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CASE REPORT

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Parapharyngeal Acinic Cell Carcinoma: A Case Report of a Rare Extra-Parotid Occurrence

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Abstract

Parapharyngeal space (PPS) tumors are among the most challenging tumors of the head and neck region to diagnose and treat. Acinic cell carcinoma (ACC) is an extremely rare extra-parotid malignant neoplasm of the PPS. Herewith, we report a case of extra-parotid, low-grade solid pattern ACC diagnosed in a 55-year-old male who presented with a left parapharyngeal bulge. Despite its rare occurrence, ACC of the PPS should be included in the differential diagnosis of PPS tumors.(**International Journal of Biomedicine. 2023;13(3):162-164.**)

Keywords: pharapharyngeal space • acinic cell carcinoma • extra-parotid

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Introduction

Acinic cell carcinoma (ACC) is a malignant neoplasm of the salivary glands, most commonly occurring in the parotid gland. ACC of the minor salivary glands is rare, constituting between 3% and 12% of salivary gland ACCs, with the majority presenting in the minor salivary glands of the palate.(1) ACC is slightly more common in women in their 50s and 60s than in other groups in the population. Previous radiation exposure and familial predisposition are among the most common risk factors for ACC.(1) ACC of the PPS separate from the parotid gland is extremely rare.

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Case Presentation

A 55-year-old male patient presented with a complaint of a left peritonsillar mass. He had no risky habits, and his family history was irrelevant. On clinical examination, a left parapharyngeal bulge was present. Neck and nasal examinations were unremarkable. On laryngoscopy examination, the larynx was unremarkable as well. Computed tomography (CT) revealed a regular, well-defined mass with a slightly heterogeneous enhancement on the left post-styloid PPS (Figure 1A). The transcervical approach was chosen for surgical excision. Tumor excision revealed a well-encapsulated, soft, rubbery, grayish-white, homogenously solid mass measuring 6×4.5×2.3cm. No invasion of the surrounding structures was seen. Representative sections were taken for histological examination. On histological examination, hematoxylin and eosin-stained slides revealed

a low-grade malignant epithelial tumor composed of well-differentiated basophilic acinar cells with granulated to vacuolar cytoplasm and eccentric nucleus (Figure 1B). There was no vascular or perineural invasion. Surgical margins were free of the tumor. The final histological diagnosis was low-grade ACC solid type.

Discussion

PPS tumors have a nonspecific clinical presentation. They can present as a mass or swelling in the neck or throat, with difficulties in swallowing, breathing, changes in voice or speech, pain in the ear or jaw, or facial weakness.[2] Imaging studies such as CT scans and MRI are used for initial diagnosis; nevertheless, a biopsy followed by surgical resection is necessary to confirm the diagnosis.[2] PPS tumors constitute approximately 0.5% to 1.5% of all head and neck tumors.[3,4] The most common tumors of the PPS are benign tumors such as pleomorphic adenomas of the salivary gland, followed by paragangliomas and neurogenic tumors.[5]

Furthermore, salivary gland neoplasms are the most frequently found primary malignant tumors.[5,6] Salivary neoplasms in PPS may arise from the deep lobe of the parotid gland, ectopic salivary rests, or minor salivary glands of the lateral pharyngeal wall.[5-7] ACC is a rare type of salivary gland tumor in the PPS.[6] In imaging studies, anteromedially parapharyngeal fat displacement by the tumor is a feature that favors extra-parotid tumor origin in PPS.[8]

WHO defines ACC as a malignant epithelial neoplasm of the salivary glands in which at least some neoplastic cells demonstrate serous acinar cell differentiation characterized by cytoplasmic zymogen secretory granules.[9] ACC grows slowly over time, and it usually presents clinically only when it becomes large enough to be detected. ACC is a low-grade neoplasm that was initially considered as a benign tumor entity. It has four histological growth patterns, such as solid, multicystic, papillary-cystic, and follicular.[9] The most frequent histological patterns are solid, composed of well-differentiated polygonal acinar cells with well-defined cytoplasmic borders, and microcystic with prominent cellular vacuolization and intercellular cystic change. Whereas the rarest patterns are the papillary, composed of papilla covered by hobnailed cells, intercalated duct-like cells, and cells with eosinophilic cytoplasm with indistinct cell borders, and the follicular pattern comprised of closely packed cystic spaces lined by flattened epithelium and filled with eosinophilic colloid-like material reminiscent of thyroid follicles.[9]A mixture of patterns is frequent.

