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Case Report

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Epithelial-myoepithelial carcinoma of the nasopharynx: Two case reports and a literature review

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Abstract

Background: Epithelial-myoepithelial carcinoma (EMCa) is a rare, low-grade malignant salivary gland tumor. Nasopharyngeal EMCa, which arises from the minor salivary glands of the nasopharyngeal mucosa, is even rarer clinically, with limited case reports. Due to its extremely low incidence, there is currently no unified standard treatment protocol, and clinical experience is limited. Its unique clinicopathological features and prognostic factors urgently need to be elucidated. This paper reports two cases of nasopharyngeal EMCa confirmed by pathology and, through a review of the English-language literature, analyzes and summarizes their clinicopathological features, treatment modalities, and prognostic factors, aiming to provide a reference for clinical diagnosis and treatment.

Methods: Two cases of early-stage nasopharyngeal epithelial-myoepithelial carcinoma were reported, and the relevant literature on nasopharyngeal epithelial-myoepithelial carcinoma was reviewed and analyzed.

Results: Both patients presented with early-stage nasopharyngeal epithelial-myoepithelial carcinoma and achieved consistent clinical outcomes. The two patients underwent surgical resection combined with adjuvant radiotherapy, achieving complete clinical remission. No disease recurrence or metastasis was observed during follow-up periods of 52 months and 17 months, respectively.

Conclusions: Our cases demonstrate that surgery combined with radiotherapy can achieve satisfactory symptom relief and disease control in patients with early-stage nasopharyngeal epithelial-myoepithelial carcinoma.

Keywords: Nasopharynx; Epithelial-myoepithelial carcinoma; Case report; Surgery; Radiotherapy.

1 Introduction

Epithelial-myoepithelial carcinoma (EMCa) was first reported by Donath in 1972 [1]. EMCa is a rare tumor of salivary gland origin, accounting for less than 1% of all salivary gland tumors and approximately 2% of malignant salivary gland tumors [2]. In 1991,

the World Health Organization (WHO) first classified it as a low-grade malignant tumor [3]. Although EMCa is rare overall, cases originating in the nasopharynx are even rarer. Its unique anatomical location and biological behavior present significant challenges to clinical diagnosis and treatment. In-depth study of such cases is of great value for improving our understanding of the disease.

EMCa is characterized by a biphasic tubular structure composed of varying proportions of ductal cells and surrounding clear cell-like myoepithelial cells. These cells exhibit infiltrative growth, and their relative proportion can vary, leading to different histological subtypes [4]. EMCa typically arises in the major salivary glands, with the parotid gland being the most common site, followed by the submandibular gland and minor salivary glands. It is occasionally found in the lacrimal gland, maxillary sinus, nasal septum, pituitary gland, tongue, trachea, lung, breast, and liver [5]. To date, cases of primary nasopharyngeal EMCa have been reported very rarely [6-9]. Due to its extremely low incidence, limited clinical experience, and a lack of high-level evidence-based medicine, current diagnostic and treatment protocols are largely based on individual case experiences. There is an urgent need to clarify its clinicopathological features and prognostic factors through the accumulation of more cases. To enhance the diagnostic and therapeutic standards and deepen the understanding of this rare disease, we hereby report two cases of nasopharyngeal EMCa treated in our department.

2. Case report

2.1 **Case 1**

2.1.1 **Patient Presentation and Initial Evaluation**

A 50-year-old female patient presented in October 2021 with a one-month history of a foreign body sensation in the pharynx accompanied by mouth breathing. A nasopharyngeal MRI revealed a mass in the nasopharyngeal cavity (Figure **1A**). Nasopharyngoscopy revealed a mass in the nasopharyngeal cavity (Figure **1B**). No metastases to other sites were detected preoperatively.

2.1.2 **Surgical Treatment**

On October 9, 2021, the patient underwent a "skull base tumor resection" under general anesthesia. A post-operative contrast-enhanced CT scan on October 27 showed post-surgical changes with partial absence of the nasopharyngeal roof and posterior wall, but no obvious abnormal enhancement (Figure **1C**).

