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Case Report

Parotid secretory carcinoma: a rare entity in a young patient

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ABSTRACT

Secretory carcinoma (SC) of the parotid gland is a rare malignancy of the head and neck that presents almost equally in both genders with the average age of presentation in the mid-40. SC is often misdiagnosed as salivary acinic cell carcinoma (AciCC) as they share similar pathological morphology which is difficult to differentiate based on staining alone. We present a case of a 22-year-old female with this uncommon pathology who underwent subtotal parotidectomy after a conflicting provisional diagnosis of a benign tumor based on cytology and CT scan reporting possible malignancy involving the deep parotid lobe. It was later confirmed as SC on histopathology examination.

Keywords: Parotid SC, Mammary analogue SC, Salivary gland tumors

INTRODUCTION

Secretory carcinoma in the head and neck region develops in individuals in their 40s, which is a relatively early age of onset compared to AciCC.[1,6] SC reported involves the parotid gland in about 70% of cases, with submandibular gland in about 7% of cases and less common in the soft palate, buccal mucosa, base of the tongue, and lip.[2,3] SC is a rare salivary gland tumor that share histological and genetic features with the secretory carcinoma of the breast.[1-4] SC shows a characteristic t (12;15) (p13; q25) chromosomal translocation on gene analysis that has not been identified in any other salivary gland tumor, which results in ETV6-NTRK3 gene fusion, similar to secretory carcinomas of the breast.[1,4,5]

CASE REPORT

A 22 years old female presented with a right parotid swelling for 2 years which progressively increased in size. She was asymptomatic and the swelling did not affect her daily activities. There was no pain associated with jaw movement or during chewing of food. The mass was situated at the angle of the mandible and measured 1×2 cm. It was a well-defined firm mass, was mobile with normal overlying skin. There was no facial nerve weakness and no palpable cervical lymphadenopathy.

FNAC was reported as cystic content containing of occasional foamy macrophages with no epithelial cell or atypical cells seen. Contrast-enhanced computed tomography of the neck demonstrated a well-defined hypodense lesion with peripheral wall enhancement that is seen arising from the superficial lobe of right parotid gland extending into the deep lobe of parotid measuring approximately 2.4×2.8×2 cm with attenuation value ranging from 3-20 HU with presences of thick enhancing internal septation within. The differential diagnosis includes Warthin tumor, parotid lymphoepithelial cyst or cystic teratoma.

In view of the CT scan findings of possible malignancy, a right subtotal parotidectomy was performed, and she had an uneventful post-operative period with intact facial nerve functions. However, the histopathological examination (HPE) of the tumour was reported as SC with clear surgical margins.

Post operatively, patient did not undergo adjuvant radiotherapy after discussion with the oncology team. She underwent a positron emission tomography (PET) scan 3

months after the surgery, which demonstrated no focal enhancing lesion or avidity at the areas of the parotid beds that may suggest recurrence or residual. Patient has been managed conservatively with annual PET scan for the past 2 years and showed no signs of recurrence to date.

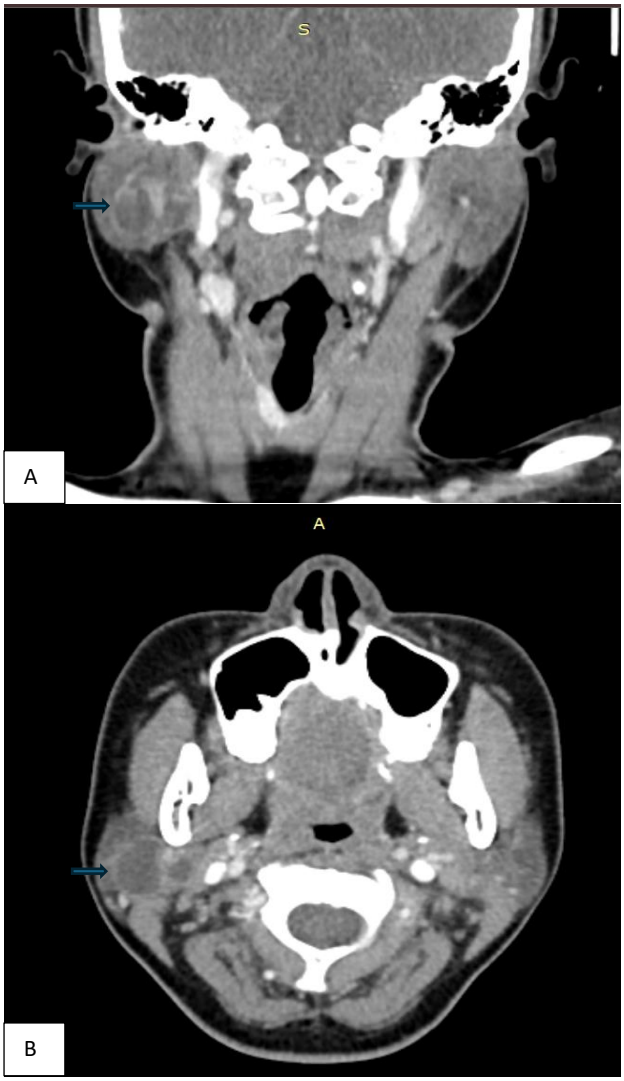

Figure 1 (A and B): An ill-defined lobulated lesion (blue arrow) in the superficial lobe extending to deep lobe of the right parotid in the coronal and axial views.

