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Giant Cervico-Thoraco-Brachial Cystic Lymphatic Malformation in an Infant: Therapeutic Strategy

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Abstract
Cystic Lymphatic Malformations (CLMs) are rare benign congenital vascular malformations characterized by slow flow and composed of abnormal cystic dilations. The most widely used classification divides them into microcystic, macrocystic, and mixed lesions. Ultrasound is the preferred initial evaluation tool for CLMs due to its non-irradiating nature and accessibility. MRI is used to classify CLMs before considering therapeutic management. Biopsy and fine needle aspiration are rarely needed to diagnose CLMs. The progression of CLMs is marked by asymptomatic periods interspersed with painful inflammatory flare-ups. Surgical treatment remains the gold standard for managing these malformations. This paper reports our experience in treating a case of giant cervico-thoraco-brachial cystic lymphatic malformation in a 4-month-old infant.

Keywords: Cystic lymphatic malformation; Infant; Cervico- thoraco-brachial; Giant; surgical resection

Introduction

Cystic Lymphatic Malformations (CLMs) are rare benign congenital vascular malformations characterized by slow flow and composed of abnormal cystic dilations. The term "cystic lymphangioma" has been abandoned [1,2,3].

According to the International Society for the Study of Vascular Anomalies (ISSVA), vascular anomalies are divided into vascular tumors and vascular malformations [4]. CLMs are classified as vascular malformations and not tumors. They fall under the category of hamartomas, along with neurofibromas and hemangiomas. This rare congenital malformation was first described in 1828 by Redenbacher and has been better understood since the reference works by Sabin in 1909 and 1912 [5]. It is considered a sequestration of lymphatic tissue that retains its growth potential.

In addition to superficial malformations, there can be deeper malformations. The most widely used classification divides them into microcystic, macrocystic, and mixed lesions. CLMs are slow-flow congenital vascular malformations that can be symptomatic from the prenatal period or later in life, most often in childhood. The distinction between macrocystic, microcystic, and mixed lymphatic malformations determines the prognosis and therapeutic approach [6].

Their diagnosis is suggested by the presence of a cervico- thoracic mass. Medical imaging can suggest the diagnosis, but only histopathological examination confirms it. The main complications are:

- Inflammatory flare-ups
- Intracystic bleeding
- Compressive manifestations, especially in the cervicofacial region (compression of the upper airways, which can be life- threatening)
- Oozing; rupture with risk of superinfection

The management of cystic lymphangiomas involves various therapies, including surgery, percutaneous sclerotherapy, and other therapeutic means depending on the type of lesions, their location, extent, and potential for progression.

Patient and observation

We report a case of a giant cervico-thoraco-brachial cystic lymphatic malformation. The patient is a 4-month-old infant girl presenting with a congenital cervico-thoraco-brachial mass diagnosed prenatally (by

prenatal ultrasound at 22 weeks of gestation) with progressive volume increase causing respiratory distress. The infant was hospitalized in the pediatric department due to respiratory distress, and given the lack of improvement with medical and positional treatment, the pediatric team urgently requested our pediatric surgery team to intervene.

On physical examination, a giant bilobed formation was observed. The first cervical mass was located on the left lateral side and at the base of the neck, measuring 25 cm longitudinally and 14 cm transversely, occupying the left supraclavicular fossa. The mass was painless, regular, renitent, mobile, and non- pulsatile **(Figure 1)**. The second enormous soft cystic thoraco- brachial mass filled the left axilla, extending beyond the thorax and elbow, measuring 23 cm by 25 cm **(Figure 2)**. There were no sensory or motor disturbances in the ipsilateral upper limb. An of Anatomical Science and Research

ultrasound showed a multicystic anechoic mass. A standard radiograph (of the neck coupled with a chest x-ray) showed a rounded opacity in the left cervical region with regular external margins, signs of compression of adjacent organs (tracheal deviation), and another large axillary opacity **(Figures 3)**.

Figure 1: The cervical mass located on the left lateral side and of the neck.

Figure 2: The cystic thoraco-brachial mass filled the left axilla.

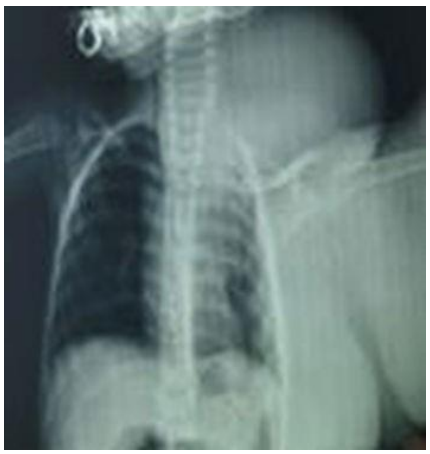

Figure 3: A standard radiograph (of the neck coupled with a chest x-ray) showed a rounded opacity in the left cervical region.

