—Case Reports—

# Epithelial-Myoepithelial Carcinoma of the Submandibular Gland: Case Report

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Epithelial-myoepithelial carcinoma (EMC) is a rare malignant salivary gland tumor that occurs mostly in the parotid gland. We report a case of EMC of the submandibular gland in a young man. The patient was aware of a slow-growing mass in the right submandibular gland for 1 year. Clinical examination and ultrasound confirmed a right submandibular mass, 2.5 × 3 $cm^2$ in size. Ultrasound-guided fine-needle aspiration indicated a diagnosis of pleomorphic adenoma, which was also suggested by magnetic resonance imaging. The submandibular gland tumor was excised. Immunohistochemical analysis showed carcinoma ex pleomorphic adenoma with a major epithelial-myoepithelial component. The patient was not treated with radiotherapy after surgery. No recurrence was observed during 24 months of follow-up. Because the morphology of EMC is similar to that of a benign tumor, it is frequently initially misdiagnosed. Surgery is always the most effective diagnostic and therapeutic measure for salivary gland tumors, especially those that grow slowly. Resection with negative margins is the treatment of choice for EMC; use of adjuvant radiotherapy is controversial. (J Nippon Med Sch 2021; 88: 238–241)

**Key words:** case report, epithelial-myoepithelial carcinoma, pathological diagnosis, salivary cancer, submandibular gland neoplasm

## Introduction

Epithelial-myoepithelial carcinoma (EMC) is a rare, low-grade, malignant salivary gland tumor that occurs mostly in the parotid gland. It was first described by Donath et al. in 1972[1] and is slightly more common in females. Ultrasound (US) is always recommended for assessment. However, because the morphology of EMC is similar to that of a benign tumor, it is frequently initially misdiagnosed. Surgery is the treatment of choice for salivary gland EMC. A more accurate diagnosis of the disease can be achieved by histological and immunohistochemical studies[2]. A malignant tumor often requires multidisciplinary treatment. Radiotherapy is effective for patients with EMC whose tumors cannot be completely resected. We report a rare case of EMC of the submandibular gland in a young man. The clinical manifestations, examinations, and pathological diagnosis are discussed, and the therapeutic strategy for EMC is summarized.

## Case Presentation

A 32-year-old Chinese man was admitted to our Department of Otorhinolaryngology in February 2017 for assessment of a right submandibular gland mass. He had been aware of the tumor for 1 year and reported that it grew slowly. He had no local pain, dyspnea, or dysphagia and no family history of such a condition. Neither systemic disease nor a history of psychosocial was present. He reported no history of relevant past interventions.

The patient's maxillofacial appearance was asymmetrical. Clinical examination showed a right submandibular mass (2.5 × 3 $cm^2$), with a smooth surface that was fixed and adhered to surrounding tissues. The mass was hard on palpation. Although most submandibular gland masses are benign tumors, palpation suggested the possibility of a malignant tumor. His skin showed no redness, and skin temperature was not elevated. Mouth opening and occlusal function were not affected. Intraorally, the mucosa appeared normally moist. The submandibular gland duct was normal. No neck lymph nodes were found. The results of laboratory testing were within normal ranges. US revealed a cystic mass with predominantly solid lesions. US-guided fine-needle aspiration

