Journal of
Medical Case Reports

CASE REPORT Open Access

Mucoepidermoid carcinoma of the base of the tongue: a case report and review of the literature

Peng Wang[1], Haiyan Liao[2], Feng Wang[1], Yufan Wang[1] and Hongyu Yang[1*]

Abstract

Background Mucoepidermoid carcinoma originates from reserve cells present in ducts of salivary glands and is the most common malignancy of the salivary glands. It is commonly found in the parotid gland, followed by the palatal and buccal mucous membranes. However, mucoepidermoid carcinoma occurrence in other intraoral sites, including the tongue base, is extremely rare.

Case presentation A 33-year-old Chinese man presented with a progressively enlarging mass at the base of his left tongue. Contrast-enhanced computed tomography and magnetic resonance imaging revealed an augmented soft tissue mass in the left jaw region with indistinct boundaries, enlargement of cervical lymph node of uncertain etiology, and no evidence of distant metastasis. A transoral needle biopsy from the mass pathologically revealed low-grade mucoepidermoid carcinoma. Complete transoral excision and cervical lymph node dissection were performed, followed by reconstruction with an anterolateral thigh free flap. Examination of the obtained surgical specimen confirmed low-grade mucoepidermoid carcinoma with *MAML2* gene fusion in the base of the tongue. The tumor was removed with negative margins, and the cervical lymph nodes were free of disease. The patient had an uneventful recovery and showed no evidence of recurrence or metastasis at 40 months of follow-up.

Conclusion We present a rare case of mucoepidermoid carcinoma at the base of the tongue. Furthermore, we review related literature and discuss its clinical features, histopathological characteristics, and treatment strategies.

Keywords Mucoepidermoid carcinoma, The base of tongue, *MAML2* rearrangement, Reconstruction

Introduction

Mucoepidermoid carcinoma (MEC) is one of the most frequently diagnosed malignant salivary gland tumors, accounting for 10–15% of all salivary tumors and usually affecting the parotid and minor salivary glands [1]. However, MEC originating in the base of the tongue is uncommon, with only 145 cases found in the SEER database from 2004 to 2016, as reported previously [2]. In addition, there are rare case reports on the treatment and follow-up of MEC at the base of the tongue. Histologically, MEC comprises a variable percentage of epidermoid squamous cells, mucous cells and intermediate cells. According to the proportion of these three types of cells and cellular differentiation degree, MEC is classified as low-, intermediate-, or high-grade. According to the World Health Organization (WHO) classification criteria, low-grade MEC usually forms large mucous-filled cysts, while higher-grade tumors show fewer mucinous cells and a more solid morphology [3]. Moreover, recent

*Correspondence:
Hongyu Yang
hyyang192@hotmail.com
[1] Department of Oral and Maxillofacial Surgery, Stomatological Center, Peking University Shenzhen Hospital, No. 1120, Lianhua Road, Futian District, Shenzhen 518036, China
[2] Department of Oncology, Shenzhen Xinhua Hospital, Shenzhen 518131, China

evidence has indicated that the specific *mastermind-like 2* (*MAML2)* rearrangement is present in more than 75% of MEC cases and may help to distinguish MEC from adenosquamous carcinoma [4, 5]. To our knowledge, *MAML2* gene fusion is thought to be associated with low-grade tumors [6]. Herein, we present an unusual case of low-grade MEC with *MAML2* gene fusion at the tongue base and discuss its clinical features, histopathological typing, and treatment modalities.

Case report

A 33-year-old Chinese man presented with an asymptomatic mass at the left-side tongue base for a 2-year duration, and it progressively enlarged for 2 months with an ulcer and bleeding. He was referred to our department in December 2020 complaining of pain, progressive dysphagia to solids, and blurry pronunciation. The patient had a history of using tobacco and betel nut for 7 and 15 years, respectively, but denied alcohol consumption. There was no family history of MEC or past medical history. Physical examination revealed the presence of an ulcerated, firm, exophytic, oval mass measuring approximately 2.0 × 2.5 cm on the dorsal surface of the tongue base(Fig. 1), mainly in the left posterior-lateral region, crossing the midline.

