

Cystic hygroma of the neck: single center experience and literature review

C. DAMASKOS^{1,2}, N. GARMPI¹, M. MANOUSHI³, A. GARMPI⁴, G.-A. MARGONIS⁵, E. SPARTALIS², C. DOULA⁶, C. MICHAEL-STANTZIA⁷, N. PATELIS⁸, D. SCHIZAS⁹, A.-T. PAPACHRISTOU¹⁰, N. ANDREATOS⁵, G. TSOUROUFLIS¹, N. ZAVRAS¹¹, K. MARKATOS¹², K. KONTZOGLU¹, E.A. ANTONIOU¹

¹Second Department of Propedeutic Surgery, Laiko General Hospital, National and Kapodistrian University of Athens, Medical School, Athens, Greece

²N.S. Christeas Laboratory of Experimental Surgery and Surgical Research, Medical School, National and Kapodistrian University of Athens, Athens, Greece

³First Pediatric Surgery Department, Aglaia Kyriakou Children's Hospital, Athens, Greece

⁴Internal Medicine Department, Laiko General Hospital, University of Athens Medical School, Athens, Greece

⁵Department of Surgery, The Johns Hopkins University School of Medicine, Baltimore, Maryland, USA

⁶Plastic Surgery Department, Nicosia General Hospital, Cyprus

⁷Department of Pathology, Aglaia Kyriakou Children's Hospital, Athens, Greece

⁸Vascular Unit, First Department of Surgery, Laiko General Hospital, Medical School, National and Kapodistrian University of Athens, Athens, Greece

⁹First Department of Surgery, Laiko General Hospital, Medical School, National and Kapodistrian University of Athens, Athens, Greece

¹⁰Medical School, National and Kapodistrian University of Athens, Athens, Greece

¹¹Third Department of Surgery, Attikon University Hospital, National and Kapodistrian University of Athens, Medical School, Athens, Greece

¹²Biomedical Researches Foundation of the Academy of Athens, Athens Greece.

Christos Damaskos and Nikolaos Garmpis equally contributed to this work

Abstract. – OBJECTIVE: Malformations of the lymphatic system are recognized as benign congenital tumors that affect infant and children in the perinatal era. In children, these abnormalities usually found in the neck and the axillary region, but they can present in other parts of the body such as mediastinum, pelvis, retroperitoneum as well as in solid organs (e.g., adrenal glands, pancreas, stomach). Our aim is to report our experience on cystic hygromas via two cases and review the literature.

MATERIALS AND METHODS: Herein we present two cases of cystic hygroma, the first of female children and the second of a female adult patient respectively. Both of these patients underwent surgical excision of the masses.

RESULTS: After the procedure, both patients have recovered well, and no recurrence of the lesion has been noted during the follow-up period.

CONCLUSIONS: Surgical treatment remains the gold-standard treatment for these tumors, while other modalities have been used with mixed results.

Key Words

Cystic, Hygroma, Lymphangioma, Lymphatic, Tumor.

Introduction

One of the most commonly presented lymphatic malformation is cystic lymphangioma or cystic hygroma (CH). Lymphangiomas are divided in three groups; lymphangioma simplex, cavernous lymphangioma, and CH¹. CH is a misnomer for a benign lesion that appears as an enlarged mass of the neck and clavicle. CH originates in embryonal life, and it is leading to a failure of communication and lymph drainage into the venous system².

CH concerns mainly the pediatric surgeons because the incidence of this particular pathology is considerably higher among children. Most commonly it is presented in the neck, clavicle and axillary areas³. However, there is evidence of late manifesta-

tions in early adulthood and adult patients, especially as far as the other than neck and head forms are concerned. It can occur in the mediastinum, in the abdomen, in the pelvis as well as in solid abdominal organs such as the adrenal glands and the pancreas⁴.

The management of CHs is mainly surgical⁵ in order to excise the enlarging lesion, relieve the patients' symptoms and exclude other pathologies. Investigation of the protruding mass includes laboratory examination, imaging studies; fine needle aspiration biopsy (FNAB) is a feasible diagnostic tool. The imaging modalities used are ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) scans to further appraise the imaging results. Herein, we present two cases of CHs, successfully treated by surgical excision. All patients' data with CHs characteristics and procedures details are presented in Table I. A brief literature review is discussed.

