Scientific Research Publishing

International Journal of Otolaryngology and Head & Neck Surgery, 2025, 14(3), 145-151
https://www.scirp.org/journal/ijohns
ISSN Online: 2168-5460
ISSN Print: 2168-5452

Low-Grade Myofibroblastic Sarcoma of the Tongue: Case Report and Review of Literature

Hua-Wei Shih[1], Chia-Jung Lee[1,2*]

[1]Department of Otolaryngology, Shin Kong Wu-Ho-Su Memorial Hospital, Taipei City
[2]School of Medicine, Fu Jen Catholic University, New Taipei City
Email: *gialon12@gmail.com

How to cite this paper: Shih, H.W. and Lee, C.J. (2025) Low-Grade Myofibroblastic Sarcoma of the Tongue: Case Report and Review of Literature. *International Journal of Otolaryngology and Head & Neck Surgery*, **14**, 145-151.
https://doi.org/10.4236/ijohns.2025.143017

Received: April 7, 2025
Accepted: May 17, 2025
Published: May 20, 2025

Abstract

Introduction: Low-grade myofibroblastic sarcoma (LGMS) is one of the ultra-rare malignant soft tissue tumors, with an annual incidence of less than 1 per 1,000,000. However, due to its rarity, there is currently no standardized treatment guideline. The therapeutic approaches remain under investigation with the ongoing accumulation of clinical experience. **Case Report:** A 55-year-old female with no significant medical history presented with a two-week history of a tongue mass and associated numbness. Physical examination revealed a 1.4 cm, firm, non-tender lesion on the ventral surface of the tongue. An excisional biopsy was performed, and the tumor was identified as an LGMS, characterized by spindle-shaped cells infiltrating adjacent skeletal muscle. Immunohistochemistry showed positive staining for α-SMA and negative results for desmin, cytokeratin, P63, CD34, S100, and Ki67. A PET-CT scan revealed focal FDG uptake in the tongue, likely related to postoperative changes, with no distant metastasis. The clinical stage was T1N0M0. In this report, a review of the literature was conducted, summarizing the interpretation of immunohistochemistry and discussing relevant studies on the treatment of LGMS. **Conclusion:** LGMS should be considered in the differential diagnosis of tongue tumors, with biopsy being crucial for confirming the diagnosis. Due to its rarity, there are no standardized treatment protocols, but surgical excision remains the primary approach. Given the risk of recurrence, regular follow-up is recommended.

Keywords

Low-Grade Myofibroblastic Sarcoma, LGMS, Ultra-Rare Malignant Soft Tissue Tumors, Malignant Neoplasm of Tongue

1. Introduction

Low-grade myofibroblastic sarcoma (LGMS) is classified as an ultra-rare soft tissue malignant tumor. Ultra-rare sarcomas, defined as an annual incidence of less than 1 per 1,000,000, poses challenges for well-powered prospective studies due to its rarity [1].

This tumor has been reported on various sites of the body. According to previous studies, the most common sites of LGMS vary, with some studies identifying the extremities and trunk as the most frequent location [2], while others suggest a higher prevalence in the head and neck region [3].

The diagnosis of LGMS depends on pathological examination of surgical specimens. Due to its rarity, there is no current standardized treatment guideline for LGMS. According to the literature, proposed treatment modalities include surgical intervention, radiotherapy, chemotherapy, and target therapy [4]-[6].

Here, we describe the clinical, histological, immunohistochemical, and radiographic features of an LGMS of the tongue and review the different managements of LGMS.

2. Case Report

A 55-year-old female with no significant systemic diseases presented to our clinic with a tongue mass accompanied by a numbness sensation for two weeks. She denied experiencing pain, and there was no discharge from the lesion. She reported no history of smoking, betel nut chewing, or alcohol consumption. She came to our hospital for help and physical examination revealed a palpable 1.4 cm tumor on the ventral surface of the tongue. The lesion was firm in consistency and nontender.

