MANAGEMENT OF CERVICOFACIAL CYSTIC LYMPHANGIOMA IN CHILDREN

DR MOUANGUE-MBONJO LOUISE[1*]
*[*1]Maxillofacial Surgery Department, Yaounde Central Hospital, Department of Otorhinolaryngology and Cervico-maxillofacial Surgery, Yaounde Central Hospital, Faculty of Medicine and Pharmaceutical Sciences, University of Douala, Cameroon,*

DR DIALLO MAMADOU MOUCTAR RAMATA[2]
[2]Regional Hospital of Mamou, Department of Otorhinolaryngology and Cervico-maxillofacial Surgery, Mamou, Guinea,

DR MINKA NGOM ESTHELLE GENEVIÈVE STÉPHANIE[3]
[3]Otorhinolaryngology Unit of the Douala General Hospital, Faculty of Medicine and Pharmaceutical Sciences, University of Douala, Cameroon,

DR BADANG AMBADIANG FLORENT DIMITRI[4]
[4]ENT Department and CervicoMaxillo-Facial Surgery, Regional Military Hospital No. 2 Douala, Faculty of Medicine and Pharmaceutical Sciences of the University of Douala, Cameroon.

PR NJIFOU NJIMAH AMADOU[5]
[5]Faculty of Medicine and Pharmaceutical Sciences, University of Douala, Cameroon.

***Corresponding Author : Dr Mouangue-Mbonjo Louise**

ABSTRACT

Introduction: *Cystic lymphangioma is a rare, benign, tumor of the lymphatic vessels. It has varying locations, with the commonest being the cervicofacial region. This study aimed to describe the clinical characteristics and outcomes of patients with cervicofacial cystic lymphangioma. Materials and Methods: This was a retrospective study of 24 patients who underwent surgery for head and neck lymphangioma between 2013 and 2023 in XX hospital. The studied variables included age, sex, symptoms, imaging findings, results, and complications. Results: Most of the patients were male (58.3%). The median patient age was 4.5 (2.0–6.7) years. Fifty percent (n=12) of patients had visible lesions at birth, and 16 (66.7%) had left-sided lesions. The shoulder (n=3; 12.5%), clavicle (n=2; 8.3%), and mandible (n=2; 8.3%) were the main sites of extension. Imaging examination was performed in nine (37.5%). Eighteen (87.5%) patients underwent total excision of the tumor. Furthermore, six (25.0%) patients had received preoperative sclerotherapy. Postoperative complications were observed in 25.0% of the patients, with lymphedema (n=6) being the commonest. Esthetic sequelae were the commonest sequelae in 37.5% (n=9) of patients. No death was registered. Conclusion: Cystic cervicofacial lymphangioma is a complex pathology requiring multidisciplinary management. Sclerotherapy leads to good outcomes in compressive forms.*

Keywords: *Cervicofacial cystic lymphangioma, diagnosis, treatment, surgery.*

DOI:10.5281/zenodo.13892285 **Manuscript ID # 177**

INTRODUCTION

Cervical cystic lymphangioma is a benign malformation of the lymphatic system. (1-3). It was first described in 1828 and classified in 1843 by Redenbacher (4) and Wernher et al., respectively (5). They can present as capillary, cavernous, or cystic lymphangiomas. Based on size, they are classified as microcystic (volume, <2 cm^3), macrocystic (>2 cm^3), or mixed lymphangiomas (6-8). Its incidence is 1 in 2,000 to 4,000 live births (6). Ninety percent of cases in children aged below 2 years occur in the cervicofacial region. Other sites include the axilla, superior mediastinum, retroperitoneum, pelvis, and limbs (9). It manifests as a progressively increasing acute cervical mass, causing the child's parents or guardians to panic. Radiology assists in clinical diagnosis, extent of the lesions, complications, image-guided percutaneous treatment, and monitoring of treatment response. Resection of extensive cervicofacial lesions is difficult; this is due to the proximity of the lesion to nerves and vessels. The treatment of choice is surgical. Diagnostic confirmation is mainly histological (10). Cervical cystic lymphangiomas in children have been the subject of few recent studies, especially in their exclusively facial location with a slightly larger number of cases (11-12). This study aimed to describe the clinical characteristics and treatment outcomes of patients with cervicofacial cystic lymphangioma based on a 10-year experience.

