Scientific Research Publishing

Surgical Science, 2023, 14, 486-495
https://www.scirp.org/journal/ss
ISSN Online: 2157-9415
ISSN Print: 2157-9407

Unusual Presentation of a Large Multi-Septated Cystic Hygroma: A Case Report and Comprehensive Literature Review

Md. Abir Tazim Chowdhury[1*], Sohail Ahmed[1], Md. Zulfiqur Haider[1], Niaz Ahmed[2], A. K. M. Fazlul Haque[3], Nasrin Aktar[1], Arup Khan[1], Md. Abdullah-Al-Mahammud Kabir[2], Md. Sifat Uddin[2], Abrar Bin Ahsan[2], Trishna Sarker[1], Md. Khaled Hossain[1], Munama Magdum[4]

[1]Department of Cardiothoracic and Vascular Surgery, Evercare Hospital Dhaka, Dhaka, Bangladesh
[2]Department of Cardiothoracic Anesthesia, Evercare Hospital Dhaka, Dhaka, Bangladesh
[3]Department of Plastic and Reconstructive Surgery, Evercare Hospital Dhaka, Dhaka, Bangladesh
[4]Department of Community Medicine, Shahid Monsur Ali Medical College, Dhaka, Bangladesh
Email: *chowdhuryabir0@gmail.com

How to cite this paper: Chowdhury, Md. A.T., Ahmed, S., Haider, Md.Z., Ahmed, N., Haque, A.K.M.F., Aktar, N., Khan, A., Kabir, Md.A.-A.-M., Uddin, Md.S., Ahsan, A.B., Sarker, T., Hossain, Md.K. and Magdum, M. (2023) Unusual Presentation of a Large Multi-Septated Cystic Hygroma: A Case Report and Comprehensive Literature Review. *Surgical Science*, **14**, 486-495.
https://doi.org/10.4236/ss.2023.147053

Received: June 20, 2023
Accepted: July 17, 2023
Published: July 20, 2023

Abstract

Background: Cystic hygromas are primarily found in the cervicofacial, thoracic, and abdominal regions, with limited occurrences in other areas. Despite existing literature on this condition, comprehensive descriptions and MRI findings of cystic hygromas in the extremities are rare. **Aim:** This case report aims to present a unique instance of a cystic hygroma in the left thigh. The objective is to provide detailed insights into the characteristics of this atypical presentation. **Case Presentation:** The case involves a 2-year-10 month-old girl with a cystic hygroma in her left thigh. The report includes a comprehensive description of the lesion's clinical features and diagnostic evaluation, emphasizing the MRI findings to enhance understanding of this rare occurrence. **Conclusion:** This case report highlights the rarity of cystic hygromas outside the cervicofacial, thoracic, and abdominal regions, explicitly focusing on the occurrence in the left thigh. By presenting detailed insights into the clinical features, MRI findings, histopathological findings, and the surgical approach employed, this report contributes to the existing knowledge on this condition in atypical locations and informs future treatment strategies.

Keywords

Cystic Hygroma, Lymphangioma, Multi-Septated Cystic Hygroma, Unusual Presentation, Vascular Tumors, Benign Soft Tissue Tumors

1. Introduction

Lymphangiomas are typically classified into capillary, cavernous, and cystic lymphangiomas. They can also be categorized based on the size of the cysts they contain as microcystic, macrocystic, or mixed lymphangiomas. Microcystic lymphangiomas have cysts smaller than 2 cm, while macrocystic lymphangiomas have cysts larger than 2 cm. Mixed lymphangiomas have cysts of varying sizes, some exceeding 2 cm and others measuring less than 2 cm [1] [2]. Cystic lymphangioma, also called cystic hygroma, is a congenital abnormality that affects the lymphatic system. The term "hygroma" comes from Greek and means a tumour filled with water [1]. About 50% of these malformations are visible at birth, with approximately 80% to 90% becoming apparent by age two [1] [2] [3] [4]. It's been observed that cystic hygromas do not prefer a particular gender [5]. However, if someone experiences an infection in their respiratory tract or skin or suffers trauma to their head and neck region, it can cause cystic hygroma to grow due to the buildup of lymph or blood. It is important to note that no reported cases of lymphangiomas turning into malignancies exist. Cystic hygromas are typically found in the cervicofacial regions, accounting for 80% of cases. They can also occur in other areas like the axilla, mediastinum, groin, tongue, and certain organs like the liver, spleen, kidney, and intestine. However, it is rare for them to develop in the limb [3] [6]. We have written a case report about a rare occurrence of cystic hygroma in the lower extremity. We provide detailed information on its diagnosis and treatment and a brief review of the available literature.

