

Oral manifestations and dental care management of a young patient with lymphangioma of the tongue: A case report.

Farah Chouchene¹, Fatma Masmoudi¹, Ahlem Baaziz¹, Fethi Maatouk¹, and Hichem Ghedira¹

¹University of Monastir Faculty of Dental Medicine of Monastir

January 26, 2021

Abstract

Lymphangiomas of the tongue are very rare tumors usually diagnosed in infancy and early childhood. Children with Lymphangiomas of the tongue may require multidisciplinary care and a close collaboration between physician and dentist to be able to eradicate any sources of dental infections before undertaking medical treatment.

Title Page:

Title: Oral manifestations and dental care management of a young patient with lymphangioma of the tongue: A case report

Short title: Oral rehabilitation in case of lymphangioma of the tongue.

1-Farah Chouchene, Assistant professor in Pediatric Dentistry. Pediatric and Preventive Dentistry Department, Faculty of Dental Medicine. ABCD F Laboratory of Biological, Clinical and Dento-Facial Approach, University of Monastir, Monastir, Tunisia. (Corresponding author)

ORCID ID: <https://orcid.org/0000-0003-0256-913X>

2- Fatma Masmoudi, Professor in Pediatric Dentistry. Pediatric and Preventive Dentistry Department, Faculty of Dental Medicine of Monastir, Tunisia. ABCD F Laboratory of Biological, Clinical and Dento-Facial Approach, University of Monastir, Monastir, Tunisia.

<https://orcid.org/0000-0002-7339-3349>

3-Ahlem Baaziz. Professor in Pediatric Dentistry. Pediatric and Preventive Dentistry Department, Faculty of Dental Medicine of Monastir. ABCD F Laboratory of Biological, Clinical and Dento-Facial Approach, University of Monastir, Monastir, Tunisia.

<https://orcid.org/0000-0003-0673-9079>

4-Fethi Maatouk. Professor in Pediatric Dentistry. Pediatric and Preventive Dentistry Department, Faculty of Dental Medicine of Monastir. ABCD F Laboratory of Biological, Clinical and Dento-Facial Approach, University of Monastir, Monastir, Tunisia.

<https://orcid.org/0000-0001-7320-8425>

5- Hichem Ghedira. Professor in Pediatric Dentistry. Pediatric and Preventive Dentistry Department, Faculty of Dental Medicine of Monastir. ABCD F Laboratory of Biological, Clinical and Dento-Facial Approach, University of Monastir, Monastir, Tunisia.

<https://orcid.org/0000-0002-1594-646X>

Correspondent author: Farah Chouchene
Faculty of Dental Medicine of Monastir. Tunisia
Address: Rue Hedi Chekir. Hiboun 5111 Mahdia
Phone: 0021622821412

Email: farah.pedo@gmail.com

Oral manifestations and dental care management of a young patient with lymphangioma of the tongue: A case report

Abstract:

Lymphangiomas of the tongue are very rare tumors usually diagnosed in infancy and early childhood. Children with Lymphangiomas of the tongue may require multidisciplinary care and a close collaboration between physician and dentist to be able to eradicate any sources of dental infections before undertaking medical treatment.

Keywords: Benign congenital tumors, Lymphangioma, Macroglossia, Sirolimus, Dental management, Case report.

Key Clinical Message: A multidisciplinary approach, a good awareness of the patient's medical history and the oral manifestations of the underlying conditions are imperative during the oral care of children with tongue lymphangioma.

