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Case Report

Primary leiomyosarcoma of tongue: A relatively uncommon entity occurring at an exceedingly rare site

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ABSTRACT

Primary leiomyosarcoma (LMS) of the tongue is an extremely rare, highly aggressive malignancy associated with poor survival outcome. We report a case of 60 years old male who presented with growth left lateral border of tongue since 6 months. Histopathological examination of the excised growth confirmed the diagnosis of LMS. An exhaustive panel of immunohistochemical markers helped in ruling out other malignant spindle cell differentials.

1. Introduction

Leiomyosarcoma of the tongue is a relatively rare non-epithelial highly malignant sarcoma of myogenic origin with high propensity for local recurences and metastasis.[1]

Leiomyosarcomas accounts for 10-15% of all soft tissue sarcomas, and are divided into three major categories for therapeutic and prognostic purposes: somatic soft tissue LMS, cutaneous LMS, and vascular LMS.[2–4] Primary LMS of tongue is an extremely rare tumor associated with high mortality and poor survival outcomes. So far very few cases have been reported in the literature. Thus, we are reporting this very rare case of primary LMS of the tongue as this neoplasm poses a significant diagnostic challenge for both clinicians and pathologists.

2. Case History

A 60-year-old male presented with a short term history of slurring of speech and difficulty in chewing due to a large growth at left lateral border of the tongue to the otorhinolaryngology outpatient department of our institute. Initial biopsy of the lesion was non conclusive. Later on patient underwent partial glossectomy with wide free margins and specimen was sent for histopathological examination. Grossly the specimen revealed a large proliferative growth at the left lateral border of the tongue with infiltrative margins. Cut surface of the tumor was homogenously greyish –white with foci of nerosis (Figure 1).

Microscopic examination showed squamous epithelium which was focally ulcerated and replaced by dense fibrinopurlent exudate with enmeshed bacterial colonies. Underlying subepithelial tissue srevealed a malignant spindle cell tumor arranged in fascicles & swirls.

Neoplastic cells were having plump spindled, fusiform nuclei with vesicular chromatin, single to multiple prominent nucleoli and moderate amount of eosinophilic cytoplasm. Tumor showed marked nuclear pleomorphism, brisk mitosis, areas of coagulative necrosis and mixed inflammatory cell infiltrate (Figure 2 a, b).

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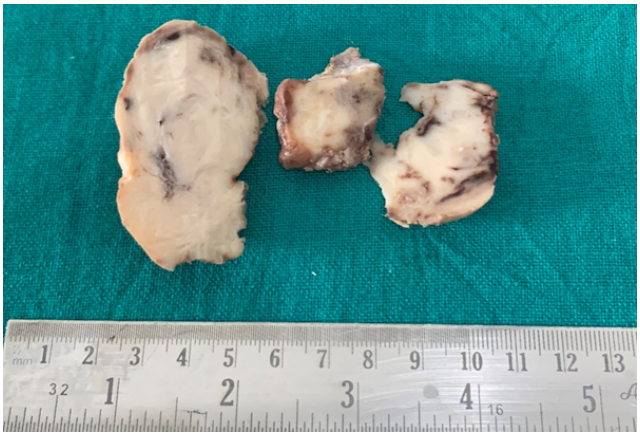

Fig. 1: Gross photograph showing a large homogenous greyish-white growth at the lateral border of tongue with few foci of coagulative necrosis

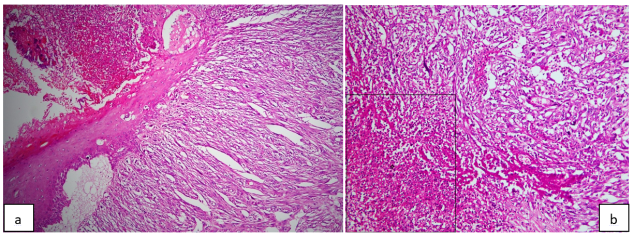

Fig. 2: a): Photomicrograph revealing a malignant spindle cell proliferation just beneath the overlying stratified squamous epithelium which is focally ulcerated; **b):** higher magnification showing marked nuclear pleomorphism and coagulative necrosis (inset), H&E x 400

