

## A Case Report On Surgical Encapsulation Of A Large Cervical Cystic Hygroma.

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### Abstract:

Cystic hygroma is a medical condition that occurs when the lymphatic system develops abnormally, resulting in the expansion of lymphatic vessels called lymphangiomas.

Cervical Lymphangioma is commonly found in the posterior triangle of the neck and can affect vital structures like the sympathetic chain, carotid artery contents, and branches of nerves such as hypoglossal, lingual, and facial.

This case report aims to outline the management of large cervical cystic hygroma in 27 years old female patient and its Surgical treatment.

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Cystic hygroma is a condition that results from the abnormal development of lymphatic vessels known as lymphangioma. [1,2]. Lymphangioma (LM) is thought to arise for the following reasons: [3]

- 1- Sequestration of lymphoid tissue from lymph sacs during development,
- 2- Failure of these tissues to communicate with the lymphatic or venous system,
- 3- Tissue dilatation leading to cystic morphology

Abnormal lymphatic system development can result in the growth and infiltration of lymphatic vessels into surrounding structures, ultimately leading to canalization. This creates areas of secretion accumulation due to the absence of a drainage pathway, resulting in the formation of cystic components. The type of lymphangioma that develops is dependent on the surrounding tissue environment, with LMs commonly forming in loose areolar tissue, while capillary and cavernous forms are more prevalent in muscles.

The investigations used in the cell proliferation indicators have shown that lymphangioma expansion is accompanied by engorgement more than cell proliferation. Molecular studies advocate that vascular endothelial growth factor C (VEGF-C) and its receptors may contribute to the development of LMs [4]. It is found primarily in the head and neck, but may also be present in other anatomical sites such as the thorax, shoulder, abdomen, pharynx, and mediastinum. The size of these cysts varies from a few millimeters to several centimeters in diameter [5,6].

The most common location of the tumor is the posterior triangle of the neck. Vital structures such as the sympathetic chain, the contents of the carotid artery, and branches of the hypoglossal, lingual, and facial nerves can be affected [7]

Cystic hygromas can be classified as septated or non-septated (multiloculated or nonlocalized) solitary cavities respectively. The incidence in adulthood is rare and the cause is uncertain, although trauma and upper respiratory tract infection have been suggested as possible triggers for the onset [8,9]. Cystic hygroma of the neck presents as a large, deep, diffuse swelling [10,11].

Giguère et al suggested classified lymphangiomas according to the size of the cystic contents [12]:

- Microcystic - Cystic spaces < 2 cm.
- Macrocystic - Cystic spaces ≥ 2 cm.
- Mixed lesions.

De Serres et al presented a staging system of LMs in the head and neck [13]:

- Stage I - Unilateral infrahyoid (17% complication rate)
- Stage II - Unilateral suprahyoid (41% complication rate)

- Stage III - Unilateral and both infrahyoid and suprahyoid (67% complication rate)
- Stage IV - Bilateral suprahyoid (80% complication rate)
- Stage V - Bilateral infrahyoid and suprahyoid (100% complication rate).

Although some authors have reported watchful waiting for lymphatic malformations (LMs), this should be considered only in patients who are asymptomatic. Medical treatment of LMs consists of the administration of sclerosing agents, such as OK-432 (an inactive strain of group

*A Streptococcus pyogenes*), pure ethanol, bleomycin, sodium tetradecyl sulfate, and doxycycline. [14] Guidelines for the use of percutaneous sclerotherapy to treat LMs of the head and neck have been developed by the Society of NeuroInterventional Surgery (SNIS). [15]

This case report aims to outline the management of cervical cystic hygroma in 27 years old female patient and its Surgical treatment.

### **I. Case presentation:**

A 27-year-old woman was referred to the Oral and Maxillofacial Surgery Department at Al-Ribat Police Hospital from General Surgery Department. She had been suffering from swelling on the left side of her neck for 11 years. As a temporary treatment, the patient underwent percutaneous drainage to have fluid drained from the lesion before being referred to the Oral and Maxillofacial Surgery Department. The Lesion was a small, solitary mass, and the patient had no difficulty swallowing or breathing. There was no history of upper respiratory tract infections or trauma during this period. On examination, there was a large swelling on the left side of the neck measuring 8 x 10 cm behind the ramus of the mandible inferiorly and occipital bone posteriorly and reaching supraclavicular inferiorly from the lower border of the mouth superiorly. The swelling was covered with normal skin color and on palpation, restricted neck movement on the left side was noted. It was soft, lobular, nonpulsatile, fluctuating, non-tender on the surface, shiny translucent, and extending into both the anterior and posterior triangles of the neck (Figure 1). Magnetic Resonance Images (MRI) showed a well-circumscribed, non-enhancing,