Materials and Methods

This was a retrospective study. Patients with lymphangioma who were admitted in the otorhinolaryngology department of Hospital XX between January 2013 and December 2023 were enrolled. Of 200 patients with lymphangioma, 35 had cystic lymphangioma. However, patients who were above 15 years old were excluded. The requirement for consent was waived due to this study's retrospective nature. Therefore, 24 patients were included into this study (Figure 1). Patients' data were collected from medical records. The variables of interest included sex, age, visibility of lesions at birth, age at diagnosis, absence/presence of recurrence, sidedness, symptoms, extent, imaging, and evolution. Qualitative variables are expressed as number (percentage), while quantitative variables are expressed as mean (standard deviation). Data were analyzed using SPSS version 23.0.

Results

We included 24 children with cystic lymphangioma (Figure 1).

Distribution of cystic lymphangioma cases during the study period

The distribution of the total number of cases of cystic lymphangioma according to year at diagnosis is shown in a linear curve in Figure 2. Of the total number of cases during the study period, the lowest number of cases one case (4.2%) was registered in 2014 and 2016 and the highest four case (16.7%) in 2018.

Patients' sociodemographic characteristics

The median age of the patients was 4.5 (2.0–6.7) years, with a range of 1–11 years. Fifty-percent (n=12) of the patients were aged 2–5 years and were males (n=14, 58.3%) Figure 3.

The patients' sociodemographic characteristics are shown in Table 1.

Table 1: Distribution of the study population by age and sex

Variables	Number of patients (N=24)	Frequency (%)
Age (Years)		
<2	4	16.7
2–5	12	50.0
6–9	5	20.8
10–14	3	12.5
Sex		
Male	14	58.3
Female	10	41.7

Tumor Characteristics at diagnosis

Fifty percent (n=12) of the children had visible lesions at birth. The age at diagnosis was <2 years in 13 (54.2%) patients. Only one child (n=2, 4.2%) had recurrent disease. The patients' clinical characteristics at the time of admission are shown in Table 2.

Table 2: Tumor characteristics at the time of diagnosis

Variables	Number of patients (N=24)	Frequency (%)
Visibility of lesions at birth		
Yes	12	50.0
No	12	50.0
Age at diagnosis, years		
<2	13	54.2
≥2	11	45.8
Recurrence		
Yes	1	4.2
No	23	95.8

Clinical features of lesions

Table 3 shows that most patients (n=16, 66.7%) had left-sided unilateral lesions. Twenty-one (87.5%) patients presented with a neck swelling. Nine (37.5%) patients had extensive lesions. The main sites of extension of the cystic lymphangiomas are shown in Figure 4.

Table 3: Characteristics of the lesions

Variables	Number of patients (N=24)	Frequency (%)
Sidedness		
Left	16	66.7
Right	8	33.3
Symptoms		
Swelling	21	87.5
Suppuration	3	12.5
Extension		
Yes	9	37.5
No	15	62.5

Imaging features of the lesions

Nine (37.5%) children underwent computed tomography (Figure 5). ultrasound shows a hollow mass of cystic cavities with hypoechoic fluid content, of variable sizes and walls. fine. The CT scan when it was carried out showed a homogeneous, hypodense tumor, with thin partitions, not enhanced by contrast.