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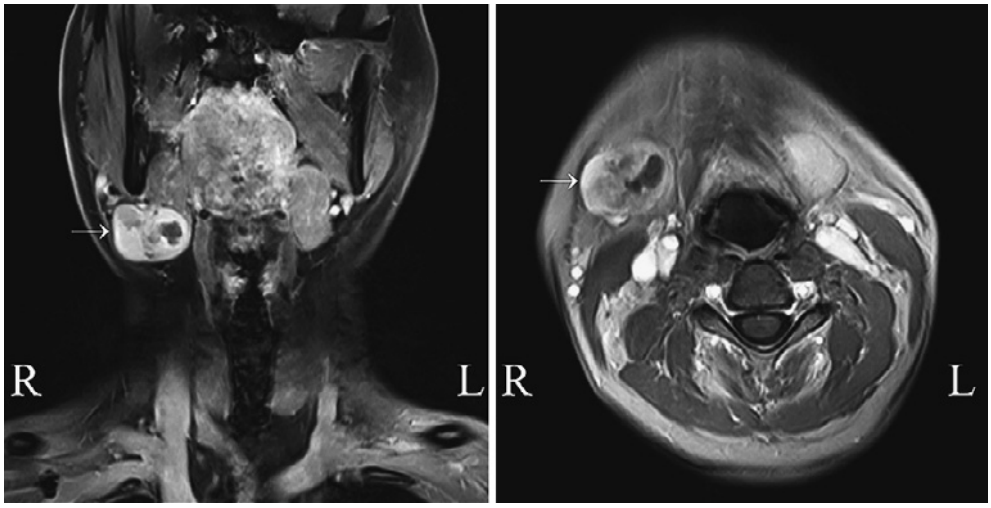


Fig. 1 Enhanced MRI showing a right submandibular gland mass with a smooth margin and uneven enhancement.

Table 1 Summary of preoperative laboratory findings

| Items | Results |
|---|---|
| US | Cystic mass with predominantly solid lesions |
| FNA | Pleomorphic adenoma |
| MRI | Possible pleomorphic adenoma without lymphatic metastasis |
| Blood chemistry, liver and kidney function, electrolytes | Normal |

(FNA) of the mass was performed, and the FNA diagnosis was pleomorphic adenoma. Magnetic resonance imaging (MRI) also indicated pleomorphic adenoma without lymphatic metastasis (**Fig. 1**). **Table 1** summarizes the preoperative laboratory findings. Imaging findings and FNA indicated a diagnosis of benign tumor.

Submandibular gland tumor excision was performed under general anesthesia. After the lower edge of the submandibular gland was separated from the digastric muscle, the gland and tumor were exposed. The tumor was bluntly separated from the submandibular gland and completely removed along with some affected glands. The hypoglossal and lingual nerves were intact and located behind the tumor. Gross evaluation of the specimen revealed a well-circumscribed nodular mass measuring 2.8 × 2.5 × 2.2 $cm^3$ with a grayish-yellow cut surface. An area of the nodule appeared to be cystic and had red-brown liquid inside. Analysis of a frozen section obtained during surgery showed basal cell adenoma. The diagnosis, confirmed by immunohistochemical analysis, was carcinoma ex pleomorphic adenoma with a major epithelial-myoepithelial component (CXPA-EMC). More than 0.15 cm of the tumor capsule and salivary gland were infiltrated. The tumor cell was positive for AE1/AE 3, vimentin, S-100, CD34, p63, and Bcl-2 and focal for EMA, CK8/18, CK5/6, CD10, SMA, and p53. Ki67 was positive in 5% of neoplastic cells (**Fig. 2**). Because the margins were tumor-free, the patient received no further treatment, such as radiotherapy, after surgery. He underwent follow-up examinations at 1 week, 6 months, and 12 months postoperatively. The last follow-up was at 2 years postoperatively, and neck MRI showed no recurrence.

## Discussion

EMC is a rare biphasic tumor that accounts for fewer than 2% of salivary gland malignancies. It presents as an indolent, asymptomatic, multinodular mass that may be present for several years before diagnosis but sometimes presents with ulceration of overlying mucosa[3,4]. Most cases involve the parotid gland (83.74%), followed by the submandibular gland (13.01%) and minor salivary glands[1]. It has also been reported in the trachea, hypopharynx, maxillary sinus, and lacrimal glands[5–8]. EMC has a slight female predominance, and peak occurrence is in the sixth decade of life[1,9]. However, in the present case, it occurred in the submandibular gland of a young man, which is rare.