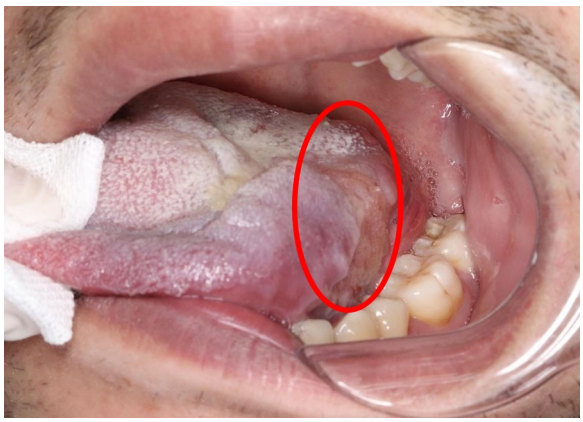

Fig. 1 Intraoral photograph showing an ill-defined mass on left lateral border of tongue, crossing the midline, covered with ulcerated lesion

Investigations

A computed tomography (CT) scan of head demonstrated a huge irregularly shaped mass that centered at the base of the left tongue, crossing the midline, infiltrating the muscle and involving the whole thickness of the tongue (Fig. 2A). Similarly, magnetic resonance imaging (MRI) revealed an heterogeneously enhancing, ill-circumscribed soft tissue mass, 4.5 × 3.0 × 3.1 cm in size, originating from the left side of the tongue base and involving the retropharyngeal space (Fig. 2B). Suspiciously, the MRI scan of the neck showed multiple enlarged bilateral cervical lymph nodes with unknown features. There was no distant metastasis found on a CT scan of the chest. Subsequently, the patient underwent needle biopsy on the mass at the base of the tongue, and the histopathological results were suggestive of low-grade MEC.

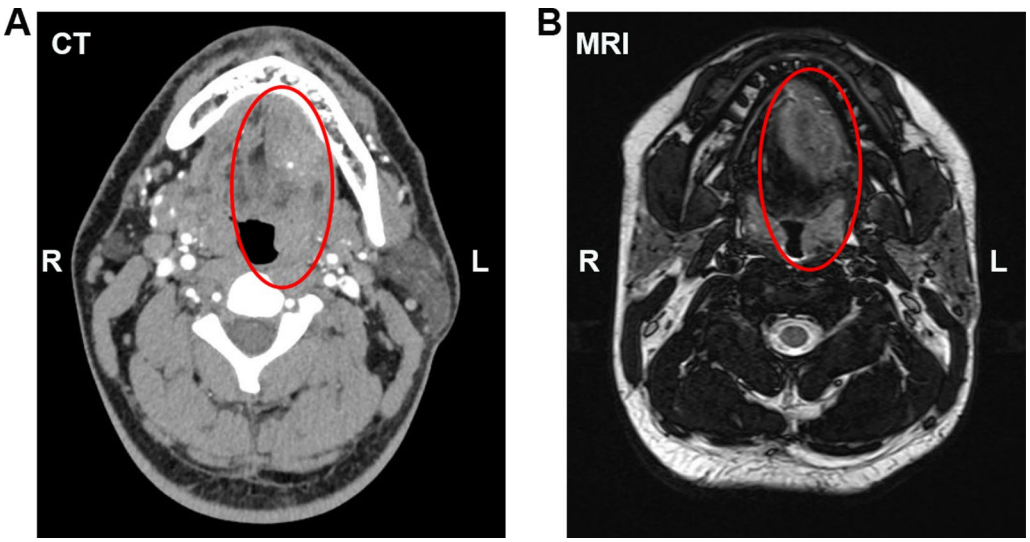

Fig. 2 Preoperative axial computed tomography (**A**) and MRI (**B**) scan showing a 4.5 × 3.0 × 3.1 cm soft tissue mass at the base of the left tongue that infiltrated the muscle and involved the whole thickness of the tongue

Treatments and follow-up

Complete transoral excision and bilateral neck dissection were performed under general anesthesia (Fig. 3A). For the resection, the patient's tongue was totally excised with a 20-mm safety margin around the tumor of the tongue base, along with the left-side posterior area of molar, lingual gingival, and oral floor tissues. The resection specimen was 7.5×7.0×3.5 cm in size (Fig. 3B). Left level I–IV and right level I–III cervical lymph node dissections were performed with sternocleidomastoid and spinal accessory nerves preserved, following by a immediate reconstruction using the left anterolateral thigh flap (Fig. 3C, D) for the defects of tongue and mouth floor.

