CASE REPORT

Lymphangioma of the oral cavity

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Abstract

Lymphangiomas are uncommon congenital hamartomas of the lymphatic system, usually diagnosed in infancy and early childhood. Commonly located at head and neck, they are rarely situated in the oral cavity. Preferred site of oral involvement is the tongue. In the absence of proper therapy, lymphangiomas of the tongue are extremely recurrent, leading to serious complications such as hemorrhage or obstruction of the upper respiratory airways. The authors present the case of eleven years old boy with pseudo-vesicles, and smooth, glossy lesions on the tongue, and a red prominent pulsative sublingual mass located at the base of the tongue. Both the macroscopic structure and the histological aspect sustain the diagnosis of lymphangioma. CT established that is a profound lymphangioma with a narrow communication with the superficial planes. *Conclusions*. Though rarely met in the oral cavity, lymphangiomas are an eventuality to take into consideration by the clinician. Early recognition is of utmost importance to initiation of proper treatment, and avoiding serious complication.

Keywords: **.**

Background

Lymphangioma is a benign hamartomatous tumor of lymphatic vessels with a marked predilection for the head and the neck region [1].

Described for the first time by Redenbacher in 1828, currently the lymphangiomas are classified as malformations and not as neoplasms [2].

Lymphangiomas represent about 6% of the total number of benign tumours of the smooth tissue in patients aged less than 20 years [3].

The oral localization of lymphangiomas is less usual, and the most common site in this case is the tongue, especially the 2 third anterior [1].

Lymphangiomas usually affect the ventral part of the tongue and are solitaire or circumscribed. Occasionally, they are associated with cystic hygroma [4].

We present a case of superficial lymphangioma of the tongue associated with a profound lymphangioma situated at the base of the tongue.

Patient and methods

At presentation in the Paediatric department, the patient was 9 years of age and reported abdominal pain, nausea, vomiting, and inconstant dyspeptic tools. At the same time he accused pain in the tongue at the contact with some aliments, leading sometimes to difficult alimentation. The parent informed us that in early childhood (at the age of 1) appeared little, asymptomatic prominences on the ventral part of the tongue (the first third and the tip). Then the lesions extended, and became more evident after ingestion of warm aliments. He was examined several times by physicians, and the lesions were diagnosed either as candidosis, either oral papilomatosis. Different antifungal treatments could not prevent the extension of the lesions.

The patient had no history of systemic disease, and the familial background excluded the existence of the similar clinical manifestations at any other family member.

At physical examination, the child was in good general condition and good nutrition state, without signs of obstruction of the superior respiratory tract. There were no cardio-pulmonary abnormal findings. Abdominal tenderness was present at superficial palpation of the epigastric and umbilical regions. Recently, the child reported difficult appetite, vomiting and dyspeptic stools.

Dermatological examination showed an enlarged reddish tongue, with numerous granular lesions, smooth, glossy, vesicle-like conglomerated in a placard affecting the two anterior thirds of the tongue including the tip, dorsal and lateral surfaces, with lesser involvement of the inferior one. The color of the lesion ranged from yellow to purple (Figures 1 and 2).

The mycological exam was positive for Candida infection, and recommended the administration of the antifungal medication.

Histological examination was performed on fragments of the lesions from the tip of the tongue showing squamous epithelium with acanthosis, papillomatosis and parakeratosis, and solitaire and clustered cystic spaces. Enlarged, clustered cystic spaces of the papillary derma presented a sheath of endothelial cells and included eosinophilic material. Lymphatic vessels, some of them with the thick walls were present in the profound derma and subcutaneous (Figures 3 and 4).

Computerized tomography showed at the base of the tongue the presence of a well delimitated mass of tissular density, protruding in the oropharynx, with slight inhomogeneous contrast substance retention. The CT aspect suggested profound lymphangiomatous tumor with a narrow communication with the superficial planes (Figure 5).

Local ultrasound showed normal Doppler arterial pattern in the common and internal carotid artery, billateral. There were no arterio-venous fistulas. Abdominal ultrasound was done to exclude hepatic or pancreatic lymphangiomas and showed no modification. Digestive endoscopy also excluded the involvement of gastrointestinal tract. The clinical and paraclinical data sustain the diagnosis of oral lymphangioma.

