignant salivary gland cancers in 2017 [4]. Tumors can develop in both major and minor salivary glands. The parotid glands account for 80% of major salivary gland tumors, whereas most minor salivary tumors present in the palate. It was reported that Adenoid Cystic Carcinoma (ACC) is the most common histology in submandibular and minor salivary glands followed in turn by Mucoepidermoid Carcinoma (MEC), Carcinoma Ex-Pleomorphic Adenoma (CExPA), and less commonly by acinic cell carcinoma, salivary duct carcinoma, and epi-myoepithelial carcinoma [1,5]. The cause of salivary gland cancer is unknown; however, it is thought to be multifactorial with many risk factors suggested and reported in the literature such as radiation, viruses (EBV and HIV), immunosuppression, smoking, alcohol intake, -UV light exposure, and genetics [6].

Although salivary gland tumors typically manifest as a single unilateral salivary gland mass, there are extremely rare cases in which tumors with different characteristics develop and coexist [7]. Approximately, salivary gland tumors with different histopathology of the ipsilateral side account for 0.3% of all salivary gland tumors [8]. Most cases of synchronous or metachronous masses of bilateral major salivary gland tumors described in the literature appeared in the parotid gland, and majority were Warthin tumors [8].

To our knowledge, there are a few cases in the literature that involve the presence of two primary malignant salivary gland neoplasms with different histologic types. We present a patient

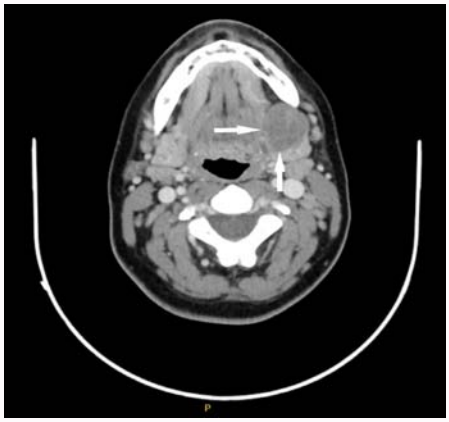

Figure 1: CT image of submandibular ACC tumor (indicated by white arrows).

with two primary metachronous tumors consisting of adenoid cystic carcinoma of left submandibular gland and mucoepidermoid carcinoma of the hard palate in a young adult patient with relevant radiologic and histopathologic findings. English-language literature on SGC is reviewed. Cases representing the involvement of different salivary glands with different histopathology were included in this study.

Case Presentation

We report a 38-year-old female, a known case of low grade mucoepidermoid carcinoma of the hard palate status post excision in 2019, she first presented to a local hospital in 2019 as a case of an asymptomatic patient with detected bluish area on routine oral exam at the junction of the hard and soft palate. She did not have any family history of cancer or a history of radiation exposure. Physical examination showed 3 cm × 4 cm bluish swelling at the junction of the hard and soft palate with no pulsation, and clear neck exam. CT with contrast showed limited lesion to the hard palate and soft palate junction with no extension to the bony structure. Soon after that, a wide excision of the lesion was performed with a local flap to close the defect. Histopathological investigation showed a low-grade mucoepidermoid carcinoma. In January 2023, the patient presented to our facility with left-sided submandibular swelling for the past 7 years, it was non-painful, not increasing in size, with no obvious skin changes. She did not complain of any otorhinolaryngological symptoms, and no significant weight loss was reported by the patient. Physical examination revealed a soft left-sided neck swelling, with no enlarged lymph node and a clear oral examination. Cranial nerves were intact bilaterally, and endoscopic examination of the nose revealed a clear nasopharynx. She underwent a Computerized Tomography (CT) scan and a Fine Needle Aspiration (FNA). The CT scan revealed a well-defined hypodense mass measuring 24 mm × 27 mm × 23 mm occupying most of the left submandibular salivary gland with no evidence of focal enhancement, septations or calcifications. No lymph node involvement was noted (Figure 1). An FNA was thus warranted. FNA revealed a salivary gland neoplasm consistent with pleomorphic adenoma, Milan System Category IV-A. The patient was planned to undergo surgical excision of the left submandibular gland. The mass was excised completely with no postoperative complications. The final surgical pathology report of the mass revealed an Adenoid Cystic Carcinoma (ACC) with tubular and cribriform patterns, with negative margins and no evidence of lymphovascular or perineural invasion. Immunohistochemistry was positive for SMA CAM5.2, cytokeratin AE1/AE3, and focally for CD117, and was negative for S-100. The pathological staging was thus pT2 Nx. Later on, the case was discussed in our facility tumor board regarding the best treatment plan for the patient and the decision was to follow with oncology for radiotherapy sessions.