Case Presentation

Case 1

The first case was about a lovely girl aged one and a half (1.5) years old. She was referred to our Department due to an asymptomatic lesion of the left side of the neck that seemed to enlarge over the past two months. Her previous medical history was free, with no perinatal problems. The lesion measured 7 cm in maximum diameter and was located to the left of the cervical region. Clinical examination revealed a soft, painless, non-pulsatile and transilluminable mass. Clinical suspicion of lymphatic malformation was initially sustained by the US; then, the patient underwent a MRI scan to better examine the nature of the lesion and to identify its anatomic correlation with the surrounded tissues (Figure 1A, 1B). The resection of the mass

was done under general anesthesia from transverse cervical incision. The lesion was located under the level of the hyoid bone, seated upon the left sternocleidomastoid muscle and in deep relation to the left jugular vein and carotid artery. After meticulous dissection, the lesion was resected completely intact. A vacuum drain was inserted which was removed on the second post-operative day and the patient discharged home. On gross pathology the mass had a macrocystic appearance and filled with bloody-colored serous fluid. The lesion was classified as Stage I CH of the cervical, according to Serres et al⁶ system recommended, both by MRI imaging and pathology report⁶. The patient is under close follow up thereafter showing no evidence of recurrence 18 months post-operatively.

Case 2

A 53 years old female was referred to our Department, because of a large mass located to her right cervical side (Figure 2A), and complaining for tension and numbness. The mass was first perceived four years earlier and was progressively enlarging during the last month. It was thought to be a benign lipoma and thus no further investigation was performed. Her medical history was free other than tobacco smoking of 20 packs/year for the past 20 years. Clinical examination revealed a palpable mass, agile from the surrounding tissues to the touch mass measuring 10 cm. Laboratory examinations were within normal limits. Following these, the patient underwent cervical US which revealed a hypoechoic lesion of 9.5 cm in its maximum dimension was revealed. A FNAB of the lesion; was not diagnostic. A CT scan disclosed a multilobular cystic lesion 15x10 cm, attached to the carotid bundle, compatible with a cystic formation of the cervical. A CT Angiography (Fig-

Table I. Clinical and pathologic features of the reported cases of cystic hygroma.

CH N=2	Age/ Sex	Size (cm)	Site	Imaging	Operation	Macroscopy	Histology	Follow-up
<i>Pt 1</i>	1,5 y / f	7	Neck-Lef	US/MRI	Resection from transverse cervical incision	Macrocystic appearance and filled with bloody-colored serous fluid	Cystic walls containing fibrous tissue surrounded by lymphoid cells; and fat follicles	18 m
<i>Pt 2</i>	53 y / f	15	Cervical side-Right	US/CT/DSA	Resection from right cervical incision	Multilobular mass	Cystic walls containing fibrous tissue surrounded by lymphoid cells; and fat follicles	27 m

Abbreviations: CH: cystic hygroma; Pt: patient; y: years; f: female; US: ultrasound; MRI: magnetic resonance imaging; CT: computed tomography; DSA: digital subtraction angiography; m: months.

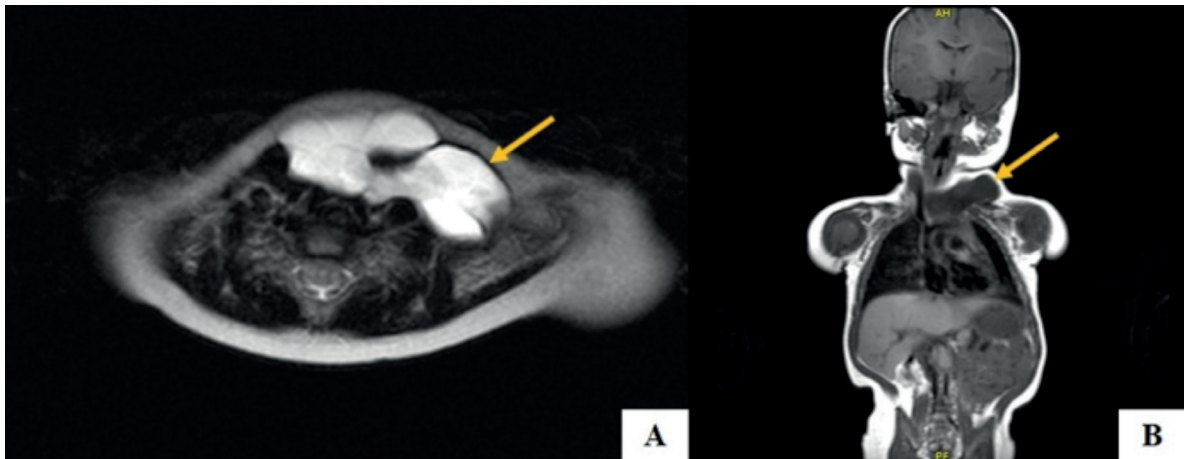


Figure 1. A-B, Magnetic Resonance Imaging (MRI) describing a 7cm lesion located to the left of the cervical region (Case 1).