The pathology of intraoperative frozen biopsy confirmed wide tumor-free surgical margins and negative cervical lymph nodes metastases. Additional microscopic examination identified that the tumor was of a mainly cystic dilatation structure with a large number of mucus cells (Fig. 4A). There was no evidence of neural or vascular involvement. Furthermore, the immunohistochemical staining revealed positive results for tumor protein 63 (P63) and cytokeratin 7 (CK7) (Fig. 4B, C, respectively). Periodic acid–Schiff (PAS) staining was positive in mucous cells of MECs (Fig. 4D). Fluorescent *in situ* hybridization (FISH) results were positive for *MAML2* gene rearrangement (Fig. 4E). On the basis of these findings, the definitive diagnosis was confirmed as low-grade MEC of the base of the tongue (T4aN0M0).

Outcome and follow-up

The patient had an uneventful operative course and postoperative recovery. He did not receive the adjuvant radiotherapy or chemotherapy owing to the low grade pathological features with low malignancy and negative metastatic cervical lymph nodes. At 9 months post operation, his healing in the reconstructed site was uneventful (Fig. 5A, B), and axial CT (Fig. 5C) and MRI (Fig. 5D) scans of the oral cavity did not indicate any evidence of recurrence or metastasis. To date, the patient has been followed up for a period of 40 months, and he is free of disease (Fig. 6A, B).

Discussion

MEC is a malignant, locally infiltrating tumor of the salivary gland, accounting for 10–15% of all salivary gland neoplasms and 30% of all salivary malignancies, and it is mainly found in the parotid gland [1]. Its occurrence in the base of the tongue is extremely rare, with less than 30 related cases reported when searching in the PubMed and CNKI databases (Table 1). In a retrospective study over a 50-year period, 145 MEC cases originating from the tongue base were found in the SEER database.

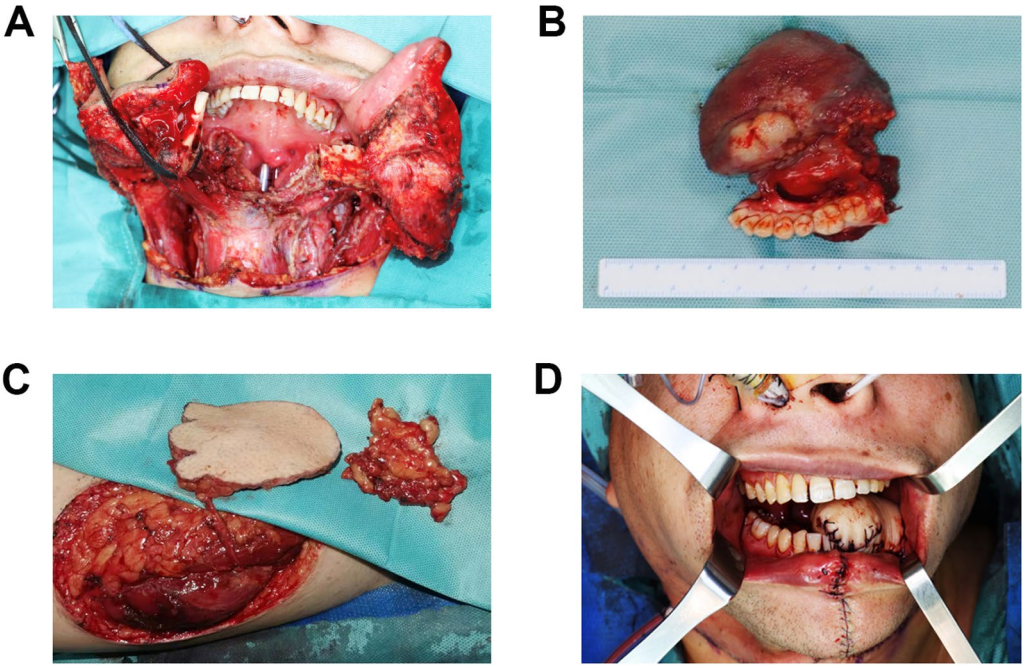

Fig. 3 Intraoperative photograph. **A** Intraoperative image of complete transoral excision and bilateral cervical lymph node dissection involving left levels I–IV and right levels I–III; **B** the close view of excised specimen; **C** a left anterolateral thigh free flap was harvested for defect reconstruction; **D** final intraoperative reconstructive appearance