We performed the operation of excisional biopsy under local anesthesia. The tumor measured approximately 1.4 × 1.0 × 0.8 cm in size. It appeared white tan and was firm to the touch. Microscopically, the tumor was composed of spindle-shaped cells with slightly enlarged nuclei. The tumor cells were arranged in fascicles and infiltrated into the adjacent skeletal muscle. The mitotic index was 3 per square millimeters. Further immunohistochemical staining was performed for differential diagnosis. The tumor cells were positive for alpha-smooth muscle actin (α-SMA). However, they were negative for desmin, cytokeratin (CK), P63, CD34, S100, and Ki67 (**Figure 1**). Based on these pathological features, a diagnosis of LGMS was favored.

The patient experienced only mild postoperative pain with no other complications. A whole-body positron emission tomography-computed tomography (PET-CT) scan was performed for cancer staging (**Figure 2**). The imaging report only indicated focal increased fluorodeoxyglucose (FDG) uptake in the tongue, likely due to postoperative changes. There was no evidence of distant metastasis. The clinical stage was T1N0M0. Unfortunately, the patient had lost follow-up at our hospital.

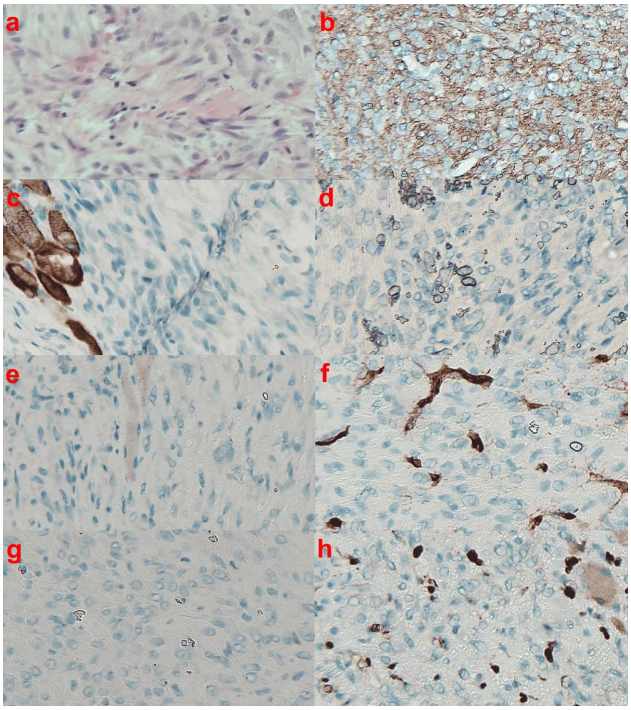

Figure 1. Histological staining and Immunohistochemistry. (a) Hematoxylin and eosin staining demonstrated that the tumor primarily consisted of spindle-shaped cells; (b) Immunohistochemically, LGMS showed positive staining for α-SMA and negative staining for; (c) desmin; (d) CK; (e) P63; (f) CD34; (g) S100; (h) Ki67 (a-h: ×200).

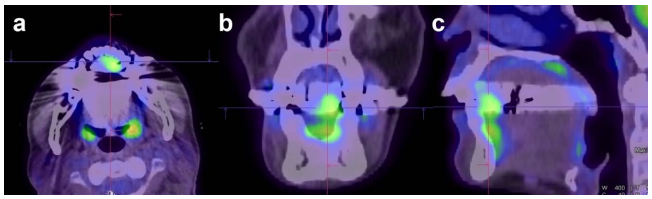

Figure 2. PET-CT reported clinical stage of T1N0M0. (a) Axial; (b) Coronal; (c) Sagittal view.

3. Discussion

LGMS is an ultra-rare malignant soft tissue sarcoma with a low prevalence [1]. It most commonly arises from the head and neck region or the extremities [2] [3] [7] [8]. LGMS can occur across a wide age range, with case reports documenting cases from 2 to 85 years old [9]. Clinically, it typically presents as a painless mass [10]. Definitive diagnosis requires histopathological examination through excisional biopsy.