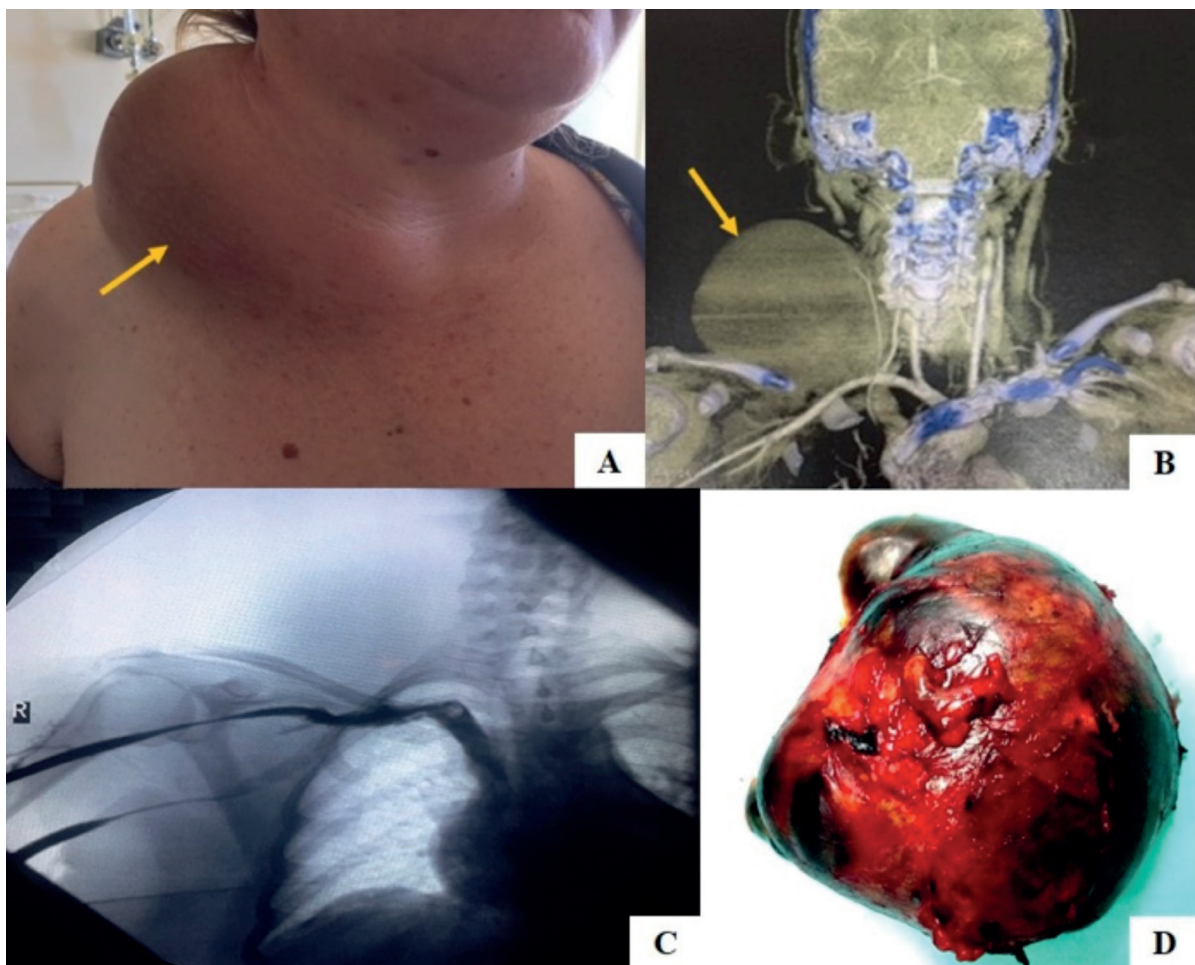


Figure 1. A, Macroscopic view of the lesion of cystic hygroma of the neck region; **B,** CT Angiography – 3D reconstruction; **C,** Digital Subtraction Angiography (DSA) excluding the possibility of communication with the major cervical vessels; **D:** Macroscopic view of the resected surgical specimen removed with its bowel (Case 2).

ure 2B) and a Digital Subtraction Angiography (DSA) scan (Figure 2C) excluded communication with the major cervical vessels. Because of the location of the mass, and the underlying symptoms of the patient, she was planned for a surgical excision of the cervical mass. She underwent a right cervical incision and the mass was removed with its capsule intact (Figure 2D). Pathology reported a multilobular mass, 15x15x10 cm long that was proved to be a CH of the cervical (Figure 3). Post-operative course was uneventful and she was discharged 5 days later. She remains healthy and free of recurrence after 27 months.