**(Figure 1) preoperative assessment with a large lesion seen on the left side**



lobulated, hypodense, multi-localized, and thin-walled cystic lesion (Figure 2). LMs appear hyperintense on T2-weighted images and hypointense on T1-weighted images. Based on the anamnesis, examination, and the results of the MRI, the initial diagnosis of “cystic hygroma in adults” was reported. The cyst involves vital structures such as sternocleidomastoid muscles, mastoid carotid sheath, internal jugular vein, facial, accessory, and hypoglossal cranial nerves.

(2,3,4,5). Laboratory tests were within the normal range. Chest x-ray was done and it revealed shadow of soft tissues beneath the left clavicle. Fig (6)

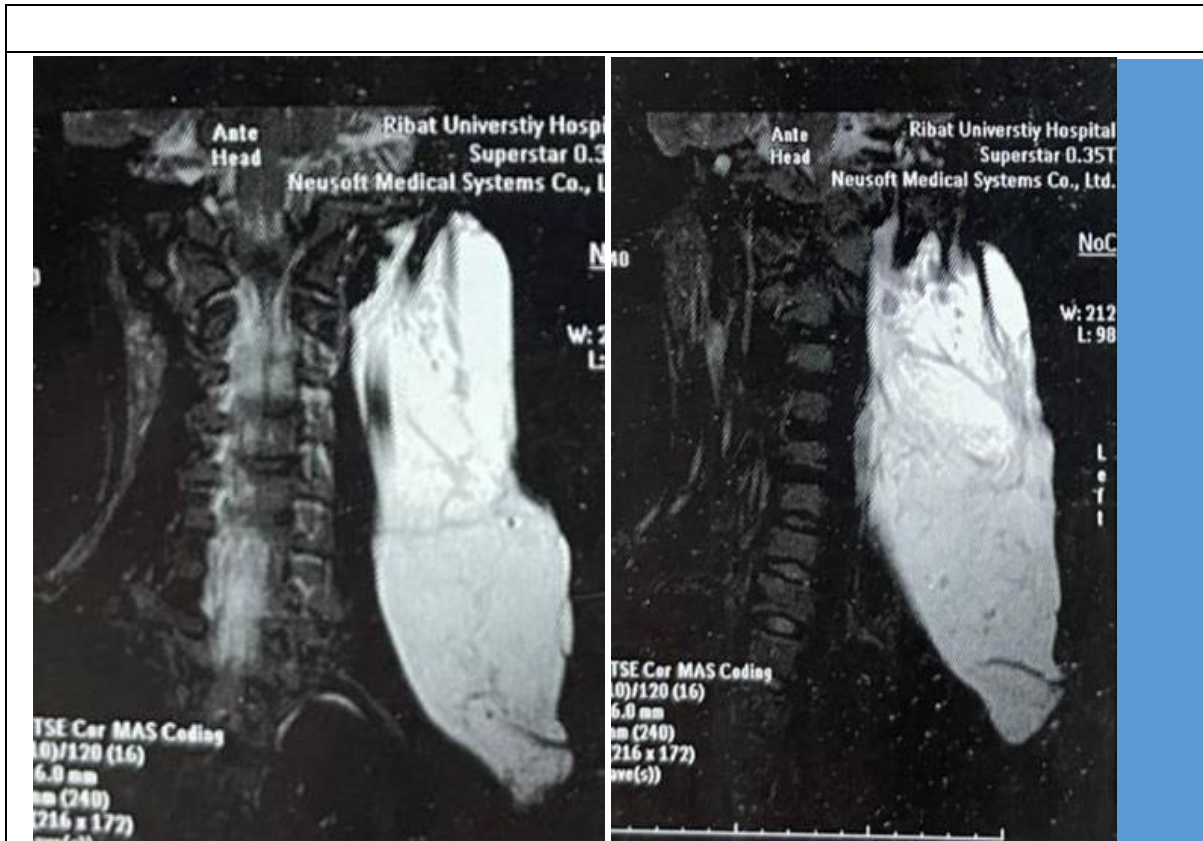
### **II. Surgical Management**

The patient went surgery under general anesthesia. The mass was explored and exposed through around 8 cm vertical incision. Dissection was done till reaching the cervical jugular lymph level III, with careful detection of close attachments around the cyst wall and adjacent structures, including the left sternocleidomastoid muscle,

carotid artery, internal jugular vein, spinal accessory nerve, and posterior cervical spinal nerves, they were all were observed and preserved.