2. Case Summary

We present a case of a girl aged two years and ten months who was presented with a soft tissue mass located on the medial aspect of her left thigh. According to the parents, the swelling exhibited slight tenderness upon touch and had been present for two months. The size of the mass had progressively increased since its initial observation, without any history of trauma to the affected area. During the physical examination, a mass measuring 8 × 3 cm in diameter was observed on the medial aspect of the thigh, approximately 3 cm proximal to the left knee. The overlying skin displayed a notable prominent vein in the center of the mass. The mass was soft, slightly tender, and not adherent to the overlying skin, although no distinct mobility could be discerned from the underlying structures. The swelling exhibited incomplete compressibility, lacked pulsations, and showed no signs of a thrill or bruit. The transillumination test yielded positive results. No other swellings were observed in the patient's body, and there was no lymphadenopathy in the surrounding areas. Furthermore, no evidence of neurovascular compromise was identified. However, the patient does not exhibit any weight loss or history of fever. Routine laboratory investigations yielded the expected results. During an ultrasound, a tubular cystic lesion was discovered on the inner part of the lower left thigh close to the great saphenous vein. The lesion

measures approximately 8.2 × 2.6 cm and shows no abnormal blood flow or connection to the surrounding veins. Good arterial function was observed in the femoral, superficial femoral, and popliteal arteries, as well as the distal tibial arteries, with no signs of narrowing or dilation. The blood flow was found to be tri-phasic, indicating healthy arterial function. In ultrasonographic images, **Figure 1** shows the lesion and the vascular structures around it.

The magnetic resonance imaging (MRI) (**Figure 2**) results indicated a cystic hygroma, or lymphatic malformation, in the subcutaneous tissue of the left thigh, measuring 5.3 × 2.2 cm and displaying multiple septae with fluid intensity. The area was independent of any blood vessels. A post-gadolinium-DTPA scan revealed a cystic appearance. The left thigh muscles, neurovascular structures, and femur appeared normal, while no abnormalities were found in the right thigh.

Based on the clinical and imaging findings, a diagnosis of lymphatic malformation was suspected. Subsequently, the patient underwent surgical excision of the mass. During the surgical procedure, following the administration of general anaesthesia, the patient was subjected to a thorough physical examination by the surgical team. Additionally, ultrasonic imaging was performed, which confirmed the previously observed findings and aided in delineating the affected region. A longitudinal incision was made, with particular attention given to the centrally marked elliptical area where the prominent vein was identified. Subsequently, an incision was made over the marked area and extended through the subcutaneous tissue, and the sac was carefully opened, revealing a sac containing approximately 50 - 100 millilitres of serous fluid. Following the removal of the fluid, the mass exhibited shrinkage. The mass was primarily located beneath the skin, with a protrusion resembling a finger extending towards the underlying muscles. Despite this, its anatomical limits were not distinctly defined. The surgical procedure was thorough and precise. The surgeon took great care to protect the surrounding nerves and vessels, except for one venous tributary. **Figure 3(a)**, depicts the dissection plane, while **Figure 3(b)** illustrates the excised mass from the thigh, and **Figure 3(c)**, demonstrates the tumour-free area of the thigh.

After excision, a closed wound drainage device under negative pressure was placed in the cavity, and the wound was closed. After the removal, the mass was examined under a microscope to determine its composition. The specimen comprised fibrofatty tissue fragments covered in skin, measuring 7 × 4 × 2 cm. Upon closer inspection, the tissue contained large vascular spaces lined with endothelial cells. These spaces contained thick eosinophilic exudate, mature lymphocytes, and some red blood cells. Although there were lymphoid aggregates present, no cancer was detected. In **Figure 4**, tissue is displayed that represents a defining characteristic of cystic hygroma. The patient was discharged successfully, and we followed up for two months. We found that the wound had healed well, with no signs of residual swelling or neurovascular impairment.