Discussion

Lymphangiomas are uncommon congenital hamartomas of the lymphatic system, usually diagnosed in infancy and early childhood as lobular masses or cystic lesions [5].

They may be present anywhere on the skin and mucosa. The most usual locations are the head and neck, followed by the proximal extremities, buttocks and trunk. Sometimes they can be located at intestinal, pancreatic and mesenteric level. Lymphangioma rarely affect the oral cavity. Affected sites in the oral cavity may include the tongue, palate, gingival and oral mucosa, lips, and alveolar ridge of the mandible [1].

The classifications of the lymphangioma are not standardized. Usually, they are divided depending on depth and size of the abnormal lymphatic vessels in two groups: a superficial one, including the circumscribed angyoma and another more profound, represented by cavernous lymphangioma and cystic hygroma. Many authors consider cystic hygroma as a cavernous lymphangioma variant. Lymphangioma circumscriptum is a localized congenital hamartoma of lymphatic vessels consisting of deep cavernous cisterns within the subcutaneous tissue, with secondary formation of more dilated lymphatics [6].

Two major theories have been proposed to explain the origin of lymphangiomas [2].

The first theory is that the lymphatic system develops from five primitive sacs arising from the venous system. Concerning the head and the neck, endothelial outpouchings from the jugular sac spread centrifugally to form the lymphatic system. Another theory proposes that the lymphatic system develops from mesenchymal clefts in the venous plexus reticulum and spread centripetally towards the jugular sac. Finally, lymphangioma develop from congenital obstruction or sequestration of the primitive lymphatic enlargement [7, 8].

These complex abnormal lymphatic patterns running from the cisterns to the skin surface, adopting a meandering course with frequent anastomosis and branching, were firstly described by Whimster IW *et al.* [9]. On the base of the three-dimensional lymphatic models rebuild *post-mortem*; he demonstrated the concept of "iceberg" in the case of lymphangiomas, the lymphatic involvement being much more important in depth than in surface. The lesions are composed from ectopic lymphatic system of lymphatics separated from the normal network of lymphatic vessels, yet communicating with the superficial lymphatics that become dilated by the continuous rise and fall of the pressure that is transmitted from the muscular walls of the deep cisterns. Thus, the blockage can determine the growth of the hydrostatic pressure with consecutive expansion of the lesions, untill an equal pressure with the contiguous tissues will be realized. The importance of the contiguous tissues is evident, as microcystic lesions are more common on the tongue, whereas macrocystic lesions predominate in the relatively compliant tissues of the neck [8].

Clinically, lymphangiomas of the oral cavity have a characteristic aspect. Usually, there is a plaque constituted from small vesicles with thin walls, translucide like frog eggs. Part of the vesicles are full with clear content (lymph), part has a blood content suggesting co-existence of the involvement of the lymphatic anomalies with abnormalities of the blood vessels. Sometimes, like in the case we present, they are associated with a diffuse, profound lymphangioma that appear like a submucous mass. This association explains the frequent recurrence, almost constant after excision [10]. Although lymphangiomas of the tongue are rare, this must be recognized early for optimal therapeutic result [11, 12].

The histopathology of lymphangioma circumscriptum is characterized by solitary and grouped dilated cystic spaces in the papillary derma. The cystic spaces are lined by endothelial cells often containing red blood cells, as well as lymphatic fluid. In the deep derma and subcutaneous fat, dilated lymphatic vessels are also seen, some of it containing thickened muscular walls [8].

A classification of the lymphangioma of head and neck on the base of the spread the anatomical involvement had been proposed by De Serres LM [13]:

1. Stage/class I – infrahyoid unilateral lesions;
2. Stage/class II – suprahyoid billateral lesions;
3. Stage/class III – suprahyoid or infrahyoid unilateral lesions;
4. Stage/class IV – suprahyoid bilateral lesions;
5. Stage/class V – suprahyoid or infrahyoid bilateral lesions;
6. Stage/class IV – infrahyoid bilateral lesions.

Ricciardelli LJ and Richardson MA [14] demonstrated that suprahyoid lymphangiomas had a significantly higher rate of recurrence than infrahyoid lymphangiomas [15].

