

## What about the Intra-parotid cystic lymphangioma in children?

Reda Hejjouji, Jawad Lahma, Safaa Rokhssi, Redallah Zrarqi, Razika Bencheikh, Mohammed Anas Benbouzid, Abdelilah Oujilal and Leila Essakalli.

ENT Department, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco.

### ARTICLE INFO

#### Article history:

Received: 04 January 2018;

Received in revised form:  
13 March 2018;

Accepted: 23 March 2018;

#### Keywords

Cystic lymphangiomas,  
Parotid gland,  
Children,  
Parotidectomy.

### ABSTRACT

Cystic parotid lymphangiomas are benign vascular tumors that rarely appear in adults; they are usually detected in the first two years of life. Their appearance is due to an abnormal development of the lymphatic system during embryogenesis. Herein we report on two cases of intra-parotid cystic lymphangioma that occurred in teen age. A six year girl and a young girl of 13 years age were admitted in our unit because of an indolent and renitent isolated mass occupying the right parotid region. The diagnostic of an intra-parotid cystic lymphangioma was clinically advocated on echographic and CT imaging findings and then definitely proven after histopathologic studies of the ablation fragments. A total parotidectomy was made, removing the entire tumor. The parotid localization of cystic lymphangioma makes their surgical management difficult because of the development of lymphangioma between the branches of the facial nerve. The question of the benefit / risk ratio makes the therapeutic decision more complex because the aggressiveness of the radical tumor resection encounters a benign tumoral origin. It opposes two fundamental principles, first that of being radical on the tumor which is very recurrent and on the principle of remaining functional while preserving the function of the facial nerve.

© 2018 Elixir All rights reserved.

### Introduction

Cystic lymphangiomas of the parotid gland are benign vascular tumors occurring rarely in the adult population. They can be seen anywhere in the body, but most often in the cervicofacial region, especially in the posterior triangle of the neck [1, 2]. They are more frequent during the first 2 years of life and both sexes are affected equally. Their occurrence is due to an embryonic developmental anomaly of the lymphatic system [2]. Herein we report on two cases of intra-parotid cystic lymphangioma that occurred in teen age.

### Case reports.

#### Case 1.

A six year girl was admitted in our unit because of an indolent and renitent isolated mass occupying the right parotid region, present at birth and characterized by iterative inflammatory episodes. The diagnostic of an intra-parotid cystic lymphangioma was clinically advocated on echographic and CT imaging findings (no MR imaging because of lack of financial resources) (figure 1) and then definitely proven after histopathologic studies of the ablation fragments. A total parotidectomy was made, removing the entire tumor. Post-operatively, the child presented again with peripheral facial paralysis, but no recurrence occurred in the course.

#### Case 2.

A young girl of 13 years age, with no specific history, presented with an 8-month course firm and indolent mass (4 cm) in the right parotid, with no another clinical sign nor anomaly. Computer tomography studies exhibited a cystic structure in the parotid with intra-cystic trabeculas (figure 2).

A right parotidectomy was done together with a flap positioning (SMAS). Post-operative course was uneventful and

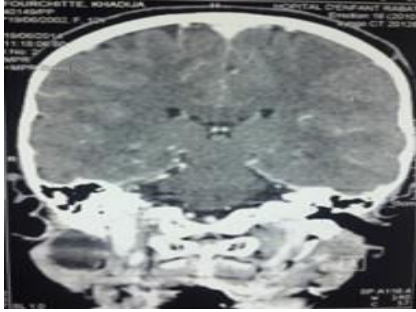
after a follow-up period of 28 months, the patient still remains in good condition.



Figure 1. Cystic structure in the right parotid.

Tele:

E-mail address: [redahejjouji@gmail.com](mailto:redahejjouji@gmail.com)



**Figure 2. Cystic structure in the parotid with intra-cystic enhanced trabeculas.**

#### Discussion.

The origin of lymphangiomas is not clearly understood. Most authors favor the theory that dysplastic lymphatic tissue is sequestered in a target tissue during the fetal development. 6 lymphatic sacs (2 jugulars, 2 iliac, 1 root of mesentery and 1 abdominal aorta) develop in the 8th week of gestation. Later, communications are established between lymphatics of various regions and with the venous system. It is suggested that the majority of lymphangiomas arise from parts of lymph sacs that fail to establish these connections.<sup>1</sup> The disconnected lymphatic structures may dilate and evolve to a cystic formation. Lymphangiomas may also be acquired after surgery or trauma,<sup>2</sup> infection, or chronic inflammation [1, 3, 4].

Lymphangiomas are divided into three categories: capillary, cavernous, and cystic type. Cystic types have the potential for extensive infiltration of surrounding tissues and lead to surgical difficulties. Therapeutically, they are classified into macrocystic, microcystic or mixed lymphangiomas [5].