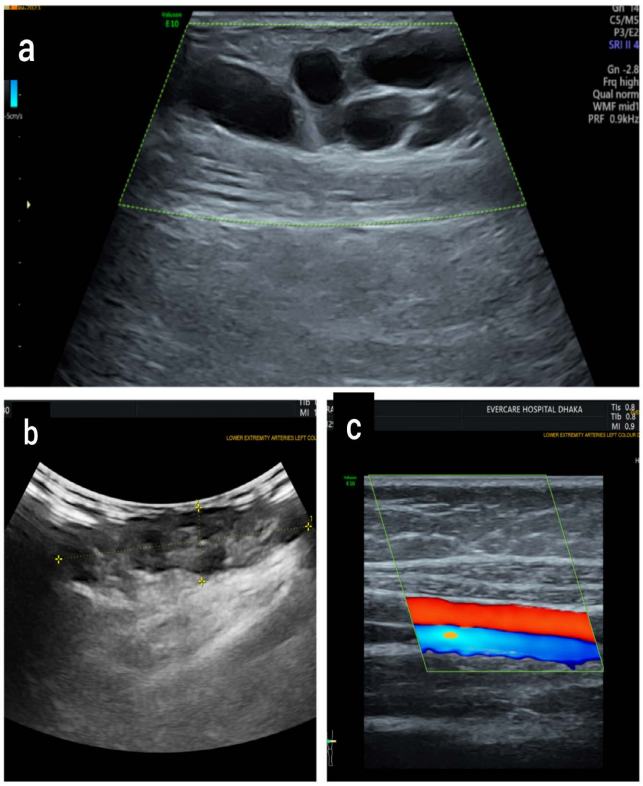

Figure 1. Ultrasound of the affected area demonstrated multi-cystic lesion with internal septatations (a), measuring 8.2 × 2.6 cm (b), and healthier vascular flow pattern pattern in relation to the lession (c).

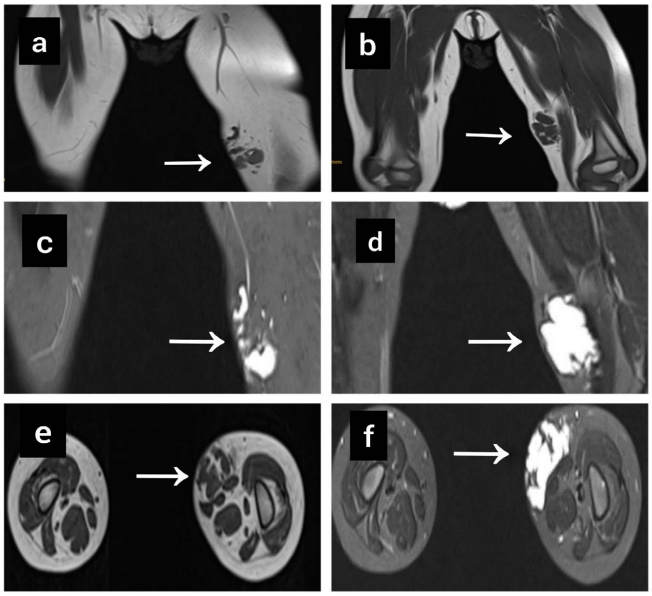

Figure 2. The MRI images depict a cystic hygroma in the left thigh, situated above the muscle and below the skin. In the T1-weighted image, a fluid-filled cavity with an irregular wall is visible, along with multiple septations (shown by a white arrow). The T2-weighted image reveals that the cystic hygroma appears hyperintense (also indicated by a white arrow) without impacting the surrounding structures. The images are labelled with coronal sections (a)-(c) and axial sections (e) (f).

