Primary mucinous adenocarcinoma of the parotid gland: A case report

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ABSTRACT
Introduction: Primary colloid carcinoma or mucinous adenocarcinoma (MAC) of the salivary glands, is an extremely rare neoplasm. Only six cases of major salivary glands origin have been occurred. Case Report: A case of MAC of parotid gland in a 55-year-old male and its diagnostic approach is given here. Conclusion: MAC is a high grade malignancy with a significant risk of recurrence and metastasis that need special treatment considerations.

Keywords: Mucinous adenocarcinoma, Colloid carcinoma, Parotid, Cytokeratin 7 (CK7)

INTRODUCTION
Primary colloid carcinoma or mucinous adenocarcinoma of the salivary glands, is an extremely rare neoplasm. The WHO defines mucinous adenocarcinoma as a malignant tumor composed of epithelial clusters with large pools of extracellular mucin [1]. Only 21 cases with major or minor salivary gland involvement have been reported to date [2, 3]. Only three cases in the parotid gland, two in the submandibular gland and one in the sublingual gland have been occurred [1, 4]. Mucinous adenocarcinoma of minor salivary glands belongs to a high-grade category with a significant risk of local recurrence, lymph node metastasis and fatal outcome.

We described the clinicopathologic findings of a MAC case arising in parotid gland. Our goal is to comprehend the profile of MAC in the salivary glands and the approaches leading to definite diagnosis.

CASE REPORT
A 55-year-old man was admitted to our clinic with complaint of a painless swelling in his left side of the face, parotid area, for 18 months. The patient’s medical history was unremarkable. A complete physical examination revealed no obvious systemic problems.

The preliminary diagnosis was tumor of salivary glands. The mass was excised surgically beneath the parotid capsule and on gross examination, the specimen measured 5×3×1.5 cm and appeared to be multilobular with areas of gelatinous consistency. Samples of the tumor were embedded in paraffin. Sections were cut and stained with hematoxylin and eosin.

Microscopically, the tumor was composed of circumscribed large pools of extracellular mucin in direct contact with the stroma. The mucin lakes surround small cluster and nests of malignant epithelial cells with bland to pleomorph and hyperchrome nuclei. The nests were solid or formed secondary lumens. A few cells contained small amounts of intracellular mucin, but signet-ring cells were not observed. Mitotic figures
were sparse (Figure 1). Colloid component was more than 50% and no cystic spaces was seen.

Periodic acid-Schiff (PAS) and mucicarmin staining confirmed its mucinous origin (Figure 2). Small sections of serous salivary gland tissue was observed in close relationship to tumor.

Complete clinical work-up and whole body scan was done which showed no trace of metastasis. Immunohistochemical staining for cytokeratin 7 (CK7) and 20 revealed only CK7 positivity which ruled out the metastasis of other malignancies and confirmed the salivary gland origin of the lesion (Figure 3A–B).

These findings identified the lesion as a primary mucinous adenocarcinoma of the parotid gland. Postoperative radiation administered to prevent future recurrence and regional metastasis.

**DISCUSSION**

The real frequency of MAC remains unknown; it comprises less than 0.1% of epithelial salivary gland tumors in Armed Forces Institute of Pathology (AFIP) files [5]. In Chinese population, MAC represented 0.1% of all salivary gland tumors and 0.4% of carcinomas. Its incidence was also estimated to be 0.07% of malignant salivary gland tumors in West China [6]. Recent study of minor salivary gland tumors from the United States indicated that MAC accounted for 0.2% of all tumors and 0.4% of malignancies [5]. MAC is more common in the intraoral minor salivary glands, with an approximately 2:1 predilection for the minor over major glands. It occurred primarily in the palate, followed by cheek, floor of the mouth and base of the tongue. MAC can also arise in the stomach, lower gastrointestinal tract and lacrimal glands [4].

Patient ages have ranged from 42 to 86 years (mean 66.6 years), and the tumor was slightly more common in males, with a male to female ratio of 10:8. The most frequent presenting symptom was a painless mass of 4 weeks to 11 years duration. The histopathologic feature of this tumor is nearly identical to mucinous eccrine carcinoma of the skin, mucinous carcinoma of the breast and colloid carcinoma of the intestine [2, 5]. Alternatively, definitive diagnosis of MAC as a primary salivary gland tumor is achieved by exclusion of metastatic diseases [5, 7]. Distinction between primary MAC from metastatic tumor is impossible, based on histologic features alone. The immunotyping of CK7/CK20 can aid to determine the tumor origin beside careful clinical history and evaluation. CK7(+) /CK20(-) phenotype may show a salivary primary tumor, whereas CK7(-)/CK20(+) profile may be a clue to an intestinal origin [8].

Most literature on mucinous adenocarcinoma of the salivary glands is in the form of single case reports [3]. The differential diagnostic consideration consist of mucinous cystadenocarcinoma (MCAC), mucoepidermoid carcinoma (MEC), mucin-rich salivary duct carcinoma (SDC), signet–ring cell adenocarcinoma, metastatic tumors and mucin extravasation phenomenon (MEP) [4]. MCAC has prominent cystic spaces with malignant epithelial lining. An MEC has typical areas of squamous, intermediate and mucin–secreting cells. An MEP shows inflammatory
changes and fibrosis with no evidence of neoplastic epithelium. Quantities of mucinous component in others are not as extensive as in MAC [4].

As mucinous adenocarcinoma of the salivary glands is a rare entity, it is difficult to compare the behavior of them to a similar tumor in a different location. Tumors of the breast and stomach are associated with a better prognosis, whereas those of the colorectum are worse.

The prognosis of salivary gland tumors varies with location, histological type and grade. The clinical stage of the disease is considered the most prognostic factor [9].

Gneppe stated that primary colloid carcinoma of the salivary glands is an aggressive tumor with a significant risk of recurrence and metastasis therefore should be considered as a high-grade carcinoma which needs surgical excision with free margins, cervical lymph node dissection and adjuvant radiotherapy depending on tumor stage [4].

CONCLUSION

Mucinous adenocarcinoma of the salivary glands is a rare entity, it is a high grade malignancy with a significant risk of recurrence and metastasis that need special treatment considerations. So one should make distinction between primary and metastatic tumors by immunotyping.

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Author Contributions
Ali Dehghani Nazhvani – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Reza Tabrizi – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Sara Amanpour – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES