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Cervical Cystic Lymphangioma in an Adult in His Fifties: A Case Report

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Abstract

Lymphangiomas are rare benign vascular tumors of lymphatic origin, of unclear pathogenesis and variable localization. Lymphangiomas are often found in young people but are exceptional over 50-years-old. Diagnosis is based on clinical and imaging findings and confirmed by anatomopathological examination of the surgical specimen. Treatment is surgical, and recurrence is frequent. We report a clinical observation of a 56-year-old patient who presented with a large cervical mass that had been evolving for XX years. Treatment was surgical. Pathological examination was in favor of a cystic lymphangioma. There was no recurrence at 1-year postoperative follow-up.

Keywords

Large, Right Later Cervical Mass, Elderly Subject

1. Introduction

Cystic lymphangiomas, also known as lymphatic malformations Cystic lymphangiomas, also known as lymphatic malformations, are rare benign dysembryoplasias of the lymphoganglionic system, responsible for a tumor syndrome caused by angiolymphatic proliferation leading to lymphatic sac filling (Miloundja et al., 2007). Their anatomical location is almost exclusively cervicofacial and their clinical onset is generally very early (Perkins et al., 2010). Diagnosis is suspected on the basis of clinical and imaging findings and confirmed by patho-

logical examination after surgical excision. It mainly affects children under 2 years of age (90% according to authors), of whom 50% - 65% are diagnosed at birth and 75% in the first year of life (Zainine et al., 2012). We report an exceptional case of cervical cystic lymphangioma in a 56-year-old adult.

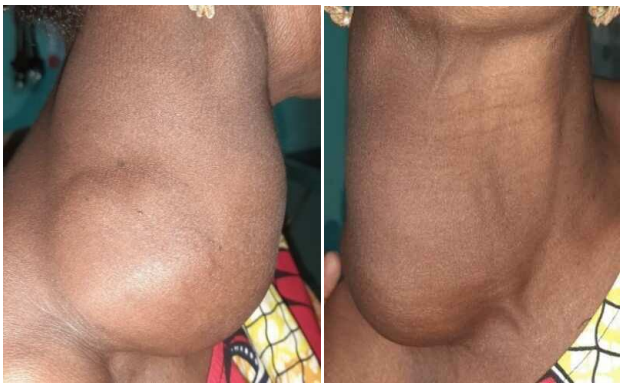
2. Observation

This was a 56-year-old female patient, with no tare or pathological history reported, who consulted for a voluminous right laterocervical swelling that had progressed over 7 years without signs of compression.

Physical examination revealed a voluminous right laterocervical mass measuring 21 cm in vertical long axis and 14 cm in transverse short axis, taking in the right jugulocarotid region, extending superiorly into the sub-maxillary region, posteriorly into the retro-spinal region, and bordering inferiorly in the supra-clavicular region and anteriorly in the pre-tracheal region (**Figure 1**). The mass was soft, irregularly surfaced, painless, mobile in relation to the deep planes and non-pulsatile. There were no sensitivomotor disturbances in the homolateral upper limb.

A cervical CT scan revealed a voluminous, thin walled, multicompartmental cystic mass on the right laterocervical side, pushing the right jugular vein medially and anteriorly, and the common carotid artery medially, and remaining in contact with their walls (**Figure 2**).

We diagnosed a cystic lymphangioma and decided to perform surgery. The operation consisted of excision by dissection with a large starfish incision beforehand, removing a large quantity of skin tissue. The aim was to facilitate exposure and skin adhesion on both sides of the mass. We found a multi-loculated mass with pockets of clear liquid content and other hematic. There were a few ruptured pockets. The excision was considered total and complete. The postoperative course was straightforward. Histology confirmed the diagnosis of cystic lymphangioma (**Figure 3**). There was no recurrence after 1 year of clinical and ultrasound monitoring (**Figure 4**).



(a) (b)

Figure 1. Right cystic lymphangioma with side view (a) and front view (b).

