Massive Myoepithelial Carcinoma Originating from the Submandibular Gland That Was Successfully Treated with Surgical Excision, Using a Part of the Lengthened Skin as a Local Flap

Summary: Myoepithelial carcinoma is rare and mostly originates from the major salivary glands. Sometimes, it is difficult to differentiate the benign from the malignant histologically, and its clinical behavior and histological features may vary. Here, we describe the case of a 55-year-old woman who presented with a massive myoepithelial carcinoma, which hung like a temple bell from her right side of the jaw, and she refused to go to the hospital for 3 years. Based on its size and location, we initially thought that, before surgical resection, neoadjuvant therapy would be necessary to reduce the tumor volume. However, after careful evaluation of the tumor characteristics (low-grade histology with outward expansion and little invasion of the adjacent tissues) and imaging findings, we decided that excision was possible. The tumor was encapsulated and had a clear border; it weighed 10.5 kg. By setting the incision line posterior to the equatorial plane and using the lengthened skin posterior to the tumor as a large local flap for the skin defect, we successfully reconstructed the skin defect without harvesting additional flap from other areas. No additional treatment was administered because a sufficient surgical margin was maintained, pathologically. She regained her daily life without recurrence or distant metastasis for 2 years. When treating a massive tumor, careful consideration of its characteristics and location is important, and in this case, we were able to use a simpler and less invasive treatment than we initially envisioned.

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Myoepithelial carcinoma is rare, accounting for 0.45% of all salivary gland tumors. Seventy-five percent of these tumors arise in the major salivary glands, and about half originate from precursor lesions, such as pleomorphic adenoma or benign myoepithelioma. Sometimes, it is difficult to differentiate benign and malignant lesions, histologically. However, the presence of tumor infiltration into the adjacent tissues and high cell proliferative activity (>7 mitoses per 10 high-power fields or a Ki-67 labeling index of >10%) are the most notable criteria for myoepithelial carcinoma.

We present a case of myoepithelial carcinoma that grew to a humongous size as a submandibular lesion. The lesion was successfully treated by surgical resec-
tion without the need for neoadjuvant therapies or use of a large skin flap from another body part.

**CASE REPORT**

A 55-year-old woman had noticed a subcutaneous tumor on the right submandibular region 3 years ago, which began to grow gradually. However, she had a very strong aversion to seeking medical attention; she was stubborn and refused to visit a hospital. As the tumor increased in size, she remained at home, becoming increasingly reclusive. However, she eventually presented at the hospital because part of the tumor became ulcerated.

Upon admission, the tumor diameter was 40 cm, and the periphery was >80 cm. The base of the tumor seemed to stem from the right mandible to the lower edge of the sternal bone, and it hung down like a temple bell, expanding laterally and inferiorly (Fig. 1). Although the lateral lower edge was ulcerated, the patient did not feel pain or complain of dyspnea when lying down. She had noticed hoarseness in her throat a week earlier, but there were no signs of brachial plexus neuropathy.

On magnetic resonance imaging, the tumor border around the submandibular and parotid glands was unclear; however, the rest of the border was clear (Fig. 2).

On a computed axial tomography (CAT) scan, no mandibular bone invasion was evident, and the jugular artery and vein were compressed but did not show signs of invasion. Angiography showed that the tumor was vascularized mainly by 3 branches from the occipital, superficial temporal, and cervical thyroid arteries.

We performed a biopsy, and the tumor was edematous and sticky. The specimen revealed a myxoid stroma background, and the tumor cells developed in a trabecular or sheet pattern. The degrees of nuclear atypia and mitosis were low (6/10 high-power fields; Fig. 3). Immunohistochemical staining revealed that the tumor was positive for p63 and S-100, and the Ki-67 index was 18.5%. Based on these findings, myoepithelial carcinoma was diagnosed.

The tumor gradually increased in size even after admission. Initially, we thought that it was too large to resect and considered neoadjuvant therapies to reduce its volume. However, after considering the tumor’s characteristics (ie, outward expansion and compressed adjacent tissues without invasion) and pathological grade, we decided that resection was possible. At first, we planned to resect the tumor from the bottom and reconstruct the skin defect using an occipito-cervico-dorsal flap. However, we thought that the skin posterior to the equatorial plane of the tumor could be preserved because it seemed to be lengthened by the hanging tumor, but was not invaded.

We made an incision line posterior to the equatorial plane, keeping at least a 5- to 6-cm margin from the border of the invaded skin that was characterized by skin color change or nodule protrusion (Fig. 4). The tumor was encapsulated and had a clear border without invasion or adherence to the adjacent tissues, except for the submandibular gland from where it originated. The main vessels and nerves were preserved by alleviating the compression caused by the tumor, and the tumor was smoothly resected. The tumor weighed 10.5 kg.

To reconstruct the defect, we used the preserved skin, which was abundant around the caudal side of the defect, as a rotation flap, and we transferred the sternocleidomastoid muscle flap to fill the dead space in the submandibular area. We closed the skin defect, without needing to harvest any additional flaps, and performed multiple Z-plasty to prevent scar contracture on the neck (Fig. 4).

