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Cervicofacial Cystic Lymphangiomas and Review of the Literature: About 2 Cases at Donka National Hospital

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Abstract

In this retrospective study, conducted over 7 years (2009-2016) at the ENT and Head and Neck Surgery Department of the Donka National Hospital, we report 2 cases of cervicofacial cystic lymphangiomas. They were a 28-month-old girl and a 2-year-old boy. The symptomatology was noted after their birth. Dyspnea and dysphagia were found in the boy. They had a satisfactory general condition. ENT examination noted a cystic-like tumor syndrome. Imaging showed evidence of a cystic lymphangioma of the cervicofacial region. Pathological examination confirmed the diagnosis. All patients underwent exeresis cervicotomy. We found adhesion of the lymphangioma cyst to the internal jugular vein in the children. The boy presented a paralysis of the chin branch of the facial nerve after the surgery. We did not find any tumor recurrence. However, cystic cervicofacial lymphangiomas are a particular aspect of surgical pathology in children in Africa. In spite of the advent of sclerosing products, surgery remains for us the treatment of choice.

Keywords

Cystic Lymphangioma, Management, Literature Review, African Setting

1. Introduction

Cystic lymphangiomas are rare benign dysembryoplasias of the lymphoganglionic system, responsible for a tumor syndrome by angiolymphatic proliferation.

This condition was first reported in 1828 by Redenbacher and has been better known since the landmark works by Sabin in 1909 and 1912 [1]. Nowadays, prenatal diagnosis of cystic lymphangiomas is possible by fetal ultrasound [2]. Their anatomical localization is almost exclusively cervicofacial and their clinical revelation is generally very early in the neonatal period [3]. Their postnatal diagnosis, in our context, is most often evoked from clinical and intraoperative data [4]. The seriousness of these tumor formations in children is due, on the one hand, to their evolutionary potential that may compress and invade the upper aerodigestive tract, and on the other hand, to the classic difficulty of their removal [5]. This therapeutic difficulty can be explained by the relative ignorance of the natural history of this condition, despite numerous etiopathogenic hypotheses. In Guinea, as in some sub-Saharan countries, cervicofacial cystic lymphangiomas are rare. They have been little studied. Therefore, the aim of this retrospective study was to correlate these 2 observations with the literature review in order to share our experience in their management in the African setting.

2. Presentation of the Cases

2.1. Case 1

This was a 28-month-old girl, with no particular history, from a sibling group of 2 brothers, admitted to an ENT consultation for a right laterocervical mass (**Figure 1(a)**). According to the parents, the onset of the disease was due to the appearance of a submohyoid mass that had increased in volume in a non-febrile context. They sent him to several health facilities in the area where the diagnosis and treatment received are not documented. Given the worsening of the symptoms, she was referred to the department for treatment.

(a) (b)

Figure 1. (a) Infant with right cervical cystic lymphangioma; (b) Infant with simple postoperative course.

Examination at entry noted satisfactory general condition, clear consciousness and good coloration of the integuments and conjunctivae. Vitals: Pulse = 110 beats/min; Respiratory rate = 22 cycles/min; Weight = 6.2 Kg; SO_2 = 99%. At the ENT sphere, there was a soft, painless, smooth surface, fixed in relation to the deep plane, healthy skin opposite, measuring 22 cm of the long axis, irreducible and non pulsatile. The transillumination test was positive. The lymph nodes were free. The rest of the ENT examination and other devices were without abnormalities. The exploratory puncture returned a clear fluid. Our diagnostic hypothesis was cervical cystic lymphangioma.

Cervical ultrasound revealed hypoechoic or anechoic, multiclustered masses occupying the right laterocervical region and not vascularized by Doppler. Cervicothoracic CT scan showed an isodense, multi-partitioned, well-limited mass occupying the right supra and infra omohyoid region. Needle cytopuncture revealed a cystic lymphangioma.

She underwent a cervicotomy for tumor removal at the age of 6 months. The exploration noted a soft, cystic-like mass occupying the supra- and submohyoid triangles and not hemorrhagic. It was adherent to the right internal jugular vein. The resection was complete. Closure was performed with a Redon Jost drain. Intraoperative accidents and incidents were absent. The specimen was sent to the laboratory for histological study. The postoperative care consisted of the administration of analgesics, antibiotics and corticosteroids. Dressings were regular. The drain was removed on the 2nd postoperative day. We observed complete healing on the 18th postoperative day (**Figure 1(b)**). The postoperative period was simple. Anatomical pathology was received on the 21st postoperative day and concluded to be a cervical cystic lymphangioma. She was monitored for 8 months.

