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CASE REPORT

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Atypical spindle cell lipomatous tumor of the tongue: A rare entity arising in an unusual location

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Abstract

Atypical spindle cell/pleomorphic lipomatous tumor (ASCPT) constitutes an emerging entity of lipomatous tumors. It is a benign tumor. It occurs typically in limbs and limb girdles. The occurrence in oral cavity is unusual. The diagnosis of this neoplasm is challenging. Herein, we report a case of ASCPT arising in the tongue.

KEYWORDS

atypical spindle cell/pleomorphic lipoma, differential diagnosis, genetic aberration, molecular biology, tongue

1 | INTRODUCTION

Atypical spindle cell/pleomorphic lipomatous tumor (ASCPT) is a confused and an evolving entity.[1] It is characterized by the presence of variable proportions of mild to moderately atypical spindle cells, adipocytes, lipoblasts, pleomorphic cells, multinucleated giant cells, and a myxoid or collagenous extracellular matrix.[2] It has been considered as "spindle cell liposarcoma".[3] Recently, thanks to the development of molecular analysis and the contribution of genetic studies, ASCPT is emerging as a distinct group.[4] This tumor predominates in the limbs and the limb girdles. The head and neck region is an exceptional site, only 23 cases were reported in the English literature.[4] The patient has generally an excellent prognosis with low rate of local recurrence after incomplete excision.[2]

Herein, we describe a new case of ASCPT of the tongue and discuss its clinical and pathological characteristics.

2 | CASE REPORT

A 77-year-old female patient admitted to the Department of Maxillofacial Surgery of Habib Bourguiba Hospital, Sfax, Tunisia, in September 2019 complaining of a painless soft mass in the margin of the tongue, first noticed one month previously. The patient had a medical history of arterial hypertension. She denied other major systemic diseases and drug or food allergies. She has no history of smoking or alcohol usage. There was no family history of similar tumors. An intraoral examination revealed a movable mass in the left lateral border of the tongue with intact surface. It measured 15 mm in its largest diameter with the clinical impression of a benign tumor. Overall examination was normal; especially there were no palpable lymph nodes. A clinical intra-oral photograph of tongue was not available. A magnetic resonance imaging of the tongue revealed a well circumbscribed mass measuring $18 \times 12 \times 8$ mm without expansion to vascular vessels. Signal intensity was slightly high with T1- and

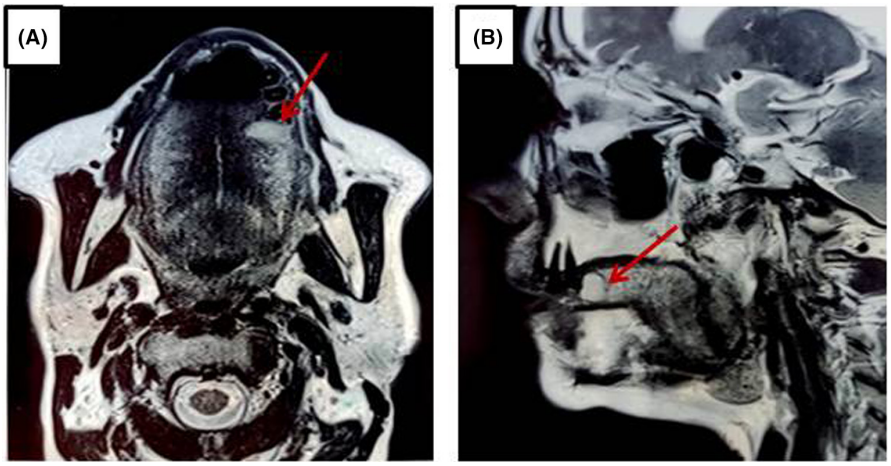

FIGURE 1 (A) Axial view showing a well circumbscribed mass of the tongue T1-hyperintense. (B) Sagittal view showing a T2-hyperintense mass without extention to the buccal floor muscle

