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CASE REPORT OPEN ACCESS

BILATERAL LYMPHANGIOMA ON THE TONGUE IN AN ADULT PATIENT: AN UNUSUAL CASE REPORT

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ABSTRACT

Introduction: Lymphangioma is a benign, hamartomatous, non-encapsulated growth malformation, consisting of an enlarged and cystic lymphatic system. **Case report:** Female, 36 years-old, melanoderma, complaining of "balls" on her tongue. In the anamnesis, she reported having noticed an injury on the tongue for about 08 months, asymptomatic, slow growth and bleeding to the trauma. In the intraoral examination, a lesion in the form of a papulomatous plaque sensitive to touch affecting the region of the lateral border of tongue, measuring approximately 2.3 cm x 3 cm in diameter. Microscopically, it was observed the presence of large vessels located in the subepithelial region, covered by a layer of flattened endothelial cells that, inside, had suggestive eosinophilic material and the presence of proliferative lymphocyte nests. Thus, histopathological diagnosis was lymphangioma. After 15 days, he returned with a healed surgical area. The patient was referred for surgical removal and the present case has been under follow-up for 02 years. **Conclusion:** These lesions in adult patients are lesions that arose in childhood and the diagnosis is late, but they have a better prognosis due to its slow growth.

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INTRODUCTION

Lymphangioma is a benign, hamartomatous, non-encapsulated growth malformation, consisting of an enlarged and cystic lymphatic system that does not directly communicate with the normal lymphatic drainage system (Smith, 2019; Abe, 2018 and Hwang, 2017). They have a predilection for the head and neck region, being rare in the oral cavity and in adult patients, due to their presence since birth or within two years of age (Abe, 1932; Hwang, 2017 and Lerat, 2016). In the oral cavity, it mainly affects the tongue (Abe, 2018; Hwang, 2017; Usha, 2014), with a clinical aspect ranging from a pink or yellowish nodular plaque to an increase in volume (Nammour, 2016). Microscopically, it consists of lymphatic vessels covered by a thin endothelial lining with marked dilations in which it may appear empty or consist of protein and leukocyte material, with three histological groups: simple (capillary), cavernous and cystic (Abe, 1932). Treatment varies from surgical excision to laser therapy, sclerotherapy, radiotherapy, electrocautery, cryotherapy, ligation, embolization and steroid administration (Abe, 1929 and Usha, 2014). The recurrence rate varies from 10% to 50% of cases of surgical removal (Catalfamo, 2012). The aim of the study is to report a case of lymphangioma diagnosed at the Oral Diagnosis service of a reference center and relate it to the findings in the literature.

Case report

Female patient, 36 years old, melanoderma, married, showed up at the clinic of the Oral Diagnosis Clinic with a complaint of "balls" on her tongue. In the anamnesis, she reported having noticed an injury on the tongue for about 08 months, asymptomatic, slow growth and bleeding to the trauma. Her main complaint was the fear of being cancer. She reported systemic comorbidity, controlled hypertension. No family history of cancer. Of medication used routinely, reported use of contraceptives and medication to control blood pressure. In the extra oral exam, she did not present any noteworthy alterations. In the intraoral examination, a lesion in the form of a papulomatous plaque sensitive to touch, but usually

painlesswas observed, affecting the region of the lateral border of the right tongue more, measuring approximately 2.3 cm x 3 cm in diameter (Figure 1), focal areas of the lingual belly and lateral border left in the lingual belly, of sessile implantation, exophytic growth, stony texture and pale pink color with the presence of whitish areas and unsatisfactory periodontal condition in the lower anterior dental elements. The clinical diagnosis was papilloma. An incisional biopsy of the lesion was performed and sent for histopathological analysis. Microscopically, it was observed the presence of large vessels located in the subepithelial region, covered by a layer of flattened endothelial cells that, inside, had suggestive eosinophilic material and the presence of proliferative lymphocyte nests (Figure 2). Immunohistochemistry was performed with markers CD34 and D2-40 (podoplanin), with diffuse positive marking for vessel walls by both and D2-40, in addition to marking lymphatic vessels, had a positive and diffuse membrane marking on epithelial basal cells and parabasal layers of the oral epithelium (Figure 2). Thus, histopathological diagnosis was lymphangioma. After 15 days, she returned with a healed surgical area. The patient was referred for surgical removal, however the present case has been under follow-up for 02 years.

