Nodular lesion in the tongue: a rare case of low grade myofibroblastic sarcoma

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SUMMARY

Low grade myofibroblastic sarcoma is an atypical myofibroblastic malignant neoplasm that rarely affects the oral cavity and presents a challenging histological diagnosis. Here we report the case of a 38-year-old woman who had a nodular and symptomatic lesion on the lateral border of the tongue. After the biopsy, the histopathological analysis revealed a spindle cell lesion with fascicular and storiform pattern. We performed immunohistochemical analysis that showed intense labeling for vimentin, smooth muscle actin and moderate labeling for calponin and S-100. We emphasize the rarity of low grade myofibroblastic sarcoma such as a nodular lesion on the tongue and the need for accurate diagnosis.

KEY WORDS: *Immunohistochemistry, Sarcoma, Diagnosis, Rare, Tongue, Neoplasms.*

INTRODUCTION

Low grade myofibroblastic sarcoma (LGMS) is a malignant mesenchymal neoplasm of myofibroblastic origin that is atypical and rare and mainly affects the head and neck region (1,2). LGMS can affect individuals in any age group, with a propensity for males of fourth decade of life and frequently presents as a painless volume increase (3). Because it is a myofibroblast lesion, the diagnosis is relatively difficult, since neoplastic myofibroblasts present a variable immunophenotype and express both muscle and myoepithelial markers (4). LGMS usually has a locally infiltrative-destructive pattern (5,6). Here we report a case of LGMS at right lateral border of the tongue with unusual clinical presentation, with emphasis on its clinical-pathological characteristics.

CASE REPORT

A 38-year-old woman, non-smoker, non-alcoholic, sought dental service due to a painful nodule on her tongue that was present for four months. The previous medical history did not present contributing information. Recent biochemical and hematological laboratory tests were within normal limits. There were no alterations in extraoral examination.

Intraoral evaluation showed a nodular lesion on the right side of the lateral border of the tongue. The lesion was hardened, reddish and sessile. Although well delimited, it presented with an infiltrative and diffuse growth pattern (Figure 1). The presumptive clinical diagnosis was oral squamous cell carcinoma and an incisional biopsy was performed.

Histopathological examination revealed the intense proliferation of myofibroblasts with fusiform morphology, with vesicular and wavy nuclei and with condensed chromatin (Figure 2a). These cells were present either forming discontinuous bundles, sometimes in a storiform pattern (Figure 2b). Pleomorphism and mitoses were uncommon. Immunohistochemical analysis showed intense labeling for vimentin and alpha smooth muscle actin (Figure 2c) and the sample was positive-focal for S-100 and Calponin (Figure 2d). Ki-67 was <5% (Figure 2e), while CD-34, CD99, p63 (Figure 2f) and desmin were negative.

Based on these findings, the diagnosis of LGMS was established. The patient was referred to a specialized cancer service for further treatment.

