East African Scholars Journal of Medicine and Surgery
Abbreviated Key Title: EAS J Med Surg
ISSN: 2663-1857 (Print) & ISSN: 2663-7332 (Online)
Published By East African Scholars Publisher, Kenya

Volume-2 | Issue-3 | Mar-2020 | DOI: 10.36349/EASJMS.2020.v02i03.004

OPEN ACCESS

Case Report

Cystic Lymphangioma of the Chest Wall in Childhood: A case report

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Article History
Received: 28.02.2020
Accepted: 25.03.2020
Published: 30.03.2020

Journal homepage:
http://www.easpublisher.com/easms/

Quick Response Code

Abstract: Cystic hygroma or lymphangioma, is an uncommon congenital, benign tumour of the lymphatic system. The atypical localization of a lymphangioma in the chest wall is very rare. We report a case of cystic lymphangioma found in the chest wall of a 2-years-old boy. Parents discovered the mass at birth. The tumor was not painful. Ultrasonography and CT scan showed a heterogenous cystic mass in the left chest. The patient underwent surgery for mass removal. Histopathological examination showed a cystic lymphangioma.

Keywords: lymphangioma, 2-years-old boy, tumor, Histopathological examination.

INTRODUCTION

Cystic hygroma or lymphangioma, is an uncommon congenital malformation of lymphatic system. There are several classifications of CL. the current classification divides them into micro-cystic, cystic, and mixed lesions (Herbreteau, D. *et al.*, 1992). Their incidence in pediatric population rises an average of 1new case per 12000 births, while lymphangiomas represent almost 5-6% of all tumors in childhood (Faul, J. L. *et al.*, 2000). Several hypotheses or theories have been developed to explain the pathogenesis of this condition, principally the role of Vascular Endothelial Growth Factor-C (Enjolras, O. *et al.*, 2007; & Callahan, A. B., & Yoon, M. K. 2012). Physical examination, ultrasonography, CT scan and MRI substantially contributes to the diagnostic approach of a chest wall cystic lymphangioma (Yokoigawa, N. *et al.*, 2014; Patoulias, D. *et al.*, 2017; Lu, D. *et al.*, 2015; & McAlvany, J. P. *et al.*, 1993). histopathological examination confirms the diagnosis. The most preferred modality of treating cystic hygroma remains complete surgical excision (Yokoigawa, N. *et al.*, 2014; & Patoulias, D. *et al.*, 2017). We are reporting a case of Cystic Lymphangioma of the Chest Wall in a two years old boy. The informed consent from the patient for this study was obtained.

CASE PRESENTATION

A two years old boy, whose parents discovered a left chest mass. The mass was discovered at birth and gradually increased in size. The physical examination revealed a mass not painful on palpation, but adhered to the deep plane (Figure 1). The biological examinations did not reveal any anomaly with regard to the morphology, on the ultrasound, we found a lobulated, heterogeneous mass, with tubular anechoic formations; mass not in contact with intra-thoracic and intra-abdominal contents (Figure 2). The CT scan referred to a discretely heterogeneous, undifferentiated hypertrophy of the left lateral chest wall muscle, measuring 25 x 35 x 50 mm, with no suspicious bone involvement or vertebral collapse (Figure 3).

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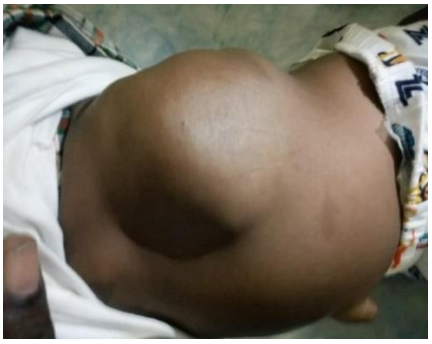