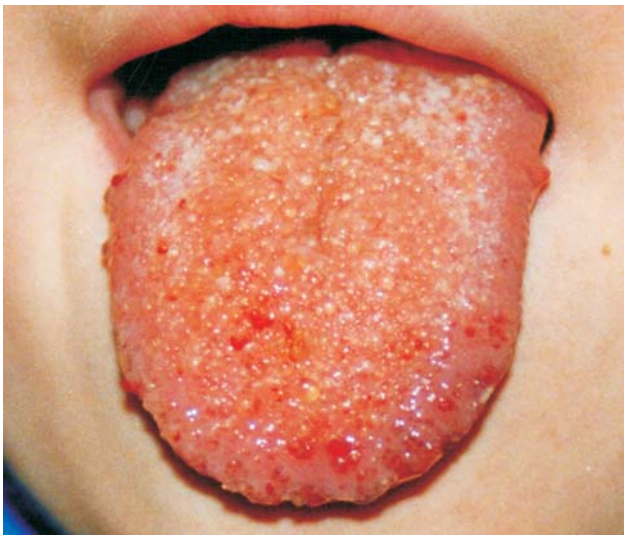

Figure 1 – ***Dorsal aspect of the tongue. Enlarged, reddish tongue with multiple granular lesions. Smooth, glowing glossy-shiny vesicle-like lesions affecting the tip and dorso-lateral surfaces of the tongue***

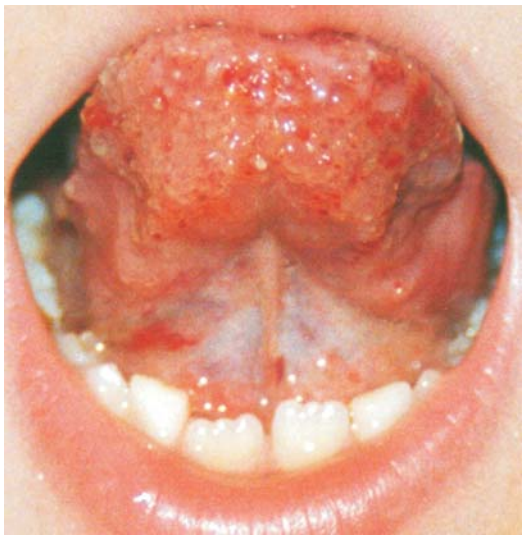

Figure 2 – ***Inferior aspect of the tongue covered by vesicle-like lesions. A reddish, prominent mass is visible at the base of the tongue***

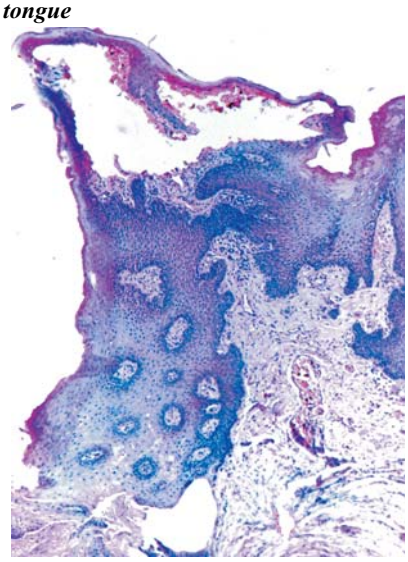

Figure 3 – ***Squamous epithelium with acanthosis, papillomatosis, parakeratosis and solitaire clustered cystic spaces***

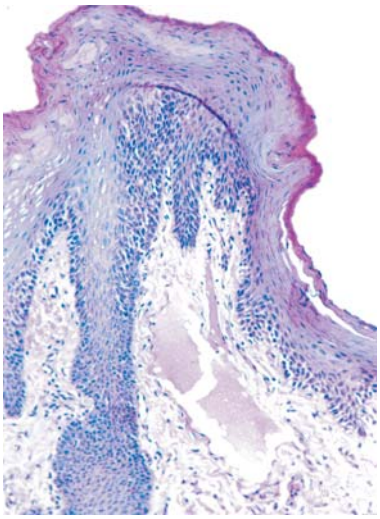

Figure 4 – ***Enlarged, clustered cystic spaces of the papillar derma, presenting a sheath of endothelial cells and included eosinophyllic material. Lymphatic vessels are present in the profound derma and subcutaneous tissue***