Postresection pathological findings showed that the tumor was encapsulated and that the capsule

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**Fig. 1.** The tumor on initial presentation. The base of the tumor appears to originate from the right mandible to the lower edge of the sternal bone, and the tumor hangs down like a temple bell, expanding both laterally and inferiorly. The diameter of the tumor is 40 cm, and the periphery is >80 cm. The lateral lower edge is ulcerated (↑).
close to the skin was partially infiltrated by round and spindle-shaped tumor cells. The tumors were attached to the submandibular gland, and a diagnosis of low-grade myoepithelial carcinoma originating from the submandibular gland was confirmed.

Radiotherapy was not performed because the tumor was encapsulated and the surgical margin was negative. The patient was discharged from the hospital 9 days postoperatively. Two years post surgery, she was no longer reclusive and her quality of life had improved greatly. She goes shopping and enjoys her life without recurrence or distant metastasis (Fig. 4).

DISCUSSION

Myoepithelioma was first described in 1943 by Sheldon,¹ and its malignant variant, myoepithelial carcinoma, was first described by Stromeyer et al in 1975. In 1991, it was included in the World Health Organization’s classification of salivary gland tumors, which recommends that malignant tumors are classified from low to high grade histologically depending on the tumor cell features (eg, mitosis and nuclear atypia).²,³,⁶ However, the clinical behaviors and histological features of these tumors vary.²,³ Even low-grade tumors can recur or metastasize.¹–³
and Hornick and Fletcher\(^6\) reported a 20% recurrence rate for low-grade tumors.

Treatment that includes wide surgical excision with a free margin is fundamental, and the role of chemotherapy and radiotherapy has not yet been established.\(^2,3\) Takayama et al\(^7\) reported on the efficacy of superselective intra-arterial chemotherapy infused with high-dose cisplatin, which could be adopted for cases that are difficult to manage surgically.

The massive myoepithelial carcinoma in our case required careful consideration to ensure proper treatment. Four cases of myoepithelial carcinoma that developed to >10 cm in size have been reported in the literature.\(^8-10\) In these cases, tumor size ranged from 13×15 cm to 20×15 cm; 3 originated from the parotid gland and 1 from the submandibular gland.\(^8-10\) Two were surgically resected, and the defects were reconstructed by free flap (ie, radial forearm and anterolateral thigh flap).\(^2\) The tumor in our case was twice the size of the largest tumor reported in the literature, and initially, we thought that some neoadjuvant therapies for volume reduction were necessary for safe manipulation around the neck area. We considered selective radiation of the tip of the tumor, superselective intra-arterial chemotherapy,\(^7\) and embolization of the branch to the distal segment of the tumor as possible treatment options for volume reduction. We had several discussions with the radiologist, oncologist, and pathologist regarding the best treatment plan for this case. With respect to the tumor characteristics, we thought that tumor could be virulent because it was massive and it kept increasing in size. However, pathologically, the degrees of nuclear atypia and mitosis were low, suggestive of a low-grade tumor. Magnetic resonance imaging and computed axial tomography scans suggested a clear tumor border and expansive tumor development without invasion because the mandibular bone and jugular artery and vein were compressed but their structure was maintained. Therefore, we deemed direct surgical excision possible, and the use of the lengthened skin as a local flap was considered as the best minimally invasive option. The tumor was encapsulated and was classified as low grade, and because the skin appeared to be lengthened by not only the tumor expansion but also the weight of the tumor, the use of this skin was deemed possible in our case. In addition, factors associated with recurrence such as tumor cell type, tumor size, and perineural and bone invasion are dependent on the tumor itself;\(^2\) therefore, as long as the tumor was successfully excised with a sufficient margin, surgical procedure for tumor removal was considered to be successful. Careful and cautious observation would still be required because of the aforementioned variations in tumor clinical behavior.\(^2,6\) Nevertheless, after careful evaluation of the tumor’s characteristics, we were able to accomplish a minimally invasive surgical intervention.

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**Fig. 4.** Intraoperative and postoperative views (A, incision design; B, view at the end of the operation; C, postoperative view). We maintained a 5- to 6-cm margin from the border of the invaded skin and designed the incision line posterior to the equatorial plane (▲) to preserve as much as possible of the intact lengthened skin (A). The skin defect was closed without harvesting any additional flaps from other body regions. Multiple Z-plasty was performed on the neck to avoid contracture (B). C, The image shows the patient at 2 years after the surgery. There is mild bulk on the submental area; however, there is no contracture on the neck, and she has been enjoying her daily life without tumor recurrence or metastasis.
CONCLUSIONS

We treated a massive myoepithelial carcinoma that was hanging from the jaw. Based on a careful evaluation of tumor characteristics and the results of imaging studies, we were able to excise the tumor directly and reconstruct the defect using the skin lengthened by the tumor as a local flap. When patients present with a massive tumor, treatment is assumed to be difficult; however, with an understanding of the nature of the tumor, regardless of its appearance, treatment can be much simpler than originally envisioned.

PATIENT CONSENT

The patient provided written consent for the use of her image.

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