Figure 1. Palpable, painful mass

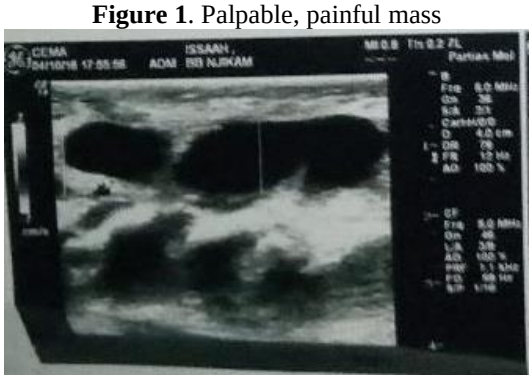


Figure 2. Ultrasonography showing the tumor with an internal hypoechoic pattern

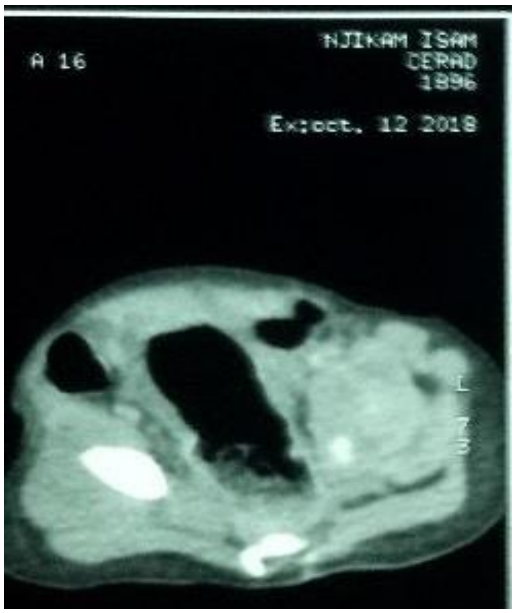
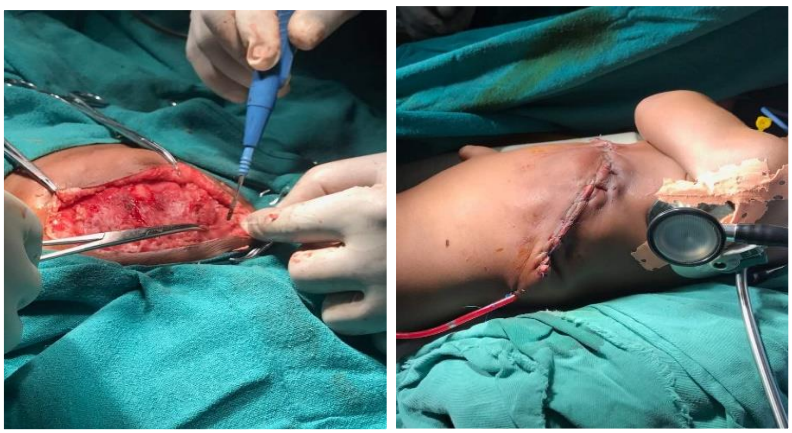


Figure 3. CT with a discretely heterogeneous, undifferentiated hypertrophy, measuring 25 x 35 x 50 mm

The patient underwent surgery for mass removal. a left thoraco-lateral incision was made. We found highlighted and extracted, a polycystic vascular mass producing a clear liquid and localized subcutaneously of the left profile chest (Figures 4 and 5).



(a) (b)

Figure 4. Procedure of lesion's excision

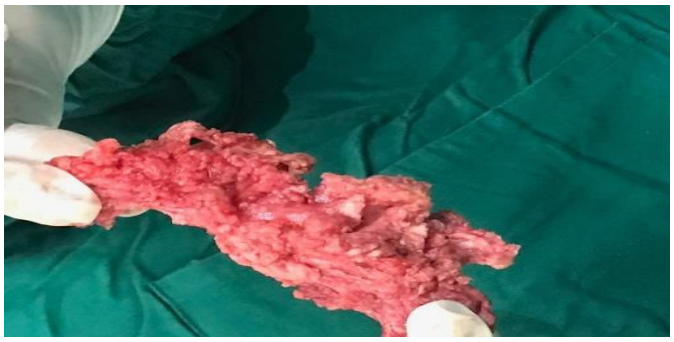


Figure 5. Total excised lesion Histopathological examination revealed a subcutaneous cystic lymphangioma.

DISCUSSION

Cystic lymphangiomas (CL) are rare benign dysembryoplasias of the lymphoganglionic system responsible for a tumor syndrome by angiolymphatic proliferation (Miloundja, J. *et al.,* 2007). They are hemodynamically inactive cystic lymphatic malformations consisting of abnormal lymphatic vessels and cysts of variable morphology (Wierzbicka, E. *et al.,* 2006).