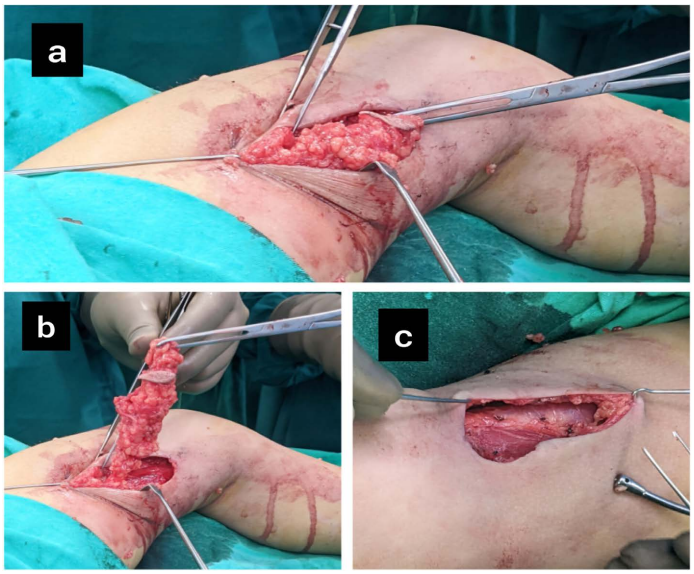

Figure 3. Figure (a) shows the dissection plane, revealing the anatomical location of the cystic hygroma in the left thigh. In Figure (b), the excised mass from the thigh is depicted, illustrating the complete removal of the cystic hygroma. Figure (c) demonstrates the tumor-free area of the thigh after successful resection.

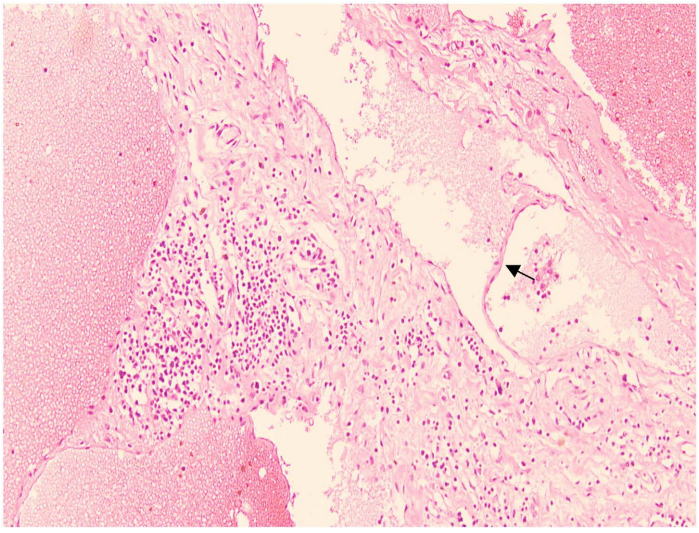

Figure 4. Histopathological image showing the characteristic features of the tissue from the cystic hygroma. The image reveals large vascular spaces lined with endothelial cells (black arrow), containing thick eosinophilic exudate, mature lymphocytes, and some red blood cells. Lymphoid aggregates are also observed, while no evidence of cancer is detected. Staining: hematoxylin and eosin stain. Magnification: 40×.

3. Discussion

In Greek, hygroma refers to a tumour that contains water. These are malformations of the lymphatic system that occur at birth. Cystic hygroma is more common than other types of lymphangioma and can consist of one or multiple large cystic lesions with limited connection to regular lymphatic channels [7]. Lymphangiomas are classified into three main types: capillary, cavernous, and cystic.

An easier way to categorize them is by the size of the cysts they contain: micro-cystic, macro-cystic, and mixed. Microcystic lymphangiomas have cysts less than 2 cm in size, while macrocystic lymphangiomas have cysts more than 2 cm in size. Mixed lymphangiomas have cysts of varying sizes, ranging from over 2 cm to under 2 cm [2] [8]. During gestation, the developing embryo has six lymphatic sacs located in different regions of the body. A network of lymphatics grows, and connective tissue infiltrates these sacs to create lymph nodes. Lymphangiomas are growths that occur due to the abnormal sequestering of lymphatic tissue from sacs during development [8].

