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CASE REPORT

PRIMARY LEIOMYOSARCOMA OF TONGUE - A RARE NEOPLASM

Avani Jain[1], Shashank Nath Singh[2], Pawan Singhal[3], Manprakash Sharma[4], Sunil Samdani[5]

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ABSTRACT: BACKGROUND: Primary leiomyosarcoma of the tongue is an extremely rare malignant mesenchymal tumour that exhibits smooth muscle differentiation. Only 23 cases of this neoplasm have been described in the literature. **CASE REPORT**: This article describes a case report of a primary leiomyosarcoma of the right lateral border of the tongue in a 35-year-old male. The diagnosis was supported by histological findings and immunohistochemical positivity for desmin, vimentin and smooth muscle actin. Hemiglossectomy with supraomohyoid neck dissection was done with three years follow-up, without evidence of local recurrence or distant metastasis. The available literature has been reviewed for a better understanding of this rare neoplasm. **CONCLUSION**: Leiomyosarcoma in this unusual site seems to have a better prognosis than elsewhere, although the number of cases are too small to draw a conclusion. Further research with longer follow-up is required to better understand this rare pathological entity.
KEY WORDS: Leiomyosarcoma, tongue, immune histochemistry.

INTRODUCTION: Leiomyosarcoma is a rare malignant mesenchymal neoplasm arising from smooth muscles. It accounts for 6-7% of all soft tissue sarcomas and arises mainly in the uterus, gastrointestinal tract and retroperitoneum[1]. Only 3-10% of leiomyosarcomas arise from head and neck region; and its occurrence in the tongue, is even lower owing to the paucity of smooth muscles in the region[2]. There are only 23 reported cases in the literature of primary leiomyosarcoma arising from the tongue[2-21]. This article describes an additional case of primary leiomyosarcoma of the tongue, its immunohistochemical features, managment and a review of literature.

CASE REPORT: A 35-year -old male presented to our tertiary care referral centre with a 4 months history of a lesion at right lateral border of the tongue. The lesion was slow growing and painful. Clinical examination revealed a reddish brown mass on right lateral border of tongue measuring approximately 2x2cm (Figure 1). It was a well circumscribed firm nodule that was tender. There were no other oropharyngeal lesions or palpable cervical lymph nodes. The patient denied any previous traumatic injury history or betel nut chewing habit. There was no relevant medical or social history.

A punch biopsy was performed and histopathology revealed a malignant mesenchymal neoplasm (Figure 2). On performing immunohistochemistry, tumour cells stained positively with smooth muscle actin (Figure 3), vimentin and desmin. Stains for epithelial marker (CK AE3/AE1), melanoma marker (S100), CD34 and factor VIII were negative. The histopathological features of the specimen in conjunction with the immunohistochemical staining pattern were consistent with leiomyosarcoma.

The chest radiograph was normal and magnetic resonance imaging indicated no disease in the neck. Right hemiglossectomy and supraomohyoid neck dissection was performed under general

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anaesthesia. Histopathological examination showed the peripheral resected margins to be tumour free and the 12 resected lymph nodes were negative for malignancy. The patient was followed up monthly for a year, 3 monthly for the next year and 6 monthly thereafter, with no evidence of recurrent disease and no further treatment in the form of radiotherapy/chemotherapy.

DISCUSSION: Leiomyosarcoma is an exceptionally rare malignant mesenchymal neoplasm in the oral cavity owing to the paucity of smooth muscles in this region. Within the oral cavity, maxilla is the most common site involved followed by the mandible, tongue, cheek and floor of mouth, in descending order[17].

The cause of leiomyosarcoma remains uncertain, although cases associated with trauma, oestrogen stimulation, ionizing irradiation and the Epstein-Barr virus have been reported[22]. Several theories have been proposed on the origins of primary leiomyosarcoma of the tongue. Since only striated muscle is found in the oral cavity, such tumours probably result from the smooth muscle of the walls of blood vessels or of the circumvallate papillae of the tongue[20]. Other proposed sites of origin are arteriovenous anastomosis and thyroglossal tissues[2, 5, 12].

