

## Case Report

# Langerhans cell sarcoma arising from the root of tongue: a rare case

Jian-Jun Ren<sup>1</sup>, Yu Zhao<sup>1</sup>, Ming-Juan Liu<sup>2</sup>, Guo Liu<sup>1</sup>, Fei Chen<sup>1</sup>

<sup>1</sup>Department of Oto-Rhino-Laryngology West China Hospital, West China Medical School, Sichuan University, Sichuan, China; <sup>2</sup>Department of Pathology West China Hospital, West China Medical School, Sichuan University, Sichuan, China

Received September 16, 2015; Accepted October 24, 2015; Epub November 1, 2015; Published November 15, 2015

**Abstract:** Langerhans cell sarcoma (LCS), a rare malignant disease with markedly malignant cytological features and poor outcome, originates from Langerhans cells and most commonly affects the lymph nodes, skin, and bone. This paper presents the case of a 58-year-old female with LCS at the root of her tongue, with neither local recurrence nor distant metastasis observed during 47 months of follow up following radiotherapy for more than one month after complete tumor resection. Histological and immunophenotypic tests revealed that the malignant tumor cells were positive for S-100 protein, CD1a, and LCA, and partially positive for CD3ε. By contrast, the tumor cells were negative for langerin, CD30, HMB45, PCK, CK5/6, and P63. Their Ki-67 proliferation index ranged from 30% to 40%. This neoplasm was diagnosed as LCS according to the classification of WHO2008. This work is the first report on LCS arising from the root of tongue. This rare case may serve as a reference for future clinical studies.

**Keywords:** Langerhans cell sarcoma, tongue, radiotherapy, immunohistochemical staining

### Introduction

Langerhans cell sarcoma (LCS), which exhibits malignant cytological features, aggressive clinical course, and generally poor prognosis, is a rare neoplastic condition. Langerhans cell tumors are classified by the World Health Organization (WHO) into LCS and Langerhans cell histiocytosis [1]. Fewer than 50 cases, which mostly occurred in lymph nodes, skin, and bone, have been reported worldwide. The average age of presentation is 50, and the overall male to female ratio is 1.3:1 [2]. In this paper, we report a rare case of LCS, which is the first case of LCS to develop at the root of the tongue.

### Case report

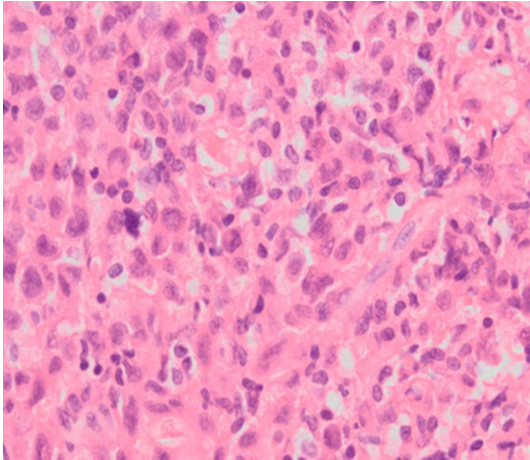
A 58-year-old woman presented to the hospital with a more than two-month history of pharyngalgia in 2011. Two months before her hospitalization, the patient experienced persistent pharyngalgia without known cause. She also experienced difficulty in swallowing and a radiating pain in the ear but without any other symptoms, such as fever, chills, and dyspnea.

The enlarged superficial lymph nodes in the patient were not palpable, and no tumor metastasis was observed. The patient was generally healthy and had no other diseases, except for high blood pressure.

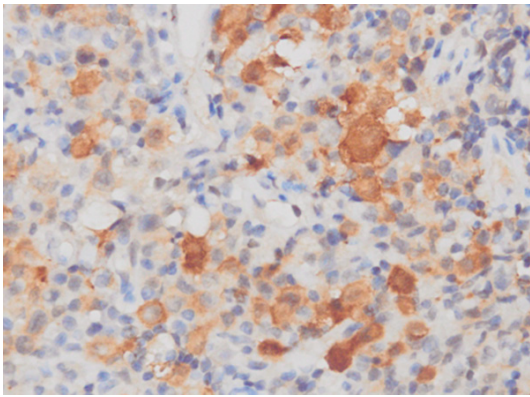
On 13 August 2011, she was diagnosed with squamous cell carcinoma of the tongue through flexible laryngoscopy and pathological examination in another hospital. However, no recovery was gained after relevant treatment. Therefore, the patient was referred to our hospital, where we performed another flexible laryngoscopy, which revealed a tongue root neoplasm. The biopsy revealed a small amount of ground tissues of the tongue root, chronic mucositis, and hyperblastosis of subcarinal lymph nodes of the mucosal epithelium, some of which were extruded. No tumor was observed. The preoperative examination was completed after the patient was hospitalized.

