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Cervical Cystic Hygroma in an Older Adult – a case study

Naczyniak limfatyczny torbielowaty u dorosłej, starszej osoby – opis przypadku

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Manuscript Preparation
E – Literature Search
F – Funds Collection

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ABSTRACT:

Introduction: Cystic hygromas are very uncommon malformations in adults. In most of the cases, they present as a painless, fluctuant swelling, usually located in the neck region. They tend to grow extensively if not removed. Cystic hygromas can range from 1 mm to several centimeters in size and are filled with clear- to straw-colored fluid. Surgical treatment remains the gold-standard treatment for these tumors. In some cases they can be misdiagnosed with branchial cysts, swollen lymph nodes or other pathologies. When this malformation is present in the prevertebral fascia its surgical treatment is challenging, and some authors advise its conservative treatment, which is in a great risk of its reoccurence. Because of potential malignant transformation in some percentage of branchial cyst cases, a similar situation was suspected.

Case report: The present report describes a case of cystic hygroma of the left side of the neck in a 92-year-old female adult and its clinical, radiological and operative features.

KEYWORDS: branchial cyst, cystic hygroma, lymphangioma, neck cyst, neck region

STRESZCZENIE:

Wstęp: Naczyniak limfatyczny torbielowaty jest bardzo rzadko spotykaną u osób dorosłych zmianą patologiczną. W większości przypadków objawia się ona jako bezbolesne, chełbocące wygórowanie tkanek miękkich, umiejscowione w obrębie szyi. Gdy guz nie zostanie usunięty, może dochodzić do jego znacznego wzrostu. Średnica, wielkość naczyniaków limfatycznych torbielowatych wynosi od 1 mm do kilku centymetrów, zaś ich wnętrze wypełnione jest płynem w kolorze od przezroczystego do słomkowego. Leczenie chirurgiczne pozostaje „złotym standardem" postępowania w przypadku tych guzów. Niekiedy mogą one zostać błędnie zdiagnozowane jako: torbiel boczna szyi, obrzęk węzłów chłonnych lub inne zmiany patologiczne. W sytuacji, gdy guz pokryty jest powięzią przedkręgową, jego leczenie operacyjne, chirurgiczne jest skomplikowane i wiąże się z podwyższonym ryzykiem uszkodzenia istotnych, sąsiednich struktur anatomicznych (naczynia, nerwy). W takich sytuacjach niektórzy autorzy zalecają zastosowanie alternatywnych, niechirurgicznych metod leczenia. Ze względu na potencjalną transformację złośliwą w pewnym procencie torbieli bocznych szyi, w opisywanym przypadku brano pod uwagę możliwość wystąpienia podobnej sytuacji.

Opis przypadku: W niniejszej publikacji opisano przypadek naczyniaka limfatycznego torbielowatego, zlokalizowanego w obrębie tkanek miękkich szyi po stronie lewej u 92-letniej kobiety, z uwzględnieniem jego cech klinicznych, obrazu radiologicznego i zastosowanego leczenia operacyjnego.

SŁOWA KLUCZOWE: naczyniak limfatyczny torbielowaty, naczyniak limfatyczny, obszar szyi, torbiel boczna szyi, torbiel szyi

ABBREVIATIONS

CH – cystic hygroma
CT – computed tomography
CTI – computed tomography imaging
FNAC – fine-needle aspiration cytology
MRI – magnetic resonance imaging
US – ultrasonography
WHO – World Health Organization

INTRODUCTION

One of the most commonly presented lymphatic malformations is cystic lymphangioma or hygroma cysticum colli, or cystic hygroma (CH). Cystic hygroma was first described in 1828 by Redenbacker and referred to as a "moist tumor" [1]. Nowadays, this benign developmental tumor is recognized as a lymphatic malformation (also known as a lymphangioma), that is the result of the collection of lymph in tissues that typically offers little resistance to expansion.