Introduction

Lymphangiomas are uncommon developmental anomalies and congenital tumors of the lymphatic system diagnosed mostly in children under the age of five years as cysts or lobulated masses, localized in the head or the neck.^{1,2} However, the involvement of the tongue, is considered very rare.²

Lymphangiomas of the tongue are caused by the formation of benign tumors on the dorsum of the tongue, but it could also occur in the palate, gingiva, buccal mucosa and, even lips.³

Tongue lymphangiomas typically demonstrated multiple blisters like a pebbly surface, nodules that appear like a cluster of translucent vesicles on the enlarged dorsal surface of the tongue, but, these lesions may also be diffuse involving large portions of the tongue causing macroglossia.^{4,5}

Aetiological factors can include maternal viral infection, maternal substance abuse, and environmental factors.⁶

Lymphangiomas are known to be associated with some syndromes such as Turner syndrome, Trisomies, fetal alcohol syndrome, cardiac anomalies, Noonan's syndrome and, familial hydrops.⁶

Histopathologically, lymphangiomas have been classified into; lymphangioma simplex which exhibits small capillary-sized vessels, cavernous lymphangioma which is composed of large dilated lymphatic vessels, and cystic lymphangioma or cystic hygroma which consists of large macroscopic cystic spaces.^{2,7}

These developmental anomalies are the most common cause of macroglossia in infancy,⁴ and patients with lymphangioma of the tongue commonly present with difficulty to retract and to move their massive tongue which may cause speech disturbances, difficulty in chewing and swallowing and compromise the mechanical plaque removal which enhances the caries risk.⁸

Complications are numerous and frequent, including air-way compromise, infection, bleeding, and dental caries.⁹

This case report aimed to discuss the most relevant oral manifestations and to describe the dental care management of a four-year-old female patient diagnosed with lymphangioma of the tongue undergoing medical treatment with Sirolimus.

Case report

This case report was prepared according to the CARE case report Guidelines.¹⁰

A four-year-old female patient was admitted to the Department of Pediatric and Preventive Dentistry at The Faculty of Dental Medicine of xxxx.

The patient was referred for general dental care by the Department of Dermatology at the Hospital of Farhed Hached xxxx, where she had been diagnostic with microcystic lymphangioma of the tongue and undergone medical treatment with Sirolimus.

The patient was born to healthy non-consanguineous parents following an uneventful full-term pregnancy.

The detailed family history revealed no relevant systemic conditions, and in the head and neck region, no other mass was detected.

The young patient's medical history revealed that the disease dates to the first day of birth when the patient presented with dyspnea and was admitted to the emergency room.

The evolution was then marked by the appearance at the age of five months of micro-vesicles in the tongue, followed by a swelling of the tongue interfering with eating and breathing.

The tongue was swollen, bluish in appearance surmounted by microvesicles with hematic content, and blackish scabs interfering with feeding and mouth closure.

The patient received several symptomatic treatments without improvement, and at the age of one and a half, the mother reported that the swelling of the tongue progressed.

At the age of two years, the diagnosis of microcystic lymphangioma of the tongue was made. The patient was treated by Sirolimus for six months with an improvement of the symptoms.

The actual general examination of the patient does not reveal any abnormalities. Only a notion of recent weight loss was reported because of the chewing disorder caused by the macroglossia.

The medical history of the patient revealed also that the patient was admitted twice to the Pediatric Intensive Care Unit for bacterial pneumonia.

On admission in our department, the patient presented with an enlarged tongue with nodular, fluffy, white, and infected pink-purple lesions on the superior and inferior aspects of the tongue (Figure 1). The patient presented also speech disturbances, swallow inability, breathing difficulty and complained of burning sensation of the tongue.

The extra-oral examination showed a convex profile with increased lower facial height.

The intraoral clinical examination revealed, poor oral hygiene with plaque accumulation, maxillary primary incisors with extensive caries, an active cavitated carious lesion on teeth 64 (Figure 2), and anterior open-bite (Figure 3).

A panoramic radiographic examination showed extensive carious lesions and root resorption in the maxillary incisors.

A mandibular deformity was also seen on the orthopantomography.

The patient presented with an anterior open bite, an increased mandibular body-ramus angle, an elongated mandibular body, and a displaced anterior teeth (Figure 4).

Since even brushing teeth was considered to be painful for the patient, the main objective of the treatment plan was to try to motivate her to change her behavior, therefore the treatment began with preventive measures including; diet counseling, basic instructions in oral health care, and a discussion on improving oral home care skills by including training on mechanically removing dental plaque and a supervised toothbrushing.