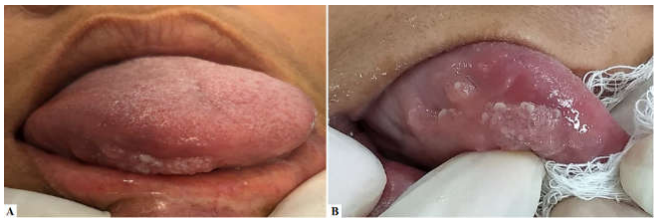

Figure 1. Clinical aspect of oral lymphangioma. A) pale pink vesicles in the tongue belly. B) Plate appearance and slightly whitish

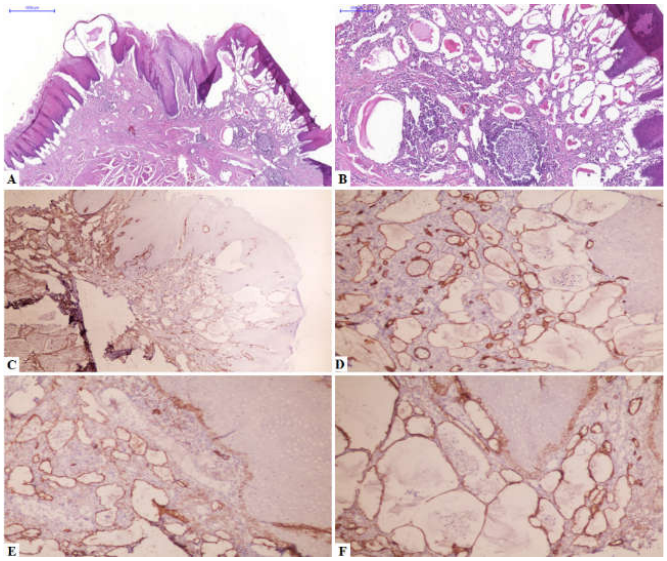

Figure 2. Morphological characteristics of oral lymphangioma. A) large lymphatic vessels adjacent to the surface epithelium (Scale bar: 1000µm); B) lymphatic vessels lined with thin endothelium and the presence of eosinophilic material inside it compatible with the lymph (scale bar: 200µm); C) Vascular immunoexpression for CD34 adjacent to the surface epithelium (100x magnification); D) Detail of the loss of immunoexpression for CD34 of some large caliber lymphatic vessels (200x magnification); E) immunoexpression of lymphatic vessels and basal layer of the surface epithelium for D2-40 (100x magnification); F) Only the endothelium of the lymphatic vessels shows immunoexpression for D2-40 (400x magnification).

DISCUSSION

The origin of lymphangioma is believed to be due to sequestrations of lymphatic tissue that does not normally communicate with the rest of the lymphatic system (Smithm 2019). Other theories have been proposed, such as the cessation of normal growth of the primitive lymphatic channels during embryogenesis and the inability of the primitive lymphatic sac to reach the venous system (Abe, 2018). They are caused by obstruction of lymphatic drainage, abnormal budding of the lymphatic tissue and sequestered lymphatic remains that retain their embryonic growth potential (Hwang, 2017). It is doubtful whether these lesions are true neoplasms.The update in the literature was performed and is related in table 1. Classification, clinical presentation and treatment planning depend on the size of the cysts and are described as macrocystic composed of cysts-like spaces measuring 2 cm in diameter or more; microcysticcomposed of smaller vascular channels measuring less than 2 cm in diameter; and mixed composed of the combination of macrocystic and microcystic spaces (Smith, 2019; Abe, 2018 and Hwang, 2017). Subtypes are probably variants of the same process and the size of the vessels may depend on the nature of the surrounding tissues. Microcystic lesions are more frequent in the mouth, where the surrounding dense connective tissue and skeletal muscles limit vascular expansion (Hwang, 2017). The present case is an example of a microcystic lesion.
The incidence is estimated at one of the 2000-4,000 live births, with no difference in race or sex, more frequent in the head and neck region and affects mainly children (Smith, 2019; Lerat, 2016 and Usha, 2014).

