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CASE REPORT

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Well-differentiated liposarcoma of tongue: A case report

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Key Clinical Message

It is important to consider WDLS as a potential cause of tongue lesions and include it in the list of differential diagnoses. When performing surgical intervention, it is crucial to remove enough tissue around the lesion, and regular follow-up is necessary due to the high risk of recurrence, despite its rarity, when margins are positive.

Abstract

Liposarcoma (LS) is the most common soft tissue sarcomas (STSs) that arise from embryonic mesenchymal tissue. Though these sarcomas commonly arise at retroperitoneal locations and extremities, the appearance of these tumors in the head and neck region is rare, with the tongue as a preferred site. As per WHO 2020, LS is classified into four subtypes based on morphology, namely, Well-differentiated liposarcoma (WDLS), Dedifferentiated liposarcoma (DDLS), Myxoid liposarcoma (MLS), and Pleomorphic liposarcoma (PLS). WLS is the most common variant among all. Here, we had a case of 55 years old male with the complaint of swelling in the left lateral border of the tongue with the preliminary diagnosis of pleomorphic adenoma. The patient underwent a left partial glossectomy with adequate margins. Further evaluation of the lesion revealed a clear cell tumor that was ultimately confirmed as liposarcoma on immunohistochemistry that showed tumor cells positive for S100, CDK4, and MDM2 with 2% Ki-67. Postsurgical status of the patient was evaluated by F18 FDG PET CTscan, which was normal. Currently, the patient is under regular follow-up.

KEYWORDS

liposarcoma, pleomorphic adenoma, tongue, well differentiated

1 | INTRODUCTION

Liposarcoma (LS) is the most common soft tissue sarcomas (STSs) that usually arises in retroperitoneal area and extremities.[1] It's rare to find the occurrence of these sarcomas in head and neck region, including oral cavity.[2] The recent classification of WHO has divided liposarcoma into four subtypes, namely Well-differentiated liposarcoma (WDLS)/ Atypical lipomatous tumor (ALT), Dedifferentiated liposarcoma (DDLS), Myxoid liposarcoma (MLS), and Pleomorphic liposarcoma (PLS).[3] WDLS, the most common variant among the four, is mostly indolent and has a lower tendency to metastasize.

2 | CASE REPORT

A 55 year male presented to the head and neck oncology department with complaints of swelling in the left lateral border of the tongue for the last 4 years along with a cytopathology report of FNAC suggesting the diagnosis of Pleomorphic Adenoma of the left lateral border of tongue. To begin with, the swelling initiated as a small nodule which gradually increased to its present size over the period of 4 years. There was no history of trauma, pain, burning sensation, difficulty in swallowing and change in speech quality. There was no significant past and family history.

The vitals were normal at the time of arrival. On examination, there was a firm nodular swelling of 2 × 1.5 cm size located in the left lateral border of the tongue with normal mucosa (Figure 1). There was no ulceration and no signs of inflammation over the swelling. Similarly, lymph nodes were not palpable in the neck.

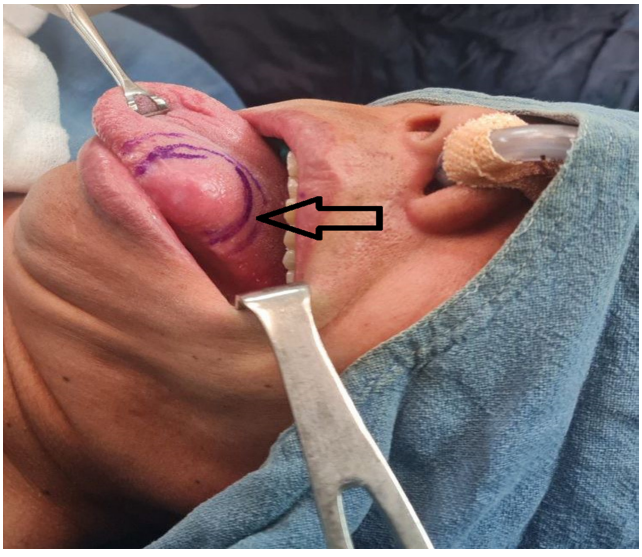

FIGURE 1 A nodular lesion located at left lateral border of tongue.