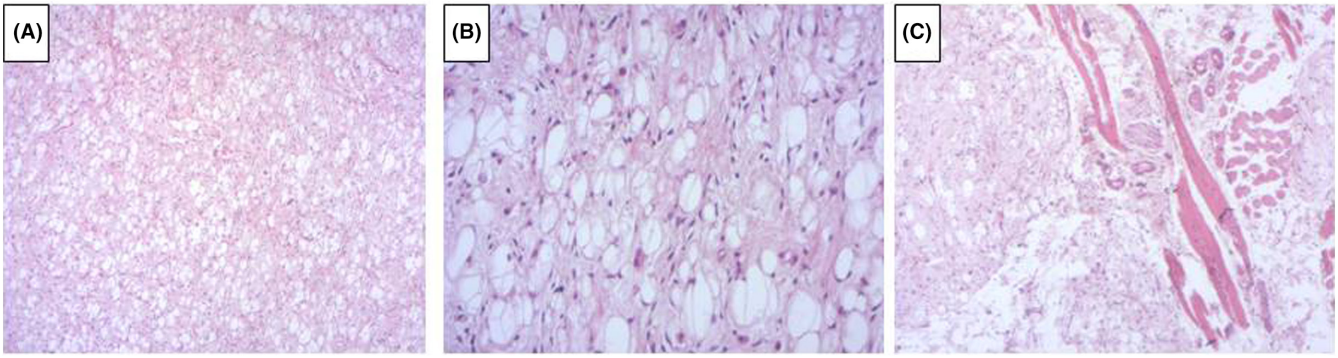

FIGURE 2 Atypical spindle cell lipoma. (A) Spindle cells are intimately admixed with variably vacuolated fat cells in a collagenous stroma (HE × 50). (B) Note the marked variation in adipocyte size and shape (HE×200). (C) The tumor infiltrates striated muscle fibers of the tongue (HE×50)

T2-weighting (Figure 1). We evoked the diagnosis of lipoma of the tongue. The patient underwent local surgical excision. The surgical procedure was uneventful. The specimen was sent for pathological examination. Gross examination showed a yellow nodular lesion partially encapsulated measuring 13 mm in maximum dimension with soft texture. Histopathologic examination showed that this lesion consisted of relatively uniform spindle cells separated by a variable number of mature adipocytes setting in a collagenous stroma (Figure 2). The spindle cells show minimal cytonuclear atypia. Lipoblasts were absent. The proportion of spindle cells and adipocytes varied in different areas. Necrosis and mitoses were absent. The tumor showed an infiltrative growth and involved the margins. Immunohistochemical studies revealed that spindle cells were diffusely positive for CD34 but are negative for smooth muscle actin (SMA) and S-100 protein (PS100). By contrast, mature adipocytes were focally positive for MDM2 (Figure 3). The above findings suggested the diagnosis of ASCPT.

Consequently, molecular biology was recommended. Molecular study for MDM2 gene showed absence of MDM2 amplification. RB1 deletion and CDK4 amplification were not tested. Thus, the diagnosis of ASCPT was made.

The post-operative follow-up showed local recurrence of the lesion after 3 months. Patient underwent tumor resection. There were same pathologic findings. The patient was well. There was neither recurrence nor metastasis at 1-year follow-up evaluation.

3 | DISCUSSION

Lipomatous tumors are frequent in clinical practice.[1] Oral lipomas represent only 0.27%–1.7% of all oral lesions.[4] Their occurrence in the tongue is extremely rare.[5] They typically arise in patient after the 4th decade of life with a slight predilection for man.[6] Several risk factors are involved, including age and hereditary. Genetic aberrations are also known as a risk factor of solitary lipomas. The most usual aberration includes translocations between 12q13 and 15 and several other chromosomes, most frequently chromosomes 1, 2, 3, and 21.[6]

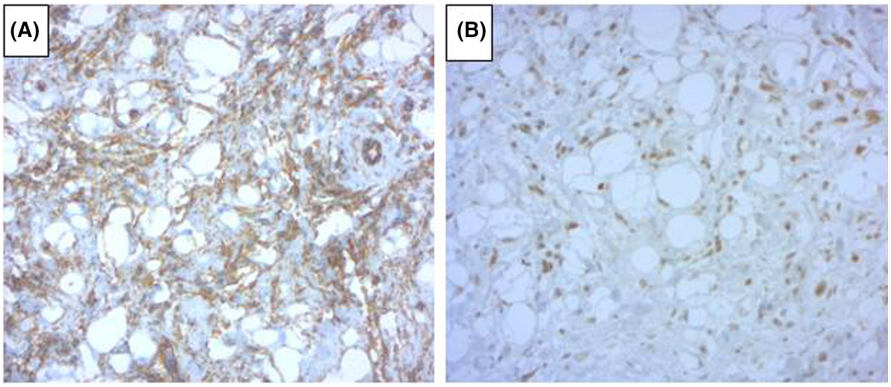

FIGURE 3 (A) Diffuse immunohistochemical expression of CD34 in spindle cells (×200). (B) Adipocytes are focally positive for MDM2 (×200)

Lipomatous tumors showing spindle cell component and atypical histologic findings called atypical spindle cell/pleomorphic lipomatous tumor (ASCPT) are not common and have various morphological features making their diagnosis challenging.[7] Several studies were carried out to identify their clinicopathological, immunohistochemical, and genetic characteristics. They are classified as benign tumor according to the 2020 World Health Organization (WHO) classification of soft tissue and bone tumors.[2]

ASCPT are characterized by indistinct margins and the presence of different proportions of mild-to-moderate atypical spindle cells, lipoblasts, adipocytes, pleomorphic cells and multinucleated giant cells. The extracellular matrix is typically myxoid or collagenous.[2]