There are certain chromosomal abnormalities that have been associated with cystic lymphangiomas, such as trisomies 13, 18, and 21, Noonan syndrome, Turner syndrome, and Down syndrome [9]. These congenital tumors may experience delayed growth. However, lymphangioma may also occur in adults due to external causes like infection, surgery, local trauma, or radiation therapy [10]. It is worth noting that our patient did not develop the condition due to any acquired etiology. Cystic lymphangioma is typically diagnosed before age two, with a prevalence of 1.2 - 2.8 cases per 100,000 children [11]. It is less commonly observed during adulthood. In this case report, the patient was a child who was two years and eight months old. Cystic hygromas can appear in different body parts, including the neck, armpit, chest, groin, and under tongue. They can also occur in rare cases in organs such as the liver, spleen, kidney, and intestine. Similar lesions can be found in the omentum and intestine mesentery [3] [4] [5] [6]. In a study of 1331 benign soft tissue tumors, Myhre-Jensen discovered 19 cases of lymphangiomas (1.4%), among which seven were identified in the neck. However, the location of the remaining 12 cases was not discussed [12]. Meanwhile, Caro *et al.* evaluated 14 cases of lymphangiomas in children and found that only one case occurred outside the neck, chest, or abdomen. The outlier case involved lymphangiomas along the inner wall of the right pelvis [13]. It is rare to come across a cystic hygroma in other locations, which can prove to be a challenging diagnosis at times [14] [15]. It is rare to come across a cystic hygroma in other locations, which can prove to be a challenging diagnosis at times. We had a similar experience when we first saw the case and identified it as a hemangioma through differential diagnosis. In this case, we addressed a lesion located on the left thigh. To the best of our knowledge, this is a seldom-documented instance in the literature concerning the thigh [16]. However, the involvement of the other parts of the extremities is rarely mentioned in the literature, and it occurs infrequently [17]. The usual presentation of cystic hygroma apparent at birth is a painless mass with worries and queries from the parents about the lesion. The other modes of presentation are related to the complications or effects of cystic hygroma, such as respiratory distress (in cervicofacial involvement), feeding difficulty, fever, a sudden increase in the size and infection in the lesion [7] [8]. In our case, the patient had swelling for two months and experienced slight pain and tenderness alongside it. These lesions are easily distinguishable during clin-

ical examinations due to their soft, compressible, non-tender, and transluminant nature, without any presence of a bruit [1]. Except for the pain and tenderness findings, above mentioned findings correlate with our case. Regarding investigation tools, Ultrasound is a great non-invasive option to consider as a first choice. An ultrasound of the lesion will typically reveal a multicystic structure with internal septations. Additionally, no blood flow will be detected on colour Doppler ultrasonography. In the case presented case report, the lesion was also large and followed this pattern [1]. In order to obtain a more accurate picture of the lesion, alternative methods such as CT scans and MRIs can be used. These methods can effectively determine the size of the lesion and its relationship to nerves and blood vessels [1] [18] [19]. These cysts can produce different types of fluids, such as milky, serous, serosanguinous or straw-coloured when aspirated using a wide-bore needle [6]. However, we did not perform pre-surgical aspiration to study the fluid as we confirmed the lymphatic origin of the lesion through radiological examination. In certain cases, to diagnose lesions occurring in uncommon areas like the larynx, inside the mouth, or in the orbit, a biopsy correlation may be necessary [20]. However, we aim for a simple approach of surgically removing the lesion and examining the tissue sample for diagnosis. Surgical removal is the most effective treatment for cystic hygroma, but sclerosant agents have shown promising outcomes as a management option. Other treatments like drainage, aspiration, radiation, laser excision, radio-frequency ablation, and cauterization have mixed results [20] [21]. After the surgical removal of cystic hygroma, patients may experience complications such as wound infection, bleeding, hypertrophied scar, and lymphatic discharge from the wound. The lesion may even recur despite complete excision in some cases (about 20%). Despite these possible complications, surgical removal is still the preferred option for life-threatening lymphangiomas and cases of spontaneous bleeding [1] [21]. Lymphangioma is identified under a microscope by enlarged lymphatic vessels in either a fibrotic or loose stromal background. Additionally, areas of papillary endothelial proliferation have been observed alongside the typical histological appearance. One should be cautious not to mistake cystic hygroma for cavernous hemangioma, as lymphangioma can be distinguished by the presence of lymphatic spaces with thin walls containing fibrous tissue, smooth muscle, and lymphoid aggregates. Lymphangioma is a benign tumor that can be treated through surgical excision [22].

