

se logra únicamente con el estudio histopatológico. Enfatizamos en la importancia de considerar diversos diagnósticos diferenciales cuando tratemos con nódulos labiales, así como el envío a estudio histopatológico de todo tejido removido, a pesar de tener la confianza de un diagnóstico clínico inicial.

Clinical case

Acquired lymphangiectasia of lower lip. A clinical case report

Diana Antonieta Flores Flores,* Alicia Rumayor Piña,*
Karla Vértiz Félix,* Minerva Galindo Rocha,*
Sandra Cecilia Esparza Díaz,* Arlene Iñigo Berumen*

* Faculty of Dentistry, Autonomous University of Coahuila.

ABSTRACT

Background: The best-known pathology of lymphatic vessels is lymphangioma, which is considered a developmental malformation rather than a true neoplasia. Oral lymphangiomas are more common in children with half of the cases affecting tongue as clusters of translucent vesicles. However, single, circumscribed, and superficial lesions may appear intraorally. They are considered as focal ectasia of lymph vessels, known as lymphangiectasia, and can be secondary to obstruction or trauma. **Objective:** The goal of this report is to present a case of a lower lip superficial acquired lymphangiectasia and to compare the differential diagnoses on the basis of the clinical presentation. **Case presentation:** A six-year-old girl presented with an asymptomatic lower lip lesion. On examination two translucent and fluctuant small nodules were observed in lower labial mucosa. The first clinical diagnostic hypothesis was mucocele. Under local anesthesia, an excisional biopsy of both lesions was performed. A histopathological study revealed a superficial ectatic vascular space, lined by endothelium. Immunohistochemistry with D2-40 confirmed its lymphatic nature. After eight months of follow-up, the patient is well and shows no recurrence. **Conclusions:** This clinical case is an example of an acquired lymphangiectasia probably secondary to trauma to peripheral lymphatic capillaries caused by the use of a mandibular removable space maintainer. This entity is different from the deep diffuse lymphangiomas which are true vascular malformations and are generally present since birth. In oral mucosa, most lymphatic lesions are superficially located and most commonly represent lymphangiectasias and not true lymphangiomas.

Keywords: Lymphangiectasia, lower lip, mucocele.

BACKGROUND

Lymphangioma is a rare entity considered to be a congenital malformation of lymph vessels. Depending on its size, it has been classified as follows: a) simple, capillary or circumscribed, b) cavernous, and c) cystic lymphangioma. The latter occurs most often in the neck and can become large.¹ In the mouth, lymphangiomas are presented as diffuse clusters

of translucent vesicular-looking structures, mainly affecting the tongue of children, adolescents and young adults. Other affected sites are palate, jugal mucosa, gums, and lips.^{2,3} Generally, these lesions are superficial and could most likely represent ectatic or dilated peripheral lymphatic capillaries as a result of obstruction, local trauma, or postirradiation, especially in the case of superficial and circumscribed lesions. On the other hand, the expression of growth factors detected in cavernous lymphangiomas and cystic hygroma is absent in the superficial type; hence, the term lymphangiectasia has been suggested to describe these alterations.^{4,5} Some authors have reported superficial lymphangiomas using the term lymphangioma circumscriptum (LC), a well-recognized term in dermatology used for superficial lymphatic lesions of the skin and genital region.^{6,7} A key finding for diagnosis is the location of the immediately subepithelial dilated vascular spaces, superficially located in the lamina propria and lifting the epithelium.^{4,6} The first case documented and reported as LC in the mouth occurred in a postirradiated patient, affecting multiple areas of the oral mucosa.⁶ On the lower lip, there are only two reports of superficial lymphangioma, both of nodular appearance, circumscribed, of soft consistency, smooth surface, translucent, and fluctuating, mimicking mucocele. However, it was until the histopathological study that the final diagnosis was made. Noteworthy, they were documented as cavernous lymphangioma and superficial lymphangioma circumscriptum, respectively.^{8,9} This report aims to describe the case of a superficial lymphangiectasia, initially diagnosed as mucocele, in order to emphasize both the importance of considering other diagnoses on the basis of clinical characteristics and the need for histopathological analysis of all tissue removed from oral cavity.