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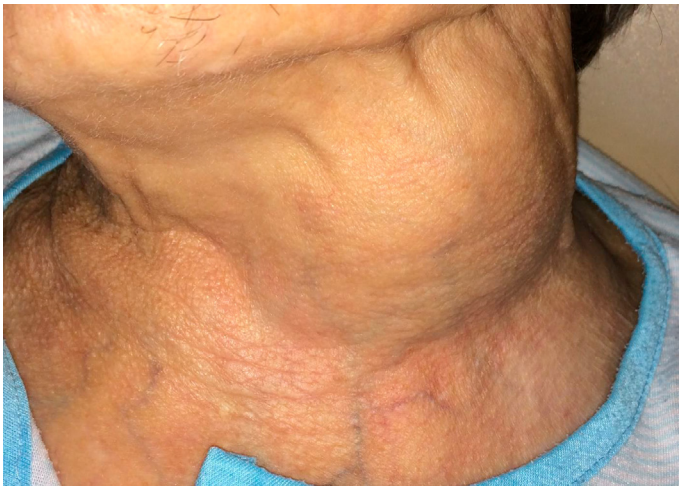

Fig. 1. Cystic mass on the left side of the neck.

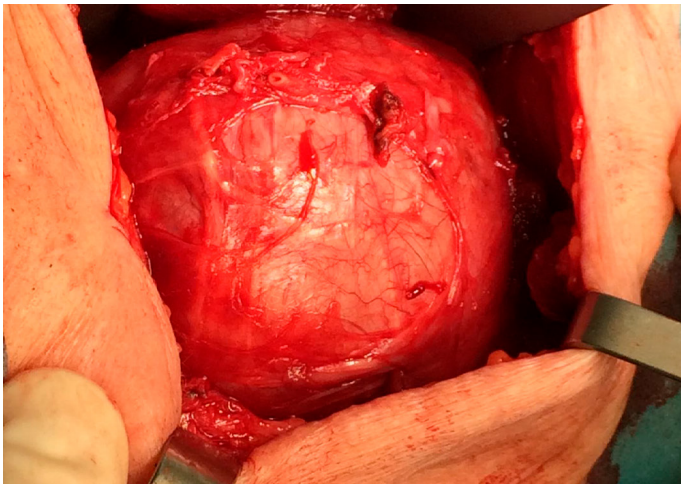

Fig. 3. Intraoperative view on well-defined cystic lesion.

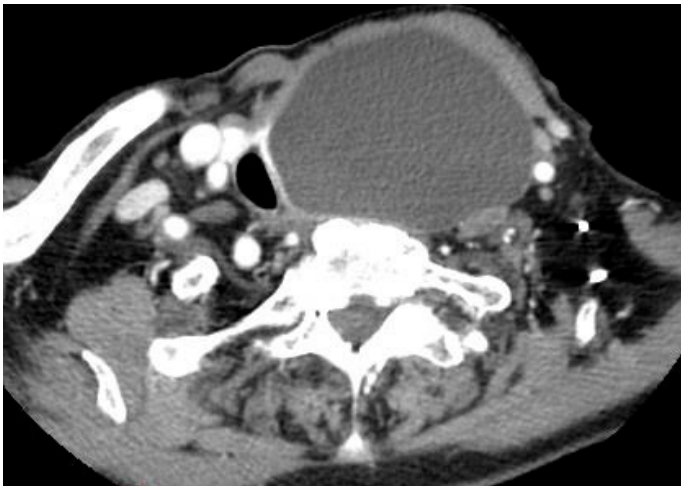

Fig. 2. Cystic mass in CT.