2.1.3 **Postoperative Pathology and Multidisciplinary Team (MDT) Discussion**

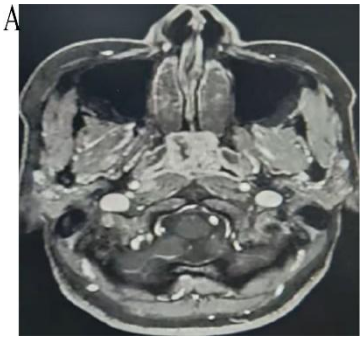
A multidisciplinary team (MDT) discussion concluded that, based on the intraoperative findings and post-operative pathology (which confirmed myoepithelial carcinoma, staged as T1N1M0, Stage II), the resection was incomplete, thus indicating a need for radiotherapy. The patient's general condition was good, with no contraindications for radiotherapy. Therefore, Intensity-Guided Radiotherapy (IGRT) was planned from November 10 to December 23, 2021, with the following regimen: PGTV 66Gy/32F, PGTVtb 60Gy/32F, and PTV 54Gy/32F (Figure 1**D,E**).

2.1.4 **Follow-Up Outcomes**

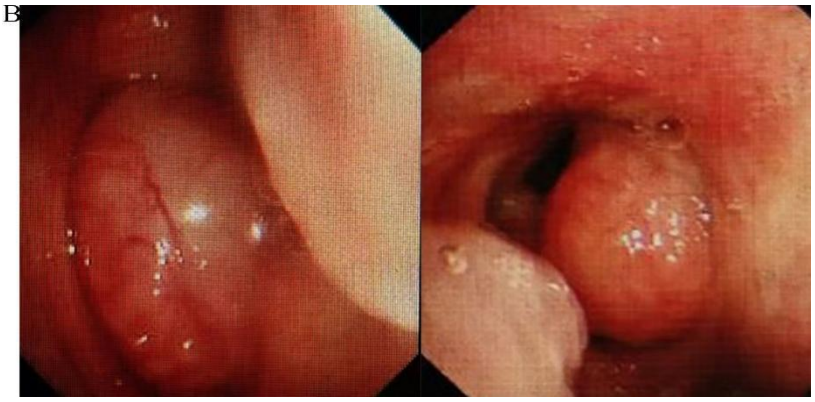
The patient was followed up regularly. As of the latest follow-up on February 12, 2026, there was no local recurrence or distant metastasis (Figure 1**F**). According to the RECIST 1.1 criteria, the patient achieved a clinical complete response (cCR). The overall survival (OS) was 54 months, and the progression-free survival (PFS) was 52 months(Figure 1M).

2.1.5 **Histopathological findings**

Microscopic examination revealed tumor infiltration into the submucosal lymphoid tissue, characterized by large nodular structures. Within these nodules, the tumor cells had abundant, clear, or eosinophilic cytoplasm and were arranged in sheet-like or papillary patterns. Focal cystic structures were also observed. Immunohistochemistry (IHC) showed: Positive (+): CK (Figure 1**G**), CK7 (Figure 1**H**), CK5/6 (Figure 1**I**), P63 (Figure 1**J**), SOX-10 (Figure 1**K**). In Situ Hybridization (ISH) (performed on a fully automated immunohistochemistry instrument): EBER (-). Special Stain: Periodic Acid-Schiff (PAS) stain was focally positive for mucin (Figure 1**L**).

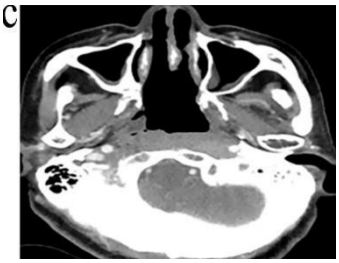


(A) Preoperative contrast-enhanced MRI: An exophytic mass in the nasopharyngeal cavity.