CLINICAL CASE

A six-year-old girl presented to the Department of Pediatric Dentistry at the Faculty of Dentistry Unit Saltillo of the Autonomous University of Coahuila for evaluation of an asymptomatic lip lesion. The lesion had appeared one month earlier. She had no pathological history of relevance. On extraoral examination no alterations were identified, while intraorally two sessile, translucent nodules of fluctuating consistency and smooth surface, located in lower lip mucosa were observed. The largest measured 0.6 × 0.5 × 0.4 cm and the smaller one had similar characteristics and was adjacent to the first.

The patient had been using a mandibular removable functional space maintainer for 1 month; both nodules coincided with the area where the appliance hooks contacted the lip mucosa (*Figure 1*).

The rest of the oral mucosa showed no alterations. The initial clinical diagnosis was mucocele. An excisional biopsy of both lesions under local anesthesia was performed and the tissues were sent to histopathological study. The hematoxylin and eosin (H&E) staining revealed a large space located in the superficial lamina propria, lifting the keratinized stratified squamous epithelium, which was atrophic without epithelial processes. This space was lined by a layer of flat cells of endothelial morphology containing dispersed slightly basophilic amorphous material, as well as few polymorphonuclear cells. Toward the base, lobes of minor salivary gland and some bundles of striated muscle of normal characteristics were observed. To determine the nature of the cells making up the lining, immunohistochemistry studies were performed using the antibody D2-40 (Dako, 1:100), which showed diffuse positivity in the lining of the cavity confirming its lymphatic nature (*Figure 2*). Abundant small lymphatic structures were also observed at the base of the lesion. The final diagnosis was superficial lymphangiectasia. The patient recovered satisfactorily and after eight months of follow-up she has shown no recurrence.

DISCUSSION

Lymphatic intraoral alterations mainly affect the tongue in the form of multiple small, diffuse and superficial vesicles; sometimes the involvement is deeper and may produce macroglossia.² Some authors have preferred the term lymphangioma circumscriptum or even lymphangiectasia when referring to superficial lesions consisting mainly of ectatic lymph vessels. On the skin, localized and superficial lesions are reported with the term lymphangioma circumscriptum.¹⁰ It has been argued that intraoral lymphangioma circumscriptum is reactive in nature and not a congenital malformation; in these cases, it is considered of acquired type.⁶ In some reports of lymphangioma circumscriptum, besides superficial lymphatic dilation, epithelial hyperplasia with papillary appearance has been found, clinically characterized as small, well-delimited, violet lesions.¹¹ Clinical reports of oral cavity using the term lymphangiectasia are scant. Recently, two cases were described in patients with Crohn's disease, one in the lower lip and the other bilaterally in the lower vestibule. Both cases

had clinical presentation of multiple vesicular lesions. The possibility of obstruction and secondary ectasia of the lymph vessels due to the presence of granulomas is discussed in these patients.¹²

This case of superficial lymphangiectasia was initially diagnosed as mucocele due to its clinical characteristics of single, localized lesions. Mucocele is a common lesion of easy clinical recognition, mostly associated with local trauma involving lower lip.¹³ Less commonly, other soft tissue lesions may occur with similar clinical appearance. If the histopathological study is omitted, confirmation of the diagnosis is not possible. Even today, some dental professionals do not consider the histopathological study as an important diagnostic tool and discard removed tissues, which can lead to serious complications to the patient. The histopathological study allowed obtaining the diagnosis of superficial lymphangiectasia. Because this entity has no major complications and the recommended treatment is surgical removal, the discussion on clinical and histological differential diagnoses is mainly academic. There are few cases in the literature, most are associated with radiation history or trauma and some with mucocele-like clinical presentation. Noteworthy, these cases are reported as lymphangioma despite the atypical clinical presentation of single nodular-looking lesions. One of the most recent studies already uses the term lymphangioma circumscriptum referring to superficial ectatic lesions; in it, of 35 cases found in the literature with this diagnosis, none was located in lip mucosa.^{8,9,14}

CONCLUSIONS

The present case of unusual clinical appearance for lesions of lymphatic origin appears to be associated with the area of rubbing of the mandibular space maintainer. Both nodules were located adjacent to the appliance hook area. Due to the clinical presentation and the histopathological findings we prefer to use the term acquired lymphangiectasia. The diagnosis confirmation is achieved only with a histopathological study. We emphasize the importance of considering several differential diagnoses when dealing with lip nodules, as well as of sending to histopathological study all tissue removed, despite having the confidence of an initial clinical diagnosis.