Discussion

The majority (45-52%) of lymphatic malformations occur in the lymphatic “rich” regions of head and neck. Nevertheless, they can also present in axillae, mediastinum, groin and retroperitoneum⁷. Worth

mentioning are the lymphatic malformations of the orbit⁸ due to the endangered vision as well as the so-called “giant” lymphatic malformations. The latter involve the tongue, base of the oral cavity, cervical region and mediastinum neighboring with vital structures and necessitating in many instances an emergency tracheostomy, staged repair, tube feeding, long term speech therapy, etc.⁹.

Lymphatic malformations are classified as: a. macrocystic, usually under the level of mylohyoid muscle which are located mainly within the anterior and posterior cervical triangle; b. microcystic, usually above mylohyoid muscle located mainly in the oral cavity, tongue, submandibular region and parotid and c. mixed type⁶.

Lymphatic malformations typically manifest clinically by age 2 years old when parents are worried and seek for a pediatric surgeon consultation. Concerns refer mainly to cosmetic reasons. Parents should be advised that these lesions do not regress spontaneously but they will grow up with the child’s size.

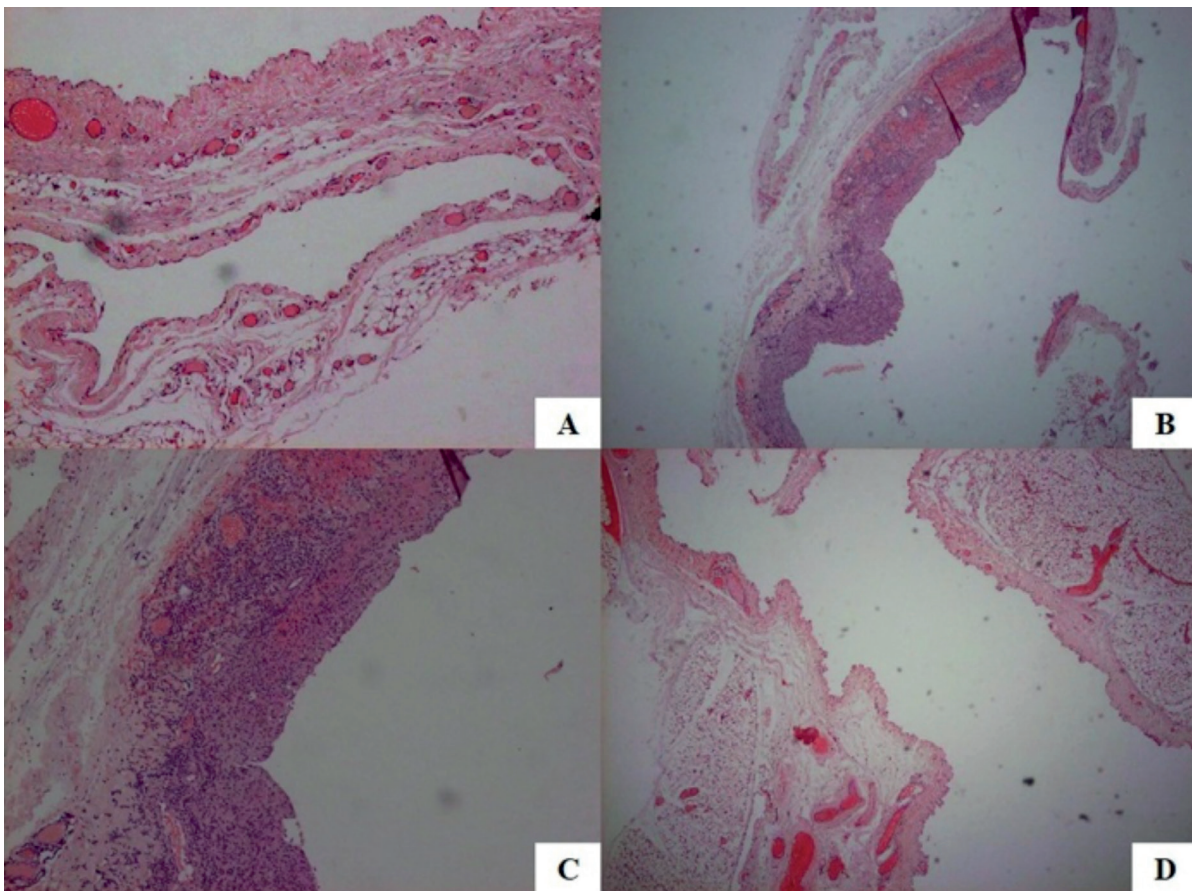


Figure 3. **A**, Cystic walls containing fibrous tissue (H&E, x100); **B**, Fibrous tissue surrounded by lymphoid cells (H&E, x40); **C**, Enlarged lymphatic vessels surrounded by lymphoid tissue (H&E, x200); **D**, Fibrous tissue and fat cells (fat follicles) containing the cystic wall (H&E, x40). (Case 2).

As far as the clinical picture, it varies from life threatening (non-patent airway), serious impact on organ function (orbit – loss of vision), to asymptomatic. Symptoms may worsen when the lesions are complicated with bleeding or infection¹⁰.

The baseline radiologic investigation is US of the lesion in addition to a plain chest x-ray which will rule out extension into the mediastinum. US can differentiate between cystic and solid tumors. The anatomic details offered by the MRI, a radiation free, child friendly tool facilitate preoperative and intraoperative decision-making and planning¹¹. Biopsy or diagnostic needle aspiration do not apply in lymphatic malformations diagnostic work up.

The therapeutic approach of these lesions was for years undoubtedly and without exception surgical. The high occurrence of recurrence as well as technical difficulties in resection due to close proximity to vital structures and poorly demarcated margins of the lesions¹² encouraged efforts to find and apply alternative methods of treatment. There is no consensus regarding lymphatic malformation treatment and no accepted guidelines exist to date¹³. It depends on the expertise of the Center and parents' preference. Needless to say that there are factors which need to be considered before any intervention and individualized care is the most appropriate approach. Among the factors are: symptomatic disease or not, size of the lesion and location (superficial or deep) as well as the type of lesion (macro- or micro- cystic)¹⁴.