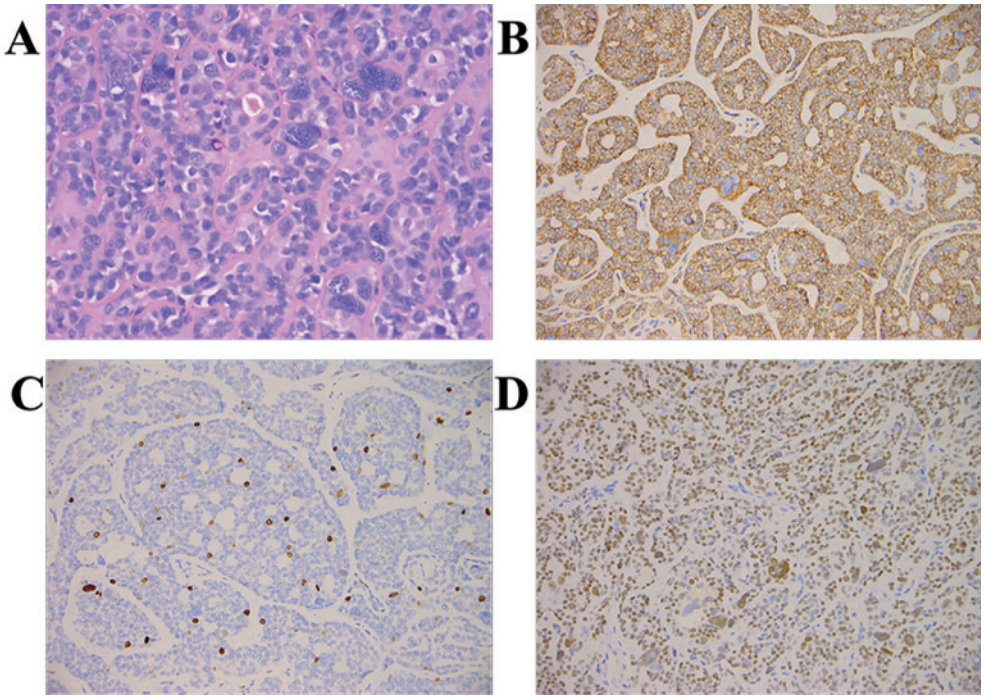


Fig. 2 (A) Presence of duct-like structures with luminal and abluminal cells (hematoxylin and eosin, 400×). (B) EMC: B-cell lymphoma-2 immunohistochemical staining (200×). (C) EMC: ki67 immunohistochemical staining (200×). D) EMC: p63 immunohistochemical staining (200×).

US is always recommended for initial radiological assessment of parotid and submandibular lesions. Because the morphology of EMC is similar to that of benign tumors, it is frequently initially misdiagnosed. Computed tomography and MRI images, although nonspecific, are useful for evaluating tumor growth and its relationship with adjacent vessels, assessing resectability, and planning the extent of neck dissection[3]. Cytological diagnosis may be challenging. A more accurate diagnosis can be achieved by histological and immunohistochemical studies[2]. In the present case, neither imaging findings nor FNA suggested malignancy.

Surgery is the treatment of choice for salivary gland EMC. Existing evidence indicates no apparent benefit with adjuvant radiotherapy[1], although a prior study suggested that it might be effective in preventing local recurrence[10]. EMC is usually deceptively multinodular and well circumscribed, and sometimes has a capsule. The histopathological appearance is distinct and always confirmed by immunohistochemical analysis with conventional light microscopy. Epithelial and myoepithelial markers include cytokeratin 7 for ductal and epithelial cells and p63 for myoepithelial cells[4,11]. EMC is considered a low-grade malignant tumor, and the 5-year survival rate is 94%. It recurs locally after resection in 30% to 50% of cases; however, the incidence of regional and distant metastasis is low. Adequate resection, with negative margins, is the treatment of choice[3].

In the present case, although preoperative examination suggested that the tumor was benign, the diagnosis could not be confirmed until surgery and immunohistochemical analysis were performed. Surgery is always the most effective diagnostic and therapeutic measure for salivary gland tumors, especially those that grow slowly. The patient received neither chemotherapy nor radiotherapy after surgery. A follow-up examination at 2 years postoperatively showed no recurrence.

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**Conflict of Interest:** The authors declare no competing interests.

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