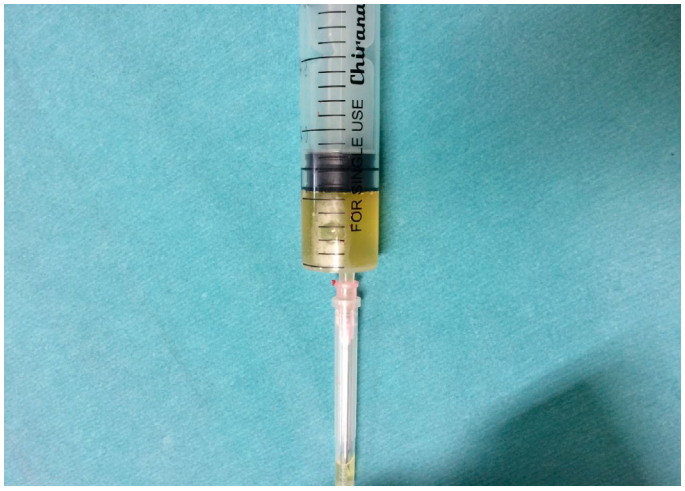

Fig. 4. Straw-colored fluid, aspirated from the cystic mass.

The incidence of cystic hygromas is estimated to be one case per 6000–16,000 live births [2]. It usually presents within 3 years of life. Approximately 50–60% of these malformations appear before one year, 80–90% before the end of the second year of life, with 95% occurring by adolescence [3]. Cystic hygromas are extremely rare malformations in adults.

Cystic hygromas usually affect the head and neck (~75%; due to their rich lymphatic drainage), with a predilection for the left side. Within the neck, the posterior triangle tends to be most frequently affected. Approximately 20% of cystic hygromas occur in the axilla. Less common locations include the mediastinum, groin, and retroperitoneum [2].

A cystic hygroma may be caused by genetic or environmental factors that cause abnormal development of the lymphatic vascular system during embryonic growth. It may occur on its own or as part of a genetic syndrome with other features, such as Turner syndrome, Down syndrome or Noonan syndrome [4]. Anomalies that are commonly associated with cystic hygroma also include cardiac defects, diaphragmatic hernia, and skeletal dysplasias [1]. Intrauterine alcohol exposure has also been associated with the development of cystic hygroma [2]. Three theories have been proposed to explain the origin of this abnormality. The first suggests that a blockage or arrest of normal growth of the primitive lymph channels occurs during embryogenesis. The second one suggests that the primitive lymphatic sac does not reach the venous system while the third advances the hypothesis that during embryogenesis, lymphatic tissue lays in the wrong area [5, 6].

The etiology of cystic hygromas in adults is controversial, but it may result from trauma, hormonal stimuli or from earlier respiratory infections [7]. In many cases, the cause is not known [4]. In the differential diagnosis of neck masses, thyroglossal duct, branchial cleft cyst, ranula, goiter, soft-tissue tumors, neck abscess, lymphadenitis, inclusion cyst of submandibular gland, haemangioma, lymphoma should be considered [2].

Cystic hygroma's treatment modalities comprise surgical excision, use of sclerosants, repeated aspiration, conservative management.

Authors present a case of a 92-year-old female patient with a large mass on the left side of the neck.

CASE REPORT

A 92-year-old female with no significant medical history presented with a large, cystic mass on the left side of the neck. Swelling had