Magnetic Resonance Imaging (MRI) of the tongue was ordered that showed a well-defined, oval-shaped, soft tissue lesion within the tongue on the left side (Figure 2A, B). This post-gadolinium-enhanced lesion measured about 18 × 19 mm in size and was seen about 19 mm distal to the tip of the tongue (Figure 2C). No evidence of restricted diffusion was noted. The lesion was seen extending to the distal edge of the tongue with no evidence of extension across the midline, into the surrounding tissue and overlying teeth and bone.

Based on the preliminary diagnosis, the patient was admitted to the hospital and scheduled for elective surgery for the removal of the lesion. The preanesthetic evaluation revealed normal vitals and examination findings. All hematology (Complete Blood Count, Prothrombin Time), biochemistry (Liver function Test, Renal Function Test) and serology (for HIV, Hepatitis B and Hepatitis C) were normal. A real-time RT-PCR was negative for SARS CoV2. The patient underwent a left partial glossectomy to remove the lesion under general anesthesia. Whole of the lesion was excised intra-orally, taking adequate margin all around. Intraoperative and postoperative periods were uneventful. Post-operatively patient was managed conservatively.

On gross examination, specimen of size 5 × 3.4 × 2.5 cm was received (Figure 3). It was well-circumscribed, gray-white, tanned solid mass with unremarkable mucosal findings. The tumor was unifocal and located at the left lateral border of the tongue with a size of 2.3 × 2 × 2.2 cm. Grossly, all mucosal, soft tissue, and deep margins (anterior, posterior, lateral, medial, superior, inferior, and deep) were uninvolved by the tumor. The distance from the closest mucosal margin was 0.7 cm (anterior), and from the deep margin was 0.2 cm.

Under microscopy, the sections showed sub-mucosal circumscribed nodular lesion composed of clear cells of variable size arranged in sheets (Figure 4A). These cells had eccentric nuclei with abundant vacuolated clear cytoplasm suggesting the diagnosis of clear cell neoplasm, which contradicted the initial diagnosis of pleomorphic adenoma. Thin-walled capillaries were observed between these clear cells (Figure 4B, C). No necrosis, increased mitosis, and atypia were appreciated. Further evaluation by immunohistochemistry showed positivity for S100, CDK4, MDM2 (Figure 5A, B, C) with 2% Ki-67 but negativity for CK favoring the histomorphological diagnosis of well differentiated liposarcoma.

On discharge, the patient was haemodynamically stable, and his wound was healing well. The post disease status of the patient was evaluated after the diagnosis of