On 9 September 2011, neoplasm resection was performed using suspension microlaryngoscopy. Incisional biopsy was also performed during the surgery. The operation revealed a 2 cm × 3 cm soft ulcerative neoplasm located at

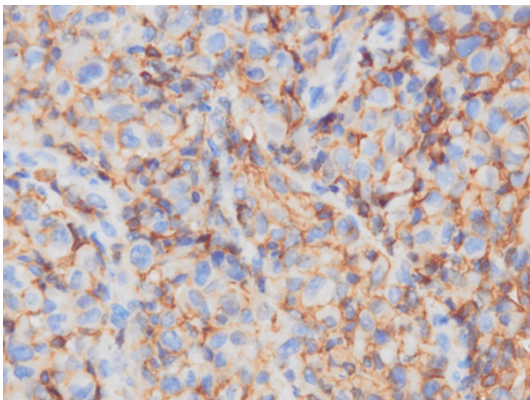
## Langerhans cell sarcoma arising from the root of tongue



**Figure 1.** Hematoxylin-eosin staining of LCS (100×).

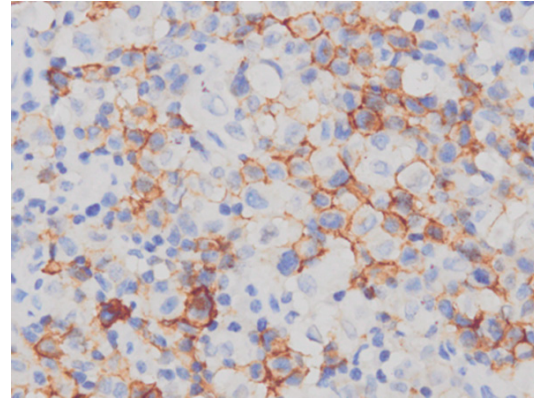


**Figure 2.** LCS tumor cells expressed S-100 protein (400×).

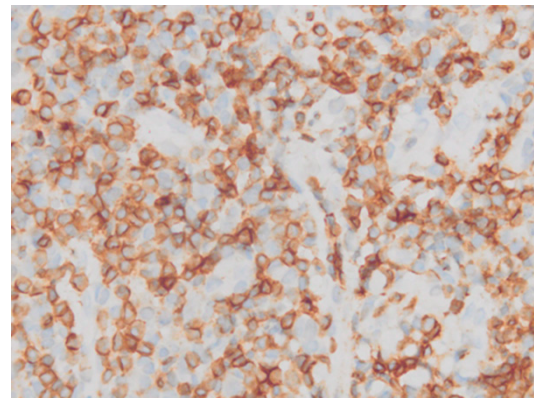


**Figure 3.** LCS tumor cells expressed LCA (400×).

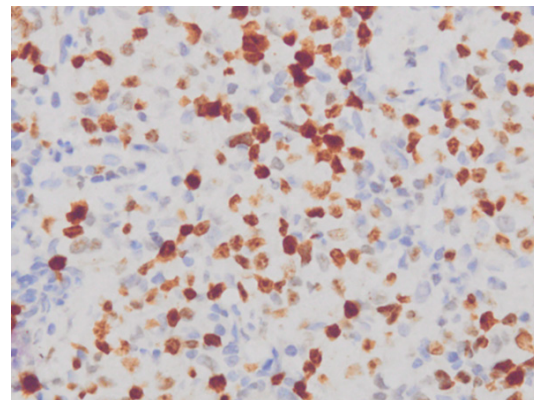
the left side of the tongue root, with poor border and slight bleeding. This neoplasm was removed completely, and thorough hemostasis was conducted.



**Figure 4.** LCS tumor cells expressed CD1a (400×).



**Figure 5.** LCS tumor cells partially expressed CD3ε (400×).



**Figure 6.** Ki-67 proliferation index ranged from 30%~40% (400×).

We adopted postoperative anti-inflammatory treatment. One month later, the patient was transferred to the oncology ward for radiotherapy. Enhanced computed tomography scan of the chest, enhanced magnetic resonance

## Langerhans cell sarcoma arising from the root of tongue

imaging of nasopharyngeal carcinoma, and abdominal color Doppler ultrasound were performed. Positioner radiotherapy against the disease was applied. The patient did not report any discomfort and was discharged on 10 October 2011. Regular re-examination was conducted. The results of telephone follow up indicated that the patient's health and mental status were good, and no evidence of local or distant tumor recurrence or metastasis was observed for nearly four years.

