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Congenital Cervico-Mandibular Cystic Lymphangioma in Pediatric Surgical Setting in Guinea

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Abstract

Introduction: Cystic lymphangiomas are rare benign malformative tumors of the lymphatic system of obscure etiopathogenesis. The cervico-facial location remains the most common (75%). Although benign, these tumors remain potentially fatal, due to possible compression of the upper aero-digestive tract. The aim of this work is to study the epidemiological, diagnostic and therapeutic characteristics of cervico-mandibular congenital cystic lymphangiomas in the pediatric surgery department of the Donka National Hospital (HND) Conakry. **Patients and methods:** This is a retrospective and descriptive study of 13 files lasting 7 years from January 2015 to December 31, 2021. The files of children whose age is less than or equal to 15 years operated on cervical tumor with histological evidence of cystic lymphangioma were retained. The data were analyzed using SPSS statistical software 21 and anonymously. **Results:** The incidence of this study was 1.86 cases per year and a sex ratio of 0.62 in favor of girls. The average age was 8 months 19 days. In the antecedents, we only find poorly monitored pregnancies. The average size of the tumors was 11.85 cm. Cervical ultrasound and standard x-ray of the cervical mass were the only examinations performed. Total surgical excision of the cervical tumor was performed in all patients. The mass was polycystic on exploration. The histological examination of the surgical specimens was in favor of a cystic lymphangioma. The surgical consequences were simple in 11 patients (84.62%) and complicated by parietal suppuration in 2 cases (15.38%). There were no cases of recurrence after one year of follow-up.

Conclusion: Cervico-mandibular cystic lymphangiomas are the most frequent locations of congenital lymphangiomas in children. Their severity is linked to the risk of compression of the aero-digestive tracts. Their diagnosis must be confirmed by the histology of the surgical specimen. Despite the therapeutic arsenal, excision of the cystic mass remains the only effective alternative in our socio-economic conditions to avoid recurrences and loss of follow-up of patients.

Keywords

Congenital Cystic Lymphangioma (LKC), Child, Pediatric Surgery Guinea

1. Introduction

Congenital cystic lymphangioma (CLC) is a rare benign malformative tumor of the lymphatic system. These malformations consist of cysts lined with vascular endothelium and filled with lymph and sometimes blood and connective tissues to varying degrees, explaining why some consider them to be hamartomas and not cystic tumors [1] [2]. This condition was first reported in 1828 by Redenbacher and better known thanks to the reference work carried out by Sabin in 1909 and 1912 [3] [4] [5]. This malformation represents 2.6% - 5% of congenital cervical masses in children. Their location is ubiquitous and the cervico-mandibular or facial regions remain the most frequent locations (75%) [6] [7]. The etiopathogenesis of this tumor remains obscure; the centrifugal theory is the most widespread. According to this theory at the neck level, there is a primitive jugular sac (embryonal lymphatic bags) interposed between the internal jugular vein and the thoracic duct on the left or the large lymphatic vein on the right and these lymphatic formations will gradually lose their connection with the vein. These formations will gradually lose their connection with the internal jugular vein to give rise to lymph nodes and ducts. It is the persistence of one of these connections with the internal jugular vein which is responsible for the formation of cystic lymphangioma [8]. The clinical revelation of cystic lymphangiomas is generally early during the neonatal period. The seriousness of these malformations is due, on the one hand, to their evolving potential capable of compressing and invading the upper aerodigestive tracts, putting the child's vital prognosis at risk, and on the other hand to the classic difficulty of their excision [3] [9]. Few studies have been carried out on cervico-mandibular LKC in children in Africa and in particular in our country [2] [3] [7] [10] [11]. The neonatal discovery of a cervico-mandibular mass with deformation of the child's face making it frightening, the survival of the child with bad luck mentioned by the children's parents as well as the difficulty of taking this malformation into our African country, motivated this study in our department, the main aim of which is to discuss: the epidemiological, diagnostic and therapeutic aspects and to review the literature.