LGMS exhibits diverse immunophenotypic profiles, including smooth muscle actin (SMA) positivity with desmin negativity, desmin positivity with SMA negativity, or co-expression of both markers [11]. On immunohistochemistry, the tumor cells of our patient were positive for α-SMA but negative for desmin, CK, CD34, and S100. We conducted a literature review on tongue LGMS and summarized the immunohistochemical staining results (**Table 1**). Desmin was positive

in 4 of 10 cases, and *α*-SMA was positive in 9 of 10 cases.

Table 1. Immunohistochemical summary of previously reported cases of LGMS in tongue.

Year	Location	Gender	Age	Size (cm)	α-SMA	desmin	CK	CD34	S100	References
2006	tongue	Female	24	2	+	-				[12]
2007	tongue	Male	41	1.5	+	+		-	-	[13]
2007	tongue	Male	53	2	+	-				[14]
2009	tongue	Female	61	2	+	-				[8]
2010	tongue	Male	56	4.2	-	+		-	-	[15]
2015	tongue	Male	74	4.1	+	-	-		-	[16]
2018	tongue	Female	38	1.5	+					[17]
2019	tongue	Male	41	2	+	+				[18]
2022	tongue	Male	73	4	+	+			+	[19]
2025	tongue	Female	55	1.4	+	-	-	-	-	Present case

LGMS are more likely to recur locally than metastasis to distant sites [20]. Although LGMS typically exhibits indolent growth, clinical follow-up is still necessary due to its associated recurrence rate. Maruyama *et al.* reported local recurrence and distant metastasis rates of 29% and 2%, respectively [21].

Because LGMS is very rare, a standardized treatment guideline has not yet been established. According to several previous studies, surgical excision may be the most effective therapy [4] [6] [7]. A 2021 study reported that surgical intervention was associated with significantly improved overall survival in LGMS patients, whereas the absence of surgery was a poor prognostic factor [6]. The role of radiotherapy and chemotherapy in LGMS is still uncertain. Some reports have documented notable clinical improvement with adjuvant chemotherapy in cases with metastases, leading to extended progression-free survival [22]. One systematic review concluded that adjuvant radiation therapy showed no benefit in reducing recurrence rates [23]. It has also been recommended to avoid radiotherapy after the resection of LGMS, as it may contribute to tumor recurrence [21]. Besides, target therapy with apatinib may be a potential treatment for LGMS [5]. Additional studies, incorporating long-term follow-up data and comprehensive clinical information, are needed to establish new treatment protocols for LGMS.

4. Conclusion

When encountering a tongue tumor, LGMS should be considered as a potential diagnosis, and a biopsy is essential to confirm the pathological diagnosis. There are no standardized treatment protocols for LGMS due to its rarity. According to current research, surgical excision is the primary treatment. Furthermore, given the potential for recurrence, it is recommended that patients undergo regular follow-up.

Ethical Approval

The study was reviewed and approved by the Ethics Committee of Shin Kong Wu-Ho-Su Memorial Hospital (Number: PJ2025032901).

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Stacchiotti, S., Frezza, A.M., Blay, J., Baldini, E.H., Bonvalot, S., Bovée, J.V.M.G., *et al.* (2021) Ultra-Rare Sarcomas: A Consensus Paper from the Connective Tissue Oncology Society Community of Experts on the Incidence Threshold and the List of Entities. *Cancer*, **127**, 2934-2942. https://doi.org/10.1002/cncr.33618

[2] Chan, J.Y.K., Gooi, Z., Wong, E.W.Y., Ng, S.K., Tong, M.C.F. and Vlantis, A.C. (2016) Low-Grade Myofibroblastic Sarcoma: A Population-Based Study. *The Laryngoscope*, **127**, 116-121. https://doi.org/10.1002/lary.26146