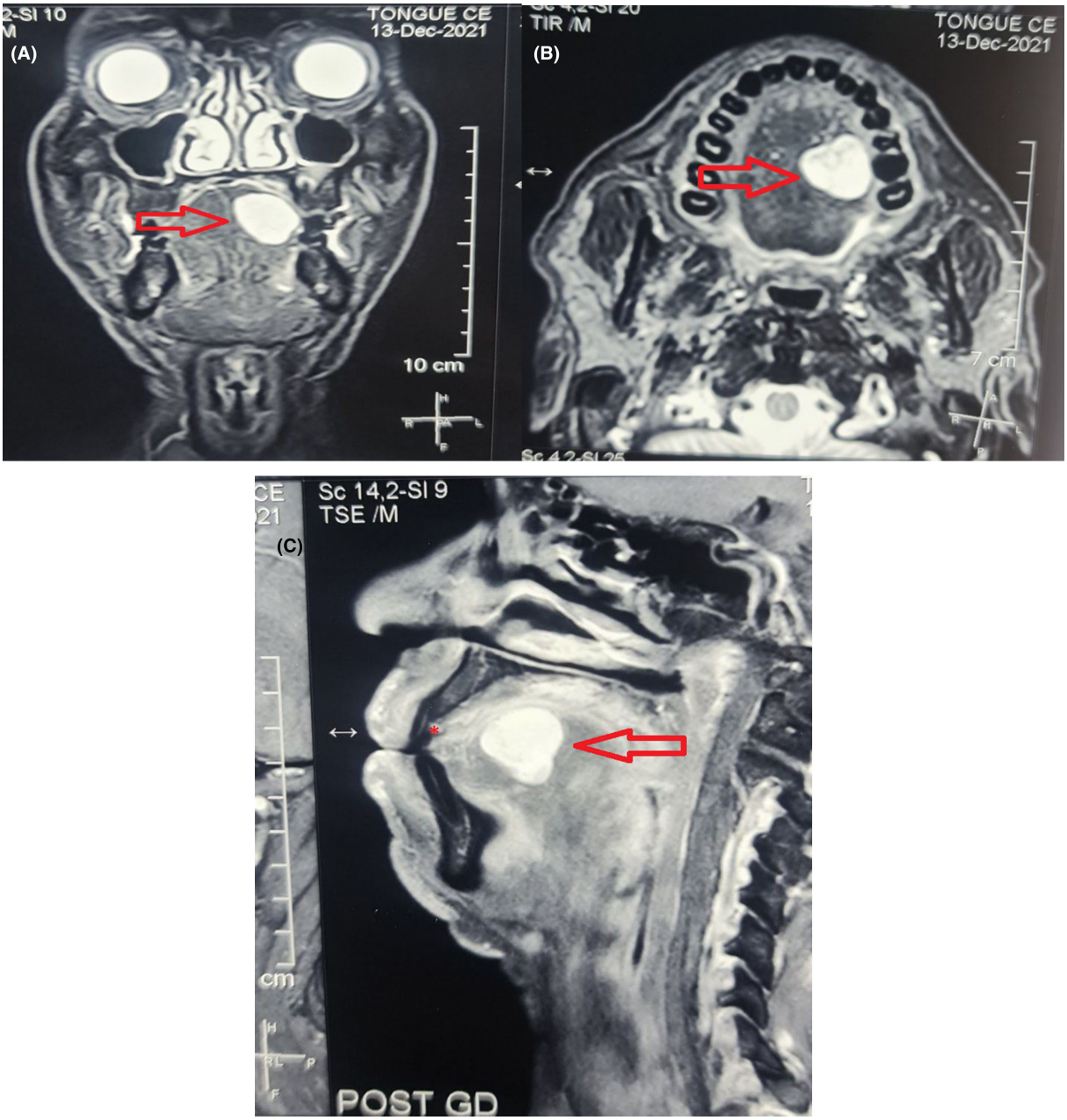

FIGURE 2 (A, B) Magnetic Resonance Imaging (MRI) of the tongue showed a well-defined, oval-shaped, soft tissue lesion within the tongue on the left side. (C) Post-gadolinium-enhanced lesion of 18 x 19 mm size seen about 19 mm distal to the tip of the tongue(Red Arrow).

WDLS by F18 FDG PET CT Scan, which was within the normal limit.

3 | DISCUSSION

Liposarcoma (LS), the tumor of embryonic mesenchymal origin, is the most common STSs, comprising 15% of all STSs. The usual site of presentation of this tumor is retroperitoneal (45%), followed by all extremities (24%).[1] However, it is uncommon to see it in the head and neck region, particularly in the oral cavity. In the review of 23 cases of LS of the oral cavity, the tongue was the preferred site with an incidence of 52%, whereas LS occurred in the buccal mucosa in 39% of cases, in another series of 18 cases of LS of the oral cavity and salivary glands.[2]

As per WHO 2020, LS is classified into four subtypes based on morphology, namely WDLS/ALT, Dedifferentiated liposarcoma (DDLS), Myxoidliposarcoma (MLS), and PLS.[3] Different LS variants have varied aggressive potentials because of their morphologic diversity. DDLS, high-grade MLS, and PLS all show a high propensity for metastasis, but

WDLS does not metastasize without dedifferentiation, and MLS has a more indolent clinical behavior and a reduced metastatic potential.[4]

WDLS, also known as ALT, is the most common variant accounting for about 40%–45% of all LA with the peak age of occurrence between 40 and 60 years.[5] Though the oral cavity, specifically the tongue, rarely gets involved by LS, often it is WDLS when there is involvement (75% of the cases).[2] As per Moritani et al., only 33 cases of WDLS/ALT were recorded globally from 1976 to 2008.[6] The condition often presents as a slowly growing painless mass without bleeding, dysphagia, dysgeusia, difficulty in articulation, and paraesthesia of the tongue. On examination,

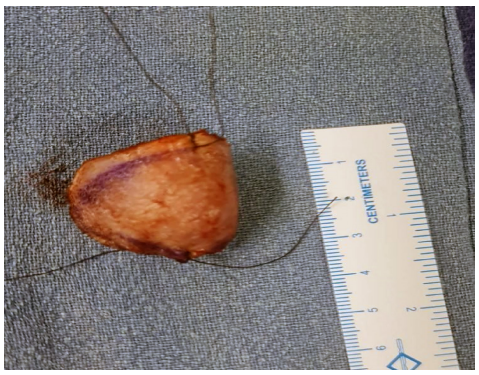

FIGURE 3 Gross specimen showing a nodular lesion and is oriented with sutures.

