Myoepithelial carcinoma of the vallecula: A case report and review of the literature

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Abstract. Myoepithelial carcinoma (MEC) of the vallecula is relatively rare, and there is no specific surgical guideline for resecting MEC in the vallecula. To the best of our knowledge, the current study reports the first case of MEC involving the vallecula of a 48-year-old female patient. In the present study, the lesion was correctly diagnosed as MEC and successfully managed using lateral pharyngotomy and by the construction of a sternohyoid myofascial flap. The patient was free of local recurrence or metastasis 18 months after surgery. The present study briefly reviews the current knowledge concerning the diagnosis and treatment of MECs in the head and neck area and offers suggestions for managing MEC in the vallecula. To conclude, MEC of the vallecula may be successfully managed by surgical treatment without evident post-surgery complications.

Introduction

Myoepithelial carcinomas (MECs), also termed malignant myoepitheliomas (1), are composed almost exclusively of tumor cells with morphological and immunohistochemical myoepithelial differentiation and clear-cut tumor infiltration into adjacent tissue. MECs account for 1-4% of all salivary gland tumors and arise predominantly from the major glands (1). Occasionally, MECs occur in the intraoral minor salivary glands and most frequently involve the palate (2). In the 2005 edition of the World Health Organization (WHO) histological classification of salivary gland tumors, MECs were considered to be low-grade tumors with a low tendency for local recurrence and metastasis (3). MECs most commonly affect patients in their third to fifth decades of life, with a slight female predilection (2). The clinical prognosis of MECs is not well characterized, however, certain studies have suggested that MECs should be recognized as high-grade malignancies with a poor prognosis (4). Extensive resection with free margins is the recommended treatment for MEC lesions (5). To the best of our knowledge, there is no published literature illustrating MECs of the vallecula, which makes the vallecula a rare location for MECs and region that is challenging to access for surgeons (6). The present study reports the case of a patient with MEC of the vallecula, which was successfully managed using a lateral pharyngotomy approach and the construction of a sternohyoid myofascial flap.

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Case report

A 48-year-old female patient was referred to the Department of Oncology, West China Hospital of Stomatology (Sichuan University, Chengdu, China) from the Sichuan Provincial People's Hospita (Chengdu, China) in April 2014, due to an 8-month history of a foreign body sensation in the pharynx and a 1-month history of intermittent hemoptysis. Upon physical examination, a firm mass was palpable near the tongue base, while the posterior margin of the mass was too deep to reach. No other significant manifestations in the oral cavity and neck region were observed. The endoscopic examination revealed an exophytic tumorous lesion with tortuous vessels on the surface. The majority of the mass was located in the vallecula and involved the base of the tongue and the epiglottis (Fig. 1). Magnetic resonance imaging (MRI) demonstrated a soft tissue mass measuring 41x35x31 mm in size, which extended from the anterior wall of the epiglottis to the base of the tongue, but did not affect the parapharyngeal space (Fig. 2). The mass was well enhanced by gadolinium administration. The pertechnetate thyroid scan single-photon emission computed tomography imaging differentiated the mass from the ectopic thyroid gland of the tongue base. Chest radiographs and ultrasounds of the abdomen showed no sign of distant metastasis.

Since the patient had a history of intermittent hemoptysis, the surgical excision was performed without a preoperative biopsy. Intraoperative frozen sections were used to diagnose the lesion. Under general anesthetic, the mass was removed using the lateral pharyngotomy approach (7). A sternohyoid myofascial flap was used to reconstruct the defect at the base of the tongue. A tracheostomy and primary suture were also performed. The trunks of the ipsilateral hypoglossal nerve and lingual artery were identified and protected prior to surgery in the pharynx

at the safe mucosal margins. The mass, which involved the epiglottis, tongue base and vallecula, was fully excised. The intraoperative frozen sections indicated a preliminary diagnosis of MEC, and the surgical margins were free of disease. The sternohyoid muscle on the right side was elevated up towards the superior margin of the thyroid cartilage, and the superficial fascia was fixed to the underlying muscle. The external surface of the flap and fascia was remodeled into the pharynx for the reconstruction of the tongue base, prior to the fixation of the residual larynx to the newly formed tongue base. The patient was discharged 10 days subsequent to the surgery, without postsurgical chemotherapy or radiotherapy. The patient received regular follow-ups, and exhibited no signs of local recurrence or lymph node or distant metastasis in the 18 months following surgery.