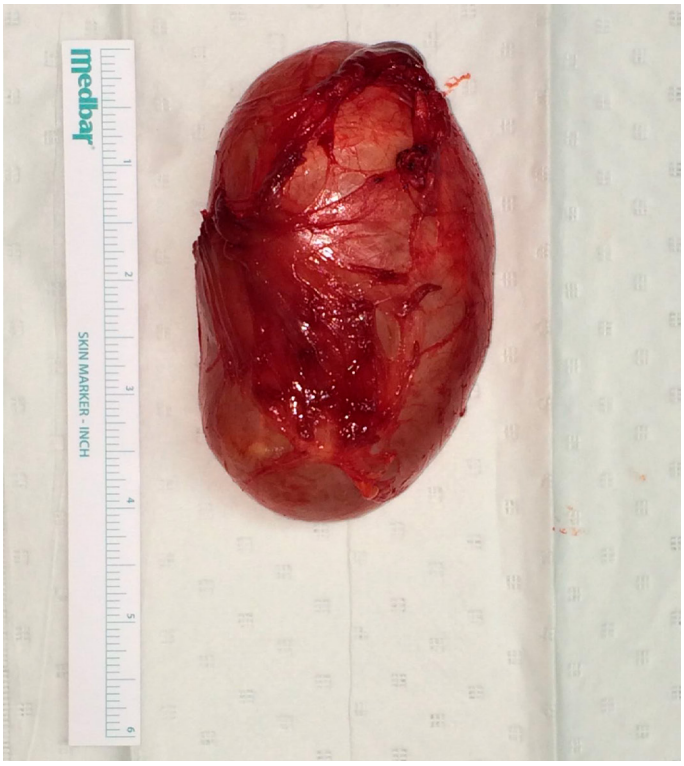

Fig. 5. Excised cystic mass.

been present on the left lateral side of the neck for 2 years and no particular pain was associated with the swelling. Computed tomography imaging (CTI) findings showed a large, homogeneous focal lesion with a size of approximately 11 x 6.5 x 8 cm, located on the left side and in the median part of the neck, extending from the left side of the body of the mandible and left mandibular angle to the superior thoracic aperture; in the lower part, the lesion extended to the prevertebral space, reaching the anterior margin of the cervical spine. The CT scan also revealed compression, deformation of the oropharynx, larynx, trachea and their displacement to the right side. At first, clinical features of the tumor associated with its size, localization and growth components were similar to a rare lateral cervical cyst with squamous cell carcinoma embedded into the cyst walls. This tumor occurrence in elderly patients and growth patterns of several years were quite similar to this rare malignant cyst transformation. A fine-needle aspiration cytology (FNAC) was later performed, which revealed clear lymphoid fluid. Clinical examination revealed a soft, painless, non-pulsatile, compressible, large cystic mass on the left side of the neck.

Considering the size of the lesion and slowly developing airway obstruction, patient was subjected for surgical procedure under general anesthesia. The resection of the mass was done under general anesthesia from vertical, slightly oblique cervical incision. During surgery, adherence of the cyst wall to adjacent structures, including left sternocleidomastoid muscle, was observed. After meticulous dissection, the lesion was resected completely intact. A vacuum drain was inserted which was removed on the second post-operative day. Post-operative period was uneventful. On the third post-operative day the patient was discharged from hospital. Histopathological evaluation described the tumor as cystic hygroma. The patient remains healthy and free of recurrence after 6 months.

DISCUSSION

The World Health Organization (WHO) recognizes three types of lymphangiomas: capillary, cavernous, and cystic. Central to any discussion of cystic hygroma is the understanding that it is synonymous with macrocystic lymphatic malformation and cystic lymphangioma [2].

Giguère et al. proposed categorizing lymphangiomas on the basis of the size of the cystic component, as follows [8, 9]:

- macrocystic – cystic spaces ≥ 2 cm; usually under the level of mylohyoid muscle which are located mainly within the anterior and posterior cervical triangle,
- microcystic – spaces < 2 cm; usually above mylohyoid muscle located mainly in the oral cavity, tongue, submandibular region and parotid,
- mixed lesions.

The microcystic form tends to predominate over macrocystic lymphatic malformation in the oral cavity and oropharynx. Microcystic lymphatic malformations commonly appear as clusters of clear, black or red vesicles on the buccal mucosa or tongue. Macrocystic lymphatic malformations tend to predominate below the mylohyoid muscle and can involve both the anterior and posterior triangles of the neck. The cysts are typically large and thick-walled and have little involvement of surrounding tissue. The overlying skin can take on a bluish hue or may appear normal [2].

Cystic hygromas often present after a sudden increase in size secondary to infection or intralesional bleeding. Spontaneous decompression or shrinkage is uncommon [2].

Potentially life-threatening airway compromise that manifests as noisy breathing (stridor) and cyanosis is a possible symptom of lymphangiomas [2].

Cystic hygromas are composed of large irregular sinuses with a single layer of flattened epithelial lining and fibrous adventitial coats. The thickness of the vessel wall varies, with both striated and smooth muscle components [2].