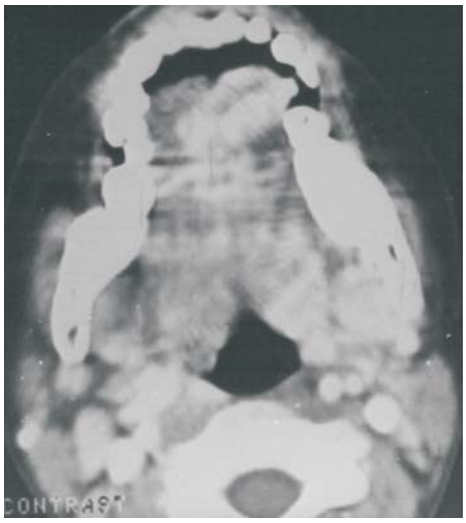

Figure 5 – ***Well-delimitated mass with tissue density protruding in the oropharynx, with discrete contrast substance retention***

Lymphatic malformations have a growing trend with the age of the child and rarely regress; clinically, their location determines the symptoms. While oral cavity is a specialized structure for speaking, deglutition, mastication and respiration, those lymphatic malformations can affect all these functions. Many patients may have no other symptoms except some cosmetic deformity.

The commonest complication of the lymphatic malformations is infection, associated with the growth of the lesion. Lymphocytopenia has been documented in all these patients, although a clear correlation with the infection risk must be established [16].

The rapid growth of lymphangiomas can be associated with hemorrhage or can lead to obstruction of the upper respiratory tract [15], 50% from the children with these lesions requiring tracheotomy [17].

The diagnostic of lymphangiomas is not difficult, being mainly clinical. Sixty percent of lymphangiomas are diagnosed at birth and 80–90% in the second year of life [2].

Lymphangioma may resemble to a number of oral lesions including hemangioma, teratoma, dermoid cyst, thyroglossal duct cyst, heterotopy of gastric mucosal cyst and granular cell tumor [18]. The differential diagnosis must also include, like in the case we presented, oral florid papillomatosis.

For inexperienced clinicians or in lesions with atypical clinical features a definitive diagnosis should be made trough biopsy and histopathologic examination [1].

Imagistic studies are important for the confirmation of the diagnosis. Prenatal ultrasound may be use for identifying cystic hygroma and also can monitored regression or recurrence. MRI is important for differentiate complex vascular malformations. Fetal MRI evaluates oral and cervical lesions previously detected by prenatal ultrasound, being also used to determine the potential of airway obstruction [19].

CT is superior to MRI for evaluation of the bone deformation and in detecting febolites caused probably by anterior intralesional hemorrhage.

Therapeutically, many approaches have been proposed. Spontaneous regression of the lesion is rarely encountered. Aspiration of the cystic content is temporary used as a measure to relieve airway obstruction, paying the price for the risk of introducing infection. Sclerozing agents are ineffective, probably as a result of the discontinuous basement membrane of the lymphatic vessels [20].

Neodymium–yitrium–aluminum Garnet (Nd-YAG) laser surgery has become widely preferred because of its advantages as less bleeding and edema versus standard methods of surgical resection [21].

Surgical excision is the usual treatment of lymphangioma. However, some clinicians do not recommend surgery for non-enlarging lymphangiomas of the tongue because of difficulties in removal and the high recurrence rate [1].

In choosing the ablative therapy of lymphangiomas the exact knowledge of the anatomy and spread of lesions are very important. In this case, in which CT proved the existence of a narrow communication between the lymphangioma from the base of the tongue and the superficial planes, the risk of recurrences after a surgical simple excision are high. We left to the surgeons the choice of the most adequate procedure.

Conclusions

Although rarely encountered in the oral cavity, lymphangiomas represent a condition that must be recognized. Their early recognition allows proper initiation of treatment and prevents the occurrence of the complications.

References

[1] JAMAROON A., PONGSIRIWET S., SRISUWAN S., KRISANAPRAKORNKIT S., *Lymphangiomas of the tongue*, Int J Pediatric Dentistry, 2003, 13:62–63.