Briefly noted beneath are the names of alternative methods applied such as radiofrequency ablation (RF) ablation¹⁵, CO₂ laser while a more extensive reference regarding sclerotherapy follows¹⁶. However, fistula formation or infection and local recurrence have been reported, so the results of these therapies remains still to be examined¹⁷.

Percutaneous treatment of lymphatic malformations is an established alternative technique which improves itself and gains acceptance among the efficient treatment methods under specific indications. The principle of percutaneous sclerotherapy is based on the chemical reaction produced by specific substances upon the endothelium of the lesion vessels. The inflammation process leads to thrombosis, obstruction of the lumen, scar formation and downsizing of the lesion. The approved substances used are OK-432¹⁸ produced by *Streptococcus sp.*, Bleomycin¹⁹ and Doxycyclin²⁰. The latter mandates sedation due to the amount of discomfort and pain caused. In all cases the cyst liquid should be aspirated before instilling the sclerosant. One of the most serious

side effects is allergic reaction. The method of percutaneous sclerotherapy has proved to be effective in macrocystic disease. Studies suggest that probably is equally effective to surgery for macrocystic lesions of the head and neck²¹.

Microscopically, CH 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. Cavernous hemangioma is the entity with similar histopathological findings complicates the differential diagnosis of CH. CH consists of thin walls containing fibrous tissue, smooth muscle and lymphoid tissue and lined with epithelium; guiding to the diagnosis of its particular pathology²².

In summary it can be claimed that surgical therapy remains to date the mainstay of treatment²³. Appropriate good knowledge of the anatomy, and experience may result in successful and cosmetic results. For emergency indications moreover, operation is the only indicated intervention. Nevertheless, given the disappointing rates of recurrence even in expert hands (15-53%), it is worthwhile considering the alternative techniques as an intraoperative adjunct²⁴. There are still many studies to be designed and conducted in order to conclude upon the best option for lymphangioma management.

Conclusions

Lymphatic malformations are a common cause of benign nature masses found at the neck and clavicle area of infants and children of perinatal age. CHs are lymphatic malformations most often observed in children; few cases of adult-onset of the disease have been described in the English literature. Provided that the mass is completely excised, an excellent prognosis is of great possibility. In both of our cases a complete resections was achieved, with specimen wall removed intact; no post-operative complications nor recurrence has been occurred. Further studies should examine the incidence of CH in adult patients to better elucidate the early diagnosis and the best treating course of the disease.

Conflict of Interest

There is no conflict of interest among the authors of this paper.

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