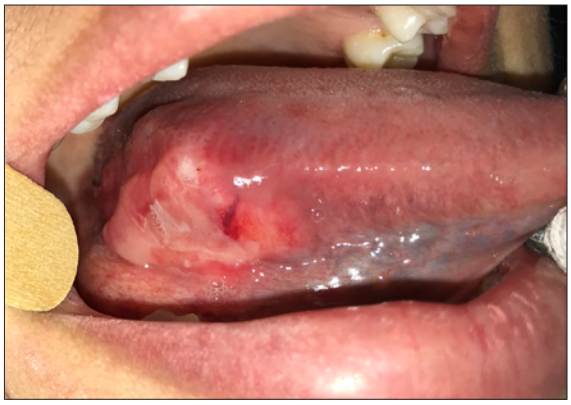

Figure 1. Initial aspect of the lesion

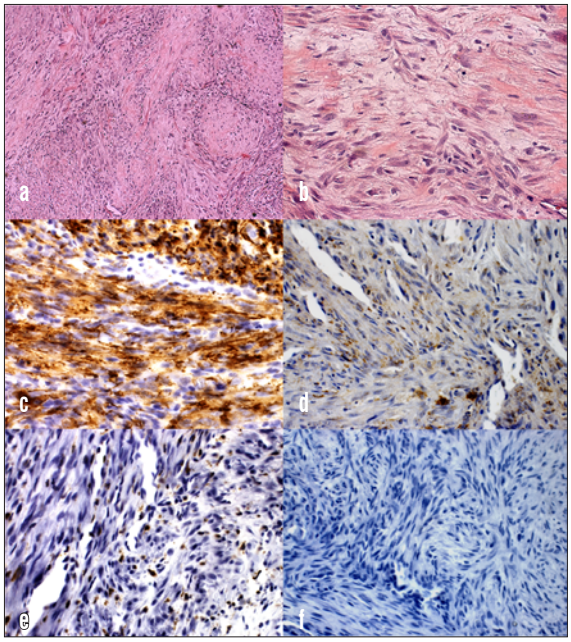

Figure 2. Histopathological (hematoxylin & eosin, HE) and immunohistochemical (IHC) aspects of LGMS: a) (HE 100×) Spindle cell lesion with fascicular and storiform pattern; b) (HE 400×) Spindle cells with wavy nucleus, eosinophilic cytoplasm and discrete pleomorphism; c) (IHC 400×) Intense positivity for smooth muscle actin; d) (IHC 400×) Focal and discrete immunopositivity for calponin; e) (IHC 400×) Immunostaining for Ki-67; f) (IHC 400×) Absence of immunostaining for p63

Arch Oncol 2020; 26(2):35-6

Published Online
October 25, 2019
https://doi.org/10.2298/AOO190827009T

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Received **2019-08-27**
Received in revised form **2019-09-23**
Accepted **2019-10-04**

DISCUSSION

Within the group of myofibroblastic lesions LGMS is a condition that most frequently affects the deep soft tissues of the head and neck (1,3). Just a few cases have been reported in the oral cavity, with less than eight cases described throughout the literature (6). This condition was first described by Mentzel et al. in 1998 (2), and it has been recently included in 2017 in the fourth classification of head and neck tumors of the World Health Organization (WHO) (3).

LGMS is characterized by fusiform and star-shaped cells with eosinophilic cytoplasm and poorly defined cell boundaries. Its nuclei are undulating with small nucleoli. These cells acquire a fascicular and storiform arrangement, immersed in a collagenous stroma of variable density. Mitotic activity is low and necrosis is usually insufficient (7). These characteristics were also found in this case.

The diagnosis of lesions with predominance of spindle cells is usually challenging and differential diagnosis should include benign conditions such as nodular fasciitis; lesions of intermediate biological behavior, such as the inflammatory myofibroblastic tumor; and malignant neoplasms like leiomyosarcoma (8).

The nodular fasciitis hypothesis was ruled out because it is a self-limiting reactive process characterized by proliferation of young myofibroblasts in a myxoid stroma that resembles histologically a granulation tissue or cells in culture. Myofibroblastic inflammatory tumor was also ruled out because it is a borderline lesion with presence of spindle cells arranged in a fascicular pattern and with foci of inflammatory lymphoplasmacytic infiltrate. Finally, leiomyosarcoma was discarded, as this is a malignant neoplasm of smooth muscle origin, whose nuclei present blunted appearance, with frequently hyalinized stroma and high number of mitoses (2,8). LGMS may cause focal recurrences and metastasis, and it has a relatively indolent course. The recurrence is the highest when tumor is sited in nasal cavity/paranasal sinus, involving deep tissue space, with size over 3-cm (8). Due to the scarcity of published works on LGMS, the surgical treatment with free margin is still the most used therapy, whereas radiotherapy and chemotherapy are not well established treatments (1,3). The patient was referred to a specialized cancer service for treatment.

The present case of LGMS was the first seen in our oral pathology service, which has dealt with over 95,000 biopsy specimens. LGMS is a rare condition that is difficult to place within the scope of differential diagnosis of nodular lesions in the tongue. Thus, we emphasize the need for accurate histopathological and immunohistochemical analyzes that aim at the correct diagnosis and prioritize for the appropriate treatment.

Declaration of Interests

Authors declare no conflicts of interest.

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