REFERENCIAS / REFERENCES

1. Neville BW, Damm DD, Allen CM, Bouquot JE. *Oral and maxillofacial pathology*. 3^a ed. Elsevier, 2009.
2. Usha V, Sivasankari T, Jeelani S, Asokan GS, Parthiban J. Lymphangioma of the tongue - a case report and review of

- literature. *J Clin Diag Res*. 2014; 8 (9): ZD12-ZD14. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4225989/>.
3. Brennan TD, Miller AS, Chen SY. Lymphangiomas of the oral cavity: a clinicopathologic, immunohistochemical, and electron-microscopic study. *J Oral Maxillofac Surg*. 1997; 55 (9): 932-935. <https://www.ncbi.nlm.nih.gov/pubmed/9294501>.
 4. Itakura E, Yamamoto H, Oda Y, Furue M, Tsuneyoshi M. VEGF-C and VEGFR-3 in a series of lymphangiomas: Is superficial lymphangioma a true lymphangioma? *Virchows Arch*. 2009; 454: 317-325. <https://link.springer.com/article/10.1007/s00428-008-0720-8>.
 5. Bin Woo S. *Oral pathology. A comprehensive atlas and text*. 2ª ed. Elsevier. 2017.
 6. Poh CF, Priddy RW. Acquired oral lymphangioma circumscriptum mimicking verrucous carcinoma. *Oral Oncology Extra*. 2005; 41: 277-280. <http://www.sciencedirect.com/science/article/pii/S1741940905000579>.
 7. Patel GA, Schwartz RA. Cutaneous lymphangioma circumscriptum: frog spawn on the skin. *Int J Dermatol*. 2009; 48 (12): 1290-1295. <http://onlinelibrary.wiley.com/doi/10.1111/j.1365-4632.2009.04226.x/abstract>.
 8. De Carvalho FK, Pinheiro TN, Bezerra da Silva RA, de Queiroz AM, Bezerra da Silva LA, Nelson-Filho P. Lymphangioma of the lower lip mimicking a mucocele in children. *J Dent Child*. 2015; 82 (2): 116-119. <https://www.ncbi.nlm.nih.gov/pubmed/26349801>.
 9. Rodríguez RF, Báez PJ, Muriel CP. Linfangioma en labio inferior: una presentación inusual. *Av Odontostomatol*. 2008; 24 (3): 203-210. http://scielo.isciii.es/scielo.php?script=sci_arttext&pid=S0213-12852008000300002.
 10. Chakravarti A, Bhargava R. Lymphangioma circumscriptum of the tongue in children: successful treatment using intralesional bleomycin. *Int J Pediatr Otorhinolaryngol*. 2013; 77 (8): 1367-1369. <https://www.ncbi.nlm.nih.gov/pubmed/23732020>.
 11. Alrashdan MS, Hammad HM, Alzumaili BAI, Alorjani M. Lymphangioma circumscriptum of the tongue, a case with marked hemorrhagic component. *J Cutan Pathol*. 2018; 45 (4): 278-281. [dx.doi.org/10.1111/cup.13101](https://doi.org/10.1111/cup.13101).
 12. Galvin S, Flint SR, Toner ME, Healy CM, Ekanayake K. Oral lymphangiectasias and Crohn's disease: two case reports. *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2018; 126 (1): e31-e34. <https://doi.org/10.1016/j.oooo.2017.12.014>.
 13. Lewandowski B, Brodowski R, Pakla P, Makara A, Stopyra W, Startek B. Mucoceles of minor salivary glands in children. Own clinical observations. *Dev Period Med*. 2016; 20 (3): 235-242. <http://www.medwiekurozwoj.pl/articles/2016-3-12.pdf>.
 14. Katsoulas N, Tosios KI, Argyris P, Koutlas IG, Sklavounou A. Lymphangioma circumscriptum, angiokeratoma, or superficial vascular ectasia with epithelial hyperplasia? *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2014; e1-e5. <http://www.sciencedirect.com/science/article/pii/S221244031301688X>.

*Dirección para correspondencia /
Mailing address:*
Alicia Rumayor Piña
E-mail: ali84rum@hotmail.com