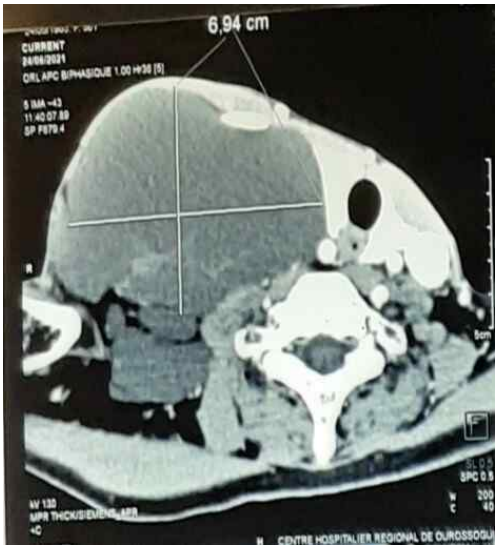


Figure 2. Sonographic cross-sectional image of the neck passing through the sixth cervical vertebra, with neighbouring structures to the left.

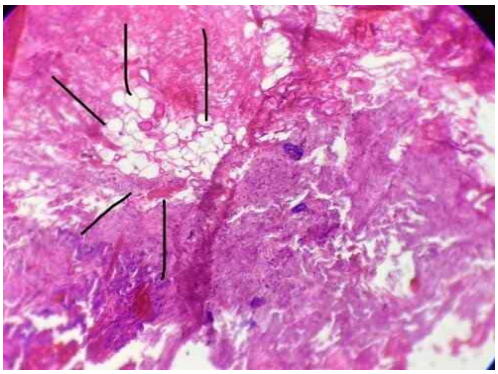


Figure 3. Proliferation of lymphatic vessels lined with flattened epithelial cells without atypia.

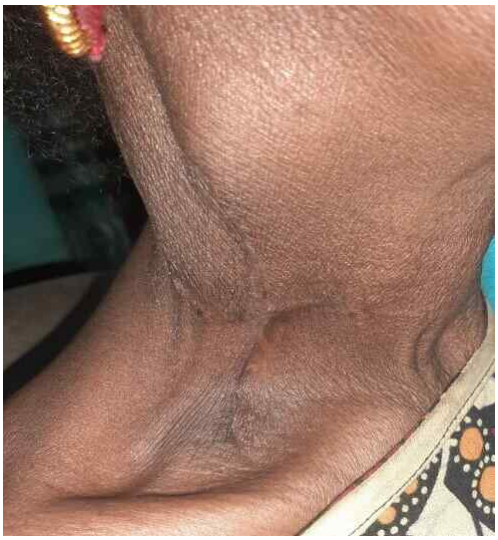


Figure 4. Post-op image at 1 year.

3. Discussion

Lymphangiomas are rare benign tumors. They can be found in the abdomen, thorax, and cervical region. The cervicofacial region is the preferred site in 75% of cases (Zainine et al., 2012). Cystic lymphangioma is a malformative lesion usually presenting in infancy (Owono et al., 2021). It accounts for 5% to 25% of vascular tumors and 6% of benign tumors in children. All ages combined, the incidence would be 0.8% of benign tumors and 0.1% of benign cervico-facial tumors (Oosthuizen et al., 2010). A few rare cases in young adults have been described between 20 and 30 (Owono et al., 2021; Sakthivel et al., 2018; Mukakala et al., 2020). However, discovery beyond the age of 50 remains exceptional, as shown by Bentebbiche's series in which the mean age of patients was 19.9 years, with extremes of 2 months and 59 years. The age group over 50 is the least representative, with 6% (Bentebbiche et al., 2020). Our patient was 56 years old and presented with a large cervical cystic lymphangioma that had been evolving for 7 years. The pathophysiology of lymphatic malformations remains poorly understood, despite very recent advances in biology and knowledge of the lymphatic system (Lerat et al., 2019).