2. Patients and Methods

This is a retrospective and descriptive study of the files of children treated at the pediatric surgery department of the HND of the Conakry University Hospital from January 2015 to December 31, 2021 (7 years). Thirteen files of cervico-mandibular LKC were collected. Children aged less than or equal to 15 years operated for cervico-mandibular mass and whose surgical specimen returned in favor of a cystic lymphangioma were selected for this study. Our exclusion criteria were the absence of histological proof of the surgical specimen, incomplete medical files and cases of cervical masses who died before the intervention (3 cases) during the study period. We collected a total of 13 files for this study. The variables retained were age of diagnosis, sex, history, characteristics of the tumor, site, side, standard radiography and imaging, surgical exploration, procedure, duration of hospitalization, the histology of the surgical specimen and the surgical aftermath. Standard radiography and ultrasound of the cervical mass were the only examinations performed. The scanner was not carried out due to lack of financial resources and the MRI due to availability of the device. All children were placed on antibiotics, analgesics and short-term corticosteroid therapy with the advice of the pediatrician. Data were collected from medical records entered by world 2013 and analyzed using SPSS statistical software 21. Patient anonymity was respected.

3. Results

During the study period we recorded 2190 patients hospitalized in the department, which makes a hospital frequency of 0.59% of cervico-mandibular LKC. The incidence in this study was 1.86 cases per year. Eight girls and 5 boys were identified, giving a sex ratio of 0.62 in favor of girls. The average age of the patients was 8 months 19 days with an age range of 8 days and 3 years. Eleven children were aged less than 1 year (84.62%) and 92.31% of children were aged less than or equal to 2 years, including three newborns or 27.27%. In the antecedents, we find only poorly monitored pregnancies (13 cases), with 8 cases of home birth, 3 at the basic health center, 2 in regional hospitals, 8 obstructed low births, no documented obstetric ultrasound [Table 1]. The age of appearance of the cystic mass was at birth in our series. All patients consulted for unilateral cervical-mandibular swelling with a predominance on the left side (8 cases) compared to 5 cases on the right [Figure 1]. The associated signs were dyspnea with bronchial congestion (10 cases) and swallowing disorder (3 cases). Clinically, the size of the cervical masses varied from 7 cm to 17 cm on the long axis with an average of 11.85 cm [Figure 2]. The unilateral swelling was painless, compressible, but irreducible and non-pulsatile with a soft and hard consistency in places. The mass did not increase upon coughing [Figure 3(a)]. The standard cervical radiograph of the mass taking the thorax face and profile showed an oblong opacity, of water tone with blurred internal contour with deviation of the trachea [Figure 3(b)]. Cervical ultrasound of the mass is not performed only in

Table 1. Some Epidemiological characteristics of our patients.

Characteristics		N (%)
Year	**Hospitalization (H)**	
2015	245	1 (0.41)
2016	223	1 (0.45)
2017	238	1 (0.42)
2018	475	2 (0.42)
2019	309	3 (0.97)
2020	311	2 (0.64)
2021	389	3 (0.77)
Age Bracket (month)		
[0 - 6]		7 (53.85)
[6 - 12]		4 (30.77)
[12 - 24]		1 (7.69)
[24 - 37]		1 (7.69)
Distribution according to sex		
Fille		8 (61.54)
Garçon		5 (38.46)
Background		
Accouchement à domicile		8
Centre de santé de base		3
Hôpital regional		2
Accouchement dystocique		8
Echographie fœtale		0

H: Hospitalization of all patients during the study period; N: number of LKC cases per year; Hospital frequency of cervico-mandibular LKC in the department is 0.59%.

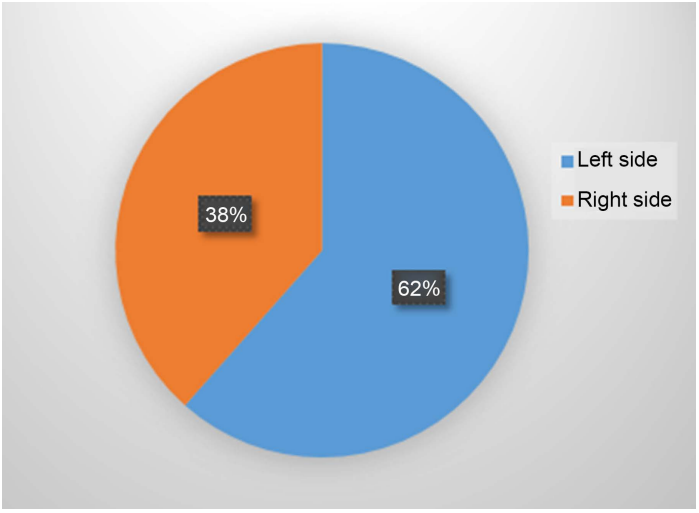


Figure 1. Distribution of the tumor according to the side.