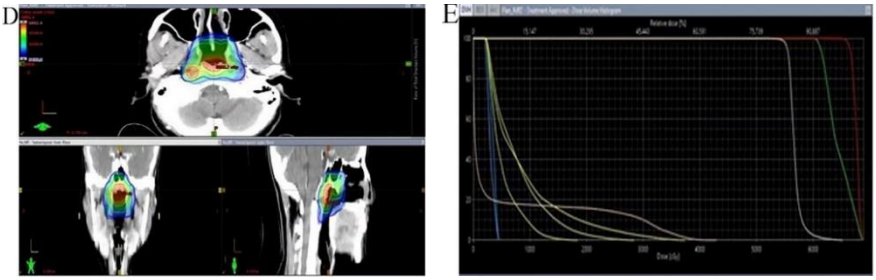
(B) Preoperative nasopharyngoscopy: A mass in the nasopharyngeal cavity obstructing the bilateral posterior choanae.

C



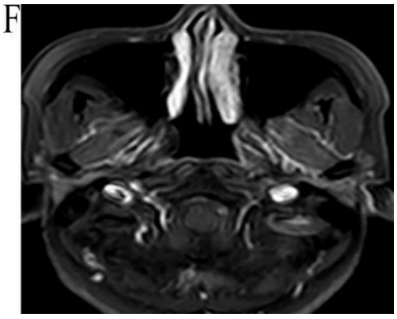
(C) Post-operative contrast-enhanced CT (Oct 27, 2021): Post-operative changes and mucosal defect in the nasopharynx. No abnormal enhancement observed.

D E

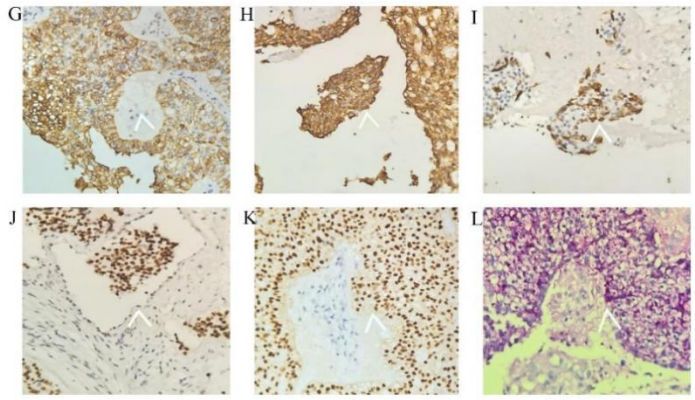


(D, E) Radiotherapy plan: (D) Dose distribution. Axial, sagittal, and coronal views showing the 95% isodose line (red) conformally covering the planning target volume (PTV) while sparing the Organs at Risk (OARs). (E) Dose-volume histogram (DVH). Quantitative analysis of radiation dose to the PTV and key OARs.

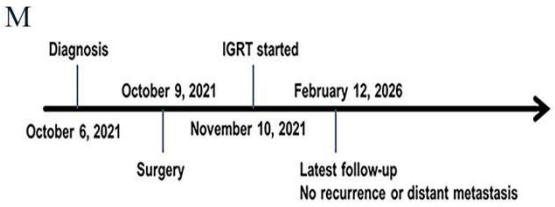
F



(F) Latest follow-up (T1 axial post-contrast MRI): No definite recurrence in the nasopharynx.



(G) Immunohistochemical staining showed positive staining for CK.(magnification, x40)，(White arrow). (H) Immunohistochemical staining showed positive staining for CK7. (I) Immunohistochemical staining showed positive staining for CK5/6. (J)Immunohistochemical staining showed positive staining for p63. (K)Immunohistochemical staining showed positive staining for SOX-10. (L)Special Stain: Periodic Acid-Schiff (PAS) stain was focally positive for mucin.



(M) Treatment timeline of the patient 1.