In the oral cavity, it mainly affects the tongue, the two most common anterior thirds, followed by the lips, buccal mucosa, soft palate and floor of the Mouth (Abe, 2018). The diagnosis of lymphangioma in adults is a rare occurrence (Usha, 2014; Ganesh, 2013).Its clinical aspect demonstrates a stony surface that resembles a group of translucent vesicles and there may be secondary hemorrhage within the lymphatic spaces and make these “vesicles” purplish, or whitish due to constant trauma, when located superficially, or in the case of injuries deep, present as ill-defined softened swelling (Abe, 2018 and Eren, 2017). Rapid growth can occur secondary to infection or hemorrhage, presumably due to increased lymph production, blockage of lymphatic drainage or secondary infection of the lesion (Usha, 2014). In the present study, the lesion affected the two thirds of the tongue bilaterally and part of the lingual belly, differing slightly from the involvement of the lingual dorsum seen in studies (Catalfamo, 2012 Ganesh, 2013 and Eren, 2017).It is noteworthy that the time of evolution of this lesion varies from months to years in adult patients, with cases over 10 years old and others over 3 months old, with the patient looking for due to a rapid growth of the lesion, in which it is sometimes possible to associate a traumatic or infectious factor and at other times no factor is associated. In the present case, it is believed that the patient already had the lesion, but due to some factor she felt that the lesion had grown and sought help. This shows that in adult patients, most of these malformations are well tolerated.The diagnosis of lymphangioma is based on clinical, imaging and microscopic characteristics (Eren, 2017). For superficial malformations, ultrasound and magnetic resonance are useful to detail the size, extent, components, postoperative monitoring and detection of recurrence (Smith, 2019; Abe, 2018; Nammour, 2016).

Table 1. Cases of lymphangioma in the oral cavity in adult patients

Study / year	Nº	Gender	Age	Evolution time	Anatomical site	Simptomatology	Imaginology	Histopatologic classification	Treatment	Outcome
Ikeda et al. [12]	1	Male	75 years	Up to 27 years	Apex of the Tongue	Dysphonia	Computed tomography	Cystic lymphangioma	Surgical removal	Five years of follow-up without recurrence
Hwang et al. [3]	1	Male	19 years	11 years	Back of Tongue	Bleeding	Magnetic resonance imaging	NR	Sclerotherapy with bleomycin 0.75 U / kg, three injections with two weeks of intralesional interval.	One year of follow-up, without recurrence
Catalfamo et al. [7]	9	5 Male and 4 Female	19 to 25 years	03 months	Tongue without other specification	Discomfort	Magnetic resonance imaging	nodular lymphangioma	Surgical removal	Six months of follow-up, without recurrence
Ganesh et al. [8]	1	Female	29 years	17 years	Back of Tongue	Increase in size	Ultrasound	Circumscribed lymphangioma	NR	NR
Eren et al. [9]	1	Female	29 years	18 years	Tongue without other specification	Bleeding and moderate pain	Did not perform	Microcystic lymphangioma	Patient did not want to undergo surgical treatment	Did not perform
Babu et al. [11]	1	Male	60 years	6 months	Left buccal mucosa	Asymptomatic	Did not perform	Circumscribed lymphangioma	Surgical removal	One year of follow-up, without recurrence
Eren et al. [9]	1	Male	27 years	Since childhood	Back of Tongue	Color change of the tongue	Did not perform	NR	Surgical removal	Regular follow-up with no sign of recurrence
Kolay et al. [10]	1	Female	32 years	NR	Left buccal mucosa	Increase in size	Did not perform	cavernous lymphangioma	NR	NR
Abe et al. [2]	1	Female	68 years	07 months	Left buccal mucosa	Increase in size	Magnetic resonance imaging	Microcystic lymphangioma	Surgical removal	Two-year follow-up, without recurrence
Carlan et al.	1	Female	36 years	08 months	Bilateral alveolar ridge and tongue belly	Bleeding	Did not perform	Microcystic lymphangioma	Surgical removal	Six months of follow-up, without recurrence