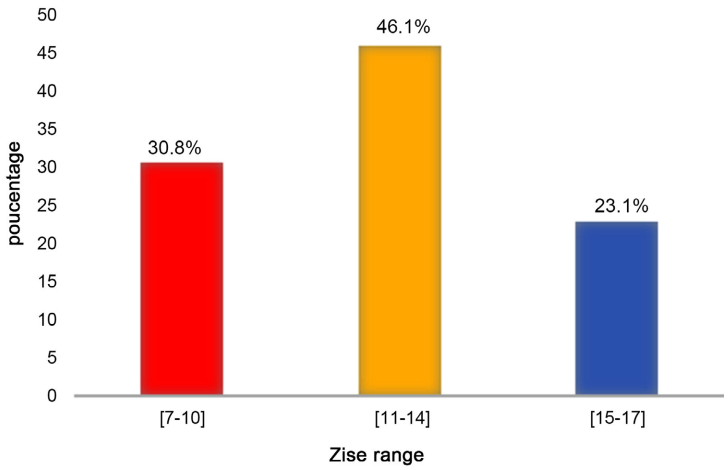
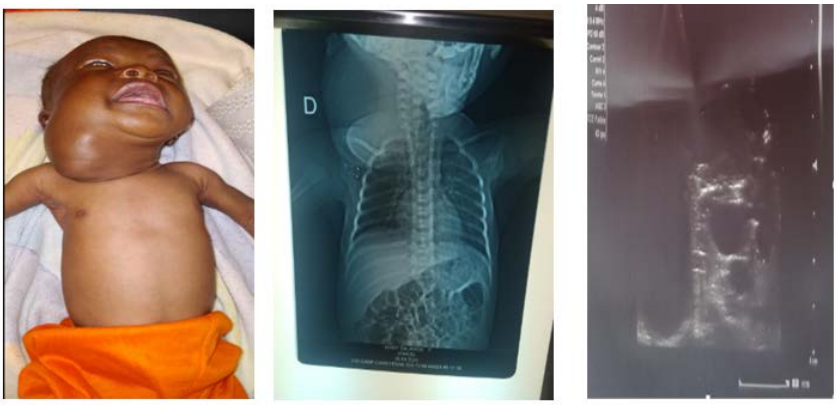


Figure 2. Distribution of the tumor according to size in cm.

12 cases; she mentioned a multiloculated cystic lymphangioma comprising hypo and anechoic macrocysts in 10 cases [**Figure 3(c)**], and 2 cases of intracystic hemorrhage separated by thin partitions, without vascularization on Doppler. CT and MRI were not performed. The alpha fetoprotein assay performed in 4 cases was negative. Complete surgical excision of the cervical masses was carried out in all our patients by a transverse approach to the lower cervical fold (10 cases) and an orange wedge (3 cases). Surgical exploration revealed a polycystic mass with clear gelatinized content (10 cases) and hematic content (3 cases). It was located in the postero-inferior triangle of the neck between the trapezius and the sternocleidomastoid muscle (SCM) contracting an adhesion with the internal jugular vein on the left and the large lymphatic vein on the right and compression of the trachea (8 cases) without desaturation of the child. Excision of the mass was complete but laborious. Closure of the compartment on a suction drain or delbet [**Figure 4**], [**Figure 5**]. Histological examination of the surgical specimens was in favor of a cystic lymphangioma and showed a conglomerate of dilated lymphatic spaces, bordered by a single layer of endothelial cells resting on a connective stroma and coexisting with lymph node formations; the vascular wall contains smooth muscle cells associated with thickened fibrous tissue in 4 cases. The postoperative course was simple in 11 of our patients (84.62%); wall suppuration with a favorable outcome in 2 cases (15.38%). No death was noted nor recurrence after follow-up. The average length of hospitalization was 9.26 days with extremes of 6 days and 14 days. No cases of recurrence after one year of follow-up.

4. Discussion

The limitations and difficulties of this study were linked to the poverty of the patients to carry out all the additional examinations, the high cost of the intervention, poor record keeping, insufficient technical support and the small size of the sample. Despite this situation we have reached results which we will discuss and comment on.



(a) (b) (c)

Figure 3. 2-month-old female infant: (a): Clinical presentation of a right cervico-mandibular congenital cystic lymphangioma; (b): Blurred right cervical opacity with slightly deviated trachea to the left; (c): Hypoechoic multiloculated cystic macrocyst formation separated by thin septa on ultrasound.