2.2 Case 2

2.2.1 **Patient Presentation and Initial Assessment**

A 56-year-old female patient was admitted to our hospital after a nasopharyngeal mass was incidentally found for one day. In September 2024, she developed nasal congestion without an obvious cause, accompanied by bilateral orbital distension and pain, and xerostomia. She denied epistaxis, tinnitus, hearing loss, facial numbness, diplopia, headache, fever, or night sweats. A CT scan performed at the Second People's Hospital of Nankang District on October 31, 2024, revealed a mass in the nasopharynx, with a suspected diagnosis of nasopharyngeal carcinoma.

2.2.2 **In-Hospital Examination and Diagnostic Findings**

The patient presented to our hospital on the same day. An electronic nasopharyngoscopy revealed a mass on the roof of the nasopharynx, obstructing both choanae. The bilateral pharyngeal recesses and torus tubarius were not visualized (Figure 2**A, B**). A contrast-enhanced MRI of the nasopharynx and neck showed a nasopharyngeal mass suspicious for malignancy, along with slightly enlarged bilateral level II cervical lymph nodes (Figure 2**C**). Pre-operative workup did not reveal metastasis to other sites.

2.2.3 **MDT Discussion and Initial Treatment Plan**

After a MDT consultation, the following treatment plan was recommended: 1. Surgical resection; 2. Adjuvant radiotherapy to be determined based on surgical margins. After being informed of the consultation results, the patient and their family decided to discharge themselves against medical advice.

2.2.4 **Re-admission and Surgical Treatment**

The patient was re-admitted on November 23, 2024, and underwent endoscopic nasocranial base tumor resection, bilateral level II lymph node dissection, nasal septal mucosal flap repair, and bilateral partial resection of the middle and inferior turbinates under general anesthesia. Post-operative pathology confirmed nasopharyngeal EMCa, staged as T1N0M0 (Stage I).

2.2.5 **Postoperative Imaging Assessment**

A post-operative contrast-enhanced CT scan one month after surgery showed post-surgical changes with a local mucosal defect but no abnormal enhancement (Figure 2**D**).

2.2.6 **Postoperative Adjuvant Radiotherapy**

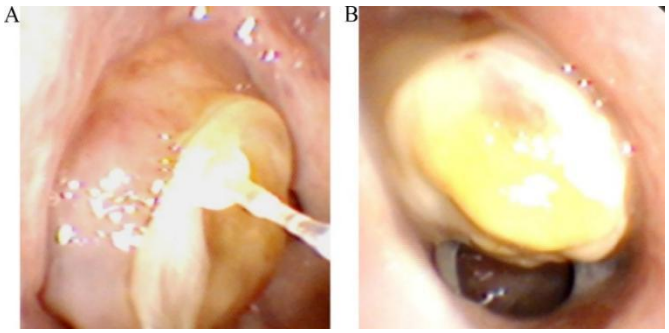
Considering the risk of recurrence, post-operative radiotherapy for the nasopharynx and neck was administered from January 7 to February 18, 2025, using a VMAT (Volumetric Modulated Arc Therapy) technique. The radiation plan was as follows: DT: PGTVnx 70Gy/33 fractions, PGTVnd-right 66Gy/33 fractions, PGTVnd-left 60Gy/33 fractions, PCTV1 60Gy/33 fractions, and PCTV2 55Gy/33 fractions (Figure 2**E, F**).

2.2.7 **Follow-Up Outcomes**

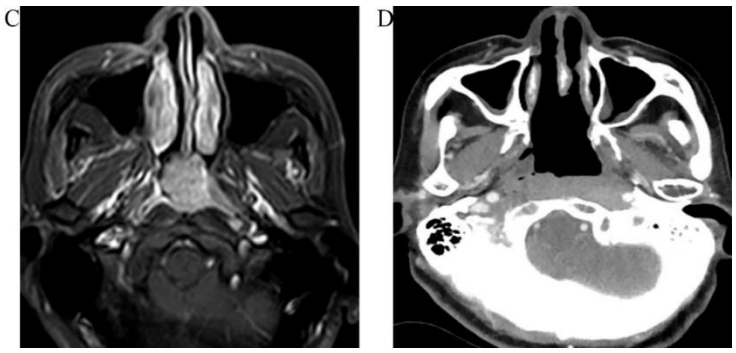
As of the latest follow-up on March 24, 2026 (Figure **2G**), there was no evidence of local recurrence or distant metastasis. According to the RECIST 1.1 criteria, the patient achieved a cCR. The OS was 18 months, and the PFS was 17 months(Figure **2N**).