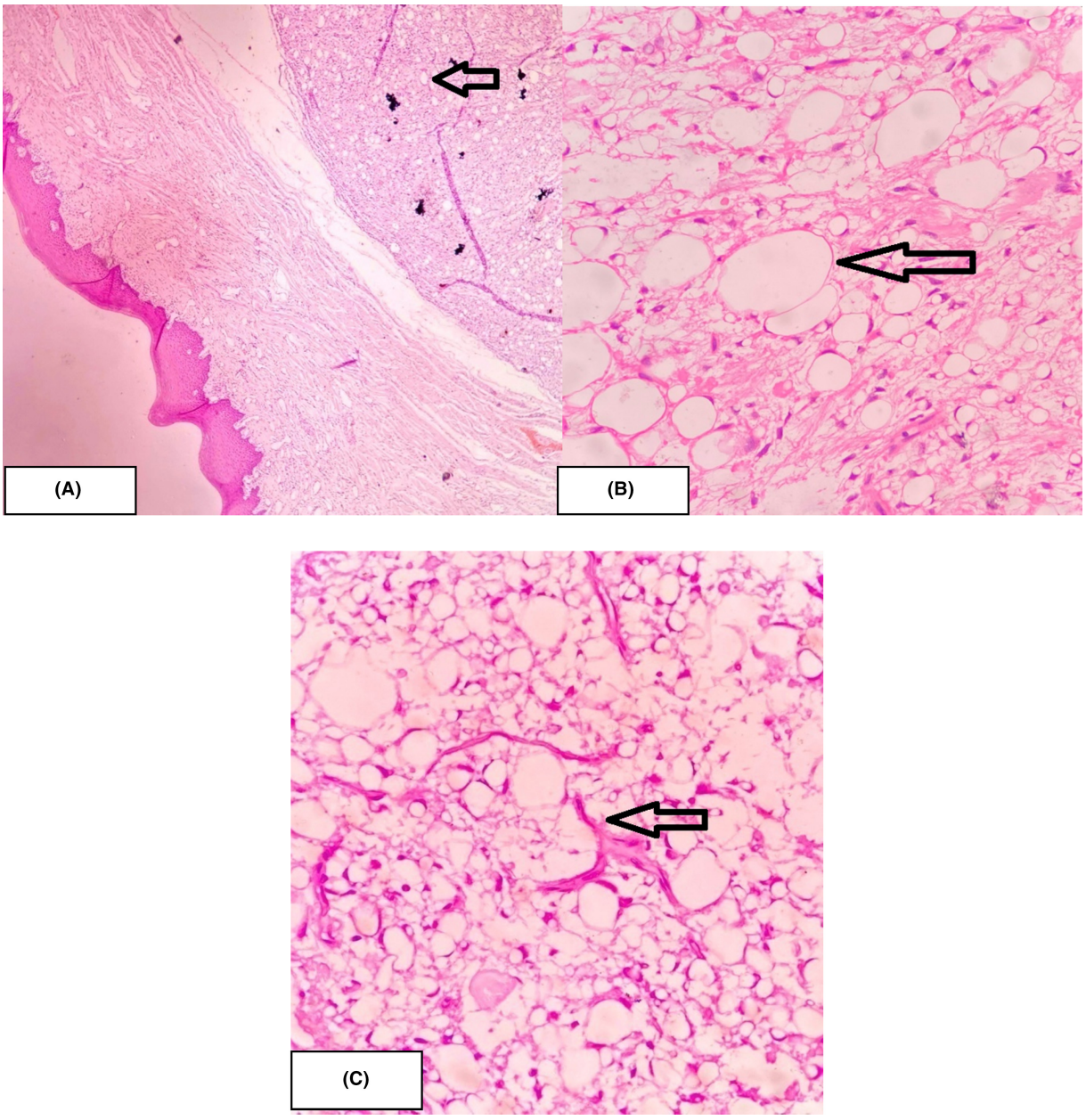

FIGURE 4 (A) H and E stained section revealing tumor(black arrow) below the mucosal epithelium. (B) H and E stained section revealing tumor cells composed of variable sized adipocytes. (black arrow). (C) Sections show chicken wired capillaries(black arrow) in between the adipocytes.