Two pathogenic theories are mentioned in the literature:

- The traumatic theory explains the occurrence of these cysts by lymphatic obstruction or contusion; but this theory is rarely confirmed by clinical history (Handa et al., 2004).
- The congenital theory is currently the most widely accepted. Lymphangiomas are thought to arise from sequestration of an embryonic lymphatic sac, which gradually fills with lymphatic fluid (Anne et al., 1992). If this theory explains our case (in which the mass gradually increased in size), how can we explain the long latency, given that 50% of cysts are present at birth and 90% are diagnosed within the first two years (Giguère et al., 2002).

Cervico-facial cystic lymphangiomas are most often discovered following the appearance of a cervical mass (50% - 67% of cases (Chen et al., 2009). On physical examination, the swelling is soft, fluctuating, mobile and non-adherent to the superficial and deep planes, with positive transillumination, increasing in volume on crying and coughing. Size can vary from 1 to 30 cm. The swelling may be uni or bilateral, and outside infectious outbreaks the skin is healthy (Jean-Gilles & Brice, 2022). The left side is often the most affected (Sjogren et al., 2017; Bentebbiche et al., 2020) whereas in our patient, it was a right later cervical mass.

Ultrasound and computed tomography provide more information on the topographical features and contents (liquid, pseudo-liquid, cavernous) of this tumor (Oosthuizen et al., 2010). However, in addition to ultrasound findings, CT and MRI provide an initial pre-surgical work-up, highlighting infiltration of surrounding soft tissues and relationships with large vessels (Guruprasad & Chauhan, 2012).

Some authors maintain that MRI remains the examination of choice, espe-

cially in small children, and enables the diagnosis to be made, the extension of the cyst to be seen, and helps in the choice of treatment (Bentebbiche et al., 2020).

Histologically, it is a proliferation of small, thin-walled lymphatic vessels associated with fibrous tissue (Bahl et al., 2016).

Current management involves either surgery or sclerotherapy (Zainine et al., 2012). Our patient was treated surgically, and the follow-up was straightforward, with no sign of recurrence at 1-year follow-up. However, even after 4 years of follow-up, the risk of recurrence still requires monitoring (Zounon et al., 2022).

4. Conclusion

Cystic lymphangioma in adults in their fifties is a rare malformation. It is most frequently found in the cervix, but rarely on the right side of the cervix. Diagnosis is radiological, with histological confirmation. Management is often surgical. Long-term post-operative surveillance is essential, however, as this is a recurrent lesion.

Conflicts of Interest

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

References

Anne, F., Hurtier, O., Garcia, J. F., Filippini, J. F., & Piriou, A. (1992). Lymphangiome kystique du cou de l'adulte apport de l'échographie et de la tomodensitométrie: a propos d'un cas. *Annales de radiologie, 35,* 212-215.

Bahl, S., Shah, V., Anchlia, S., & Vyas, S. (2016). Adult-Onset Cystic Hygroma: A Case Report of Rare Entity. *Indian Journal of Dentistry, 7,* 51. https://doi.org/10.4103/0975-962X.179374

Bentebbiche, T., Daoud, F., Bencheikh, M., & Rous, I. (2020). Les lymphangiomes kystiques cervico-faciaux étude rétrospective sur une série de 16 cas. *Journal de la faculté de médecine de Blida,* 9-11.

Chen, W. L., Zhang, B., Wang, J. G., Ye, H. S., Zhang, D. M., & Huang, Z. Q. (2009). Excision chirurgicale des malformations lymphatiques macrokystiques géantes cervico-faciales chez le nourrisson et l'enfant. *Revue internationale d'oto-rhino-laryngologie pédiatrique, 73,* 833-837.