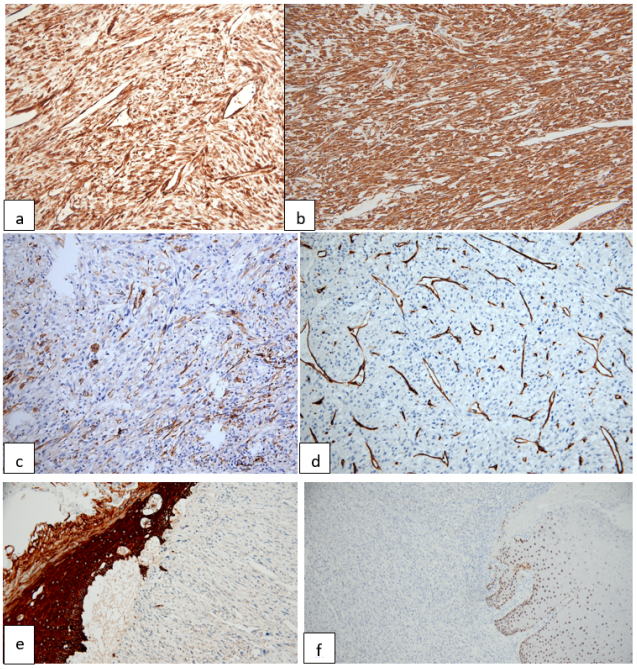

Fig. 3: Photomicrograph showing diffuse & strong staining for SMA (**a**), Vimentin (**b**), Focal h- Caldesmon positivity (**c**), rich vasculature of the tumor beautifully highlighted by CD34 immunostain (**d**), and tumor cells show complete absence of AE1/AE3 & p63 expression (**e & f**)

On immunohistochemistry (IHC), neoplastic spindle cells showed strong and diffuse positivity for Vimentin and SMA whereas variable staining was observed for Desmin, h-Caldesmon, and S-100. Neoplastic cells were completely negative for AE1/AE3, CK7, HMB-45, and p63. CD 34 beautifully highlighted the rich vasculature of the tumor (Figure 3 a-f).

Ki-67/MIB-Index was high reaching approximately 45% in the most proliferative areas (Figure 4).

The diagnosis of primary leiomyosarcoma of the tongue was confirmed on the basis of characterstic histopathological features in conjuction with a panel of IHC markers and the patient was referred to the radiation oncology unit of the hospital for necessary neoadjuvant therapy/palliative radiotherapy. Patient was lost to follow-up thereafter.

3. Discussion

Primary leiomyosarcoma of the tongue is an exceedingly rare, highly aggressive malignancy associated with higher mortality and poor survival outcomes. Within the oral

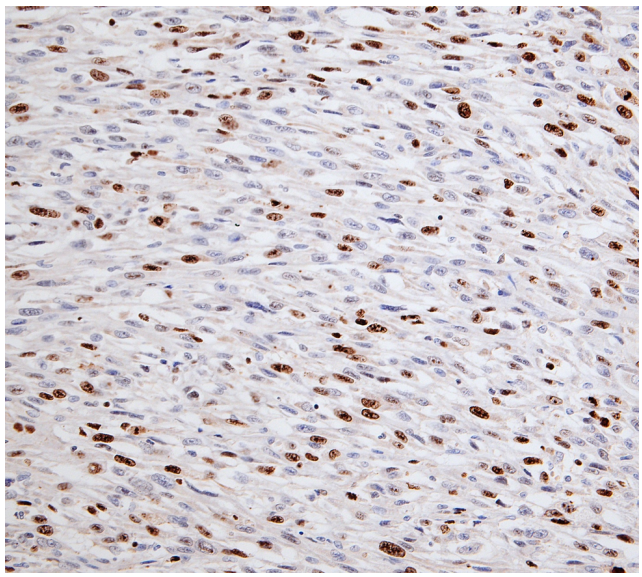

Fig. 4: High Ki-67/MIB-1 index reaching approximately 45% at hot spots

cavity buccal mucosa is the common site and tongue is very rare site of occurrence.[5] Barnes et al. reported that the most common sites of head and neck region are oral cavity (22%), the sinonasal tract(19%), and the facial subcutis(17%).[6] Primary LMS of tongue poses a significant diagnostic challenge because of its rarity at this site, however extensive sampling with multiple section examination in cojuction with a panel of immunohistochemical markers helps in conforming the accurate diagnosis. The chief differentials include spindle variant of squamous cell carcinoma, malignant peripheral nerve sheath tumor (MPNST), rhabdomyosarcoma, malingnant melanoma and monophasic synovial sarcoma. The characterstic histopathological features of leiomyosarcoma include sweeping fascicles and whorls of highly atypical spindled cells having fusiform, plump vesicular nuclei with single to multiple prominent nucleoli and moderate amount of eosinophilic cytoplasm along with many foci of coagulative necrosis and brisk mitosis.

IHC panel of markers including PANCK, HMB-45, S-100, SMA, Desmin, Vimentin, EMA, p63, p40 and proliferation marker KI-67 index helped in ruling out the differentials in our case with results discussed in the case history, thus confirming the diagnosis of LMS tongue.

The only effective treatment of this aggressive tumor is complete resection with free margins and post-operative chemotherapy/radiotherapy if needed.[7–10]

4. Conclusion

Our case concluded that because of extreme rarity of LMS of the tongue it can pose a diagnostic challenge to both clinicians and pathologists, however in the hands of keen pathologist in conjuction with a panel of IHC markers definitive diagnosis is possible; thus guiding the management plan of the patient without undue delays.

5. Source of Funding

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6. Conflicts of Interest

None.

7. Ethics Approval

Approved by the institutional ethical commiettee.

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