Treatment and outcomes

All patients underwent surgery; 18 (87.5%) patients underwent total excision of the tumor. Six (25%) patients had received sclerotherapy preoperatively. Postoperative complications were observed in 6 (25.0%) patients; all six patients had lymphedema. Regarding sequelae, they were present in 9 (37.5%) patients, with the commonest being esthetic sequelae. We did not find other sequelae such as dysphagia, even less laryngeal dyspnea. (Table 4).

Table 4: Patient outcomes

Variables	Number of patients (N=24)	Frequency (%)
Complications		
No complication	18	75.0
Lymphedema	6	25.0
Sequelae		
No sequelae	15	62.5
Esthetic	9	37.5

Discussion

This is a study of the clinical characteristics and outcomes of pediatric patients with cystic lymphangioma. Cystic lymphangiomas are rare benign dysembryoplasias resulting from a defect in lymphatic vessels, causing a large fluid-filled sac to form in the nuchal region (11). Most cases occur in the posterior triangle of the neck (4). The prevalence worldwide is approximately 0.2–3% (12). According to existing literature, approximately 50% of cases have visible lesions at birth, and 90% of cystic lymphangiomas develop before the age of 2 years old (10, 13); we had similar findings in this study. We registered 24 cases in ten years, consistent with the finding of Lin et al.; they registered four cases in four years (14). However, Ozen et al. found less cases 17 cases in 20 years (15).

The International Society for the Study of Vascular Anomalies calls this disease macrocystic lymphatic malformations (16). It is commonly associated with aneuploidy (e.g., Turner, Down, Patau, and Edward syndromes) and other genetic syndromes (Alpert's, Fryns', and Noonan syndromes, congenital anomalies) (17). The abnormal expression of vascular endothelial growth factor (VEGF)-C and VEGF receptor type 3 (VEGFR3), as well as PIK3CA mutations, have been implicated in the pathogenesis of intrauterine lesions (16). We have not recorded comorbidities or associated genetic syndromes. Both sexes are affected in a similar manner, regardless of whether the patients are adults or children. There is a slight predominance of boys, as seen in our study (18-19); this is unlike the female predominance reported by Donka et al. (1). The average age of our population was 4.5 years, which is almost same as the reported average age in existing studies (2,3).

Regarding the clinical presentation, all our patients presented with soft, compressible, painless, transluminal, regular, well-defined, and noiseless lesions. Cystic lymphangioma manifests as slow-growing lesions that can regress spontaneously sometimes. Intralesional hemorrhage is common and explains the sudden increase in the

volume of the lesion (18). Large-volume tumors cause compression of vital structures such as the sympathetic chain, components of the carotid sheath, and branches of the hypoglossal, lingual, and facial nerves, resulting in dysphagia and dyspnea. We observed no signs of compression. Most patients in this study had left-sided lesions, similar to the observations by Ozen et al; they found left-sided lesions in 53% of cases (20).

Few patients had suppurative lesions at the time of diagnosis. These have been described in the literature (21). Hag et al. reported suppurative lesions in 35% of 20 patients in Khartoum (22). Suppurative lesions can be detected in utero with ultrasonography performed during the first or second trimester (23). They are seen as bilateral thin-walled cystic lesions, with a septa or not, in the fetal occipito-cervical region on the sagittal and axial views (7). Differential diagnoses include hematoma, abscess, lymphocele, and soft tissue sarcomas. In our study, no antenatal screening was performed. Additional imaging is needed to strengthen the diagnosis and assess the relationship of the tumor with nearby structures and vessels, especially when anticipating possible surgery. On ultrasonography, the tumor appears as hypoechoic or anechoic lesions with sediment or fine internal echoes and reinforced posterior echoes. On computed tomography, it may be seen as a mass with a low fluid density (10–36 HU) with visible septa seen sometimes on contrast-enhanced images. Albumin lymphography and/or lymphoscintigraphy are useful in examining the lymphatic tracts; however, these examinations are rarely performed in practice and cysts are usually not opacified (7). Magnetic resonance imaging is the second-line imaging option. It helps to better visualize the content of the cyst and assess the perivascular extension of the lesion very well. Cystic lymphangioma is hyposignal in T1 and hypersignal in T2. Partitions are hyposignal in T1 and T2. With gadolinium injection, little or no parietal and septal enhancement is observed (21, 24). In our study, only nine children had undergone a medical imaging examination.