2.2.8 **Histopathological findings**

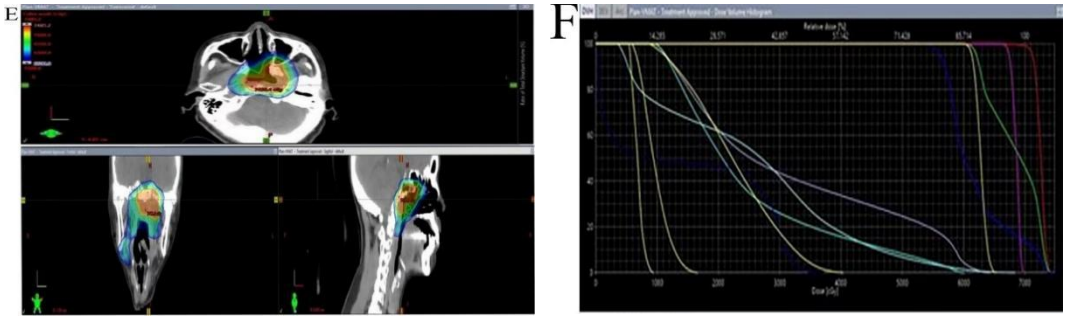
Microscopic examination revealed extensive necrosis and abundant carcinoma tissue. The cancer cells exhibited clear cytoplasm, round nuclei with visible nucleoli, and were arranged in a diffuse, sheet-like pattern. Papillary and glandular/acinic structures were also observed within the tumor. IHC showed the following results: Positive (+): CK (Figure **2H**), CK7 (Figure **2I**), CK5/6, (Figure **2J**), P63 (Figure **2K**), SOX-10 (Figure **2L**), EGFR (2+), P16 (focally), EMA. In ISH: EBER (-). Special Stain: PAS stain was positive for mucin (Figure **2M**).



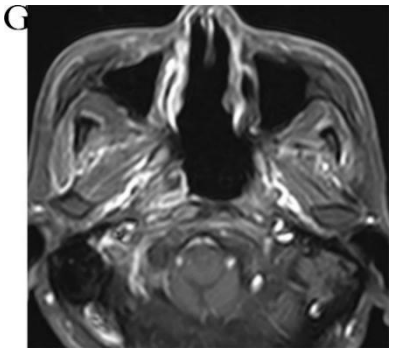
(**A)**Nasopharyngeal mass obstructing the left choana. **(B)**Nasopharyngeal mass obstructing the right choana



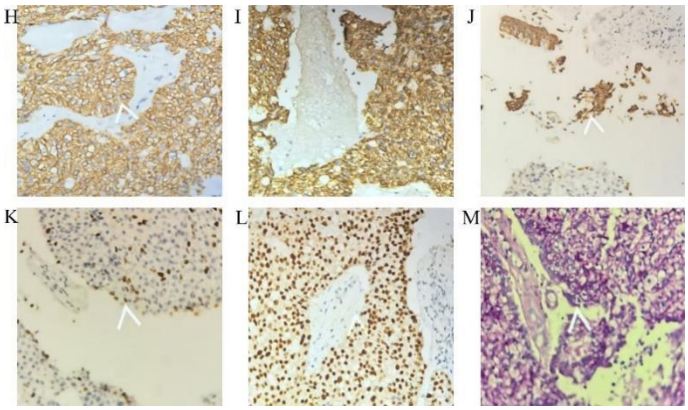
(C) Contrast-enhanced T1-weighted axial MRI (Nasopharynx and Neck): Enhancing nasopharyngeal mass suspicious for malignancy, with slightly enlarged bilateral level II lymph nodes.**(D)** Follow-up contrast-enhanced CT (Dec 30, 2024): Post-surgical nasopharyngeal changes with local mucosal defect and no abnormal enhancement.