Microscopically, cystic hygroma is characterized by enlarged lymphatic vessels in a fibrotic and loose stromal background. It is characterized by the presence of lymphoid tissue in various formations; even lymphoid follicles can be observed. This feature can be misleading in favor of diagnosing atypical lymphoid proliferations [10].

The main histopathological differential diagnosis of cystic hygroma is cavernous hemangioma in which blood filled large cystic spaces, described to be similar to lymphangioma. The lining endothelium of cavernous hemangioma shows positivity with pancytokeratin and Factor VIII, immunohistochemically. The presence of lymphatic spaces with thin walls containing fibrous tissue, smooth muscle, and lymphoid aggregates favors the diagnosis of lymphangioma [11, 12].

Although most cystic hygromas are multicystic, a unilocular cyst is found in approximately 10% of cases. Cysts can range from 1 mm to

several centimeters in size and are filled with clear- to straw-colored fluid, which is eosinophilic and protein-rich [2].

Magnetic resonance imaging (MRI), computed tomography (CT), and ultrasonography (US) are all helpful in outlining the nature of a cystic neck mass.

MRI is the consensus study of choice, it provides the best soft-tissue detail and can delineate the relation of the lesion to underlying structures. Contrast can be used to differentiate hemangiomas from lymphangiomas.

CT carries the risk of radiation exposure, and detail is lost if the cystic hygroma is surrounded by tissue of similar attenuation. Contrast helps to enhance cyst-wall visualization and the relation to surrounding blood vessels.

US is the least invasive study. It is useful in demonstrating the relationship of LM to the surrounding structures. It can be used to detect LM in utero [2].

Treatment options for a cystic hygroma depends on the size, location and symptoms present. Due to difficult and challenging surgical procedure owing to the multicystic nature of the lesion with thin wall and proximity to vital neurovascular structures, many nonsurgical options have been advocated. Treatment options may include surgery (recommended when possible), percutaneous drainage, sclerotherapy, laser therapy, radiofrequency ablation. These different treatment options may be used in various combinations. In very rare cases, cystic hygroma will shrink or go away without treatment (spontaneous resolution) [4].

Sclerotherapy consists in the administration of sclerosing agents, such as OK-432 (an inactive strain of group *A Streptococcus* pyogenes, which produces an inflammatory reaction when applied with intracystic injection and then causes the destruction of endothelium, sclerosis, and cicatricial contraction of cyst wall), bleomycin, pure ethanol, bleomycin, sodium tetradecyl sulfate, and doxycycline [13].

An infected LM should be treated with intravenous antibiotics, and definitive surgery should be performed once the infection has resolved. Incision and drainage or aspiration results in only temporary shrinkage, and subsequent fibrosis can further complicate the resection [2, 6, 14].

As stated above, the mainstay of treatment of cystic hygromas is radical surgical excision [15].

However, it should be acknowledged that cystic hygroma is a benign lesion with no malignant transformation and if acute infection occurs prior to resection, surgery should be delayed at least 3 months [2, 5, 9, 14].

The surgical team should attempt to remove the cystic hygroma completely or, failing that, to remove as much as possible, sparing all vital neurovascular structures. Cystic hygromas are ideally removed in a single procedure because secondary excisions are complicated by fibrosis and distorted anatomic landmarks [2].

Signs of airway obstruction necessitate surgical evaluation at the time of diagnosis. In emergency situations, aspiration with a needle may prevent from performing an emergency tracheostomy.

Many authors have suggested the use of nonsurgical management in asymptomatic cases only. A study by Alqatein reported spontaneous regression is often associated with recurrence and hence is not a valid treatment for cystic hygroma and concluded that surgical excision of the lesion should be considered in all cases [16, 17, 14].

Unlike hemangiomas, cystic hygromass do not commonly resolve spontaneously.

The long-term outlook (prognosis) associated with a cystic hygroma may depend on when the cystic hygroma is detected, the size and location of the lesion, whether complications arise, and whether an underlying syndrome or chromosome abnormality is present.