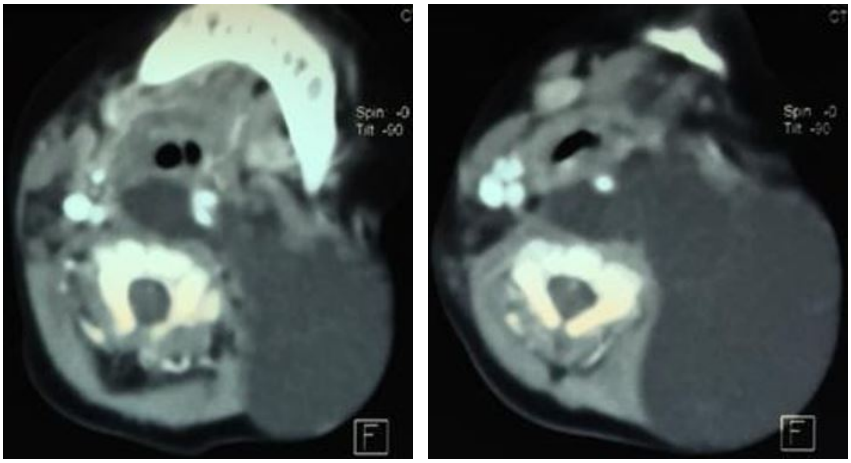
2.2. Case 2

This is a 2-year-old boy with a history of recurrent rhinitis from a sibling group of 4 children. He was admitted for a high left laterocervical mass. The history goes back to birth with the appearance of a left subdigastric mass that had increased in volume. The parents sent him to a health facility where he was diagnosed as having adenomegaly and received undocumented treatment. The evolution was marked by the increase in volume of the mass accompanied by dyspnea and dysphagia in a non-febrile context. Thus, the child was referred to the department for better management.

The examination at entry noted a good general condition, clear consciousness, good coloration of the integuments and conjunctiva. Vitals: BP = 100/60 mmHg; Pulse = 126 beats/min; Respiratory rate = 22 cycles/min; Weight = 9.5 Kg. In the ENT sphere, we noted a soft, painless mass, extending from the left parotid region to the right submental and submaxillary region, measuring 25 cm of the major axis, fixed in relation to the deep plane, healthy skin in front of it, regular border and accompanied by compression signs. We retained a compressive cervicofacial cystic lymphangioma.

Further investigations were requested. Cervical ultrasound found hypoechoic, polylobed, non-vascularized masses on Doppler and was in favor of a cervicofacial cystic lymphangioma. The cervicofacial CT scan (**Figure 2(a)** and **Figure 2(b)**) noted an isodense, multiclobed mass with a mass effect on the aerodigestive tract (pharynx, larynx and esophagus). The fine needle cytopunction came back in favor of a cystic lymphangioma.

The patient underwent a cervicotomy of exeresis by Sibileau Carréga approach. The exploration noted a mass, polylobed, non-hemorrhagic, cystic in appearance, voluminous and adherent to the left superior laryngeal, the left internal jugular vein and the left carotid bifurcation. The exeresis was laborious and complete. Closure was done on an aspirative Redon Jost drain (**Figure 3** and **Figure 4**). The intraoperative incident was marked by an involvement of the chin branch of the facial nerve. Postoperative management was based on antibiotics, analgesics, corticosteroids and dressings. The patient was discharged on the 5th postoperative day. The anatomopathological examination received on the 20th postoperative day, found a cystic lymphangioma. He was followed for 16 months.



(a) (b)

Figure 2. (a) Isodense, multi-partitioned compressive mass; (b) Isodense, multi-partitioned compressive mass.

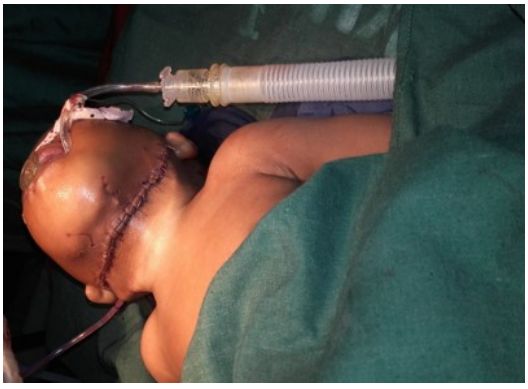


Figure 3. Cervicotomy for removal of cystic lymphangioma.

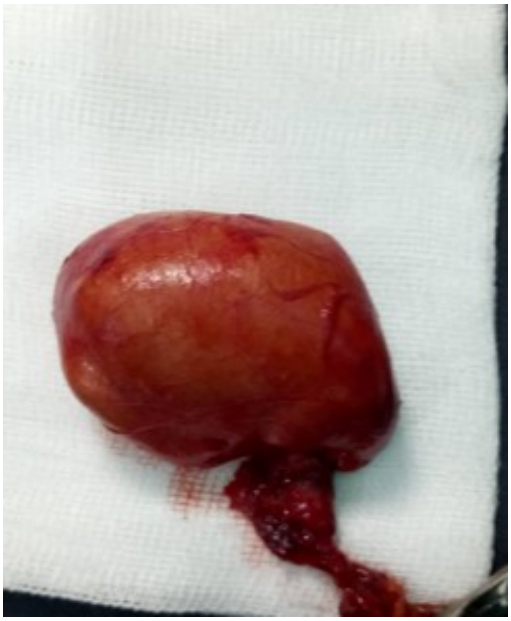


Figure 4. Surgical specimen of cystic lymphangioma.