They are more frequent in middle-aged adults (sixth to ninth decade).[8,9] They occur usually in men with sex ratio of 1,5/1.[4] ASCPT tend to arise in subcutis lower extremities, thigh, shoulder, chest wall, and paratesticular region.[8] Deep soft tissue is unusually affected.[8] Intra-cavitary and visceral locations are exceptional.[1] Rare cases of ASCPT were described in head and neck region with 23 cases reported in the English literature.[4] The occurrence of ASCLT in oral cavity and especially in tongue is extremely rare.[4] Only one case similar to this current case was found in the literature.[5]

Clinically, ASCPT presents as a subcutis indolent mass or nodule.[1] Grossly, these tumors are not completely encapsulated, their diameter vary between 0.5 and 28 cm with an average size of 5 cm. They have a nodular or multinodular architecture with infiltrative character.[4,9]

A broad spectrum of microscopic features was noted even within the same tumor, due to the variable proportion of cellular and extracellular elements.[7]

ASCPT are typically composed of the admixture of the proliferation of spindle cells, adipocytic cells, and collagenous or myxoid stroma. Ropy collagen fibers are absent.[4] The spindle cells have an eosinophilic cytoplasm with hyperchromatic ovoid nuclei. Cytonuclear atypia are generally focal. The adipocytic component shows mature adipocyte cells with considerable variability in shape and size. Lipoblasts can be seen. They vary from small univacuolated to large multivacuolated with hyperchromatic nuclei. In most cases, "pleomorphic" multinucleated and hyperchromatic cells are dispersed within spindle and adipocytic cells. Mitoses are exceptional. Necrosis was not reported in any case.[1,4]

Immunohistochemically, spindle-shaped tumor cells are positive for CD34. S-100, and desmin expression can be observed in a significant case. However, focal and weak expression of MDM2 or CDK4 can be rarely noted.[4] In our case, spindle cells were diffusely positive for CD34, some adipocytic cells stain positively for MDM2.

The amplification of MDM2 and CDK4 gene is always absent.[4] That was similar in our case. Cytogenetically, 50%–70% of ASCPT show loss of RB1 and its flanking genes RCBTB2, DLEU1, and ITM2B.[1]

The diagnosis of ASCPT is challenging due to the wide range of pathologic features and lack of specific genetic findings.[4] They can be confused with spindle cell lipoma (SCL) when they have low cellularity and mild cytonuclear atypia. However, SCL have a distinctive anatomic distribution. They are completely encapsulated and well demarcated. There is no important variation in shape and size of adipocytes. Ropy collagen bundles are often encountered.[10] In our case, the infiltrative growth pattern was against spindle cell lipoma.

Dedifferentiated liposarcoma (DDLS) should not be mistaken with ASCPT. In fact, some cases with low-grade dedifferentiation are composed of fibroblastic spindle cells with moderate cytonuclear atypia. However, low-grade DDLS is non-lipogenic.[2] The MDM2 and CDK4 gene amplification can easily make the diagnosis.[1]

Diffuse neurofibroma, dermatofibrosarcoma protuberans (DFSP), mammary-type myofibroblastoma, and morphologically low-grade malignant peripheral nerve sheath tumor (MPNST) are considered as differential diagnosis of ASCPT. In fact, these spindle cells tumors may present a marked adipocytic component and infiltrative behavior.[4]

While ASCPT is considered as a low-grade neoplasm, it has a marked infiltrative character. The rate of local recurrence is about 10%–15%.[1] No case of distant metastasis

or death from this pathology has been reported in the literature.[4] Surgical excision with clear margins is recommended to limit the risk of local recurrence.[4] Most patients undergoing complete surgical excision will have an excellent prognosis.[2]

We are reporting this case to discuss a new entity that appeared in the 5th edition of World Health Organization (WHO) classification of soft tissue and bone tumors. Our case is distinguished by an exceptional location of ASCLT. In fact, ASCLT of the tongue is extremely rare. Pathologists and clinicians should be aware of this emerging entity to avoid misdiagnosis. In fact, complete resection with free tumor margins and adequate management are recommended to avoid both recurrence and abusive re-excision.

AUTHOR CONTRIBUTIONS

Soumaya Graja involved in concept, data collection and processing, writing the manuscript, and literature search. Chiraz Chaari and Mouna Zghal involved in literature search and writing the manuscript. Morched Dhouib contributed in collecting data and writing the manuscript. Chahir Kammoun collected data and writing the manuscript. Slim Charfi involved in analysis, interpretation, literature search, and writing the manuscript. Tahya Sellami-Boudawara approved the manuscript.

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CONFLICT OF INTEREST

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

DATA AVAILABILITY STATEMENT

None.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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