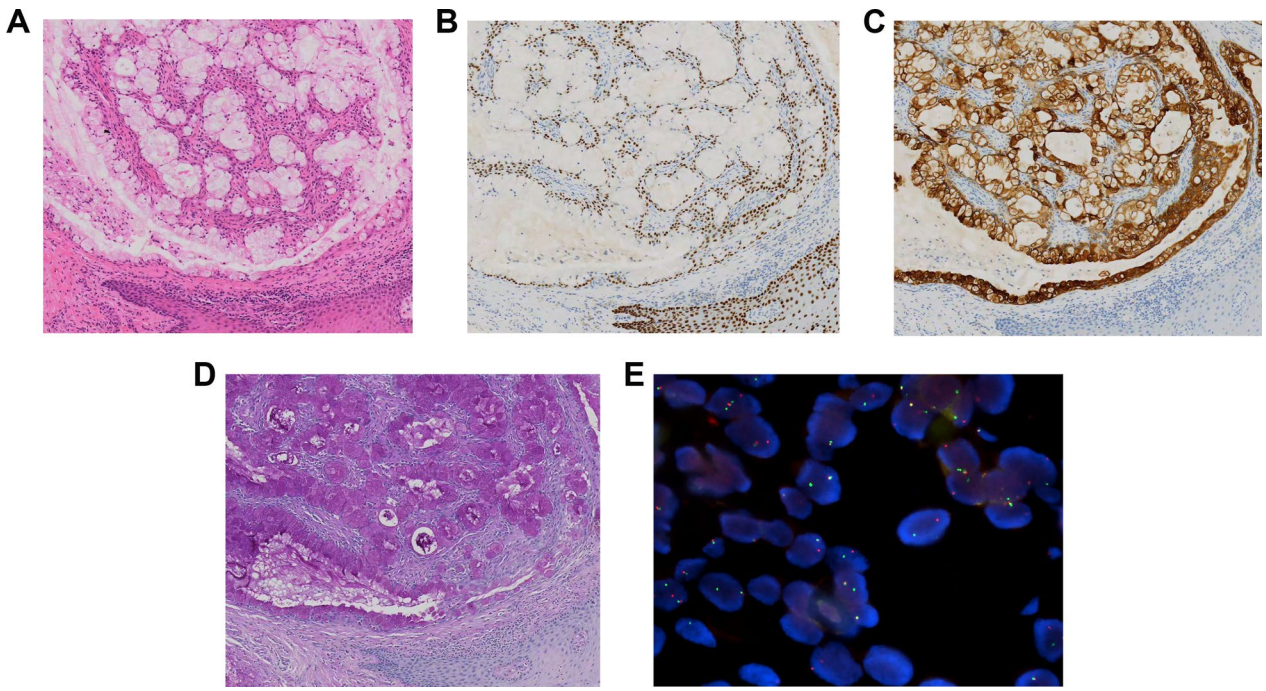

Fig. 4 Pathological and molecular analysis revealed a low-grade MEC in the base of the tongue. **A** Hematoxylin and eosin staining showed proliferation of the tumor specimen with cystic dilatation full of mucus cells (magnification, ×100); **B** immunohistochemistry demonstrating positivity for tumor protein 63 (magnification, ×100); **C** immunohistochemistry demonstrating positivity for cytokeratin 7 (magnification, ×100); **D** periodic acid–Schiff staining demonstrating positive mucous cells and the contents of microcystic spaces (magnification, ×100); **E** fluorescent *in situ* hybridization confirming the presence of *MAML2* rearrangement where the green probe and red probe are split

This study showed that MEC of tongue was more common in females and normally presented in patients over 50-years-old (approximately 75%) [2].