[3] Cai, C., Dehner, L.P. and El-Mofty, S.K. (2013) In Myofibroblastic Sarcomas of the Head and Neck, Mitotic Activity and Necrosis Define Grade: A Case Study and Literature Review. *Virchows Archiv*, **463**, 827-836. https://doi.org/10.1007/s00428-013-1494-1

[4] Kim, J., Choi, W., Cho, H.S., Lee, K.S., Park, J.K. and Kim, B. (2021) Surgical Treatment and Long-Term Outcomes of Low-Grade Myofibroblastic Sarcoma: A Single-Center Case Series of 15 Patients. *World Journal of Surgical Oncology*, **19**, Article No. 339. https://doi.org/10.1186/s12957-021-02454-5

[5] Lin, Y., Gao, X., Liu, Z., Liu, Z., Li, Y., Liang, R., *et al.* (2022) Effective Treatment of Low-Grade Myofibroblastic Sarcoma with Apatinib: A Case Report and Literature Review. *Pharmacogenomics and Personalized Medicine*, **15**, 573-582. https://doi.org/10.2147/pgpm.s359492

[6] Xu, Y., Xu, G., Wang, X., Mao, M., Wu, H., Baklaushev, V.P., *et al.* (2020) Is There a Role for Chemotherapy and Radiation in the Treatment of Patients with Low-Grade Myofibroblastic Sarcoma? *Clinical and Translational Oncology*, **23**, 344-352. https://doi.org/10.1007/s12094-020-02425-4

[7] Giraldo-Roldan, D., Louredo, B.V.R., Penafort, P.V.M., Pontes, H.A.R., Alves, A.P., Lima, F.C.A., *et al.* (2023) Low-Grade Myofibroblastic Sarcoma of the Oral and Maxillofacial Region: An International Clinicopathologic Study of 13 Cases and Literature Review. *Head and Neck Pathology*, **17**, 832-850. https://doi.org/10.1007/s12105-023-01577-3

[8] Demarosi, F., Bay, A., Moneghini, L. and Carrassi, A. (2009) Low-Grade Myofibroblastic Sarcoma of the Oral Cavity. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*, **108**, 248-254. https://doi.org/10.1016/j.tripleo.2009.03.031

[9] Yamada, T., Yoshimura, T., Kitamura, N., Sasabe, E., Ohno, S. and Yamamoto, T. (2012) Low-Grade Myofibroblastic Sarcoma of the Palate. *International Journal of Oral Science*, **4**, 170-173. https://doi.org/10.1038/ijos.2012.49

[10] Mocellin, S. (2020) Low-Grade Myofibroblastic Sarcoma. In: Mocellin, S., Ed., *Soft Tissue Tumors*, Springer International Publishing, 515-517. https://doi.org/10.1007/978-3-030-58710-9_160

[11] Mentzel, T. (2001) Myofibroblastic Sarcomas: A Brief Review of Sarcomas Showing

a Myofibroblastic Line of Differentiation and Discussion of the Differential Diagnosis. *Current Diagnostic Pathology*, **7**, 17-24. https://doi.org/10.1054/cdip.2000.0057

[12] Laco, J., Simáková, E., Slezák, R., Tucek, L., Mottl, R., Spacek, J. and Ryska, A. (2006) Low Grade Myofibroblastic Sarcoma of Tongue: A Case Report. *Ceskoslovenska patologie*, **42**, 150-153.