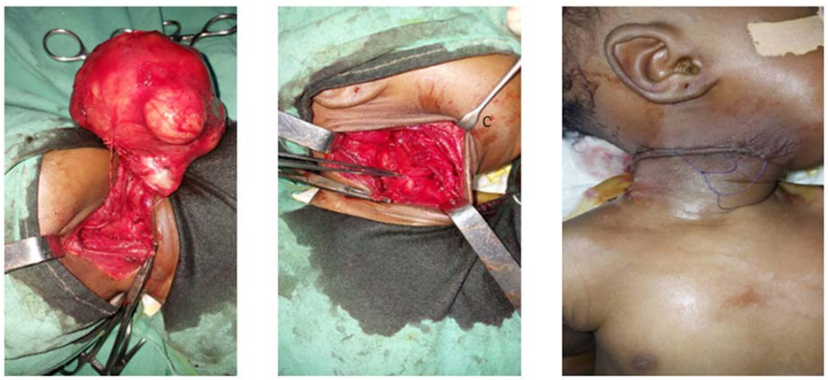
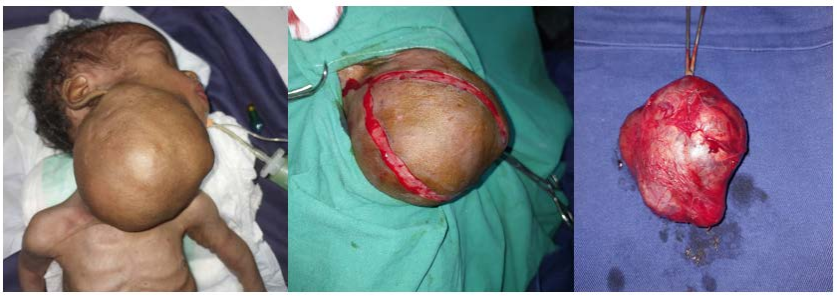


Figure 4. Exploration of a cystic mass + followed by total excision and aesthetic appearance of the neck



(a) (b) (c)

Figure 5. (a): Compressive cystic lymphangioma, 3-month-old boy; (b): Surgical approach using an orange wedge; (c): Extraction of an inflammatory cyst with hematic content.

Cystic lymphangiomas are rare benign dysembryoplasias of the lymphoganglionic system responsible for a tumor syndrome due to angiolymphatic proliferation of cervico-facial location in most cases. It is often clinical cases that are

reported in the literature. Our hospital frequency was 0.59% which agrees with the data in the literature [1] [2] [6] [7] [11]. In [**Table 2**] we compare our incidence to those of some authors from Africa. Cystic Lymphangiomas mainly affect children under 2 years old [12] as is the case in our study: 92.31% of our children were less than or equal to 2 years old. B.BA [12] in MALI, reported 52.8% of infants for the age group of 1 month to 2 years, CHARABI in Denmark cited by B.BA found 50% for the same age group; J. Miaoundja [3] in Gabon reported in his 16 cases of LKC 7 infants aged less than one year; In our study the sex ratio was 0.62 in favor of girls; In general, there is no predominance of sex, it depends on the series [3] [7]. The tumors were unilateral cervical with predominance on the left side (8/5) in our study as mentioned in the literature. This is explained by the fact that the lymphatic system predominates on the left. The diagnosis is currently often antenatal thanks to fetal ultrasound in the second trimester, visualizing a multiloculated cystic formation separated by thin septa [9] [13] [14]. Fifty percent of cysts are present at birth and 90% are diagnosed within the first two years of life [15]. All our cases are diagnosed postnatally during delivery and confirmed by ultrasound and radiography; cervical x-ray looks for tracheal deviation or mediastinal extension [16]. Cervical ultrasound provides more information [14] [17]. Ultrasound in our study made it possible to specify the size, and the multi-partitioned nature of the cysts (very suggestive of the diagnosis), to assess the contents of different pockets which were hypoechoic fluid with no flow on Doppler and fine echogenic elements (2 cases) in favor of intracystic hemorrhage. Tumor extension is better assessed with CT and MRI [14]; MRI seems to give more specific images with hyper signal in T2 reproducing the different septations. Above all, it allows a precise analysis of the extensions to the mediastinum, the parotid region and the parapharyngeal regions as well as the relationships of the cyst with the vasculonervous elements of the neck [3]. These two examinations could not be carried out because of their high cost and the poverty of the children's guardians. Surgical exploration of our surgical cases showed a cystic tumor located in the postero-inferior triangle of the neck, extending laterally (stage 1 of the SERRES classification) [6] and opposing an adhesion with the internal jugular vein at left and the large lymphatic vein on the right. The compression of the trachea due to the volume of the cysts

Table 2. Annual incidence of Cervico-mandibular LKC.

