Case Report

A case report of atypical lipomatous tumor of the tongue and literature review

Ayman FA Foad*, MD, Mohammed T Musa**, MD

Assistant Professor, University of West Kordufan, Faculty of Medicine & Health Science, Department of Pathology*, Associate Professor, University of Khartoum, Faculty of Medicine, Department of Surgery**

Summary

Lipoma-like well differentiated liposarcoma/atypical lipomatous tumor (WDL/AL) is a low grade soft tissue tumor of lipogenic origin. It has a well documented incidence in truncal and acral locations. The occurrence in the oral cavity, notably the tongue is a rare incident. We present a case of a 40-year-old male with dorsolateral mass on the surface of the tongue, after surgical excision of the lesion with a safe margin, under general anesthesia, the lesion has been diagnosed based on hematoxylin and eosin stained histopathological specimen as lipoma-like WDL/AL. Macroscopic and microscopic features of the lesion are compared with the published case reports in the literature.

Keywords: Well differentiated liposarcoma, atypical lipomatous tumor, liposarcoma circumscriptum, lipocyte, lipoblast, Myxoid liposarcoma, dedifferentiated liposarcoma, sclerosing, spindle cell.

Introduction

Well differentiated liposarcoma/atypical lipomatous tumor (WDL/AL):

Liposarcoma, an entity firstly described by Virchow, usually arises in adults, with no gender predilection (1, 2).

The World Health Organization (WHO) recognizes five variants; well differentiated/atypical lipomatous tumor (WDL/AL), myxoid, pleomorphic, dedifferentiated and mixed. The first two variants are of low grade and are the commonest (2).

Well differentiated liposarcoma/atypical lipomatous tumor comprises more than half of the low grade cases. These tumors are often incited in deep soft tissues and declared itself as a slowly growing painless mass. Acral and superficial masses are assigned as atypical lipomatous tumor, for the tribute of their resectability and low recurrence rate (50%). On the other hand, retroperitoneal and groin lesions are less amenable for complete resection, hence, de-bulking surgery is the logical resort, this results in a higher recurrence rate (90%) and higher grade transformation (28% vs. 5%).

Four variants of WDL/AL have been described by the WHO: lipoma-like, sclerosing, spindle cell and inflammatory.
Usually mixed patterns of varying proportion are declared in each variant\(^1,2\).

In spite of being the most common variant of liposarcoma family, WDL/AL is rarely encountered in the oral cavity, including the tongue. The first ever reported case was published back in 1976\(^3\). Eighty-eight percent (45/51 cases) of subsequently reported cases of lingual liposarcoma were WDL/AL, and 12% were myxoid liposarcoma.

Two cases of WDL/AL were spindle cell, and two were sclerosed WDL/AL variants. One case showed heterologous differentiation and another case had dedifferentiated element. Only 11 cases were assigned as lipoma-like, the other cases subtyping were not declared in the literature.

Case Presentation
A 40-year-old Sudanese male, presented in December 2011 to the Medical and Health Centre of the University of Khartoum. He is a farmer, married, with four children. He had a small tongue swelling for two years, which increased gradually in size. It was painless with no other symptoms referable to other systems. He had received broad spectrum antibiotic, with no obvious response. He was a known diabetic for 5 years, and he was on oral hypoglycemic agent. He is not a smoker, alcohol consumer or sniffer.

On physical examination, the swelling was a small nodule on the left border of the dorsum of the tongue; ovoid in shape, about 1.5 x 1.0 cm in dimension. The mucous membrane over the mass was intact, smooth, and yellowish and attached to the mass. The swelling wasn’t tender, and firm in consistency. No other abnormalities in the oral cavity were noticed, and the regional lymph nodes were not palpable.

Computerized tomography showed a single localized mass without regional lymphadenopathy. Routine investigations were all within normal. The nodule was widely excised with 1 cm margin under general anesthesia.

The excised mass was sent to Sudan National Laboratory. Gross examination showed formalin fixed circumscribed nodule covered by a tegument; measuring 1.5 x 1.0 x 0.7cm. The mass was bisected; the cut sections were tan colored with fatty textured bosselated surfaces. The specimen was sequentially processed, embedded in paraffin block, sectioned, mounted on a slide and stained with hematoxylin and eosin. Microscopic low power view (X4) revealed submucosal circumscribed lobulated mass sharply demarcated from the overlying para-keratotic squamous epithelium and the underlying lingual skeletal muscle bundles (Fig 1).