Malignant transformation has never been reported. The mass evolves in three ways: spontaneous regression, slow progression, rapid enlargement. Infection within the cyst is usually caused by staphylococcus or streptococcus species. Nerve paralysis by compression could be secondary to the hemorrhage within the cyst. Lymphangioma can infiltrate and cause osteolysis of adjacent bones. An ulceration or rupture of the cyst has been reported [2, 5, 6].

Lymphangiomas can easily be diagnosed from history and physical examination. Nevertheless, imaging studies play an important role to confirm the diagnosis and to exclude other possible differentials of congenital lesions namely branchial cyst, thyroglossal cyst, teratoma and lipoma [7-9]. Computed tomography and magnetic resonance imaging play a significant role in surgical management of lymphangiomas. They help to delineate extension and anatomy study before operation to avoid iatrogenic injury to vital structures [7-9].

Because of its development between the facial nerve ramifications, the occurrence of a cystic lymphangioma in the parotid gland renders its surgical management more difficult. The ratio concerning the benefits versus the risks also renders decision making process more complex because one needs to confront radical tumor ablation to the presence of a benign tumor [10, 1&].

Two fundamental principles are opposed in the management of intra-parotid lymphangioma: firstly, to have a radical approach when facing such a tumor with a high rate of recurrence; secondly, to preserve the function when preserving facial nerve function [11, 12].

Treatment is indicated in cases where potential complications are anticipated such as primarily airway compromise as well as aesthetic issue or the size of the tumor.

Surgery remains the mainstay of treatment. As lymphangiomas often infiltrate adjacent structures like great vessels and nerves, complete removal is technically challenging. On the other hand, partial removal leads to recurrence. Postoperative complications occurred in 12–33% of cases and high recurrence rate of 15–53% did occur even in expert hands [13].

Sclérothérapie has been advocated but is less used because of its potential of scar retraction, which raised aesthetic concern and also making subsequent surgery more difficult [14, 15].

#### Conclusion.

Even if lymphangiomas can easily be diagnosed from history, physical examination and imaging studies, its management is challenging and must always been done on an individual case basis, taking to account the potential risks on adjacent vital structures.

#### References.

- Bailey CM.** Cystic hygroma. *Lancet*. 1990;335(8688):511–512.
- Alqahtani A et al.** 25 years experience with lymphangiomas in children. *J Pediatr Surg*. 1999;34(7):1164–1168.
- Oak SN, Redkar RG, Kapur VK.** Posterior midline cervical fetal cystic hygroma. *J Postgrad Med*. 1992;38:93–96.
- Kumar N, Kohli M, Pandey S, Tulsu SP.** Cystic hygroma. *Natl J Maxillofac Surg.*, 1 (1) (2010), pp. 81–85.
- Ngui LX, Gopalan S, Chai CK.** A rare case of spontaneous regression of huge neck lymphangioma post primary infection. *Egyptian Journal of Ear, Nose, Throat and Allied Sciences* (2016) 17, 99–102.
- Som PM, Lidov M, Lawson W.** Hemorrhaged cystic hygroma and facial nerve paralysis: CT and MRI findings. *J Comput Assist Tomogr*, 14 (4) (1990), pp. 668–671.
- Perkins JA, Maniglia C, Magit A, et al.** Clinical and radiographic findings in children with spontaneous lymphatic malformation regression. *Otolaryngol Head Neck Surg*. 2008;138(6):772–777.
- Chisin R, Fabian R, Weber AL, et al.** MR imaging of a lymphangioma involving the masseter muscle. *J Comput Assist Tomogr*. 1988;12:690–692.
- Liu DY, Guan J, He HX.** Diagnosis and surgical treatment of cervical lymphangioma. *Zhonghua Yi Xue Za Zhi.*, 89 (48) (2009), pp. 3413–3416
- Berri T, Azizi S.** Large cystic lymphangioma of the parotid gland in the adult. *Egyptian Journal of Ear, Nose, Throat and Allied Sciences*, 2014; 15 (3): Pages 259–261.
- Oosthuizen JC, Burns P, Russell JD.** Lymphatic malformations: a proposed management algorithm. *Int J Pediatr Otorhinolaryngol*. 2010;74:398–403.
- Zhou Q, Zheng JW, Mai HM, et al.** Treatment guidelines for lymphatic malformations of the head and neck. *Oral Oncol*. 2011;47:1105–1109.
- Colbert SD et al.** Lymphatic malformations of the head and neck— current concepts in management. *Br J Oral Maxillofac Surg*. 2013;51:98–102.
- Sichel JY, Udassin R, Gozal D, et al.** OK-432 therapy for cervical lymphangioma. *Laryngoscope*. 2004;114(10):1805–1809.
- Ogita S, Tsuto T, Tokiwa T, Takahashi T.** Intracystic injection of OK-432: a new sclerosing therapy for cystic hygroma in children. *Br J Surg*. 1987;74:690–691.