ACC of the PPS separate from the parotid gland is extremely rare. In our thorough investigation, when excluding deep parotid lobe ACCs, we encountered only seven reported cases of ACC in the PPS separate from the parotid gland reported in the literature.[6,10-14]

Our case was a solid pattern ACC, presenting in a 55-year-old male. We encountered no invasion of the surrounding structures, as well as no intravascular or perineural invasion. The tumor, in our case, was separate from the parotid gland, indicating that it could be arising from the minor salivary glands of the PPS. ACC on histology comes into close differential diagnosis with other salivary and head and neck masses, such as salivary metastasis of thyroid carcinoma, which are thyroglobulin positive, salivary oncocytoma, composed of eosinophilic non–serous cells, mucoepidermoid carcinoma positive for p63 and mammary analog secretory carcinoma that lacks PAS-positive secretory granules and is vimentin and adipophilin positive.[15,16] In addition to the characteristic morphological presentation, in our case, we also report no positivity for thyroglobulin, p63, vimentin, and adipophilin. Normal salivary gland is also a potential differential diagnosis with ACC.

Nevertheless, our case lacked its normal lobular architecture. Of the reported cases in the literature, all but one case of ACC,[13] separate from the parotid in PPS, were low-grade tumors with no recurrence and metastasis. We also report a low-grade extra- parotid ACC in PPS.

Nevertheless, ACCs of salivary glands have an unpredictable course. Local recurrence has been reported, as have occasional, distant metastasis to the lungs and the bones via hematogenous spread and metastasis to regional lymph nodes. The reported mortality of ACC at 5 years is less than 10%. ACCs of the PPS, because of their rarity and clinical presentation, are challenging to be diagnosed and treat.[17]

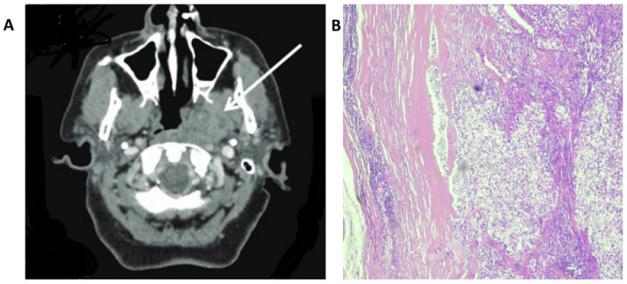

Figure 1. *A. Computed Tomography (CT) presents a regular, well-defined mass with a slightly heterogeneous enhancement on the left post-styloid PPS. B. Histological section of hematoxylin and eosin stained low-grade malignant epithelial tumor, composed of well-differentiated basophilic acinar cells with granulated to vacuolar cytoplasm and eccentric nucleus (X 100).*

Treatment consists of surgical removal of the tumor, which may require complex surgical techniques to preserve important structures, depending on the location of the tumor, including the carotid artery, jugular vein, and cranial nerves. Radiation therapy and chemotherapy may also be used in some cases, alone or in combination with surgery.[7,8] Our case was treated only with surgical resection, as we encountered no invasion beyond the tumor capsule, and the histological features were of no high grade. Moreover, there was no vascular or perineural invasion. However, there are reports of low-grade ACC behaving aggressively; therefore, we opted for treatment with radiation therapy and chemotherapy despite the low-grade morphology.[6,10-13]

In conclusion, extra-parotid ACC of PPS is an extremely rare tumor that should be considered in the differential diagnosis of PPS tumors. Surgical excision ACC is curative

in low-grade tumors with no invasion of the surrounding structures. However, its surgical management is challenging because of the location and relationship with nearby structures, and therefore radiation therapy and chemotherapy are often adjunct treatments to surgical resection.

Competing Interests

The authors declare that they have no competing interests.