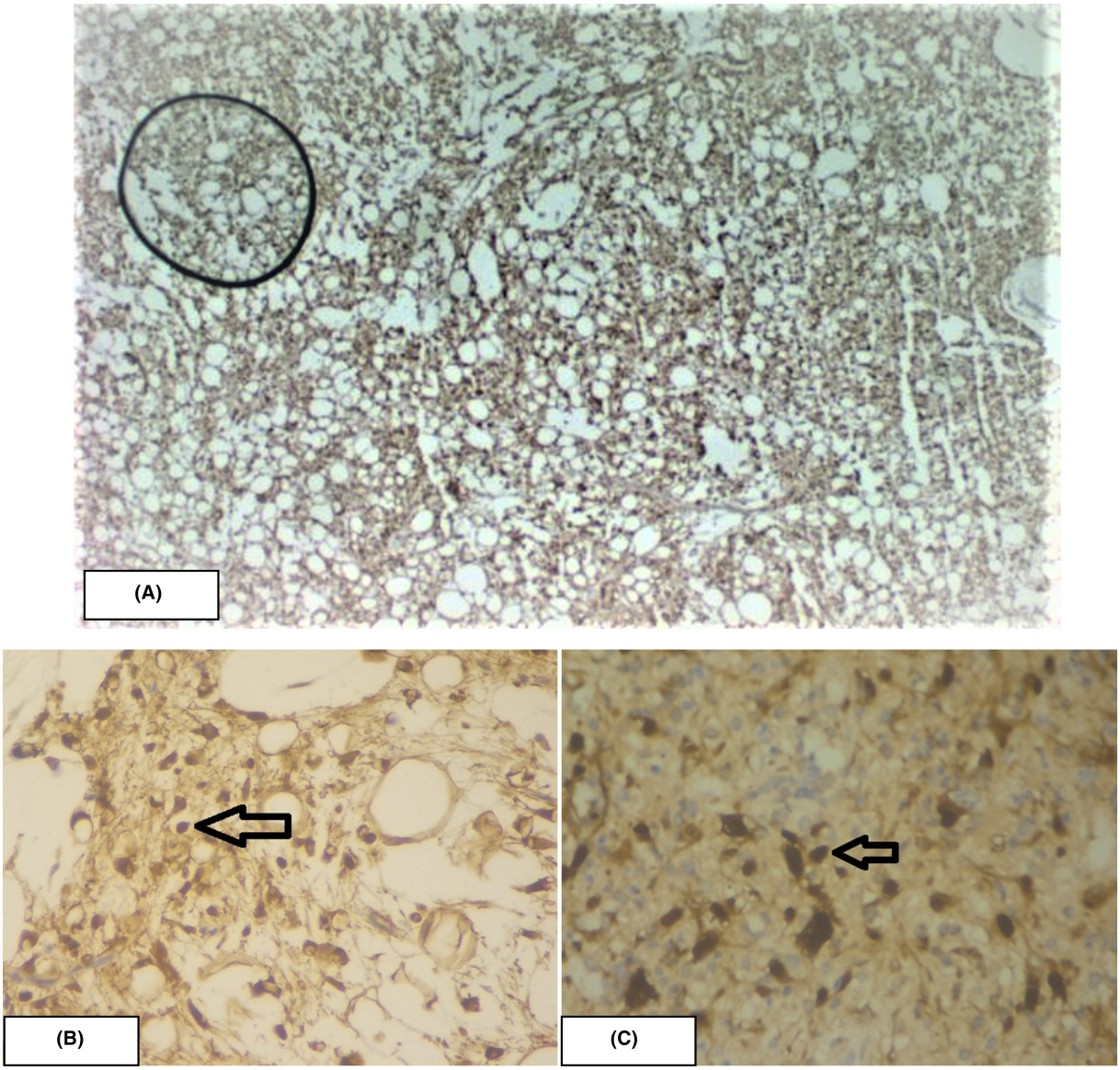

FIGURE 5 (A) S100 immunohistochemistry: Tumor cells show cytoplasmic and nuclear staining for S100. (B) CDK4 immunohistochemistry: Tumor cells show nuclear positivity for CDK4. (C) MDM2 immunohistochemistry: Tumor cells exhibit nuclear positivity for MDM2.