Clinically, leiomyosarcoma often presents as a discrete, painless, slowly enlarging mass that is firmly adherent to surrounding soft tissue[21]. Histologically, the tumour is locally aggressive and highly infiltrating[23], often leading to local recurrence and haematogenous metastasis to cervical and pulmonary regions[24]. Both histological examination and immunohistochemistry staining are essential for the final diagnosis of oral leiomyosarcoma, as histology alone cannot differentiate between the different mesenchymal tumours. The histological criteria for diagnosis include the presence of nuclear pleomorphism, bizarre cell forms, a pattern of interlacing bundles of smooth muscle cells, and a high rate of mitosis (>5 mitotic figures/high power field)[21]. Immunohistochemical staining of leiomyosarcoma has consistently demonstrated a positive response to smooth muscle actin (SMA), vimentin and desmin. Whereas, a negative response to the S100 protein and the cytokeratins is usually found[2, 7].

Once diagnosed, wide local excision of the tumour with tumour free margins and regional lymph node dissection when required seems the most appropriate treatment that may improve long term survival[21]. The benefits for treating leiomyosarcoma of the tongue with chemotherapy and radiotherapy remain uncertain and there is insufficient evidence to support their efficacy. Only one case has been reported to date, that was treated with radiotherapy alone, owing to surgically inoperable leiomyosarcoma at the base of the tongue; with 1.5 years follow-up, no local recurrence or distant metastasis was noted[7]. Chemotherapy alone for the treatment of leiomyosarcoma tongue has been reported in only one case in literature, in which owing to lung and multiple soft tissue metastasis, palliative treatment seemed more appropriate. A combination of ifosfamide and doxorubicin was used, following which there was a marked decrease in the primary tongue lesion[21]. Further research is required to determine the efficacy and prognostic benefit of adjuvant therapy in the management of leiomyosarcoma of tongue.

Table I summarizes all the 24 reported cases, including the present case, of primary tongue leiomyosarcoma[2-21]. Based on these studies, the most appropriate treatment is wide local excision of the tumour with histologically proven tumour-free margins. However, the number of reported cases of primary leiomyosarcoma of the tongue is small, and further research and long term follow-up is

required to better understand the behaviour, risk factors and predictors of treatment outcome to improve the long term survival of this rare neoplasm.

SUMMARY:

- Leiomyosarcomas are malignant tumours of smooth muscles and hence, are rare in the head and neck, especially the tongue.
- Diagnosis is confirmed by histological examination and immunohistochemical positivity for vimentin, desmin and smooth muscle actin.
- Wide local excision of the tumour with tumour free margins and regional lymph node dissection when required is the treatment of choice.
- Low reported recurrence rate and survival data suggests a better prognosis, however, further case experience and research is required to better understand this rare neoplasm.

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STUDY	YEAR	GENDER	AGE (years)	SITE	METASTASIS	TREATMENT	FOLLOW UP (years)
Yannopoulos and Stout[3]	1962	M	11mo	Tip of tongue	No	Excision	4-6
O'Day et al[4]	1964	M	3	Sublingual	Yes	Excision	2, deceased
O'Day et al[4]	1964	F	19	Sublingual	Yes	Excision and neck dissection	11
Goldberg et al[5]	1970	M	54	Sublingual	No	Excision	1.5
Lack[6]	1986	M	2.5	Base of tongue	No	Excision+ CT	4
Aydin and Dreyer[7]	1994	M	70	Base and dorsum of tongue	No	RT	1.5
Mayall et al[8]	1994	M	60	Tip of tongue	No	Excision	1
Piattelli and Artese[2]	1995	F	80	Lateral border tongue	No	Patient rejected treatment	N/A
Tandon et al[9]	1996	M	22	Base of tongue	No	Excision	5
Gorsky and Epstein[10]	1998	M	57	Lateral border of tongue	Not recorded	Excision + RT	4, deceased