4. Conclusion

We want to share a unique case of a cystic hygroma found in a child's lower limb. While cytological examinations can help diagnose, we rely on clinical and radiological evaluations before deciding on surgery to avoid invasive procedures. We later confirmed the diagnosis through histological examination. Although cystic hygroma cases involving the extremities are infrequent in literature, clinical-radiological co-relation can significantly help achieve appropriate manage-

ment and a favourable outcome.

Recommendation

Based on our clinical experience with this specific case and after reviewing several pieces of literature, we would like to offer some recommendations for readers who may encounter a similar clinical case in the future.

1) If you come across soft tissue swelling in children that do not have a bruit or thrill, it is essential to consider the possibility of cystic hygroma as a potential cause, even if it is found in unusual areas.

2) To distinguish cystic hygroma from other lesions like hemangiomas, it is best to rely on clinical findings and Ultrasound (USG) as the primary noninvasive approach.

3) If surgery is needed, it is recommended to use computed tomography (CT) or magnetic resonance imaging (MRI) to plan and carry out the procedure with greater accuracy.

4) To confirm the tissue diagnosis of cystic hygroma, it is necessary to perform histopathology as a mandatory step.

5) If you come across any cases of cystic hygroma in unusual locations during your clinical work, please report them. This helps to build the current knowledge base on the topic and improve understanding in the field.

6) Emphasize long-term follow-up due to a recurrence rate of approximately 20%, ensuring early detection and intervention if needed.

Ethics Statement

The parents of the patient provided written consent for the publication of any images or data that could potentially identify the patient in this article.

Acknowledgements

We want to express our gratitude to Prof. Dr. S. M. Mahbubul Alam for assisting us with the analysis of the histopathological slides.

Conflicts of Interest

The authors have no conflicts of interest related to the publication of this paper.

References

[1] Mirza, B., Ijaz, L., Saleem, M., Sharif, M. and Sheikh, A. (2010) Cystic Hygroma: An Overview. *Journal of Cutaneous and Aesthetic Surgery*, **3**, 139-144. https://doi.org/10.4103/0974-2077.74488

[2] Sanlialp, I., Karnak, I., Tanyel, F.C., Senocak, M.E. and Büyükpamukçu, N. (2003) Sclerotherapy for Lymphangioma in Children. *International Journal of Pediatric Otorhinolaryngology*, **67**, 795-800. https://doi.org/10.1016/S0165-5876(03)00123-X

[3] Kaur, N., Gupta, A., Amratash, and Singh, N. (2007) Giant Cystic Hygroma of the Neck with Spontaneous Rupture. *Journal of Indian Association of Pediatric Surgeons*, **12**, 154-155. https://doi.org/10.4103/0971-9261.34959

[4] Dhrif, A.S., El Euch, D., Daghfous, M., Cherif, F., Mokni, M. and Dhahri, A.B. (2008) Macrocystic Lymphatic Lymphangioma (Cystic Lymphangioma) of Upper Extremity: A Case Report. *Archives de Pédiatrie*, **15**, 1416-1419. https://doi.org/10.1016/j.arcped.2008.06.003

[5] Sichel, J.Y., Udassin, R., Gozal, D., Koplewitz, B.Z., Dano, I. and Eliashar, R. (2004) OK-432 Therapy for Cervical Lymphangioma. *Laryngoscope*, **114**, 1805-1809. https://doi.org/10.1097/00005537-200410000-00024

[6] Kocher, H.M., Vijaykumar, T., Koti, R.S. and Bapat, R.D. (1995) Lymphangioma of the Chest Wall. *Journal of Postgraduate Medicine*, **41**, 89.

[7] Manikoth, P., Mangalore, G.P. and Megha, V. (2004) Axillary Cystic Hygroma. *Journal of Postgraduate Medicine*, **50**, 215-216.