Authors	Annual Frequency
OMAR BERRADA (March 2022) [7]	17 cas/11: 1.54
J. MILOUNDJA (2007 au Gabon) [3]	16 cas/15: 1.07
RAFANOMEZANTSOA T Malgache 2020 [11]	1 cas
KEITA A [10] en 2022	2 cas/7: 0.28
T. BENTEBBICHE (Tunisie en 2020) [6]	16 cas/5: 3.2
OUR STUDY	**13 cas/7: 1.86**

in our case would explain the dyspnea and swallowing disorders of our patients; All our patients were intubated despite this situation even if a few cases of difficult intubation were reported (3 cases) in the operating protocols. They were all on antibiotic prophylaxis (clavulanic acid + Amoxicillin) and steroid anti-inflammatory drugs (3 - 5 days). We have not encountered any suprahyoid locations which fall within the framework of diffuse lymphangiomatoses; In these forms, we must look for invasion of the parotid, the cheek, the tonsillar compartment, the parapharyngeal space, the tongue or the larynx; they pose a problem of recurrence and aesthetics postoperatively [2] [3]. Excision of the mass was laborious because of adhesions but complete in our study without intraoperative incident. The postoperative course was simple in 11 of our patients (84.62%); wall suppuration with a favorable outcome in 2 cases (15.38%). No death was noted nor recurrence after follow-up. The average length of hospitalization was 9.26 days with extremes of 6 days and 14 days. J. Miloundja [3] reports 2 cases of facial paralysis (left and right jugal lymphangioma) and 1 case of serious respiratory distress (right latero-cervical lymphangioma). The surgical risk is more difficult in cavernous, microcystic and infiltrative lymphangiomas and in cases of inflammatory or hemorrhagic outbreaks. In our case it was macrocystic cystic lymphangiomas. The difficult dissection in our case was related to tissue fibrosis and the inflammatory process. This situation was confirmed by the histological examination of our surgical specimens where we found fibrous tissues within the cystic tumors (4 cases). Our histological results are identical to those of J. MILOUNDJA in Gabon [**Figure 6**]. The dissection was meticulous in our cases. Indeed, the cystic lymphangioma has intimate relationships with the cervical venous system, the thoracic duct or the large lymphatic vein, and there is no real cleavage plane; the surgical procedure must be, as much as possible, complete to avoid recurrences. However, the concept of complete excision should not expose the patient to postoperative functional and aesthetic risks for

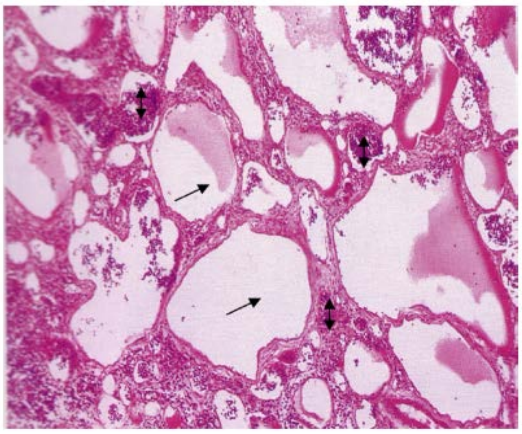


Figure 6. Histological appearance of a cystic lymphangioma with dilation of the lymphatic vessels (arrow) and vascular ectasia (double arrow). Image J. MILOUNDJA (Gabon 2007) [3].

a condition that remains benign [10]. The treatment of cystic lymphangiomas with sclerosing products has been mentioned by certain authors in the literature as first-line treatment [12] [18]. In Mali B.BA [12] reports 27 cases of serotherapy with 90˚ alcohol with 63.9% complete recovery and 36.1% of cases, sclerotherapy was associated with surgery. The average treatment duration was 1.8 months, with extremes of 21 and 120 days. For other authors [1] [3], this treatment is only effective in macrocystic lymphangiomas.

5. Conclusion

Cervico-mandibular cystic lymphangiomas are the most common locations of congenital cystic lymphangiomas in children. Their severity is linked to the risk of compression of the aero-digestive tracts and the difficulty of their excision. The paraclinical and clinical diagnosis must be confirmed by the histology of the surgical specimen. Despite the therapeutic arsenal, excision of the cystic mass remains the only effective alternative in our socio-economic conditions to avoid recurrences and loss of follow-up of patients.

Conflicts of Interest

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

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