Typically, primary tumors of MEC at the base of the tongue present as a slowly enlarging mass. At the initial stage, the tumor may be present as a painless and fixed mass, while advanced tumors may be associated with pain with nerve or bone invasion [17]. Consistently, the patient in our case had a 2-year history of a mass on the base of the tongue with no significant enlargement or pain in the previous 1.5 years. However, with the progressive enlargement of the mass, the patient presented with pain and ulceration.

MEC is a malignant glandular epithelial neoplasm characterized by mucous, intermediate, and epidermoid cells [13], and can be diagnosed and classified as low-, intermediate-, or high-grade on the basis of histopathological evidence of the proportion of these cells [28]. Low-grade MEC is characterized by low malignancy, whereas high-grade MEC is more invasive and has a poor prognosis [29]. In addition to pathological examination, detection of the *MAML2* gene rearrangement by FISH also can be performed to assist MEC diagnosis, since *MECT1-MAML2* gene fusion is considered to be MEC-specific [5, 28].

Radical resection is the primary treatment for MEC. For high-grade MEC with nerve and lymphovascular invasion, tumor-free surgery should be ensured, and chemotherapy and/or radiotherapy may be used after operation [30]. The prognosis of MEC depends on the primary site, clinical stage, surgery, radiotherapy, and chemotherapy [2, 24]. The 5-year survival rates for low- and high-grade MECs are 70% and 50%, respectively [23]. Some low-grade MECs may also metastasize to the regional lymph nodes, resulting in recurrence and poor prognosis [31]. The present case was graded histologically as low, owing to the large number of mucus cells, high proportion of cystic constituents, and lack of evidence of neural and vascular involvement. The patient showed no evidence of recurrence 40 months after the operation. Although recurrence of MEC is uncommon, some recurred cases after primary excision have been reported [11], suggesting that MEC requires close long-term follow-up.

Local disease control and good survival rates can be achieved with surgery combined with radiotherapy and chemotherapy for tongue MEC [2, 32]. However, the clinical value of radiotherapy and chemotherapy for MEC is controversial. Most studies have reported that radiotherapy and chemotherapy alone cannot cure MEC