Fig 1: A circumscribed mass of WDL/AL, both at the superficial (top- squamous epithelium) and the deep margin (bottom- lingual muscle bundles)

Multiple thick collagen septa traversing the tumor were seen (Fig 2). Higher power views (X10 & X40) showed variably sized lipocytes and lipoblasts, vividly displaying atypical hyperchromatic pleomorphic knobby nuclei with coarsely clumped chromatin and nuclear vacuolization (Fig 2). Atypical hyperchromatic stromal spindle cells were noted, often with perivascular accentuation (Figs 1,2). Mitotic figures were rare.
The above-mentioned features were consistent with the diagnosis of atypical lipomatous tumor-lipoma-like variant; the case was signed out after reviewing the literature and consulting senior colleagues.

**Discussion**

We present a case of atypical lipomatous tumor of the tongue, in a 40-year-old male. Liposarcoma of the tongue usually afflicts adults; the youngest reported subject was an 8-year-old female, and the oldest was an 86-year-old male. There is an apparent gender predilection in favor of male subjects in reported cases; 70% were males. This gender bias is not a feature of WDL/AL arising in other sites. Whether this bias is going to dissolve upon further cases accumulation, it remains to be elucidated.

Our patient lapsed two years till seeking medical advice and granted surgical management. Duration of symptoms prior to biopsy ranged from 3 months up to 10 years in reported cases. This goes with the presumed indolent behaviour of the lesion. The patient hasn’t shown any evidence of residual disease in subsequent post-operative follow-up visits until April 2012 (date of surgery December 2011). The longest published disease free survival was 14 years. The longest followed-up case, albeit with intermittent recurrences was 28 years.

Overall recurrence rate among published cases was 10%. Incomplete resection is the most pertinent factor in determining recurrence potential. Previously reported cases in the literature, indicates the indolent behaviour of the tumor and mandates a conservative surgical approach.

**Macroscopy**

On the macroscopic view of our case, the lesion appeared circumscribed, nodular and yellow tan on cut section; measuring 1.5 x 1.0 x 0.7 cm. Grossly WDL/AL appears as a circumscribed nodular or multinodular mass with bosselated surface, hence the synonym liposarcoma circumscription. The size ranged from 0.5 to 3.5 in greatest dimension in reported cases. Cut sections showed yellow to tan fatty surface, with speckled whitish pale areas in cases with prominent sclerosis.

**Microscopy**

Under the microscope, our case displayed the salient features of WDL/AL arising in conventional sites: low grade hyperchromatic neoplastic cells of varying sizes amidst spindly stroma. In reported cases of lipoma-like WDL/AL, the lesions were circumscribed at the microscopic level with pushing borders. Skeptical microscopic scanning at low power (X4) steers the pathologist to scrutinize the fields with higher power lenses (X10, X20 and X40), eventually revealing the worrisome features; variably sized lipocytes, hyperchromatic nuclei with coarse clumped chromatin, nuclear pleomorphism and vacuolization (Figs 2, 3).

Lipoblast once considered essential to diagnose liposarcoma may be sparse or absent in small incisional biopsies, furthermore it might be encountered in indolent benign lipoma variants (pleomorphic lipoma and spindle cell lipoma). Thick bands of collagen and muscular veins wall harboring atypical spindle cells is one of the constant features of WDL, that have been revealed in our case (Fig 3).
Fig (3): Variously sized lipocytes and lipoblasts warped around a blood vessel, showing hyperchromatic spindle cells percolating through its wall.

Focal amyloid deposits and stromal myxoid changes may be seen. Inflammatory foci with germinal follicles formations are sometimes noted. Occasionally, bland heterologous differentiated elements are encountered, such as cartilage (1,2). We did assigned the lesion as lipoma-like variant of WDL/AL, in the literature this variant constituted 29.4% (15/51 including our case), some of the cases didn’t declare a detailed microscopic description of the lesions. Some authors upon histological recognition of the diagnosis advice margin revision of about 0.3 cm. Tumor negative revised margins correlated with long tumor free survival (9).

Ancillary Tests
The diagnosis of WDL/AL is straightforward with careful analysis of the morphological patterns under the light microscope, though we didn’t resort to the sophisticated molecular techniques. Immunohistochemistry hasn’t much to do in aiding the diagnosis of WDL/AL. These tumors are positive for vimentin and S-100 markers. Karyotyping shows ring chromosomes and long marker-chromatin derived from 12q13-15 (10-12), corresponding to MDM2 (Murine Double Minute-2) and CDK4 (Cyclin Dependant Kinase-4) over expressions, which can be revealed with immunohistochemical studies in ambiguous cases (segregation of pleomorphic/spindle cell lipomas from WDL/AL) (13). The above-mentioned chromosomal and molecular anomalies are also seen in WDL/AL arising in other locations.

In conclusion, atypical lipomatous tumor is a well-known entity, which is rarely encountered on the oral cavity especially on the surface of the tongue. Cooperative efforts of the surgeons and the pathologists are warranted to reach such diagnosis. Follow-up is mandatory for the lack of long-term survival studies due to cases scarcity.

References