Treatment depends on the form and location. In macrocystic forms, aspiration and/or sclerotherapy are the first-line treatment. For microcystic infiltrating forms, curative treatment is often impossible. In our study, all patients had undergone surgery. In addition, 6 (25%) children had received sclerotherapy preoperatively as multiple percutaneous injection sessions of propanolol; the evolution was favorable. Other tumor reduction procedures can be combined with the help of laser or radiofrequency. Some authors suggest OK-432 intralesional therapy in patients with unresectable lymphangiomas).

The main differential diagnoses are cysts and vascular abnormalities, especially venous malformations in children (24). Diagnostic confirmation of cystic lymphangiomas is done by histopathological examination (25). To limit postoperative complications from postoperative lymphatic leakage, hemostasis should be done stepwise during cervicotomy (1). In this study, 6 (25%) patients had lymphedema. These data are similar to those of a previous study wherein the frequency of postoperative complications was 30% (18). The reported recurrence rate is 40% after incomplete resection and 17% after macroscopically complete resection for cystic lymphangiomas occurring anywhere [4, 10]. No recurrence was detected in our patients after a mean follow-up period of 32.6 months.

Conclusion

Cervical cystic lymphangioma is easy to diagnose without imaging. Surgery is the treatment of choice. However, non-surgical treatment options may be considered for large head and neck cystic lymphangiomas to avoid post-operative complications. to avoid postoperative complications. Histopathological examination of the surgical specimen is the mainstay of diagnosis.

References

1. Keïta A, Diallo I, Fofana M, Kourouma S, Diallo MMR, Diallo MA, et al. Cervicofacial Cystic Lymphangiomas and Review of the Literature: About 2 Cases at Donka National Hospital. IJOHNS 2022; 11(06): 292-9.
2. Berrada O, Beghdad M, El Krimi Z, Oukessou Y, Rouadi S, LarbiAbada R, Roubal M, Mahtar M. Cervicofacial cystic lymphangiomas in 17 childrens: A case series. Ann Med Surg (Lond). 2022 May 19;78:103835. doi: 10.1016/j.amsu.2022.103835. PMID: 35734738; PMCID: PMC9206928.
3. O. Oulghoul, FZ. Hadid, O. Benhoummad, Y. Rochdi, A. Raji. Surgical Management of Cervico-facial Cystic Lymphangioma in Children. European Journal of Medical and Health Sciences. June 2021. http://dx.doi.org/10.24018/ejmed.2021.3.3.840
4. Nadour K, Moujahid M. Cervicothoracic cystic lymphangioma: about a case. Pan Afr Med J [Internet]. 2016 [cited 2024 Jan 20];25. Available from: http://www.panafrican-med-journal.com/content/article/25/189/full/ (French)
5. Hakimi T. Giant cervical lymphangioma encompassing the neck great vessels (carotid artery & jugular vein) Case report. Annals of Medicine & Surgery [Internet]. 6 avr 2023 [cited 2024 janv 20]; Publish Ahead of Print. Available from: https://journals.lww.com/10.1097/MS9.0000000000000452
6. Mukakala AK, Banza MI, Musapudi EM, Lubosha NA, Kasanga TK, Nafatalewa DK, et al. Cystic lymphangioma of the neck in a young adult: a case study and a review of the literature. Pan Afr Med J. 2020