DISCUSSION

SC is a new entity in head and neck cancer as reported by Skalova et al based on the identification of the t (12;15) (p13; q25) chromosomal translocation which differentiates it from AciCC. It shares the histological, immunohistochemical, and genetic characteristics found in breast SC, an extremely rare neoplasm that usually affects young patients.[1-3,5] As opposed to AciCC, SC has a slightly higher prevalence in men than in women although some study reported no gender predilections.[4,5] Majority of SC cases involve the parotid gland (70%), followed by submandibular glands (7%) and less commonly in the oropharynx.[2,3]

Most of the patients will present with an indolent, painless parotid, neck, or oral cavity mass[4,5] but some cases were reported exhibit rapid growth over months.[5]

Various imaging modalities have been used for further investigations such as ultrasound (US), CT and MRI. SC demonstrates hypoechoic feature on US and appears hyperintense on the T1 phase of MRI.[1]

Microscopically, SC was found to form papillary groups on cytology, with abundant, prominent or multivacuolated cytoplasm.[2,5] Both AciCC and SC have similar cellular arrangement on cytology smears. However, SC tends to have increased extracellular and intracellular mucin compared to AciCC, and a greater variation in the size of the cytoplasmic vacuoles found.[2,5]

Histologically, SC consistently shows positivity for mammaglobin, S-100 protein, and vimentin.[1-5] It also tends to variably express pancytokeratin, CK7, CK8, EMA, STAT5a, and GCDFP15[3] while negativity for androgen receptor (AR), BRST-2, CK-20, P63, and SMA.

SC distinguishes itself by the absence of zymogen vacuoles, a distinctive feature of AciCC[-3], and the presence of strong S-100 protein positivity, which is lacking in AciCC but there is no single immunohistochemical marker that can diagnose SC alone.[1,3,5]

Hence the gold standard to diagnose SC is by fluorescence in situ hybridization identification (FISH) since this translocation has not been found in any other salivary gland carcinoma.[1,5,6]

SC showed higher rate for recurrence and disseminated disease, compared to AciCC which is evidenced by increased positive lymph nodes on dissection and a higher recurrence rate.[1,6]

Treatment of SC is either a superficial parotidectomy or a total parotidectomy, depending on the tumor location for localized and less aggressive cases, while more invasive or metastatic may need adjunctive neck dissection, radiation, or chemotherapy.[4] To date, there is no standardized treatment protocol due to the rarity of the disease, although surgical excision was uniformly performed in nearly all reported cases. In our case, subtotal parotidectomy was perfomed and no radiation or chemotherapy were given.

CONCLUSION

SC is a rare malignancy of salivary gland tumor that distinguishes itself from AciCC while closely resembling the histology and genetics of breast secretory carcinoma.

Despite exhibiting an indolent course in most patients, certain cases show a tendency for distant metastasis and mortality. While the pathology literature provides increasing data on this disease, it is still not adequately presented in the otolaryngology community as to date there is no standardized treatment protocol for this disease. More research is necessary to understand the clinical patterns and prognosis of SC.

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REFERENCES

1. Sun L, Thorson T, Zhu R, Huo J, Tong J, Rodgers WH, et al. A case report of parotid mammary analogue secretory carcinoma and reviews. Int J Surg Case Rep. 2019;55:88-91.
2. Sethi R, Kozin E, Remenschneider A, Meier J, VanderLaan P, Faquin W, et al. Mammary analogue secretory carcinoma: Update on a new diagnosis of salivary gland malignancy. The Laryngoscope. 2013;124(1):188-95.
3. Suzuki K, Yagi M, Kanda A, Kobayashi Y, Konishi M, Miyasaka C, et al. Mammary analogue secretory carcinoma presenting as a cervicallymph node metastasis of unknown primary site: a case report. Case Rep Oncol. 2017;10(1):92-198.
4. Montalvo N, Galarza D, Redrobán L. Secretory Carcinoma of the Parotid: Making the Correct Diagnosis of a Rare Salivary Gland Carcinoma When Molecular Biology Testing Is Not Available. Case Rep Pathol. 2019;2019:5103496.
5. Forner D, Bullock M, Manders D, Timothy W, Christopher JC, Liane BJ, et al. Secretory carcinoma: the eastern Canadian experience and literature review. J Otolaryngol Head Neck Surg. 2018;47(1):69.
6. Ogawa M, Yokoo S, Yamaguchi T, Suzuki K, Seki-Soda M, Shimizu T, et al. Diagnosis and treatment of secretory carcinoma arising from the oral minor salivary gland: Two case reports. Medicine (Baltimore). 2021;100(51):e28390.

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