Because the patient was currently undergoing treatment with Sirolimus to help decrease the size of the lesion, the dental treatments were carried out under antibiotic prophylaxis. This decision was made in consultation with the patient's physician.

After administering local anesthesia, tooth 64 was isolated using a rubber dam. The entire carious tissue was removed and the tooth was sealed with glass ionomer cement GIC (Riva self-cure, SDI, Australia) than the extraction of the upper incisors (Teeth: 51-52-61-62) was done.

The surgical and conservative treatments were preceded by professional prophylaxis and topical applications of 2% Neutral Sodium Fluoride Gel (Master-Dent®), USA).

Follow-up visits, every three months, in collaboration with the Dermatology Department have been scheduled for the restorative treatment re-evaluation, observation for the onset of new lesions, monitoring the tumor evolution and the size of the patient's tongue to achieve a removable partial denture to maintain the space of the incisors while correcting the functions, improving the aesthetics and correcting the anterior open bite (Figure 5).

Discussion

Lymphangiomas are a relatively rare benign congenital tumors of the lymphatic system.¹¹

Most of the cases are detected since birth and about 80% are developed before the age of 2 years.^{12,13}

In the present case report, the patient presented a history of an enlarged tongue since the age of five months, the tongue continued to swell, and thereafter caused feeding and breathing but the diagnosis of microcystic lymphangioma of the tongue was confirmed not until the age of two years.

When lymphangiomas involve the tongue, a condition frequently referred to "macroglossia" can occur and can be associated with a set of problems unique to this anatomic location.¹⁴

Macroglossia may lead to two main types of complications: it can cause dentoskeletal problems, such as anterior open bite, mandibular prognathism with an increase in mandibular length, higher gonial angle, anterior facial height and excessive proclination of the mandibular incisors; and functional deficits such as difficulty in drooling, swallowing, obstruction of the upper airway, and alterations in speech.¹⁵

In the present case report in addition to the dentoskeletal and functional problems, the patient presented poor oral hygiene associated with extensive carious lesions.

The clinical appearance of lymphangioma depends essentially on the lesion extension. The superficial lesion presents as papillary lesions with pebbly surface due to the occurrence of several translucent vesicles with the same color as that of adjacent mucosa or occasionally with a mild reddish hue. These lesions can sometimes present a particular aspect such as a tapioca pudding or frog eggs like appearance. While the deeper lesions manifest as diffuse nodules which are soft in consistency and with negligible alterations in color or texture.¹⁶

Due to infection or hemorrhage, lymphangiomas in a child can grow suddenly but they can also shrink spontaneously.⁴

These lesions can be described as localized or diffuse growth.¹²

Treatment's objectives of tongue lymphangioma are essentially the restoration of tongue size, the preservation of taste, correction of mandibular and dental deformities.⁴

These congenital malformations can be treated with radiofrequency ablation, surgery, sclerotherapy, interferon, and corticosteroids.¹⁷

Conserved approaches like surgical excision with low relapse rates, cryotherapy, and laser can be considered for the treatment of superficial and/ or localized lymphangiomas.^{13,17}

While the management of diffuse lymphangioma can be more complicated because the lesion can involve a more extensive area which was the case in the present report.¹²

Microcystic lesions do not respect tissue planes, are diffuse and generally difficult to eradicate.¹⁶

In the present report, due to the diffuse nature of the lesion, the patient was treated with Sirolimus; 1.5 mg/kg/day administered in two divided daily doses.

The Sirolimus is an immunosuppressive and antitumor agent that belong to the Mammalian target of rapamycin (mTOR) inhibitors group which plays an important role in cellular anabolism, catabolism, cell growth, and angiogenesis.^{17,18}

This molecule has been used in patients with tuberous sclerosis to decrease the size of the lesions of angiomyolipoma and lymphangioliomyomatosis (LAM).^{17,19}

Most recently, oral sirolimus, which can inhibit lymphatic vessel regeneration, invasion, LAM growth, and vascular endothelial growth factor VEGF secretion, has been reported to be efficacious for patients with lymphangioma of the tongue.^{17,20}

As an immunosuppressant, sirolimus can cause neutropenia and may predispose the patient to an increased risk of infection.²¹

Ideally, all dental procedures should be completed before immunosuppressive therapy is initiated but when the dental needs cannot be treated before therapy, priorities should be infections, extractions and periodontal care.²²

In the present case report, because the patient was undergoing treatment with Sirolimus, the dental treatment was carried out under antibiotic prophylaxis.