There are several classifications of CL. the current classification divides them into micro-cystic, cystic, and mixed lesions (Herbreteau, D. *et al.,* 1992). Wegener in 1977 proposed a classification essentially used by surgeons that classifies CL into capillary lymphangiomas (including small vessels with narrow light), cavernous (dilated, anfractuous and inter-communicable) and cystic (or cystic hygroma with large confluent cavities filled with light yellow liquid) (Flanagan, B. P., & Helwig, E. B. 1977; & Landing, B.H., & Farber, S. 1956).

Several hypotheses or theories have been developed to explain the pathogenesis of this condition: First, it should be noted that the Vascular Endothelial Growth Factor-C is the receptor ligand VEGFR-3 and VEGFR-2, which, in transgenic mice, induce the growth of lymphatic vessels in the skin without influencing the development of blood vessels. Also, since this hyperplasia is selective, VEGF-C could play a role in diseases of the lymphatic system and, therefore, be the targets of new therapeutic alternatives (Enjolras, O. *et al.,* 2007; & Callahan, A. B., & Yoon, M. K. 2012). Next, we know that the endothelial cells of cystic lymphangiomas secrete a significant amount of bFGF (Fibroblast Growth Factor), which is an inducer of angiogenesis. In parallel, the level of an angiogenesis inhibitor, thrombospondin-1, secreted by the malformation cells, is lowered. Lymphangiomas are therefore, in part the result of a runaway angiogenesis and, in the future may be, antiangiogenesis. The specific treatments could be a new set therapeutic strategy in this pathology (Callahan, A. B., & Yoon, M. K. 2012; & Wassef, M. *et al.,* 2015). Note that a theory has been developed stipulating the origin of these malformations could be acquired based on the fact that the lymphatic circulation would be interrupted, thus causing an ectasis of the vessel. But the abundant anastomoses which exist in the lymphatic system, easy to highlight, the absence of anatomical or pathological cause at the origin of this lymphatic retention, oppose this hypothetical obstruction of all the lymphatic channels (Enzinger, & Weiss, S. W. 1995).

In our case, the patient was two years old. Yokoigawa *et al.,* described a lymphagioma of chest wall in a 2-month-old girl (Yokoigawa, N. *et al.,* 2014). For Patoulias *et al.*,, the child was 5 years old [6]. Despite the presence of a cystic lymphangioma at birth in 50% of all cases and by the age of 2 years in 90% of all cases, diagnosis may be delayed significantly, like the case found by Lee *et al.*,, the patient was 20 years old and others described cases by others authors (Lee, W. S. *et al.,* 2011; & Mirza, B. *et al.,* 2010). Lu *et al.,*

presented a case of giant lymphangioma at chest wall diagnosed prenatally at 19 weeks gestation (Lu, D. *et al.,* 2015).

Physical examination substantially contributes to the diagnostic approach of a chest wall lymphangioma cystic. Clinically, cystic lymphangioma occurs as a large, soft, cystic mass. Transillumination, where possible, may also help the diagnosed lolis, confirming the presence of a cystic lesion and excluding that of a solid structure, without any bruit (Yokoigawa, N. *et al.,* 2014; & Mirza, B. *et al.,* 2010). But Patoulias decribed a case with whom a Transillumination of the lesion was not feasible (Patoulias, D. *et al.,* 2017). Some of those tumors are not paintful, like the one we presented. Others found paintful (Patoulias, D. *et al.,* 2017; & Lee, W. S. *et al.,* 2011).

Concerning the dimensions of the tumor, they varies. In our case, it was a mass approximately 25 x 35 x 50 mm. Patoulias' case was a subcutaneous cystic lesion, 2.1 x 3.2 cm in dimensions [6]. The tumor presented by Yokoigawa measured 4 × 5 × 1 cm and by Lu was 14 x 9 x 9 cm (Yokoigawa, N. *et al.,* 2014; & Lu, D. *et al*., 2015).