the lesion mainly presents as a firm or soft, elastic, nodular, and movable yellow-tan mass with variation in size depending upon the time of presentation.[2,6–9] Though the lesion is limited to submucosa at the time of presentation, as in our case, the cases with mucosal extrusion have also been reported.[6,7] Lymph nodes are typically not palpable, although neighboring lymph nodes were palpable in the case of Nunes et al.[10]

A lipoma with regressive alterations and an intramuscular lipoma is among the differential diagnoses for WDLS of the tongue. It was the initial diagnosis in the cases of Moritani et al. and Allon et al.[6,8] Other conditions that should be considered in the differential diagnosis are amyloidosis, myxoma, myxosarcoma, benign fat tumors (hibernoma), angiolipoma, fibrolipoma, pseudosarcomatous fasciitis, and malignant histiocytoma.[8,10] Amyloidosis was a preoperative clinical diagnosis in the study of Allon et al., whereas preliminary diagnosis in the study of Dubin et al.[8,11] Due to the broad spectrum of differential diagnosis, detailed histopathological examination is essential for a definite diagnosis.

The new World Health Organization categorization splits WDLS into three subclasses based on their morphologic characteristics: adipocytic, sclerosing, and inflammatory. Despite discovering three histologic variations, these subclasses have little clinical significance.[1] The tumor is yellow-tan, lobulated, and often covered by intact mucosa, giving them the appearance

of a lipoma.[2] But, in contrast to the lipoma, WDLS is made up of a relatively mature adipocytic proliferation in which significant variation in cell size is easily discernible and is admixed with fibrous connective tissues. Here, adipocyte nuclei are heavily stained in discrete areas, and unusual multinucleated stromal cells are frequently seen.[6] It presents with limited nuclear atypia and few or no lipoblasts.[12]Lipoblasts, when present, are vacuolar, multinucleated, or have highly stained nuclei.[6]It's vital to note that the existence of lipoblasts does neither guarantee nor exclude a diagnosis of liposarcoma.[13]

Immunohistological markers add to the diagnosis of liposarcoma. WDLS exhibits vimentin, S100, MDM-2, Ki-67, and CDK4 positivity. However, the positivity for the spindle cell component of WDLS is still under dispute.[2,8] The genes that code for all of these proteins are found on chromosome 12q13–15. These DNA sequences make up the majority of the supernumerary ring and giant rod chromosomes, which are the cytogenetic hallmarks of ALTs in around 93% of instances.

The treatment of choice for WDLS is wide surgical excision. Lymph node dissection depends upon the state of metastasis. Though the prognostic value of tumor size is unclear, adequate margin excision has great prognostic importance as the recurrence rate can increase from 17% to 80% due to incomplete removal of the tumor.[11] Transformation of WDLS to dedifferentiated type increases the chance of metastasis.[12] Hence, adequate margin coverage and close observation after surgery are crucial in WDLS.

4 | CONCLUSION

Though WDLS of the tongue is rare, it should always be on the list of differential diagnoses of tongue lesions. Final diagnosis is made on histopathological examination along with immunohistochemistry positivity for MDM2, CDK4, and S100. Resecting adequate margin is crucial during the surgical intervention as margin positivity has a high predilection for recurrence of WDLS. Similarly, regular follow-up is pre-vital as there is a chance of recurrence despite it being rare.

AUTHOR CONTRIBUTIONS

Moushami Singh: Conceptualization; resources; supervision; visualization; writing – original draft; writing – review and editing. **Arun R Napit:** Resources; writing – original draft; writing – review and editing. **Gunja Piya:** Resources; writing – original draft. **Prabhat Chandra Thakur:** Resources; writing – review and editing. **Sambit Mohanty:** Resources. **Sayali shinde:** Resources. **Ekta Jain:** Resources. **Hari Prasad Dhakal:** Supervision; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that there is no potential conflict of interest with respect to the research, authorship, and /or publication of this article.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient before the submission of the report.

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