References

1. Babu SS, Sunil S, Prathap A, Mathew AL. Acinic cell carcinoma of the posterior buccal mucosa: A rare case report. J Cancer Res Ther. 2020 Apr-Jun;16(3):675-679. doi: 10.4103/jcrt.JCRT_399_18.
2. Jiang C, Wang W, Chen S, Liu Y. Management of Parapharyngeal Space Tumors: Clinical Experience with a Large Sample and Review of the Literature. Curr Oncol. 2023 Jan 11;30(1):1020-1031. doi: 10.3390/curroncol30010078.
3. Galli J, Rolesi R, Gallus R, Seccia A, Pedicelli A, Bussu F, Scarano E. Parapharyngeal Space Tumors: Our Experience. J Pers Med. 2023 Feb 2;13(2):283. doi: 10.3390/jpm13020283.
4. Shahab R, Heliwell T, Jones AS. How we do it: a series of 114 primary pharyngeal space neoplasms. Clin Otolaryngol. 2005 Aug;30(4):364-7. doi: 10.1111/j.1365-2273.2005.00993.x.
5. Riffat F, Dwivedi RC, Palme C, Fish B, Jani P. A systematic review of 1143 parapharyngeal space tumors reported over 20 years. Oral Oncol. 2014 May;50(5):421-30. doi: 10.1016/j.oraloncology.2014.02.007.
6. van Hees T, van Weert S, Witte B, René Leemans C. Tumors of the parapharyngeal space: the VU University Medical Center experience over a 20-year period. Eur Arch Otorhinolaryngol. 2018 Apr;275(4):967-972. doi: 10.1007/s00405-018-4891-x.
7. Hughes KV 3rd, Olsen KD, McCaffrey TV. Parapharyngeal space neoplasms. Head Neck. 1995 Mar-Apr;17(2):124-30. doi: 10.1002/hed.2880170209.
8. Chen Z, Chen YL, Yu Q, Zhou SH, Bao YY, Shang DS, Ruan LX. Excision of tumors in the parapharyngeal space using an endoscopically assisted transoral approach: a case series and literature review. J Int Med Res. 2019 Mar;47(3):1103-1113. doi: 10.1177/0300060518816190.
9. El-Naggar AK, JKC C, Grandis JR, Takata T, Grandis J, Slootweg P (eds). WHO Classification of Head and Neck Tumours, 4th Edition, Volume 9. Lyon, IARC; 2017.
10. Yokoyama M, Nomura Y, Semba T. Acinic cell carcinoma of the parapharyngeal space: case report. Head Neck. 1993 Jan-Feb;15(1):67-9. doi: 10.1002/hed.2880150115.
11. Nguyen J, Palacios E, Horam E, Neitzschman H. Unusual parapharyngeal acinic cell carcinoma. Ear Nose Throat J. 2012 Jan;91(1):16-20. doi: 10.1177/014556131209100105. Erratum in: Ear Nose Throat J. 2012 Feb;91(2):53.
12. Lien KH, Young CK, Chin SC, Liao CT, Huang SF. Parapharyngeal space tumors: a serial case study. J Int Med Res. 2019 Aug;47(8):4004-4013. doi: 10.1177/0300060519862659.
13. Segovia VPK, Alonzo DM, Ong EAV. Parapharyngeal Acinic Cell Carcinoma: Initially Managed as a Case of Peritonsillar Abscess. Int J Head Neck Surg. 2022;13(3):88–9
14. Kumral TL, Uyar Y, Yildrim G, Berkiten G, Salturk Z. Parafarengeal Bölgede Ekstra-Parotidea Asinik Hücreli Karsinom. ENTcase. 2016;2(1):28-32
15. Tumors of the Salivary Glands, Atlas of Tumor Pathology: Third Series. Fascicle 17 G. L. Ellis and P. L. Auclair. Armed Forces Institute of Pathology, Washington D.C. ISBN: 1 881041 26 3 (Printed). 1996.
16. Kumar U. Acinic Cell Carcinoma Papillary-Cystic Variant: Diagnostic Pitfalls in Fine Needle Aspiration Cytology. J Clin Diagn Res. 2017 May;11(5):ED05-ED06. doi: 10.7860/JCDR/2017/21347.9772.
17. Cha W, Kim MS, Ahn JC, Cho SW, Sunwoo W, Song CM, Kwon TK, Sung MW, Kim KH. Clinical analysis of acinic cell carcinoma in parotid gland. Clin Exp Otorhinolaryngol. 2011 Dec;4(4):188-92. doi: 10.3342/ceo.2011.4.4.188.