Macroscopically, the excised mass was 4x3 cm in size and the cut surface was pale white in color. The surrounding muscle was infiltrated by the mass. Additional microscopic examination identified the infiltrative growth pattern and necrotic foci in the tumor. The microscopic examination also revealed that the majority of the tumor was composed of clear cells that were characterized by abundant vacuolated clear cytoplasm and displaced nuclei. Scattered spindle cells and epithelioid cells were also identified in the tumor (Fig. 3). Only a few cells were recognized with cellular atypia, and the number of mitoses was low. A hyalinized matrix was also identified, which divided the tumor cells into small nests or thin cords. There was no clinical or pathological evidence of a pre-existing pleomorphic adenoma. Cytokeratin-5, -6 and -7 (CK-5, -6 and -7) and tumor protein p63 (p63) were expressed in the tumor cells when assessed using immunohistochemical staining; however, the S-100 protein (S-100) and carcinoembryonic antigen (CEA) were not expressed. As a result of the histological examination and immunohistochemical staining, a diagnosis of MEC was made.

The present study was in compliance with the Declaration of Helsinki (8) and was approved by the Ethics Committee of the West China College of Stomatology, Sichuan University. Written informed consent was obtained from the patient for the publication of the case report and accompanying images.

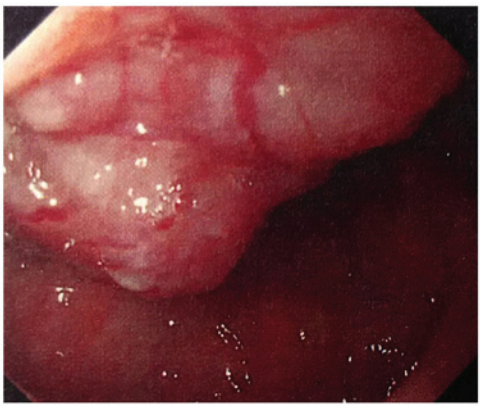

Figure 1. Endoscopic examinations revealed an exophytic tumorous lesion. The mass was mainly located in the vallecula and involved the base of the tongue and the epiglottis.

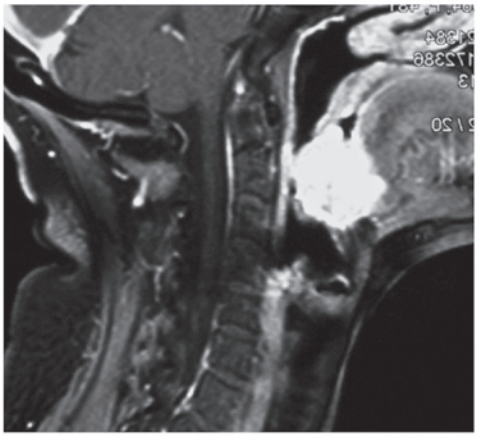

Figure 2. Magnetic resonance imaging revealed a 41x35x31-mm soft tissue mass that extended from the anterior wall of the epiglottis to the base of the tongue, but did not affect the parapharyngeal space.

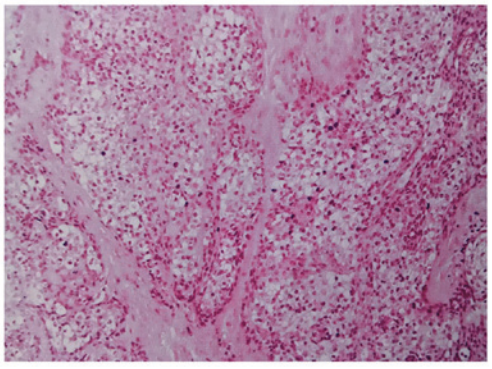

Figure 3. The tumor was mainly composed of clear cells with scattered spindle cells and epithelioid cells. Only a few cells demonstrated cellular atypia and the number of mitoses was low. A hyalinized matrix divided the tumor cells into small nests or thin cords.