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Dry et al[11]	2000	F	15	Not specified	No	Excision + CT	4
Dry et al[11]	2000	F	88	Floor of mouth	Yes	No treatment	Deceased
Lo Muzio et al[12]	2000	M	67	Lateral border of tongue	No	Excision	5
Sakamoto et al[13]	2005	M	67	Tip of tongue	No	Excision	Not recorded
Kazemian et al[14]	2005	M	32	Lateral border of tongue	Yes	Excision + MND + CT	1.5, deceased
Yang et al[15]	2006	F	57	Lateral border of tongue	No	Excision + SOND	3
Yang et al[16]	2006	F	54	Tip of tongue	No	Excision	1
Ethunandan et[17] al	2007	F	79	Not specified	No	Excision	2.5
Ethunandan et al[17]	2007	F	97	Not specified	No	Excision	4.5
Crossman et al[18]	2008	F	46	Lateral border of tongue	No	Excision	5
Yan et al[19]	2010	M	48	Floor of mouth	No	Excision + SOND	1.5, deceased
Pires et al[20]	2010	M	55	Lateral border of tongue	No	Excision	4
Ahn et al[21]	2011	F	54	Lateral border of tongue	Yes	Palliative CT	1 mo
Present case	2012	M	35	Lateral border of tongue	No	Excision + SOND	3

TABLE 1: CASESOFPRIMARYLEIOMYOSARCOMATONGUE

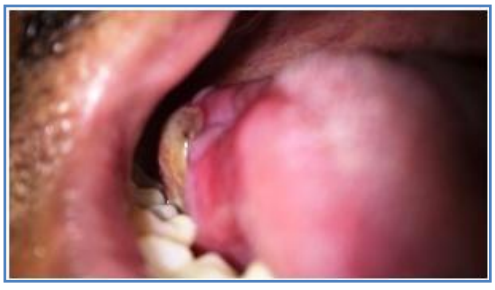

Fig. 1: Clinical photograph of the lesion involving the right lateral border of tongue.

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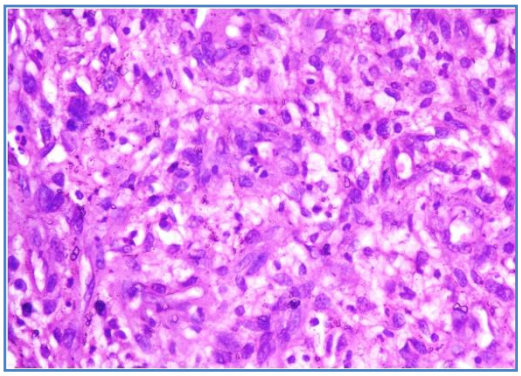

Fig. 2: Photomicrograph of the lesion showing interlacing spindle cell bundles demonstrating cellularpleomorphism and mitosis (H&E, x40).

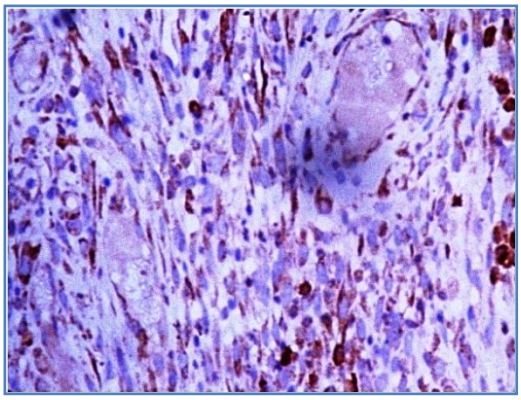

Fig. 3:Photomicrograph of the lesion showing diffuse staining with smooth muscle actin (x40).

AUTHORS:

1. Avani Jain
2. Shashank Nath Singh
3. Pawan Singhal
4. Manprakash Sharma
5. Sunil Samdani

PARTICULARS OF CONTRIBUTORS:

1. Junior Resident, Department of Otolaryngology and Head and Neck Surgery, Sawai Man Singh Medical College and Hospital, Jaipur, Rajasthan, India.
2. Assistant Professor, Department of Otolaryngology and Head and Neck Surgery, Sawai Man Singh Medical College and Hospital, Jaipur, Rajasthan, India.
3. Associate Professor, Department of Otolaryngology and Head and Neck Surgery, Sawai Man Singh Medical College and Hospital, Jaipur, Rajasthan, India.
4. Professor, Department of Otolaryngology and Head and Neck Surgery, Sawai Man Singh Medical College and Hospital, Jaipur, Rajasthan, India.
5. Department of Otolaryngology and Head and Neck Surgery, Sawai Man Singh Medical College and Hospital, Jaipur, Rajasthan, India.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr.Avani Jain,
Flat No. C-8, Tower 1,
New MotiBagh,
New Delhi – 110023, India.
Email-avanijain87@hotmail.com

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