### Pathological findings

Intraoperative frozen section examination showed that the resected specimen was likely a diffuse large cell lymphoma or poorly differentiated carcinoma. Furthermore, wax test and immunohistochemical staining showed mucosal ulceration with necrosis at the tongue root. Hematoxylin and eosin staining revealed Patchy cytological atypia cells infiltration and squamous hyperplasia, which was suspected to be a malignant tumor (**Figure 1**). Immunohistochemical studies demonstrated that the malignant tumor cells were positive for S-100 protein (**Figure 2**), LCA (**Figure 3**), and CD1a (**Figure 4**) and partially positive for CD3ε (**Figure 5**). By contrast, the tumor cells were negative for langerin, CD30, HMB45, PCK, CK5/6, and P63. The Ki-67 proliferation index ranged from 30% to 40% (**Figure 6**). Based on the histological and immunophenotypic findings and the WHO2008 classification, this neoplasm was diagnosed as LCS [3].

### Discussion

Langerhans cells, which are epithelial resident dendritic cells, serve as the "sentinels" of the mucosa and alter the immune system in response to pathogen entry and to establish tolerance to self-antigen and commensal microbes [4]. These cells are normally present in the suprabasal squamous epithelium of the skin and mucous membranes [5]. Oral mucosal Langerhans cells can engage and internalize a wide variety of pathogens [4]. Langerhans cell proliferation was first described by Langerhans in 1868. The first LCS case was reported in 1984 by Wood et al. [6]. LCS can be diagnosed through histological and immunohistological tests.

According to the literature, most cases are characterized by poor prognosis and short sur-

vival time, illustrating a lack of effective treatments. Lucas et al. [7] reported that surgery is a good option to treat isolated lesions or confined nodal disease, although the benefit of adjuvant treatment is unknown. Furthermore, they found that LCS responds well to radiotherapy, which might be useful as a local treatment, at least for pain relief and symptom control. They also suggested that irradiation is a good option in cases of symptomatic or bulky lesions. Similarly, Nakayama [8] suggested that radiotherapy is the most effective treatment for localized LCS, although further accumulation of LCS cases is unquestionably necessary to establish the optimal treatment strategy.

In the present case, the malignant tumor was solitarily localized in the root of the tongue, and the patient needed to undergo radiotherapy for local control after removal of the neoplasm. The patient had undergone radiotherapy for more than one month, with a total dose of 50 Gy. No recurrence or metastatic signs were observed for 47 months without any chemotherapy. Based on our experience, localized LCS without any lymphovascular invasion or distant metastasis should be treated with radiotherapy because it results in better curative effects compared with chemotherapy.

LCS cases occurring in the tongue root remain rare, and the case reported in this paper may serve as a reference for future clinical practice.

### Acknowledgements

This work was supported by the National Natural Science Foundation of China (Grant No. 81170898) to Yu Zhao.

### Disclosure of conflict of interest

None.

**Address correspondence to:** Dr. Yu Zhao, Department of Oto-Rhino-Laryngology, West China Hospital, West China Medical School, Sichuan University, 37 Guo Xue Alley, Chengdu 610041, Sichuan, China. Tel: +86 1398210 3388; Fax: +86 028 85422433; E-mail: yutzhao@163.com

### References

- [1] Weiss LM, Grogan TM, Pileri SA, et al. Langerhans cell sarcoma. In: Jaffe ES, Harris

## Langerhans cell sarcoma arising from the root of tongue

- NL, Stein H, Vardiman JW, editors. World Health Organisation classification of tumours. Pathology & genetics: tumours of haematopoietic and lymphoid tissues. Lyon, France: IARC Press; 2001. pp. 283.
- [2] Howard JE, Dwivedi RC, Masterson L, Jani P. Langerhans cell sarcoma: A systematic review. *Cancer Treat Rev* 2015; 41: 320-331.
- [3] Jaffe E, Harris N, Stein H, et al. Tumors derived from Langerhans cells. In World Health Organization classification of tumours: Pathology and genetics, tumours of haematopoietic and lymphoid tissues. In: Barnes L, Eveson JW, Reichart P, editors. Sidransky D. Lyon: IARC Press; 2008. pp. 278-289.
- [4] Upadhyay J, Upadhyay RB, Agrawal P, Jaitley S, Shekhar R. Langerhans Cells and Their Role in Oral Mucosal Diseases. *N Am J Med Sci* 2013; 5: 505-14.
- [5] Ferringer T, Banks PM, Metcalf JS. Langerhans Cell Sarcoma. *Am J Dermatopathol* 2006; 28: 36-39.
- [6] Wood C, Wood GS, Deneau DG, Oseroff A, Beckstead JH, Malin J. Malignant histiocytosis X. Report of a rapidly fatal case in an elderly man. *Cancer* 1984; 54: 347-352.
- [7] Lucas A, Barca EG, Servitje O. Langerhans cell sarcoma: Response to radiotherapy. *Rep Pract Oncol Radiother* 2010; 15: 107-109.
- [8] Nakayama M, Takahashi K, Hori M. Langerhans cell sarcoma of the cervical lymph node: A case report and literature review. *Auris Nasus Larynx* 2010; 37: 750-3.