Careful manipulation was done but due to the deep location of the cyst, it ruptured before taking a fluid sample via an empty syringe. The cyst wall was intact and no obvious remaining seen. without vital structural damage Fig (7,8,9), vacuum drain size 16 was inserted and secured, and the wound was closed by standard layering technique. The patient's recovery from General Anesthesia was uneventful and she was admitted to the ICU for the first 24 hours postoperatively, there were no signs of neurological dysfunction or deviation in the angle of the mouth, and she was discharged from the hospital after 7 days with oral medications. Pt follow-up continued for one month.

The histological findings showed cystic spaces that are lined with endothelium and have a small amount of surrounding stroma.



**Fig (2,3) Coronal MRI image reveals longitudinal expansion of the lesion**

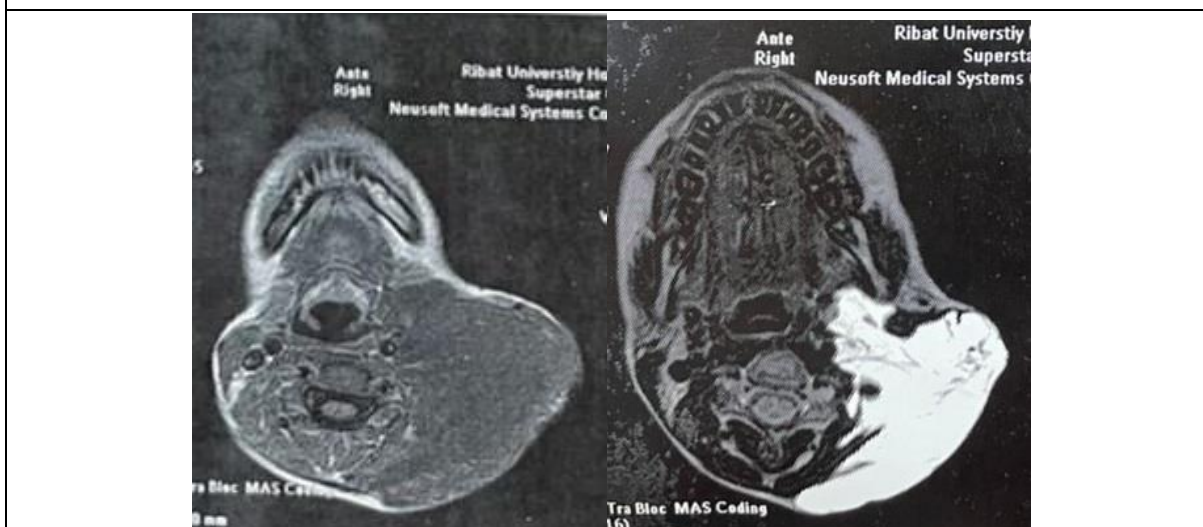


Fig (4,5) Horizontal MRI image reveals horizontal expansion of the cyst



Figure 6) Chest ray revealed soft tissues beneath the left



Fig 7) Intraoperative view of the cyst



Fig (8,9) Postoperative view of the

### III. Discussion:

The first mention of the condition was made by Redenbacher in 1828, with the term "cystic hygroma" coined by Wernher in 1834. According to the World Health Organization (WHO), there are three types of lymphangiomas: capillary, cavernous, and cystic. It is important to note that cystic hygroma can be used interchangeably with cystic lymphangioma and macrocystic lymphatic malformation when discussing the condition. Capillary lymphangiomas are often asymptomatic when they are small, while cavernous and cystic lymphangiomas share similar histological characteristics. However, cystic lesions are typically larger and more symptomatic.

Giguère et al. suggested the following categories for lymphangiomas depending on the size of the cystic component [12,13]:

- 1- Microcystic – spaces < 2 cm; usually above the mylohyoid muscle, basically in the oral cavity, tongue, submandibular region, and parotid,
- 2- 2 cm macrocystic - cystic areas; frequently below the level of the mylohyoid muscle, mainly anterior and posterior cervical triangles.
- 3- mixed lesions.