Discussion

MECs are a rare group of tumors that account for 1-4% of salivary gland tumors and 0.2-0.32% of minor salivary gland tumors (2). MECs are diagnosed following the criteria of lesions that are composed almost exclusively of tumor cells, with myoepithelial differentiation and clear-cut tumor infiltration into adjacent tissue (6). The majority of studies demonstrate that MECs are more prevalent in the major salivary glands (5,6); however, the findings of a study by Kane *et al* indicated that minor salivary glands have a greater involvement (71%) with MEC (1). MECs involving intraoral minor salivary glands tended to occur in middle to older age groups (range, 14-77 years; mean, 56.9 years), with a slight male predilection (male-to-female ratio, 0.87:1) and a predominance of palate involvement (60.7%) (2). These results were generally in agreement with the clinical findings of MECs that involved the major salivary glands.

Considering the rarity and variety of MECs, the clinical prognosis and biological behavior of the disease were not previously well-characterized. In the 1991 and 2005 editions of the WHO Histological Classification of Salivary Gland Tumors, MECs were considered to be low-grade tumors with a low tendency for local recurrence and metastasis (3,9). However, according to a study by Yu *et al* (5), MECs may be

recognized as high-grade malignancies with poor prognoses. Yang *et al* (2) analyzed MECs of the intraoral minor salivary glands and indicated that those particular MECs were likely to be low-grade malignancies. Di Palma and Guzzo (10) indicated an association between the biological behavior and the origin of the tumor, finding that *de novo* MECs tended to be a high-grade malignancies. An association between the biological behavior and the origin of the tumor was not identified in the studies by Savera *et al* (6) and Kane *et al* (1), and in the latter study, several other valuable histological features that correlated strongly with the clinical behavior of MECs were identified.

Histologically, the tumor cells of MECs are categorized according to cytological features as epithelioid, clear, plasmacytoid and spindle cells (6). The majority of the neoplasms exhibit one prevalent cell type, which blends imperceptibly with the surrounding cell types (6). Two tumor-associated matrices, consisting of myxoid and hyalinized matrices, are acknowledged, and the hyaline stroma is the most commonly observed (1). In the present study, the MEC lesion was mainly composed of clear cells with scattered spindle cells and epithelioid cells, which was divided into small nests or thin cords by a hyalinized matrix. Immunohistochemistry is essential in order to identify MECs. The diagnosis of MECs requires reactivity with the CKs and at least one of the other myoepithelial markers, including S100, vimentin, calponin, p63 or CD10 (3). In the present study, the MEC sample expressed CK-5, -6 and -7 and p63, but did not express S-100 and CEA. The majority of previous studies indicated that the MECs expressed S-100, and only 16% of MEC lesions did not express S-100, which made the diagnosis of MEC even more challenging (1,3,6). In the absence of S-100 expression, the expression of calponin, common acute lymphocytic leukemia antigen, p63 and vimentin aided the diagnosis of MECs (1). p63 is a useful marker of myoepithelial cells in salivary gland neoplasms, but is also expressed in squamous cell carcinoma and mucoepidermoid carcinoma (6). The differentiation of cells is aided by the expression of CEA at the cell luminal surface; therefore, CEA is used to exclude neoplasms with clear cells or epitheloid cells, including adenoid cystic carcinoma, adenocarcinoma and epithelial-myoepithelial carcinoma (6). In the present study, exclusive myoepithelial differentiation was confirmed using morphological and immunohistochemical examinations, while the malignancy diagnosis was supported by an infiltrative growth pattern and the presence of necrotic foci.