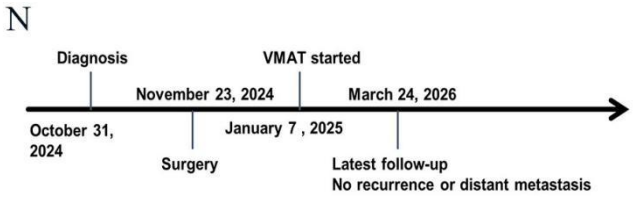
(E, F) Radiotherapy Plan: (E) Dose distribution on axial, sagittal, and coronal CT: 95% isodose line (red) conformally covering PTV while sparing OARs. (F) Corresponding DVH: Quantitative analysis of dose to PTV and OARs.



(G) Latest follow-up (T1 axial post-contrast MRI): Post-surgical changes and local mucosal defect in the nasopharynx, with no abnormal enhancement.



(H) Immunohistochemical staining showed positive staining for CK.(I) Immunohistochemical staining showed positive staining for CK7. (J) Immunohistochemical staining showed positive staining for CK5/6. (K) Immunohistochemical staining showed positive staining for P63. (L) Immunohistochemical staining showed positive staining for SOX-10. (M) Special Stain: Periodic Acid-Schiff (PAS) stain was focally positive for mucin.



(N) Treatment timeline of the patient 2.

3. Discussion

EMCa is a rare, low-grade malignant salivary gland tumor with a low incidence. It predominantly affects the major salivary glands, most commonly the parotid gland, followed by the submandibular and minor salivary glands [5]. The presence of minor salivary glands within the nasopharyngeal mucosa makes it a potential site for this tumor. EMCa shows a slight female predominance, with over 70% of cases occurring in patients over 60 and a female-to-male ratio ranging from 1.6:1 to 6:1 [5]. Although classified as a low-grade malignancy, EMCa is associated with a 35-50% risk of local recurrence and an 8.1-25% risk of distant metastasis [10, 11]. A retrospective cohort study of 246 salivary gland EMCa patients reported disease-specific survival (DSS) rates of 91.3%, 90.2% and 80.7% at 60, 120 and 180 months post-operatively [12]. We herein report two female cases presenting with nasal obstruction but without neurological symptoms like headache or facial numbness. Imaging confirmed nasopharyngeal masses without evidence of distant metastasis. Following radiotherapy, the patients have been followed for 52 and 17 months, respectively, with no signs of recurrence or metastasis. Their overall survival times are 54 and 18 months, respectively.

The typical histopathology of EMCa is characterized by a biphasic tubular structure, composed of an inner layer of eosinophilic ductal epithelial cells (ECs) and an outer layer of clear myoepithelial cells (MCs). These cells exhibit infiltrative growth, and their proportions can vary, leading to different histological subtypes [4]. This histological diversity, complexity, and heterogeneity pose significant challenges to diagnosis. Immunohistochemical (IHC) staining reveals positivity for glandular epithelial markers such as CK, CK7, and Epithelial Membrane Antigen (EMA), as well as varying degrees of expression for myoepithelial markers including S100, smooth muscle actin, P63, and Calponin [5, 13, 14]. EMC needs to be distinguished from several salivary gland neoplasms. Adenoid cystic carcinoma, although also a biphasic tumor, is composed of smaller myoepithelial cells with angulated, hyperchromatic nuclei and displays characteristic cribriform architecture along with true and false glandular lumina, whereas EMC features larger transparent myoepithelial cells lacking these features. Pleomorphic adenoma is well circumscribed with myxochondroid stroma, in which myoepithelial cells gradually merge into the stroma, and can be readily distinguished from EMC. Clear cell mucoepidermoid carcinoma, in addition to abundant clear cells, contains mucous, intermediate, and epidermoid cells; it expresses p63 but lacks other myoepithelial markers. Clear cell carcinoma is characterized by a trabecular or cord-like pattern composed of a uniform population of clear cells, with or without hyalinized stroma, and expresses CKpan and EMA but not myoepithelial markers, thereby allowing its differentiation from EMC. One study suggests that HRAS mutation is the most common molecular event in primary EMC, occurring in approximately 80% of cases, whereas EMCs arising from malignant transformation of pleomorphic adenoma lack this mutation [15]. Sanger and Next-Generation Sequencing (NGS) results indicate that HRAS mutations are predominantly located at codon 61 (Q61R), linking EMCa to mutations at this HRAS locus [15]. Furthermore, nasopharyngeal myoepithelial carcinoma has been reported to consist of oval clear cells embedded in a Periodic Acid-Schiff (PAS)-positive basement membrane-like substance [7, 8]. Related studies have also shown that SOX10 is expressed in EMCa and serves as a marker for intercalated duct differentiation [16]. In the present report, immunohistochemical staining of the two nasopharyngeal cases revealed positivity for glandular epithelial markers (CK, CK7, CK5/6) and the myoepithelial marker P63. Special stains showed positive results for PAS and SOX10. Based on these morphological and immunohistochemical findings, a diagnosis of EMCa was established.