The ultrasonography is helpful in the diagnosis of cystic hygroma and in the preoperative evaluation of the patient. Ultrasonography could show a characteristic multilobular cystic mass that contains a septum of variable thickness, highlighting the cystic structure, the hypoechoic content, the lobulated periphery, the absence of vessels, and presence of thin septula (Yokoigawa, N. *et al.,* 2014; & Patoulias, D. *et al.,* 2017). Other authors have also demonstrated solid areas in cystic hygromas (Kraus, R. *et al.,* 1986; & Sheth, S. *et al.,* 1987). Sheth *et al.,*.correlated sonographic findings with pathologic specimen and demonstrated that the echogenic component corresponded to a cluster of abnormal lymphatic channels, are too small to be resolved with ultrasound (Sheth, S. *et al.,* 1987). His importance is also in the evaluation of lymphangioma's dimensions and limits ruled out intrathoracic extension (Nakazato, Y. *et al.,* 1995). Ultrasound is very helpful in determining the extent of cystic hygromas before surgery and in assessing postoperative complications and recurrences (Kapoor, R. *et al.,* 1994). The characteristic sonographic appearance on antenatal ultrasonography is multiseptate, thin-walled cystic mass; occasionally the cystic mass may have a more complex echo texture with cystic and solid components (Mirza, B. *et al.,* 2010).

CT scan and MRI are important in order to determine the possibility of intrathoracic extension of the lymphangioma and the affinity with the adjacent anatomic structures (Lu, D. *et al.,* 2015; Kapoor, R. *et al.,* 1994; & Hoffman-Tretin, J. *et al.,* 1988). In particular, CT may be crucial for the configuration of an operative plan, highlighting the affinity of the lymphangioma with the adjacent structures, mainly the large vessels.

MRI allows a better delimitation of the lesion, in relation to the organs of neighborhood and muscles. Macro-cystic malformation appears in hypo signal in T1 and in hyper frank and homogeneous signal, on T2 weighted sequences (compared to,muscles). An intra cystic hemorrhage results in a hyper signal on the T1 weighted sequence. The intra cystic partitions are better visible after injection of gadolinium chelates (Siegel, M. J. *et al.,* 1989; & Yuh, W. T. *et al.,* 1991). MRI remains the exam of choice, to appreciate the extension in depth in the case of circumscribed forms of lymphangiomas which are, undetectable on clinical examination. It also makes it possible to assess the infiltration of the underlying tissues and thus prevent incomplete gestures (McAlvany, J. P. *et al.,* 1993). It is important to complete the diagnosis by the histological exam. In our case the diagnosis was confirmed by histopathological examination. Cystic hygromas are benign lesions and can remain asymptomatic in a patient for a long duration like Cahill *et al.,*., main observed complications were hemolytic anemia and delayed neurologic involvement (Cahill, A. M. *et al.,* 2011). Recent advances in sclerotherapy have expanded contemporary lymphangioma management options (Perkins, J. A. *et al.,* 2010). Some authors suggested either the conduction of systemic chemotherapy or the administration of interferona, when surgical intervention is not feasible, due to either the large dimensions or the increased likelihood of iatrogenic injury to adjacent anatomic structures but with poor results (Turner, C., & Gross, S. 1994; & Reinhardt, M. A. *et al.,* 1997). Sclerotherapy with intra-lesional bleomycin, as a primary treatment modality, for cystic hygroma, has been tried. Various case reports and original studies have documented good response to the therapy (Mahajan, J. K. *et al.,* 2004; & Orford, J. *et al.,* 1995). The other agent used as sclerosant is OK432, has more satisfactory results and less complications as compared to bleomycin. (Mahajan, J. K. *et al.,* 2004; & Orford, J. *et al.,* 1995; & Qin, Z. P. *et al.,* 1998). Radio-frequency ablation and laser excision of the lymphangiomas are also proposed for the treatment of the cystic hygroma. Laser has been increasingly used especially for laryngeal lymphangiomas. It causes point destruction of the lesion, thus avoiding damage to the adjacent vital structures (Selz, P. A., & Arjmand, E. M. 1998; & Bozkaya, S. *et al.,* 2006).

CONCLUSION

Cystic lymphangiomas (CL) are rare benign dysembryoplasias of the lymphoganglionic that occur principally in childhood. The mean treatment remains complete surgical excision.

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