Due to the high recurrence rate, radical surgery with free margins is recommended for the successful management of MEC lesions (5,6), and the efficacy of radiotherapy and chemotherapy to treat MEC remains controversial. Metastasis of MEC to the lymph nodes is infrequent, and since the present patient showed no evidence of lymph node metastasis, a neck dissection was not recommended. Several surgical approaches have been proposed for resecting tumors in the neck region, depending on the size and site of the tumor (7,11). Kermani *et al* presented the case of a benign myoepithelioma of the vallecula, and removed the mass using the suprahyoid approach (12). In the present study, the suprahyoid approach was not recommended due to the infiltrative growth pattern of the MEC and the involvement of the vallecula, tongue base and epiglottis, which was identified by MRI scans. Since the lateral pharynx was not affected by MEC, a lateral pharyngotomy was performed for radical resection of the mass with free margins. The wide local resection in this area is likely to induce a swallowing impairment and velopharyngeal incompetence. In order to prevent such complications, a sternohyoid myofascial flap was used to reconstruct the tongue base. No evident complications were noted following surgery, and the patient was free of recurrence for 18 months following surgery.

In conclusion, the present study presents the successful surgical treatment of a rare MEC in the vallecula. The present case requires consideration due to: i) The unusual location of the mass, as no previous case of MEC was indicated to arise from the vallecula; ii) the unusual immunohistochemical appearance, as rare MECs do not express S-100, as presented here; and iii) the unusual technique used to treat the MEC, as there are no specific surgical guidelines for resecting MECs in the vallecula. The present study demonstrates that the lateral pharyngotomy and sternohyoid myofascial flap are viable options for the successfully management of MECs of the vallecula.

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References

1. Kane SV and Bagwan IN: Myoepithelial carcinoma of the salivary glands: A clinicopathologic study of 51 cases in a tertiary cancer center. Arch Otolaryngol Head Neck Surg 136: 702-712, 2010.
2. Yang S, Li L, Zeng M, Zhu X, Zhang J and Chen X: Myoepithelial carcinoma of intraoral minor salivary glands: A clinicopathological study of 7 cases and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 110: 85-93, 2010.
3. Thompson L: World Health Organization classification of tumours: Pathology and genetics of head and neck tumours. Ear Nose Throat J 85: 74, 2006.
4. Wang C, Zhang Z, Ge Y, Liu Z, Sun J, Gao Z and Li L: Myoepithelial carcinoma of the salivary glands: A clinicopathologic study of 29 patients. J Oral Maxillofac Surg 73: 1938-1945, 2015.
5. Yu G, Ma D, Sun K, Li T and Zhang Y: Myoepithelial carcinoma of the salivary glands: Behavior and management. Chin Med J (Engl) 116: 163-165, 2003.
6. Savera AT, Sloman A, Huvos AG and Klimstra DS: Myoepithelial carcinoma of the salivary glands: A clinicopathologic study of 25 patients. Am J Surg Pathol 24: 761-774, 2000.
7. Laccourreye O, Benito J, Menard M, Garcia D, Malinvaud D and Holsinger C: Lateral pharyngotomy for selected invasive squamous cell carcinoma of the lateral oropharynx - part I: How. Laryngoscope 123: 2712-2717, 2013.
8. World Medical Association: World Medical Association Declaration of Helsinki: Ethical principles for medical research involving human subjects. JAMA 310: 2191-2194, 2013.
9. Seifert G, Sobin LH and Thackray AC: Histological Typing of Salivary Gland Tumours. 2nd edition. pp9-10. Springer-Verlag, Berlin, 1991.
10. Di Palma S and Guzzo M: Malignant myoepithelioma of salivary glands: Clinicopathological features of ten cases. Virchows Arch A Pathol Anat Histopathol 423: 389-396, 1993.
11. Mulwafu W, Fagan JJ and Lentin R: Suprahyoid approach to base-of-tongue squamous cell carcinoma. S Afr J Surg 44: 120, 122-124, 2006.
12. Kermani W, Belcadhi M, Ben Ali M, Sriha B and Bouzouita K: Myoepithelioma of the vallecula: A case report. Ear Nose Throat J 90: E9-E11, 2011.