Furthermore, a systematic search of the PubMed database from its inception to April 3, 2026, identified five previously reported cases of nasopharyngeal EMCa. In conjunction with the two cases presented herein, a total of seven cases were included for analysis, with their baseline clinical and immunohistochemical features summarized in **Table 1**. The mean age at diagnosis was 56.6 years, and the majority of patients were female (85.7%). Treatment strategies were classified by initial modality: five patients (71.4%) underwent primary surgical resection followed by adjuvant

(chemo)radiotherapy, while the remaining two (28.6%) received primary (chemo)radiotherapy. During follow-up, locoregional recurrence occurred in two patients—one from the surgery group and one from the primary chemoradiotherapy group. Additionally, one patient in the surgery group (14.2%) developed distant metastasis despite resection and multimodal adjuvant therapy. All patients were confirmed alive at the end of their respective follow-up periods. The mean age of patients with recurrence was 42.5 years.

Given the rarity of EMCa, a consensus on the optimal treatment regimen is lacking [17]. While radical surgical resection is the mainstay of treatment, evidence supporting the benefit of adjuvant radiotherapy is limited, and its true survival advantage remains unclear [12, 18]. Conversely, some studies suggest that extensive resection combined with optimal adjuvant therapy may yield better survival outcomes. Consequently, postoperative adjuvant radiotherapy is often recommended for patients with high-risk features such as tumors larger than 4 cm, positive margins, perineural invasion, lymph node metastasis, or recurrent disease [2,19]. One study posited that sequential radiotherapy could enhance the complete response (CR) rate, and for primarily unresectable tumors, radical radiotherapy with or without chemotherapy is a viable option [20]. The two cases in this report were early-stage and achieved a clinical complete response with surgery and adjuvant radiotherapy. For unresectable cases, treatment typically involves chemotherapy, radiotherapy, and immunotherapy, though the efficacy of these approaches is not well-defined. Reported chemotherapy regimens for metastatic Myoepithelial carcinoma (MC) include single-agent taxanes or platinum, as well as combinations of taxane-platinum or platinum-anthracycline. Despite these options, the prognosis for metastatic disease remains poor [21]. For instance, Johnson et al. [22] described a case of cutaneous MC with multiple bone metastases that progressed despite chemotherapy and local radiotherapy, leading to the patient's death five months post-diagnosis. Immunotherapy holds potential as a therapeutic avenue for MC. Agarwal et al. [23] and Cormier et al. [24] reported PD-L1 positivity in MC patients, correlating it with poorer survival. In a separate case report [6], two patients were treated with immunotherapy. The first achieved a clinical complete response, while the second, who had relapsed and metastasized after multiple lines of therapy, achieved disease control upon the addition of immunotherapy. These findings suggest immunotherapy could be an effective strategy for EMCa. However, despite promising results in some case reports, its definitive role in treating MC requires validation through further research.