Clamoxyl ® with a dose of 50mg/kg orally as a single dose one hour before to the procedure was prescribed for the patient.

For dental procedures, the decision regarding the need for antibiotic prophylaxis should be made in consultation with the patient's physician. Immunosuppressive therapy may cause many acute and long-term side effects especially in the oral cavity, and any existing or potential sources of dental infections can compromise the medical treatment.²²

In this report, giving the general health status of the patient, to be able to perform a removable partial denture and to correct the anterior open-bite, check-ups every three months have been scheduled to control the reduction of the size of the tongue.

Conclusion

Young patients with lymphangioma of the tongue, usually presenting with macroglossia, may not only have a significant chewing and breathing problems but also malocclusions and speech disorders, and dental problems. The enlarged tongue with partial or total immobility compromises the mechanical removal of the dental plaque which can lead to an increase of the caries risk in these children. For this reason, dental intervention must be done promptly and efficiently, with attention to the patient's medical history, treatment protocol, and health status.

Abbreviations:

mTOR: Mammalian target of rapamycin.

LAM: Lymphangioliomyomatosis.

Declarations:

Ethics approval and consent to participate: Not applicable.

Consent for publication: Written informed consent was obtained from the legal guardian of the patient (the father of the child) for publication of this case report and any accompanying images.

Availability of data and materials: All data generated and analysed which related this case report are included in this published article.

Competing interests: None of the authors has declared any conflict of interest or financial disclosures.

Funding: There was no financial support for this study.

Acknowledgments: Authors do not have any conflict of interest related to this study.

Author contribution: FC and FM performed the initial examination and collected clinical data, FC provided the clinical dental care of the patient and have continued performing regular clinical and radiographic follow-up. AB supervised the clinical dental care of the patient. FC was responsible for the literature search and wrote the paper. FM revised and edited the manuscript and figures. HG provided comprehensive judgement and assisted in editing the final version of the manuscript. All authors read and approved the final version of the manuscript prior to submission.

References:

1. Lobtiz BI, lang T. Lymphangioma of the tongue. *Pediatr Emerg Care* . 1995;11:183-185.
2. Balakrishnan A, Bailey CM. Lymphangioma of the tongue. A review of pathogenesis, treatment and the use of surface laser photocoagulation. *J Laryngol Otol* . 1991;105:924-929.
3. Farronato M, Fama A, Lanteri V, Nucci F, Farronato G, Maspero C. Lymphangioma of the tongue associated with open bite: case report. *Eur J Paediatr Dent* . 2019;20:311-314.
4. Kheur SM, Routray S, Ingale Y, Desai RS. Lymphangioma of tongue: a rare entity. *Indian J Dent Adv* . 2011;3:635-638.
5. Yaita T, Onodera K, Xu H, Ooya K. Histomorphometrical study in cavernous lymphangioma of the tongue. *Oral Dis* . 2007;13:99-104.
6. Barry S, Allotey J, Brundler AM, Duggal MS. Case report: cystic hygroma. *Eur Arch Paediatr Dent Off J Eur Acad Paediatr Dent* . 2012;13:323-325.
7. Rathan JJ, Vardhan BH, Muthu MS, Venkatachalapathy, Saraswathy K, Sivakumar N. Oral lymphangioma: A case report. *J Indian Soc Pedod Prev Dent* . 2005;23:185.
8. Cheng L-H, Lee J-C, Kao C-H, Shi Z-P, Lin Y-S. Lymphangiomatous macroglossia associated with extensive cervicomediastinal cystic hygromas. *J Chin Med Assoc* . 2013;76:653-656.
9. Padwa BL, Hayward PG, Ferraro NF, Mulliken JB. Cervicofacial lymphatic malformation: clinical course, surgical intervention, and pathogenesis of skeletal hypertrophy. *Plast Reconstr Surg* . 1995;95:951-960.
10. CARE Checklist. CARE Case Report Guidelines. <https://www.care-statement.org/checklist>
11. McDonald JS. Chapter 28 - Tumors of the Oral Soft Tissues and Cysts and Tumors of Bone. In: Dean JA, ed. *McDonald and Avery's Dentistry for the Child and Adolescent (Tenth Edition)* . Mosby; 2016:603-626.
12. de Queiroz AM, Silva RAB, Margato LC, Nelson-Filho P. Dental care management of a young patient with extensive lymphangioma of the tongue: a case report. *Spec Care Dent Off Publ Am Assoc Hosp Dent Acad Dent Handicap Am Soc Geriatr Dent* . 2006;26:20-24.
13. Erugula SR, Mvs S, Sameera A, Vujhini SK, Kandukuri MK. A rare case of oral lymphangioma of tongue. *Int J Contemp Pediatr* . 2016;3:1112-1114.
14. Morfit HM. Lymphangioma of the tongue. Treatment and appraisal of long-term results. *Arch Surg Chic Ill 1960* . 1960;81:761-767.