3. Discussion

In this series, our limitations were marked by the rarity of the cases, the low socioeconomic level of the patients to carry out all the assessments and the insufficiency of the technical platform. However, 2 cases of cervicofacial cystic lymphangiomas were identified and will be correlated with the review of the literature.

Cystic lymphangiomas are rare benign tumors that can occur in any region of the body, but cervicofacial localizations predominate [6] [7] [8]. They represent between 2.6% and 5% of benign congenital cervical masses [4]. However, we have documented two cases of cervicofacial cystic lymphangiomas over a period of 7 years. Our result is lower than those reported by Ozen (17 cases in 20 years) [8] and Triglia (three cases per year) [9]. Our finding is in line with the data in the literature.

Cervical cystic lymphangioma classically sits in the supraclavicular region [10]. This location has been reported in girls. According to François [3], anterior localizations are as frequent as posterior localizations. Indeed, suprahyoid cystic lymphangiomas are part of diffuse lymphangiomatosis whose deformities are sometimes less visible [9]. In these forms, one should look for invasion of the parotid gland, cheek, tonsil cavity, parapharyngeal space, tongue or larynx. Isolated jugal and parotid localizations are rare and pose the problem of recurrence and aesthetic sequelae postoperatively. In our case, it occupied the right parotid region extending to the submental and left maxillary region in the boy.

The cyst is present at birth in only 50% - 60% of cases, but it is expressed in 90% of cases before the age of two [3]. This was the case in our series. Some lymphangiomas do not appear until late in adulthood, as shown by the studies of Raji [11] and Diop [12]. The origin of cystic lymphangiomas is not well known.

For some authors, the formation of a lymphangioma occurs by anarchic development from the primary lymphatic sacs: this is the centrifugal theory [5]. For others, lymphangioma occurs when the lymphatic vessels do not establish connections with the rest of the lymphatic or venous system: this is the centripetal theory [3]. The centrifugal theory would tend to explain the surgical finding of connections between the cystic lymphangioma and the internal jugular vein in all our patients.

The major functional signs are those determined by the compression of the neighbouring organs when the size of the cervical mass becomes important. They motivate an immediate consultation of the patient. These may include dyspnea, dysphagia, dysphonia, neck pain and limitation of neck movement. However, we noted dyspnea and dysphagia in the boy. Physical examination revealed a mass of variable size, soft, regular or poly-lobed, covered by thinned and normal skin, sometimes pale or bluish, even angiomatous. This mass is painless, depressible, but irreducible and non-pulsatile [10]. Our patients presented with almost all of these physical signs which were cystic in appearance.

Some examinations are useful to look for deep extension. Cervical ultrasonography provides more information [4] [13] and was of great interest to us. It allowed us to specify the size, the number of cystic pockets, their multi-compartmental character, to analyze the echogenecity and to appreciate the content of different liquid pockets without flow by Doppler. The multi-compartmental aspect was very suggestive of the diagnosis. However, the tumor extension is sometimes difficult to specify on ultrasound because of the often large volume of the tumor. This information is better appreciated by CT scan and magnetic resonance imaging [5] [13]. The data provided by the CT scan have led to an adapted therapeutic method.

The treatment of cervico-facial cystic lymphangiomas is classically surgical [3] [9] [14]. However, our patients benefited from an excisional cervicotomy. The Paul-André, Sebileau-Carrega and Redon incisions allow a wider approach and control of the vasculo-nervous elements of the neck and face. Some authors [15] consider that general anesthesia is less risky when performed at six to nine months of age. On the other hand, early surgery is preferable, before inflammatory or hemorrhagic flare-ups make dissection difficult. Indeed, the cystic lymphangioma presents intimate relationships with the cervical venous system, the thoracic duct or the great lymphatic vein, and there is no real cleavage plane. The often thinned and atrophic muscles lose their role as a beacon in cervical surgery. The surgical procedure must be as complete as possible to avoid recurrence. However, the notion of complete removal should not expose the patient to postoperative functional and aesthetic risks for a condition that remains benign [9] [16].

Complications related to surgery are to be avoided. They are important when the number of repeat surgeries is multiplied and when the child is younger. This calls for caution in extended forms, in particular at the base of the tongue, at the valecula, or at the laryngeal margella where the impossibility of intubation ob-

liges to perform a tracheotomy. Local complications are responsible for long after-effects [3]. The neurological sequelae are variable according to the series and are represented by peripheral facial paralysis, paralysis of the mental branch of the facial nerve and a case of paralysis of the recurrent nerve [10] [14] [17]. Only the boy developed a paralysis of the left mental branch of the facial nerve. The main problem with cystic lymphangioma is postoperative recurrence. It is mostly seen after incomplete exeresis [3] [4] [7]. This multiplies the number of surgical procedures under even more difficult conditions. We have not noted any recurrence related to the complete removal of cystic lymphangioma.

4. Conclusion

Cervicofacial cystic lymphangiomas are benign tumor malformations of congenital origin. They are a particular aspect of surgical pathology in African children. The signs of compression can be life threatening. Histology confirms the diagnosis. Surgery remains, despite its difficulties, the exclusive treatment in our sub-Saharan countries. It must be early and complete in order to guarantee a complete cure and the absence of recurrence.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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