Jun 2 [cited 2024 Feb 3];36. Available from: http://www.panafrican-med-journal.com/content/article/36/54/full/ (French)
7. A Case Study of Pathology on a Rare Case of Scalp Lymphangioma in an Adult Patient - PMC [Internet]. [cited 2024 Jan 20]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10411358/ (French)
8. Figaro N, Rampersad R, Juman S. Doxycycline Sclerotherapy of a Cervical Cystic Hygroma: A Caribbean Institution Experience. Case Rep Otolaryngol 2020; 2020: 1-5.
9. Department of Hygiene, Victor Babeş University of Medicine and Pharmacy, Timişoara, Romania, Borugă VM, Szilagyi DN, Department of ENT, Faculty of Medicine, Victor Babeş University of Medicine and Pharmacy, Timişoara, et al. Cystic hygroma of the neck – case report. Rom J Morphol Embryol 2021; 62(3): 845-8.
10. Cystic Hygroma: An Overview - PMD. [cited 2024 Jan 20]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3047730/ (French)
11. Paediatric Vascular Abnormalities. A Clinical and Radiological Perspective - PMC. [cited 2024 Jan 20]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10723972/ (French)
12. Al-Karaja L, Ibdah MG, Tos SM, Giacaman N, Yousef AMA, Salhab R. Rapidly enlarging axillary cystic hygroma in a 6-year-old male patient: A case report and literature review. Int J Surg Case Rep 2023; 102: 107806.
13. J. Miloundja , AB Manfoumbi, R. Mba Ella. Nguema, L. N'Zouba Lymphangiome kystiquecervico-facial de l'enfant au Gabon Lymphangiome cervico-facial kystique chez l'enfant au Gabon Annales d'Otolaryngologie et de Chirurgie Cervico-faciale Volume 124, numéro 6, décembre 2007, pages 277-284 https://doi.org/10.1016/j.aorl.2007.03.006 .
14. S Chemingui, C. Amal, H. Ziedi, D. Mzoughi, S. Bouzid, et al Les lymphangiomes kystiques cutanés : une série de 16 cas. Annales de Dermatologie et de Vénéréologie – FMC Volume 2, Issue 8, Supplement 1, November 2022, Page A154 https://doi.org/10.1016/j.fander.2022.09.219 .
15. Azizi AN, Ghellab O, Ghellab I, Belkhir B. Compressive cervicofacial cystic lymphangioma at one month. (French)
16. Cystic Hygroma | Reference Article in Radiology | Radiopaedia.org [Internet]. [cited 2024 Jan 31]. Available from: https://radiopaedia.org/articles/cystic-hygroma-1 (French)
17. Rapid Expansion of Lymphatic Malformations: Imaging, Association with Asymptomatic Streptococcus Infection, Multidisciplinary Management, and Therapeutic Approach - PMC. [cited 2024 Feb 3]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10316009/ (French)
18. Lin HJ, Lin FCF, Yang TL, Chang CH, Kao CH, Tsai SCS. Cervical lymphatic malformations amenable to transhairline robotic surgical excision in children: A case series. Medicine 2021; 100(37) :e27200.
19. Surgical Treatment of Cervicofacial Cystic Hygromas in Children | ORL | Karger Publishers. [cited 2024 Feb 3].Available from: https://karger.com/orl/article-abstract/67/6/331/261099/Surgical-Treatment-of-Cervicofacial-Cystic?redirectedFrom=fulltext
20. ISSVA Classification of Vascular Abnormalities and Molecular Biology - PMC. [cited 2024 Jan 31]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8876303/ (French)
21. Massive cystic hygroma of the neck with respiratory distress in newborn. Journal of Pediatric Surgery Case Reports. 2021; 71: 101945.
22. Cervicofacial cystic lymphangiomas in 17 childrens: A case series - PMC. [cited 2024 Jan 30]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC9206928/
23. Spontaneous regression of lymphangiomas in a single center over 34 years - PMC. [cited 2024 Feb 3]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5640371/ (French)
24. Ozen IO, Moralioglu S, Karabulut R, Demirogullari B, Sonmez K, Turkyilmaz Z, et al. Surgical Treatment of Cervicofacial Cystic Hygromas in Children. ORL 2005; 67(6): 331-4.
25. Ali E, Karim N, Hicham A, Mohamed Z. Cystic lymphangioma of the oral floor extends to the submandibular region of adults. Pan Afr Med J. 2016; 24 [cited 2024 Feb 3]. Available from: http://www.panafrican-med-journal.com/content/article/24/202/full/ (French)
26. El Hag OA, Ali EAM, Osman MAK. Clinical Profile of Lymphatic Malformation from One Oral and Maxillofacial Center in Sudan. OJST 2017; 07(08): 345-52.
27. Behera S, Bawa M, Kanojia RP, Saha PK, Singh T, Samujh R. Outcome of antenatally diagnosed cystic hygroma - Lessons learnt. International Journal of Pediatric Otorhinolaryngology 2020; 138: 110227.
28. Delmas J. Cervicofacial lymphatic malformations of antenatal diagnosis. 2020. (French)
29. Bezzola T, Bühler L, Chardot C, Morel P. Surgical treatment of abdominal cystic lymphangioma in adults and children. Journal of Surgery. 2008; 145(3): 238-43. (French)