Although trauma and upper respiratory tract infection have both been suggested as potential triggers for its onset, the condition is uncommon in adults and has an unknown etiology. <sup>[17]</sup>

High-resolution ultrasound is one of the diagnostic investigations used to diagnose cystic tumors of the neck. It can show the existence of a cystic lesion with several septa and no internal vascular flow, allowing it to be distinguished from mixed vascular lesions. CT and magnetic resonance imaging (MRI), on the other hand, identify the mass's relation to the adjacent tissue more precisely.

The T1 and T2 MRI patterns assist in the detection of cleaved planes with muscles and can determine whether there is a vascular malformation. <sup>[18]</sup>

In the literature, numerous diagnostic techniques have been discussed. Previous to therapy, imaging technologies such as ultrasonography, magnetic resonance imaging, and computed tomography (CT) have been employed, although imaging choices vary depending on cost, convenience, and resolution. <sup>[19]</sup>

Complete surgical removal with preservation of the vital anatomic structures and functionality is the mainstay of treatment <sup>[20]</sup>. However, Nonsurgical treatment options for cystic hygroma include interferon alpha, laser therapy, and the infusion of intralesional sclerosing agents <sup>[21,22]</sup>

Cervical lymphangioma has a recurrence rate of about 15% <sup>[20]</sup>, although incomplete excision of the lesion might have a recurrence rate of up to 88%. <sup>[23]</sup>

#### **IV. Conclusion:**

Cystic hygromas are a rare type of malformation that can occur in adults. Failure to address cystic hygroma can lead to severe complications such as airway obstruction, difficulty swallowing, and obstructive sleep apnea. Infection and abscess formation can result from hemorrhaging within the cystic hygroma. Medical professionals use medical history, ultrasound of the neck, MRI, CECT,

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#### **Ethical approval**

As this is a case report, ethical approval has been exempted.

#### **Consent**

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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The authors declare that there is no conflict of interest.

#### **References:**

- [1]. Bloom DC, Perkins JA, Manning SC. Management Of Lymphatic Malformations. *Curr Opin Otolaryngol Head Neck Surg* 2004; 12: 500–4. Pmid:15548907.
- [2]. Naidu SI, Mccalla MR. Lymphatic Malformations Of The Head And Neck In Adults: A Case Report And Review Of The Literature. *Ann Otol Rhinol Laryngol* 2004; 113: 218–22. Pmid:15053205
- [3]. Mirza B, Ijaz L, Saleem M, Sharif M, Sheikh A. Cystic Hygroma: An Overview. *J Cutan Aesthet Surg*. 2010 Sep. 3 (3):139-44. [Qxmd MEDLINE Link].
- [4]. Perkins JA, Manning SC, Tempero RM, Cunningham MJ, Edmonds JL Jr, Hoffer FA, Et Al.
- [5]. Lymphatic Malformations: Current Cellular And Clinical Investigations. *Otolaryngol Head Neck Surg*. 2010 Jun. 142 (6):789-94. [Qxmd MEDLINE Link].
- [6]. Veeraraghavan G, Denny C, Lingappa A. Cystic Hygroma In An Adult; A Case Report. *Libyan J Med*. 2009;4(4):160–1. [PMC Free Article] [Pubmed] [Google Scholar] [Ref List]
- [7]. Hamoir M, Plouin-Gaudon I, Rombaux P, Francois G, Cornu AS, Desuter G Et Al. Lymphatic Malformation Of Head And Neck, A Retrospective Review And Support For Staging. *Head Neck*. 2001;23(4):326–37. [Pubmed] [Google Scholar] [Ref List].
- [10]. 7-. Antoniadis K, Kiziridou A, Psimopoulou M. Traumatic Cervical Cystic Hygroma. *Int J Oral Maxillofac Surg*. 2000;29(1):47–48. Doi: 10.1016/S0901-5027(00)80124-1. [Pubmed] [Crossref] [Google Scholar] [Ref List]
- [11]. 8- Schefter RP, Olsen KD, Gaffey TA. Cervical Lymphangioma In The Adult. *Otolaryngol Head Neck Surg* 1985; 93: 65–69. Pmid:3920626.
- [12]. 9. Antoniadis K, Kiziridou A, Psimopoulou M. Traumatic Cervical Cystic Hygroma. *Int J Oral Maxillofacial Surg* 2000; 29: 47–8. Doi:10.1034/J.1399-0020.2000.290111.X
- [13]. Shafer WG, Et Al. *A Textbook Of Oral Pathology*. 4. Philadelphia, PA: WB Saunders Co; 1983. Pp. 159–160.
- [14]. Barnhart RA, Brown AK., Jr Cystic Hygroma Of The Neck. *Arch Otolaryngol*. 1967;86(1):74–
- [15]. 78. Doi: 10.1001/Archotol.1967.00760050076016. [Pubmed] [Crossref] [Google Scholar]
- [16]. Giguère CM, Bauman NM, Smith RJ. New Treatment Options For Lymphangioma In Infants And Children. *Ann Otol Rhinol Laryngol*. 2002 Dec. 111 (12 Pt 1):1066-75. [Qxmd MEDLINE Link].
- [17]. De Serres LM, Sie KC, Richardson MA. Lymphatic Malformations Of The Head And Neck. A Proposal For Staging. *Arch Otolaryngol Head Neck Surg*. 1995 May. 121 (5):577-
- [18]. 82. [Qxmd MEDLINE Link]