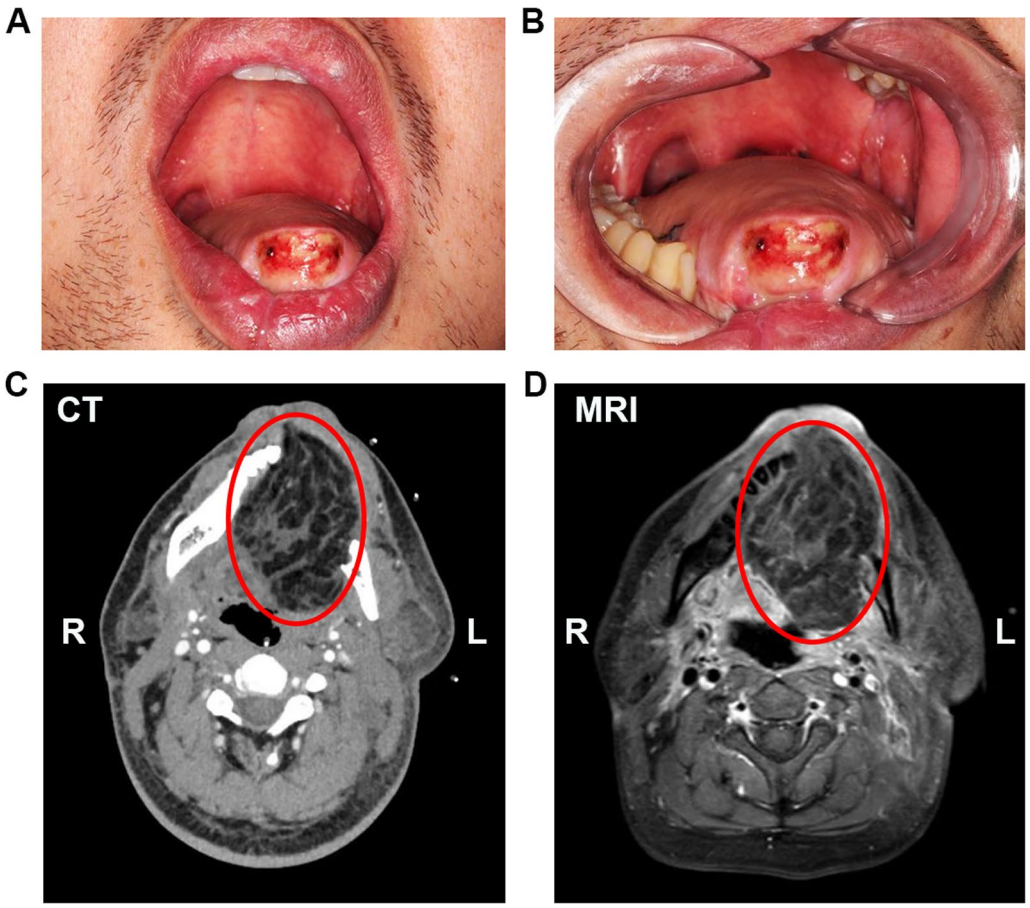

Fig. 5 Physical examination and imaging scans of the patient at 9-month follow-up showed no evidence of recurrence. **A**, **B** Postoperative photograph demonstrating well-healed reconstructed site; computed tomography (**C**) and magnetic resonance imaging (**D**) scan did not show any evidence of recurrence in 9 months postoperative

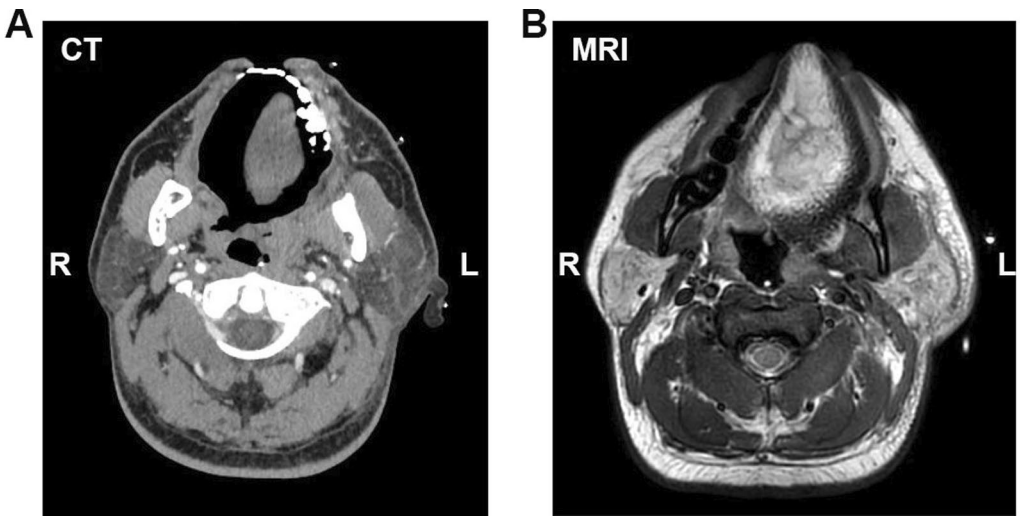

Fig. 6 Imaging scans of the patient at 40-month of follow up showed no evidence of recurrence. Computed tomography (**A**) and magnetic resonance imaging (**B**) scans did not show any evidence of recurrence at 40 months postoperative

Table 1 Summary of the case reports of mucoepidermoid carcinoma of base of the tongue

Years	Authors	Gender	Age	Tumor grade
1973	Heidelberger and Batsakis [7]	F	–	High grade
1975	Adkins and Putney [8]	M	–	Low/intermediate
1985	Pickell [9]	M	46	Low grade
1997	Pfendler [10]	F	59	Low grade
2000	Varghese *et al.*[11]	M	–	Low grade
2003	Pires *et al.*[12]	F	40	Low grade
2007	Andrews and Eveson [13]	M	38	High grade
2007	Leong *et al.* [14]	M	27	High grade
2009	Liu *et al.* [15]	F	82	High grade
2011	Sobani *et al.* [16]	M	71	Low grade
2013	Martellucci *et al.* [17]	F	69	Low grade
2014	Kalogirou *et al.* [18]	M	42	Low grade
2015	Bollig *et al.* [19]	F	40	Intermediate
2015	Mesolella *et al.* [20]	F	42	Low grade
2015	Vingerhoedt *et al.* [21]	M	46	Low grade
2016	Su *et al.* [22]	M	/	Low grade
2017	Rubin *et al.* [23]	F	33	Low grade
2017	Mathew *et al.* [24]	F	45	Low grade
2017	Naseem *et al.* [25]	M	66	High grade
2018	Zahran *et al.* [26]	F	40	Low grade
2018	Y Chen *et al.* [27]	3M; 2F	27–76	4 Intermediate; 1 high grade

F female, *M* male

completely; however, postoperative adjuvant radiotherapy can improve the 5-year survival rate of patients with MEC by 38% [31], and the recommended dose is 60–66 Gy [33].