[2] PATEL J. N., SCIUBBA J., *Oral lesions in young children: Pediatric Clinics of North America*, 2003, 50(2):469–486.

[3] COFFIN C. M., DEHNER L. P., *Vascular tumours in children and adolescents: a clinicopathologic study of 228 tumours in 222 patients*, Pathol Annu, 1993, 28(Pt 1):97–120.

[4] KARMODY C. S., FORTSON J. K., CALCATERRA V. E., *Lymphangiomas of head and neck in adult*, Otolaryngol Head Neck Surg, 1982, 90(3 Pt 1):283–288.

[5] TASCA R. A., MYATT H. M., BECKENHAM E. J., *Lymphangiomas of the oral cavity*, Int J Pediatrics Otorhinolaryngol, 1999, 51:201–205.

[6] WILSON G. R., COX N. H., MCLEAN N. R., SCOTT D., *Squamous cell carcinoma arising within congenital lymphangioma circumscriptum*, Br J Dermatol, 1993, 129(3):337–339.

[7] BILL A. H., SUMMER D. S., *A unified concept of lymphangiomas and cystic hygroma*, Surg Gynecol Obstet, 1965, 120:79–86.

[8] MCALVANY J. P., JORIZZO J. L., ZANOLLI D. *et al.*, *Magnetic Resonance Imaging in the evaluation of lymphangioma circumscriptum*, Arch Drematol, 1993, 129(2):194–197.

[9] WHIMSTER I. W., *The pathology of lymphangioma circumscriptum*, Br J Dermatol, 1976, 94:473–486.

[10] LONGHIN S., DIMITRESCU A. L., WOLFSHAUT A., *Dermatologie infantilă*, Ed. Medicală, Bucureşti, 1979, 313–314.

[11] BROCK M. E., SMITH R. J. N., PAREY S. E., MOBLEY D. L., *Lymphangioma. An otolaryngologic perspective*, Int J Pediatr Otolaryngol, 1987, 14:133–134.

[12] LOBITZ B., LANG T., *Lymphangioma of the tongue*, Pediatr Emerg Care, 1995, 3:183–185.

[13] DE SERRES L. M., SIE K. C., RICHARDSON M. A., *Lymphatic malformation of the head and neck: a proposal for staging*, Arch Otolaryngol Head Neck Surg, 1995, 121:577–582.

[14] RICCIARDELLI L. J., RICHARDSON M. A., *Cervicofacial cystic hygroma patterns of recurrence and management of the difficult case*, Arch Otolaryngol Head Neck Surg, 1991, 117:546–553.

[15] HARTL D. M., ROGER G., DENOYELLE F., *Extensive lymphangioma presenting with upper airway obstruction*, Arch Otolaryngol Head Neck Surg, 2000, 126:1378–1382.

[16] TEMPERO R. M., HANNIBAL M., FINN L. S. *et al.*, *Lymphocytopenia in children with lymphatic malformation*, Arch Otolaryngol Head Neck Surg, 2006, 132:93–97.

[17] ALQAHTANI A., NGUYEN L. T., FLAGEOLE N. *et al.*, *25 years experience with lymphangiomas in children*, J Pediatr Surg, 1999, 34(7):114–118.

[18] LALWANI A. K., ENGEL T. L., *Teratoma of the tongue: a case report and review of the literature*, Int J Peditr Otorhinolaryngol, 1992, 24:261–268.

[19] YONETSU K., NAKAYAMA E., KAWAZU T. *et al.*, *Value of contrast – enhanced magnetic resonance imaging in differentiation of hemangiomas from lymphangiomas in the oral and maxillofacial region*, Oral Surg Oral Med Oral Pathol Oral Radiol Endod, 1999, 88(4):496–500.

[20] BRENNAN T. D., MILLER A. S., CHEN S. Y., *Lymphangiomas of the oral cavity: a clinicopathologic immunohistochemical and electronomicroscopic study*, J Oral Maxillofacial Surg, 1997, 55:932–935.

[21] TASAR F., TUMER C., SENER B. C., SENEIFT K., *Lymphangioma treatment with Nd-YAG laser*, Turk J Pediatr, 1995, 37(3):253–256.

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