15. Abeleira MT, Seoane-Romero JM, Outumuro M, Caamaño F, Suárez D, Carmona IT. A multidisciplinary approach to the treatment of oral manifestations associated with Beckwith-Wiedemann syndrome: a long-term case report. *J Am Dent Assoc* 1939 . 2011;142:1357-1364.
16. V U, Sivasankari T, Jeelani S, Asokan GS, Parthiban J. Lymphangioma of the tongue - a case report and review of literature. *J Clin Diagn Res JCDR* . 2014;8:12-14.
17. Akyüz C, Ataş E, Varan A. Treatment of a tongue lymphangioma with sirolimus after failure of surgical resection and propranolol. *Pediatr Blood Cancer* . 2014;61:931-932.
18. Vignot S, Faivre S, Aguirre D, Raymond E. mTOR-targeted therapy of cancer with rapamycin derivatives. *Ann Oncol Off J Eur Soc Med Oncol* . 2005;16:525-537.
19. Bissler JJ, McCormack FX, Young LR, et al. Sirolimus for angiomyolipoma in tuberous sclerosis complex or lymphangioma. *N Engl J Med* . 2008;358:140-151.
20. Fetoui NG, Boussofara L, Gammoudi R, Belajouza C, Ghariani N, Denguezli M. Efficacy of sirolimus in the treatment of microcystic lymphatic malformation of the tongue. *J Eur Acad Dermatol Venereol* . 2019;33:336-337.
21. Hammill AM, Wentzel M, Gupta A, et al. Sirolimus for the treatment of complicated vascular anomalies in children. *Pediatr Blood Cancer* . 2011;57:1018-1024.
22. AAPD. Dental Management of Pediatric Patients Receiving Immunosuppressive Therapy and/or Radiation Therapy. *Pediatr Dent* . 2018;40:392-400.

List of figure Legends :

Figure 1: Oral examination showing enlarged tongue with nodular, fluffy, white and infected pink-purple lesions on the superior and inferior aspects of the tongue. 9

Figure 2: Intraoral examination showing maxillary primary incisors with extensive caries and active cavitated carious lesion on teeth 64. 9

Figure 3: Intraoral examination showing anterior open bite. 9

Figure 4: Panoramic radiograph showing the presence of caries lesions on the maxillary primary incisors. 10

Figure 5: Appearance of the maxillary anterior region after extraction of the primary incisor at three months follow-up. 10

Hosted file

figure lymphangioma.pdf available at <https://authorea.com/users/391838/articles/505837-oral-manifestations-and-dental-care-management-of-a-young-patient-with-lymphangioma-of-the-tongue-a-case-report>