NR: Not related.

Microscopically, they are composed of lymphatic vessels that may show slight dilation (microcystic) or macroscopic structures similar to cysts (macrocystic) (Usha, 2014 and Nammour, 2016). The lymphatic vessels are located just below the surface epithelium and usually replace the conjunctive papillae, being able to observe the extension of these vessels into the deep connective tissue and to the skeletal musculature (Smith, 2019; Usha, 2014 and Nammour, 2019). Sometimes it may be necessary to use immunohistochemical markers to aid in diagnosis, such as podoplanin, vascular endothelial growth factor receptor 3, HA lymphatic endothelium receptor 1, D2-40 and Prox 1 (Abe, 2018). Kolay et al. (Kolay, 2018) used the monoclonal antibody D2-40 that identifies an oncofetal glycoprotein (M2A antigen) that is found in lymphatic endothelial cells, positively marking all their cases. The differential diagnosis consists of hemangioma, dermoid cyst, teratoma, amyloidosis, neurofibromatosis, granular tumor, neurofibroma, salivary gland tumors, gastric mucosal cyst heterotopia, meningoencephalocele (Abe, 2018 and Usha, 2014).The treatment of oral lymphangiomas varies from surgical therapy to non-surgical therapy with sclerosing agents and corticosteroids (Smith, 2019; Lerat, 2016). Management is individualized and often depends on the degree of functional impairment. Total surgical removal may not be possible in all cases, due to the large size or involvement of vital structures.

Recurrence after surgery is common, around 39%, especially for microcystic lymphangiomas of the oral cavity, due to its infiltrative nature (Smith, 2019; Lerat, 2016). Laser photocoagulation has been reported to be useful for controlling the size of the tongue and removing superficial lymphangioma in some cases (Usha, 2014).Sclerotherapy with one of these sclerotic agents: bleomycin, ethanol, acetic acid, sodium tetradecyl sulfate and OK-432, is more successful in macrocystic lymphangiomas, with the majority of patients showing clinical resolution of 75% to 100%.A lower success rate is obtained in the mixed and microcystic types [1], however, perform sclerotherapy with bleomycin 0.75 U / kg, three intralesional injections in an oral lymphangioma and obtained regression of the injury and no recurrence after one year of follow-up (Catalfamo, 2012). Intralesional steroids alter cell functions, resulting in regression of lymphatic malformation without a significant inflammatory reaction (Hwang, 2017). Conservative management, including adequate oral hygiene and regularly scheduled dental prophylaxis, should focus on reducing infectious complications (Usha, 2014).Spontaneous regression of lymphatic malformations is rare, but it has been reported in approximately 3% of cases (Lerat, 2016 and Eren, 2017). The prognosis is good for most of these patients. In the present study, surgical removal had good results, with no recurrence in a six-month follow-up, corroborating with (Catalfamo, 2012 and Eren, 2017).

It is believed that these lesions in adult patients are lesions that arose in childhood and that for a long time they did not show changes in the daily lives of these patients, making the diagnosis late. Possibly these lesions have a better prognosis than in pediatric patients and in cases of head and neck lymphangiomas. It is of fundamental importance to monitor these patients for a long period. Unfortunately, the follow-up period varies from six months to one year in the literature and clinical studies evaluating non-surgical therapies for oral lymphangiomas in adult patients are scarce, and further studies are needed.

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