In conclusion, MEC incidence at the tongue base is extremely low, with atypical clinical symptoms. Our case findings emphasize the importance of oncologic workup to determine primary tumor location and ensure accurate histopathology and molecular feature. MEC prognosis may be closely related to the adequacy of the lesion resection, histological grade, and clinical stage. Owing to the unpredictability of the disease, close lifelong follow-up is recommended, regardless of the presence of a tumor.

Abbreviations

MEC	Mucoepidermoid carcinoma
WHO	World Health Organization
MAML2	*Mastermind-like 2*
CT	Computed tomography
MRI	Magnetic resonance imaging
HE	Hematoxylin and eosin
PAS	Periodic acid–Schiff

Acknowledgements

The authors acknowledge the patient who agreed to participate in this study.

Author contributions

All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by PW, HL, FW, YW, and HY. The first draft of the manuscript was written by PW, and all authors commented on previous versions of the manuscript. All authors read and approved of the final manuscript.

Funding

This study was supported by the Natural Science Foundation of Guangdong Province (2019A1515011911), Shenzhen Fund for Guangdong Provincial High-level Clinical Key Specialties (no. SZGSP008), and the Sanming Project of Medicine in Shenzhen (SZSM 201512036, Oral and Maxillofacial Surgery Team, Professor Yu Guangyan, Stomatology Hospital Peking University) to the Department of Oral and Maxillofacial Surgery, Peking University Shenzhen Hospital, Shenzhen, Guangdong, China.

Availability of data and materials

No data set was generated for this article.

Declarations

Ethics approval and consent to participate

Not applicable. Ethical review and approval were waived for this study because it was a single case report. The participant provided written informed consent to participate in the study. All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975 as revised in 2008.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

Received: 10 October 2024 Accepted: 5 February 2025
Published online: 24 February 2025