Giguère, C. M., Bauman, N. M., & Smith, R. J. (2002). New Treatment Options for Lymphangioma in Infants and Children. *Annals of Otology, Rhinology & Laryngology, 111,* 1066-1075. https://doi.org/10.1177/000348940211101202

Guruprasad, Y., & Chauhan, D. S. (2012). Cervical Cystic Hygroma. *Journal of Maxillofacial and Oral Surgery, 11,* 333-336. https://doi.org/10.1007/s12663-010-0149-x

Handa, R., Kale, R., & Upadhyay, K. K. (2004). Lymphangiome médiastinal isolé herniant à travers l'espace intercostal. *Journal asiatique de chirurgie, 27,* 241-242. https://doi.org/10.1016/s1015-9584(09)60042-7

Jean-Gilles, P. M., & Brice, S. J. (2022). *Oro-Cervical Cystic Hygroma, Surgical Management: Clinical Case.* I-Articles Médicaux, 14.

Lerat, J., Bisdorff-Bresson, A., Borsic, M., Chopinet, C., Couloignier, V., Fakhry, N. et al. (2019). Lignes directrices (version courte) de la Société française d'oto-rhino-laryngologie (SFORL) sur les malformations lymphatiques cervicales de l'adulte et de l'enfant: diagnostic. *Annales européennes d'oto-rhino-laryngologie, maladies de la tête et du cou, 136,* 109-112. https://doi.org/10.1016/j.aforl.2018.07.003

Miloundja, J., Ngoma, A. M., Ella, R. M., Edzang, B. N., & N'Zouba, L. (2007). Lymphangiomes kystiques cervicofaciaux de l'enfant au Gabon. *Annales d'Otolaryngologie et de Chirurgie Cervico-faciale, 124,* 277-284. https://doi.org/10.1016/j.aorl.2007.03.006

Mukakala, A. K., Banza, M. I., Musapudi, E. M., Lubosha, N. A., Kasanga, T. K., Nafatalewa, D. K. et al. (2020). Lymphangiome kystique du cou chez un adulte jeune: À propos d'un cas et revue de la littérature. *Pan African Medical Journal, 36,* Article 54. https://doi.org/10.11604/pamj.2020.36.54.21758

Oosthuizen, J. C., Burns, P., & Russell, J. D. (2010). Malformations lymphatiques: un algorithme de prise en charge proposé. *Revue internationale d'oto-rhino-laryngologie pédiatrique , 74,* 398-403.

Owono, J. P., Poduermo, S., & Silvère, N. O. (2021). Lymphangiome Kystique Abdominal: A Propos de deux Cas au Centre Hospitalier Régional Amissa Bongo de Franceville. *Health Sciences and Disease, 22,* 125-127.

Perkins, J. A., Manning, S. C., Tempero, R. M., Cunningham, M. J., Edmonds Jr, J. L., Hoffer, F. A., & Egbert, M. A. (2010). Lymphatic Malformations: Review of Current Treatment. *Otolaryngology—Head and Neck Surgery, 142,* 795-803. https://doi.org/10.1016/j.otohns.2010.02.026

Sakthivel, C. K., Thangadurai, R. R., Sahoo, A. K., & Gopalakrishnan, G. (2018). Adult Cystic Lymphangioma of the Parotid Gland: An Unwonted Presentation. *Cureus, 10,* e2644.

Sjogren, P. P., Arnold, R. W., Skirko, J. R., & Grimmer, J.F. (2017). Répartition anatomique des malformations lymphatiques cervico-faciales basée sur les groupes ganglionnaires. *Revue internationale d'oto-rhino-laryngologie pédiatrique, 97,* 72-75.

Zainine, R., El Aoud, C., Sellami, M., Beltaief, N., Sahtout, S., & Besbes, G. (2012). Les lymphangiomes kystiques: A propos de 25 cas. *Tunisie médicale, 90,* 19-24.

Zounon, A. A. W. D. S., Vodouhe, U. B., Mekire, A. G., Avakoudjo, F., Adjibabi, W., & Yehouessi, B. V. (2022). Prise en Charge du Lymphangiome Kystique Intra-Parotidien Exofacial de l'Enfant: Management of childhood exofacial intraparotid cystic lymphangioma. *Health Sciences and Disease, 23,* 112-115.