[8] Fonkalsrud, E.W. (2006) Lymphatic Disorders. In: Grosfeld, J.L., O'Neill Jr, J.A., Coran, A.G., Fonkalsrud, E.W. and Caldamone, A.A., Eds., *Pediatric Surgery* (6*th Edition*), Mosby Elsevier, Chicago, 2137-2145. https://doi.org/10.1016/B978-0-323-02842-4.50136-4

[9] Sehgal, V.N., Sharma, S., Chatterjee, K., Khurana, A. and Malhotra, S. (2018) Unilateral, Blaschkoid, Large Lymphangioma Circumscriptum: Micro-and Macrocystic Manifestations. *Skinmed*, **16**, 411-413.

[10] Chung, J.C. and Song, O.P. (2009) Cystic Lymphangioma of the Jejunal Mesentery Presenting with Acute Abdomen in an Adult. *Canadian Journal of Surgery*, **52**, E286.

[11] Guruprasad, Y. and 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

[12] Myhre-Jensen, O. (1981) A Consecutive 7-Year Series of 1331 Benign Soft Tissue Tumours: Clinicopathologic Data. Comparison with Sarcomas. *Acta Orthopaedica Scandinavica*, **52**, 287-293. https://doi.org/10.3109/17453678109050105

[13] Caro, P.A., Mahboubi, S. and Faerber, E.N. (1991) Computed Tomography in the Diagnosis of Lymphangiomas in Infants and Children. *Clinical Imaging*, **15**, 41-46. https://doi.org/10.1016/0899-7071(91)90047-Y

[14] Wright, C.C., Cohan, D.M., Vegunta, R.T., Davis, J.T. and King, D.R. (1996) Intrathoracic Cystic Hygroma: A Report of 3 Cases. *Journal of Pediatric Surgery*, **31**, 1430-1432. https://doi.org/10.1016/S0022-3468(96)90847-7

[15] Hamada, Y., Yagi, K., Tanano, A., *et al.* (1998) Cystic Lymphangioma of the sCrotum. *Pediatric Surgery International*, **13**, 442-444. https://doi.org/10.1007/s003830050364

[16] Pandit, S.K., Rattan, K.N., Budhiraja, S. and Solanki, R.S. (2000) Cystic Lymphangioma with Special Reference to Rare Sites. *The Indian Journal of Pediatrics*, **67**, 339-341. https://doi.org/10.1007/BF02820682

[17] Thakur, S.K. (2010) Unilocular Cystic Lymphangioma of Thigh—An Extremely Rare Clinical Entity. *Indian Journal of Surgery*, **72**, 417-418. https://doi.org/10.1007/s12262-010-0142-0

[18] Mansingani, S., Desai, N., Pancholi, A., Parajapati, A., Vohra, P. and Raniga, S. (2005) A Case of Axillary Cystic Hygroma. *Indian Journal of Radiology and Imaging*, **15**. https://link.gale.com/apps/doc/A158965207/AONE?u=anon~21d98473&sid=googleScholar&xid=9d04bd82

[19] Arora, A., Narula, M.K., Sonkar, P. and Chadha, R. (2003) Cystic Hygroma of Chest Wall. *Indian Journal of Radiology and Imaging*, **13**, 120.

[20] Nazarian-Mobin, S.S., Simms, K., Urata, M.M., Tarczy-Hornoch, K. and Hammoudeh, J.A. (2010) Misleading Presentation of an Orbital Lymphangioma. *Oral Surgery*, *Oral Medicine*, *Oral Pathology*, *Oral Radiology*, **109**, E82-E85. https://doi.org/10.1016/j.tripleo.2009.08.042

[21] Sobol, S.E. and Manoukian, J.J. (2001) Acute Airway Obstruction from a Laryngeal Lymphangioma in a Child. *International Journal of Pediatric Otorhinolaryngology*, **58**, 255-257. https://doi.org/10.1016/S0165-5876(01)00433-5

[22] Derin, S., Şahan, M., Dere, Y., Çullu, N. and Şahan, L. (2014) Cervical Cystic Hygroma in an Adult. *Case Reports in Pathology*, **2014**, Article ID: 209427. https://doi.org/10.1155/2014/209427