Figure Legends

Figure 1: Patient selection flow chart

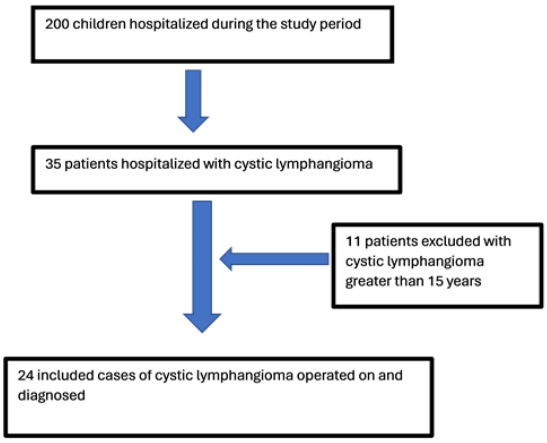

Figure 2: Distribution of cystic lymphangioma cases between 2013 and 2023

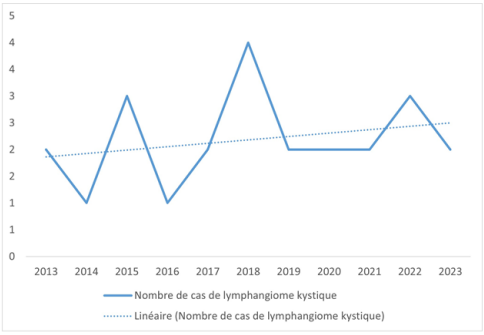

Figure 3: Extensive cystic lymphangioma manifesting as left laterocervical swelling in a male one-month child
A. Preoperative anteroposterior view of the parotid-masseter region to the left clavicle
B. Lateral view at the end of the intervention
C. Excised specimen with a rough and smooth surface measuring 7.5 × 6.5 × 1 cm

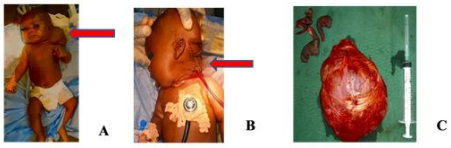

Figure 4: Patient distribution by location of tumor extension

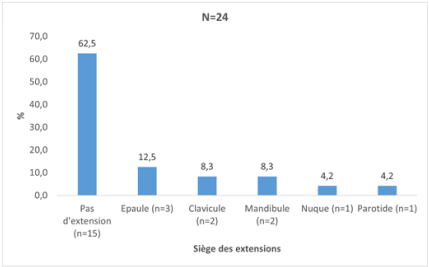

Figure 5: Representative image of polylobed isodense lymphangioma (red arrow) infiltrating the sternocleidomastoid muscle (yellow arrow) and adjacent to the carotid sheath (blue arrow)