- [19]. Acevedo JL, Shah RK, Brietzke SE. Nonsurgical Therapies For Lymphangiomas: A Systematic Review. *Otolaryngol Head Neck Surg.* 2008 Apr. 138 (4):418-24. [Qxmd MEDLINE Link].
- [20]. [Guideline] Heit JJ, Do HM, Prestigiacomo CJ, Delgado-Almandoz JA, English J, Gandhi CD, Et Al. Guidelines And Parameters: Percutaneous Sclerotherapy For The Treatment Of Head And Neck Venous And Lymphatic Malformations. *J Neurointerv Surg.* 2017 Jun. 9 (6):611-617. [Qxmd MEDLINE Link].
- [21]. Sherman BE, Kendall K. A Unique Case Of The Rapid Onset Of A Large Cystic Hygroma In The Adult. *Am J Otolaryngol Head Neck Med Surg* 2001; 22(3): 206–210
- [22]. Acevedo J.L.: Lymphatic Malformation (Cystic Hygroma). Medscape Reference. Available From: <https://emedicine.medscape.com/article/994055-overview>. Updated Jan 28, 2019. Accessed September 01, 2020. Google Scholar
- [23]. M. Biasotto, E. Clozza, And G. Tirelli, "Facial Cystic Lymphangioma In Adults," *The Journal Of Craniofacial Surgery*, Vol. 23, No. 4, Pp. E331–E334, 2012.
- [24]. Karkos PD, Spencer MG, Lee M, Hamid BN. Cervical Cystic Hygroma/ Lymphangioma: An Acquired Idiopathic Late Presentation. *J Laryngol Otol* 2005; 119(7): 561–3.
- [25]. Colbert SD, Seager L, Haider F, Evans BT, Anand R, Brennan PA. Lymphatic Malformations Of The Head And Neck-Current Concepts In Management. *Br J O Maxillof Surg.* 2013;51(2):98-102.
- [26]. Dokania V, Rajguru A, Kaur H Et Al. Sudden Onset, Rapidly Expansile, Cervical Cystic Hygroma In An Adult: A Rare Case With Unusual Presentation And Extensive Review Of The Literature. *Case Rep Otolaryngol* 2017; 2017: 1061958
- [27]. 20-. Aydin S, Demir MG, Selek A. A Giant Lymphangioma On The Neck. *J Craniofac Surg* 2015; 26: E323–5
- [28]. D. W. Kim, "OK-432 Sclerotherapy Of Lymphatic Malformation In The Head And Neck: Factors Related To Outcome," *Pediatric Radiology*, Vol. 44, No. 7, Pp. 857–857, 2014.
- [29]. M. A. Reinhardt, S. C. Nelson, S. F. Sencer, B. C. Bostrom, S. C. Kurachek, And M. E. Nesbit,
- [30]. "Treatment Of Childhood Lymphangiomas With Interferon-A," *Journal Of Pediatric Hematology/Oncology*, Vol. 19, No. 3, Pp. 232–236, 1997.
- [31]. Kaira V, Kaira P, Agarawal T. Cervical Cystic Lymphangiomas In Adults: A Case Series Of A Rare Entity With Literature Review. *Head Neck Pathol* 2021; 15: 503–8
- [32].
- [33].