4. Conclusion

These two cases underscore the rarity and diagnostic challenges of nasopharyngeal EMCa. Individualized multimodal therapy, including surgery and precision radiotherapy, proves effective in ameliorating symptoms and extending survival for patients with early-stage nasopharyngeal epithelial-myoepithelial carcinoma. Future studies are required to refine therapeutic strategies and further improve prognosis.

Limitations of this case series: First, these are single-center case reports with a very limited number of patients. Second, neither patient underwent a PET-CT scan, which was attributable to financial constraints. Third, both cases presented with early-stage disease; the absence of patients with locally advanced or metastatic disease limits our ability to report on the treatment and prognosis for these stages. Future clinical encounters with such patients should be reported to accumulate further data.

Clinical trial number

Not applicable.

Authors' contributions Lei Tan: Writing–original draft, Investigation, Methodology, Visualization, Writing–review & editing. Research design. GaoYin Zheng: Visualization, Data curation, Writing – review & editing. Wei Wu: Visualization, Writing–review & editing. Xianhua Lei: Pathological diagnosis, Writing–review & editing.

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Data availability

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation. No new sequencing data, structural data, or datasets were generated or analyzed in this case report. All relevant clinical information is included in the manuscript.

Ethics approval and consent to participate

The studies involving human participants were approved by the Medical Ethics Committee of Ganzhou Cancer Hospital (approval no 2026-125). The studies were conducted in accordance with the Declaration of Helsinki, local legislation and institutional requirements. Informed consent to participate was obtained from the patient/patient's legal guardians[if the pa tient's age is less than 16].

Patient consent for publication

Written informed consent was obtained from the patient for the publication of the present case report and any accompanying images.

Competing interests

1 The author declares that they have no competing interests.

TABLE1 Baseline Clinicopathological characteristics of reported cases of
2 nasopharyngeal epithelial-myoepithelial carcinoma.

Author/Year	Age/Sex	Tumor size	Clinical manifestation	Epithelial markers	Myoepithelial markers	Treatment	Metastasis time	Metastasic sites	Metastatis therapy	Survival outcome
Imate et al. [7]	60/F	NA	Ear pain	EMA, Ber; EP-4AE1/AE3, MNF116, S-100a and b antibodies	SMA	Surgery	NA	NA	NA	No recurrence or metastasis in 55 months
Kim et al.[8]	80/F	2.8×2.6 cm	Nose blockage, Epistaxis	CK7	SMA	CCRT + Chemotherapy	NA	NA	NA	No recurrence or metastasis in 24 months
Zhang et al.[9]	51/M	5.6×3.4 cm	wallowing pain, Ear tinnitus	CK7	P63, SMA, VIM	CCRT	NA	NA	NA	Recurrence 33 months after initial diagnosis
Xiong et al [6]	65/F	4.6×2.7 x4.5cm	Foreign body sensation, Sleep snoring	CK7	P63, SMA	Surgery + Radiotherapy + Immunotherapy	NA	NA	NA	No recurrence or metastasis 31 months after surgery
Xiong et al [6]	34/F	4.5×3.0 x5.0cm	Nasal obstruction, Epistaxis, Hyposmia Hypogeusia, Tinnitus, and Facial numbness, Diplopia, Hearing loss	Pan-CK	P63	Surgery + CCRT + targeted therapy	36 months	lung	multi-line chemotherapy therapy+Immunotherapy	Recurrence and lung metastasis 24 months after CAP regimen treatment. No recurrence or abnormally enlarged lung nodules 23 months after lung metastases.
Patient 1	50/F	NA	Pharyngeal foreign body sensation, buccal respiration	CK、CK7	P63	Surgery + Radiotherapy	NA	NA	NA	No recurrence 52 months after initial diagnosis

Patient 2	56/F	NA	Nasal obstruction, Bilateral orbital distension and pain, Thirst	CK、CK7	P63	Surgery + Radiotherapy	NA	NA	NA	No recurrence 17 months after initial diagnosis

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