[13] Jay, A., Piper, K., Farthing, P.M., Carter, J. and Diwakar, A. (2007) Low-Grade Myofibroblastic Sarcoma of the Tongue. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*, **104**, e52-e58. https://doi.org/10.1016/j.tripleo.2007.06.004

[14] Meng, G., Zhang, H., Bu, H., Zhang, X., Pang, Z., Ke, Q., *et al.* (2007) Myofibroblastic Sarcomas: A Clinicopathological Study of 20 Cases. *Chinese Medical Journal*, **120**, 363-369. https://doi.org/10.1097/00029330-200703010-00003

[15] Mori, T., Shimane, T., Hayashi, T., Uzuki, A., Ikenoya, Y., Akiyama, R., *et al.* (2010) Low-Grade Myofibroblastic Sarcoma at the Base of the Tongue. *The Showa University Journal of Medical Sciences*, **22**, 239-243. https://doi.org/10.15369/sujms.22.239

[16] Gómez-Oliveira, G., Arribas-García, I., Serrano Alvarez-Buylla, A., Sánchez-Burgos, R., Martínez-Pérez, F. and Alvarez-Flores, M. (2015) Low-Grade Myofibroblastic Sarcoma. Two Rare Tumors in Two Rare Locations. *Revista Española de Cirugía Oral y Maxilofacial*, **37**, 108-112. https://doi.org/10.1016/j.maxilo.2013.04.004

[17] Mikami, Y., Fujii, S., Kohashi, K., Yamada, Y., Moriyama, M., Kawano, S., *et al.* (2018) Low-Grade Myofibroblastic Sarcoma Arising in the Tip of the Tongue with Intravascular Invasion: A Case Report. *Oncology Letters*, **16**, 3889-3894. https://doi.org/10.3892/ol.2018.9115

[18] Kawaharada, M., Katagiri, W., Maruyama, S., Nishiyama, H., Hayashi, T., Kobayashi, T., *et al.* (2021) Low-Grade Myofibroblastic Sarcoma of the Tongue with Difficulty of Diagnosis: A Case Report and Review of the Literature. *Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology*, **33**, 93-97. https://doi.org/10.1016/j.ajoms.2020.07.011

[19] Ab Rahman, N., Harun, M.H. and Sharif, S.E.T. (2022) Myofibroblastic Sarcoma of the Oral Cavity: A Diagnostic Dilemma and Report of Two Cases. *Journal of Oral Medicine and Oral Surgery*, **28**, 1-6. https://doi.org/10.1051/mbcb/2022010

[20] Agaimy, A., Wünsch, P.H., Schroeder, J., Gaumann, A., Dietmaier, W., Hartmann, A., *et al.* (2007) Low-Grade Abdominopelvic Sarcoma with Myofibroblastic Features (Low-Grade Myofibroblastic Sarcoma): Clinicopathological, Immunohistochemical, Molecular Genetic and Ultrastructural Study of Two Cases with Literature Review. *Journal of Clinical Pathology*, **61**, 301-306. https://doi.org/10.1136/jcp.2007.048561

[21] Maruyama, T., Nakasone, T., Nimura, F., Matayoshi, A., Kawano, T., Nishihara, K., *et al.* (2017) Indolent Growth of Low-Grade Myofibroblastic Sarcoma of the Cheek Mimics Benign Lesions: A Case Report and Literature Review. *Oncology Letters*, **13**, 4307-4314. https://doi.org/10.3892/ol.2017.6020

[22] Katalinic, D. and Santek, F. (2017) Giant Low-Grade Primary Myofibroblastic Sarcoma of the Posterior Chest Wall. *World Journal of Surgical Oncology*, **15**, Article No. 96. https://doi.org/10.1186/s12957-017-1167-7

[23] Mamikunian, G., Ziegler, A., Block, A. and Thorpe, E. (2023) Risk Factors for Recurrence and the Role of Radiotherapy in Low-Grade Myofibroblastic Sarcoma: A Systematic Review. *American Journal of Clinical Oncology*, **46**, 420-425. https://doi.org/10.1097/coc.0000000000001025

Abbreviations

LGMS: Low-grade myofibroblastic sarcoma
α-SMA: Alpha smooth muscle actin
SMA: Smooth muscle actin
CK: Cytokeratin
PET-CT: Positron emission tomography-computed tomography
FDG: Fluorodeoxyglucose