References

1. Peraza A, *et al*. Mucoepidermoid carcinoma. An update and review of the literature. J Stomatol Oral Maxillofac Surg. 2020;121(6):713–20.
2. Luo X, Han H. Prognostic analysis and nomogram establishment in patients with mucoepidermoid carcinoma of the tongue: a population-based study using the SEER database. Eur Arch Otorhinolaryngol. 2022;279(3):1489–98.
3. World Health Organization Classification of Tumours Editorial Board, BlueBooksOnline - WHO Classification of Tumours Online. https://tumourclassification.iarc.who.int/aboutus, 2021.
4. White VA, *et al*. Mucoepidermoid carcinoma (MEC) and adenosquamous carcinoma (ASC), the same or different entities? Mod Pathol. 2022;35(10):1484–93.
5. Bishop JA, *et al*. MAML2 rearrangements in variant forms of mucoepidermoid carcinoma: ancillary diagnostic testing for the ciliated and warthin-like variants. Am J Surg Pathol. 2018;42(1):130–6.
6. Jee KJ, *et al*. Genomic profiles and CRTC1-MAML2 fusion distinguish different subtypes of mucoepidermoid carcinoma. Mod Pathol. 2013;26(2):213–22.
7. Heidelberger KP, Batsakis JG. Mucoepidermoid carcinoma of the tongue. J Laryngol Otol. 1973;87(12):1239–42.
8. Adkins WJ, Putney FJ. Mucoepidermoid carcinoma of the tongue. Trans Sect Otolaryngol Am Acad Ophthalmol Otolaryngol. 1975;80(5):465–7.
9. Pickell G. Chronic glossopharyngeal neuralgic pain associated with mucoepidermoid carcinoma. CMAJ. 1985;133(6):579–80.
10. Pfendler DF. Glossopharyngeal neuralgia with tongue carcinoma. Arch Otolaryngol Head Neck Surg. 1997;123(6):658.
11. Varghese BT, *et al*. Late scar recurrence in mucoepidermoid carcinoma of base of tongue. J Laryngol Otol. 2000;114(4):299–301.
12. Pires FR, *et al*. Synchronous mucoepidermoid carcinoma of tongue and pleomorphic adenoma of submandibular gland. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2003;95(3):328–31.
13. Andrews KV, Eveson JW. Myokymia (fasciculation) of the tongue as a unique presentation of mucoepidermoid carcinoma. Int J Oral Maxillofac Surg. 2007;36(1):79–81.
14. Leong SC, *et al*. Mucoepidermoid carcinoma of the tongue. Singapore Med J. 2007;48(10):e272–4.
15. Liu W, *et al*. Mucoepidermoid carcinoma arising in a background of sialadenoma papilliferum: a case report. Head Neck Pathol. 2009;3(1):59–62.
16. Sobani ZU, *et al*. Mucoepidermoid carcinoma of the base of tongue. J Pak Med Assoc. 2011;61(9):945–7.
17. Martellucci S, *et al*. Mucoepidermoid carcinoma of the tongue base mimicking an ectopic thyroid. Case Rep Otolaryngol. 2013;2013:1–3.
18. Kalogirou EM, *et al*. An asymptomatic tumor on the dorsal tongue. Oral Surg Oral Med Oral Pathol Oral Radiol. 2014;117(6):659–62.
19. Bollig, C., *et al*., Base of Tongue Mass. Mucoepidermoid carcinoma. JAMA Otolaryngol Head Neck Surg, 2015. 141(10): p. 935–6.
20. Mesolella M, *et al*. Mucoepidermoid carcinoma of the base of tongue. Acta Otorhinolaryngol Ital. 2015;35(1):58–61.
21. Vingerhoedt SI, *et al*. Elevated carcinoembryonic antigen tumour marker caused by head and neck cancer: a case report and literature study. Cancer Radiother. 2015;19(2):106–10.
22. Su R, *et al*. Mucoepidermoid carcinoma of the lingual tonsil. Am Surg. 2016;82(9):e269–70.
23. Rubin A, *et al*. Case report: recurrent mucoepidermoid carcinoma of the tongue in adult female patient with lung cancer. Clin Med Insights: Ear, Nose Throat. 2017;10:117955061772046.
24 Mathew L, *et al*. Mucoepidermoid carcinoma of the posterior-lateral border of tongue: a rare presentation. BMJ Case Rep. 2017. https://doi.org/10.1136/bcr-2017-221521.
25. Ghazali N, Flashburg A, Ord RA. Extensive regional metastasis of high-grade mucoepidermoid carcinoma of an unknown primary tumor. J Oral Maxillofac Surg. 2017;75(4):874.e1-874.e7.
26 Zahran M, Youssef A. Mucoepidermoid carcinoma of the tongue base: a case report. OTO Open. 2018. https://doi.org/10.1177/2473974X18791559.
27. Chen Y, *et al*. Mucoepidermoid carcinoma of tongue: 5 cases. Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2018;53(11):863–5.
28. Whaley RD, Gupta S, Erickson LA. Mucoepidermoid carcinoma. Mayo Clin Proc. 2023;98(9):1427–8.
29 N AK, *et al*. Mucoepidermoid carcinoma of sublingual gland: a malignant neoplasm in an uncommon region. Case Rep. 2011. https://doi.org/10.1136/bcr.02.2011.3864.
30. McHugh CH, *et al*. Prognostic factors in mucoepidermoid carcinoma of the salivary glands. Cancer. 2012;118(16):3928–36.
31. Shidnia H, *et al*. Carcinoma of major salivary glands. Cancer. 1980;45(4):693–7.
32. Yook JI, *et al*. The myoepithelial cell differentiation of mucoepidermoid carcinoma in a collagen gel-based coculture model. J Oral Pathol Med. 2004;33(4):237–42.
33. King W, Ko S, Miller D. Recurrent high-grade invasive mucoepidermoid carcinoma of larynx: a case report and review of the literature. Rare Tumors. 2016;8(2):6204.

Publisher's Note