Recurrence is rare when all gross disease is removed. If residual tissue is left behind, the expected recurrence rate is approximately 15% [2, 10, 14].

As a person with a cystic hygroma ages, morbidity is often related to cosmetic disfigurement and how the cystic hygroma affects other critical structures, such as nerves, blood vessels, lymphatics, and the airway. Complications from surgery may also occur, and depending on the location may include damage to neurovascular structures (including cranial nerves), chylous fistula or chylothorax (leakage of lymphatic fluid), or hemorrhage [2, 16, 14].

CONCLUSION

Cystic hygromas are very rare malformations in adults, which should nonetheless be considered in the differential diagnosis of head and neck cystic masses. Provided that the mass is completely excised, this disease has a good prognosis. In the presented case a complete resection was achieved, with specimen wall removed intact; neither post-operative complications nor recurrence has been reported on. Further studies should examine the incidence of cystic hygroma in adult patients to better explain the early diagnosis and the best treating course of the disease.

References

1. Rangel V., Karen H.: Cystic hygroma. J Diag Med Sonograp., 2008; 24(4): 218–222.
2. 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.
3. Zadvinskis D.P., Benson M.T., Kerr H.H.: Congenital malformation of the cervicothoracic lymphatic system: embryology and pathogenesis. Radiographics, 1992; 12: 1175–1189.
4. Lymphatic Malformations. NORD. Availabe from: https://rarediseases.org/rare--diseases/lymphatic-malformations. Updated 2019. Accessed September 10, 2020.

5. Kennedy T.L.: Cystic hygroma-lymphangioma: A rare and still unclear entity. Laryngoscope, 1989; 99: 1–10.
6. Sichel J.Y., Udassin R., Gozal D. et al.: OK-432 therapy for cervical lymphangioma. Laryngoscope, 2004; 114: 1805–1809.
7. Bahl S., Shah V., Anchlia S., Vyas S.: Adult-onset cystic hygroma: A case report of rare entity. Indian J Dent, 2016; 7(1): 51–54.
8. Giguère C.M., Bauman N.M., Smith R.J.: New treatment options for lymphangioma in infants and children. Ann Otol Rhinol Laryngol, 2002; 111(12): 1066–1075.
9. De Serres L.M., Sie K.C., Richardson M.A.: Lymphatic malformations of the head and neck. A proposal for staging. Arch Otolaryngol Head Neck Surg, 1995; 121: 577–582.
10. Damaskos C., Garmpis N., Manousi M. et al.: Cystic hygroma of the neck: single center experience and literature review. Eur Rev Med Pharmacol Scie, 2017; 21: 4918–4923.
11. Chand E.M., McNeely T.W., Freant J.L.: Pathologic quiz case: male with increasing abdominal girth. Arch Pathol Lab Med, 2000; 124(11): 1723–1724.
12. Derin S., Şahan M., Dere Y., Çullu N., Şahan L.: Cervical cystic hygroma in an adult. Case Rep Pathol., 2014; 2014: 209427.
13. Acevedo J.L., Shah R.K., Brietzke S.E.: Nonsurgical therapies for lymphangiomas: a systematic review. Otolaryngol Head Neck Surg., 2008; 138(4): 418–424.
14. Waner M., Teresa M.O.: Multidisciplinary Approach to the Management of Lymphatic Malformations of the Head and Neck. Otolaryngol Clin North Am, 2018; 51(1): 159–172.
15. Ozen I.O., Moralioglu S., Karabulut R. et al.: Surgical treatment of cervicofacial cystic hygromas in children. ORL J Otorhinolaryngol Relat Spec, 2005; 67(6): 331–334.
16. Alqahtani A., Nguyen L.T., Flageole H., Shaw K., Laberge J.M.: 25 years' experience with lymphangiomas in children. J Pediatr Surg, 1999; 34: 1164–1168.
17. Bhatia M., Mattoo A.: Cystic hygroma: A surgical challenge. Med J DY Patil Univ, 2016; 9: 758–760.

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