Discussion

Multiple salivary gland tumors are seen on rare occasions; most cases have tumors with the same histological type. The coexistence of tumors of different histological types in the salivary gland is extremely rare [9,10]. Overall, MEC is the most common SGC especially for parotid glands, followed by ACC, which accounts for almost one quarter of cases [11]. It is more common in females, affects all age groups and often arises from the minor salivary glands [11]. Most malignant salivary gland tumors involve minor salivary glands mainly in the palate followed by the parotid gland, submandibular gland, and lingual gland [12]. The definition of a second primary tumor was first described and defined in 1932 by Warren and Gates, with the criteria being. 1. Each tumor must be histopathological confirmed and confirmed to be malignant; 2. Each must be geographically separated and distinct, and the lesions should be separated by normal mucosa; 3. Probability of one being the metastasis of the other must be excluded [13].

A study by Schwartz et al. [14], assessed the incidence of synchronous and metachronous head and neck malignancies in patients with confirmed malignancy, in a review of 851 patients most had multiple risk factors for head and neck malignancies such as smoking and alcohol intake. Their results revealed that the probability of developing a metachronous malignancy in the 5-year follow-up period was around 22%. Factors that significantly affected the risk of metachronous malignancy were the primary site of the first malignancy, as well as the age of the affected patient. As the development of a second malignancy leads to a worse prognosis of such patients a well-tailored screening program should be implemented in such patients, as well as assessment for the need for chemoprevention. A large-scale systematic review assessed around 450,000 patients from multiple studies to analyze the risk of second primary tumors in the head and neck region. The risk of metachronous malignancy was found to be 9.4% [15]. Another study assessed the risk of developing a Second Primary Tumor (SPT) in patients with a head and neck tumor, they found that the most common locations for SPTs was the oral cavity, pharynx, larynx, lungs, and esophagus [16].

A review of the English-language literature revealed a limited

Table 1: Reported cases of multiple malignant salivary gland tumor with different anatomic location and histopathology (present case included).

Author	Age (Y)	Gender	Location	Histologic type	Temporality
Hosni et al. [17]	56	Male	Parotid/SMG	MEC/ACC	Metachronous
Whitt et al. [7]	57	Male	Palate/Floor of Mouth	MEC/ACC	Metachronous
Argyris et al. [18]	91	Female	Upper Lip/Buccal Mucosa	PLGA/Myoepithelioma	Metachronous
Present Study	38	Female	Palate/SMG	MEC/ACC	Metachronous

number of multiple salivary gland tumor cases of different anatomic locations and histopathology. Table 1 shows the reported cases along with their demographic data (age and gender), tumor location, histologic subtypes, and chronologic occurrence of tumors [7,17,18]. Three cases were identified, 2 patients were men and 1 was woman, with a mean age of 68 years. Two cases represented metachronous malignant tumors histologically diagnosed as MEC and ACC. The third case represented a metachronous malignant tumor histologically diagnosed as Polymorphous Low-Grade Adenocarcinoma (PLGA) and Myoepithelioma. All cases had a metachronous pattern.

This paper reports an in-depth study of a rare case of multiple salivary gland neoplasm with different histopathology and anatomic locations. Concept of extremely rare tumors developing in two primary malignant tumors with different histopathology in two distinct anatomical locations may explain the case we encountered.

Conclusion

Despite how rare it's to encounter multiple salivary gland cancers of different histopathology, regular follow-up for these patients is mandatory. Our case and the English- language literature made it clear how crucial it is to perform a thorough head and neck pre surgery examination that takes into account the patient history, location of the tumor, risk factors, histological typing, and detailed evaluation of other salivary glands including the use of radiological exams to exclude the presence of second primary.

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