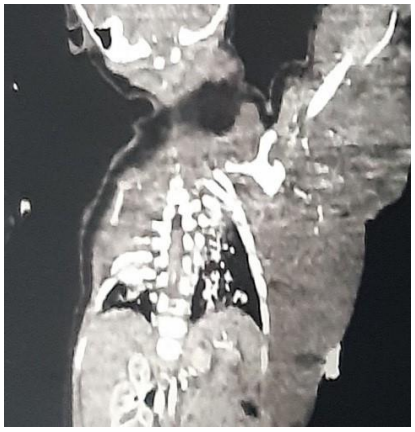

Figure 4: An MRI confirming the diagnosis of cystic lymphangioma.

Based on these findings, a diagnosis of cervico-thoraco- brachial cystic lymphatic malformation was considered. An MRI confirmed the diagnosis (Figure 4). Surgical intervention was indicated to resect the cervical mass responsible for respiratory distress. Intraoperatively, a fluid- filled mass was found with no other vascular malformations. The cervical mass was excised without incident, and the postoperative course was uneventful. The mass was sent to the laboratory for histopathological examination, which concluded it was a cystic lymphangioma. The walls consisted of fibromuscular tissue with many lymphoid follicles with clear centers, and the lymphatic channels were dilated with mixed lymph and blood content.

Discussion

Cystic Lymphatic Malformations (CLMs) are congenital malformations typically discovered in children. Histologically, they mimic the structure of lymphatic vessels, with walls composed of collagen fibers and some smooth muscle fibers. Nearly 75% are located in the head and neck, with an incidence estimated between 1.2 to 2.8 per 1000 births [7,8], and are less commonly found in the axillary, thoracic, or abdominal walls [9]. The etiology of CLMs remains poorly understood. Ultrasound is the preferred initial examination for CLMs due to its non- irradiating nature and accessibility. Before considering therapeutic management, MRI is used to differentiate between macrocystic CLMs (which can be aspirated and potentially treated with sclerotherapy) and microcystic CLMs [10]. MRI also determines the extent of the malformation, especially in mediastinal locations [11], as seen in our case. For follow-up, especially in the pediatric population, ultrasound is the primary modality, with MRI used as needed [6]. MRI requires sedation in young children (< 6 years), as movements during the scan can compromise image quality.

Biological tests are not useful for diagnosing isolated cystic lymphatic malformations but are useful for detecting complications or pre-therapeutic evaluations.

A biopsy is rarely necessary to diagnose CLMs, which are typically diagnosed based on clinical and radiological findings. However, in doubtful cases (particularly some microcystic or deep CLMs), a biopsy should be performed [6]. Large surgical biopsies can result in prolonged lymphatic leakage at the incision site [6].

Fine-needle aspiration is rarely used diagnostically but can be performed before sclerotherapy, revealing a citrine fluid rich in lymphocytes, sometimes slightly bloody.

The progression of CLMs is marked by asymptomatic periods interrupted by episodes of painful inflammatory flare-ups, superinfections, or intralesional hemorrhages, with variable frequency. These flare-ups often cause significant and sudden increases in CLM volume, although temporary. Their duration varies (from 48 hours to 1 month), averaging about ten days. They can occur following trauma or infectious episodes, particularly viral ones [12]. The macrocystic and microcystic components evolve differently over time:

Macrocystic lymphatic malformations can regress after inflammatory flare-ups, particularly following nearby infections or intralesional hemorrhages, leading to inflammatory remodeling and fibrosis, potentially resulting in spontaneous regression ("natural sclerosis").

Microcystic components tend to progress over time, becoming thicker and more bothersome.

Data on the spontaneous involution of macrocystic CLMs are as follows:

Phang et al., found that patients with spontaneous remission of their CLMs more often had congenital

cervical CLMs, with remission occurring approximately 24 months after the CLM's appearance [6].

Perkins et al., [13], in a study of 104 patients with cervicofacial lesions followed for one year, found a spontaneous regression rate of 12.5%, with regression occurring between 2 and 7 months.

Sclerotherapy (interventional radiology) is the first-line treatment for most lymphatic malformations (14). It is more effective for macrocystic [15] CLMs than for microcystic [16] ones. Sclerotherapy was not indicated due to:

The unavailability of sclerosing agents.

The need for a more potent sclerosing agent (e.g., alcohol) for larger malformations, considering the intrinsic toxicity of each sclerosing agent, which can cause local and systemic complications. Cardiac arrests have been reported with absolute alcohol.

A 2014 retrospective study on 174 patients with cervicofacial CLMs did not show the superiority of sclerotherapy over surgery, but numerous biases limit the study's interpretation [17].

Partial resection surgery ("modeling" surgery) is indicated to reduce the volume of deforming and bothersome lesions and can be repeated over time.

The treatment of cervical cystic lymphangiomas is primarily surgical, and definitive diagnosis is made through histopathological examination [18,19]. In our case, we performed a left lateral transverse cervicotomy, allowing us to resect the tumor entirely in its compressive cervical portion on the upper airways **(Figure 5A,5B,5C)**. The postoperative course was uncomplicated, without vascular or nerve sequelae, and the patient showed no recurrence at 6 months **(Figure 5D,5E)**, 1 year, and 18 months postoperatively. The surgery's difficulty lay in:

The infant's small size and short neck.

The enormous volume and poorly defined cystic formation.

A history of two inflammatory flare-ups making dissection difficult.

Lack of clear cleavage planes with many adhesions to vascular and nerve structures, making total tumor resection laborious.

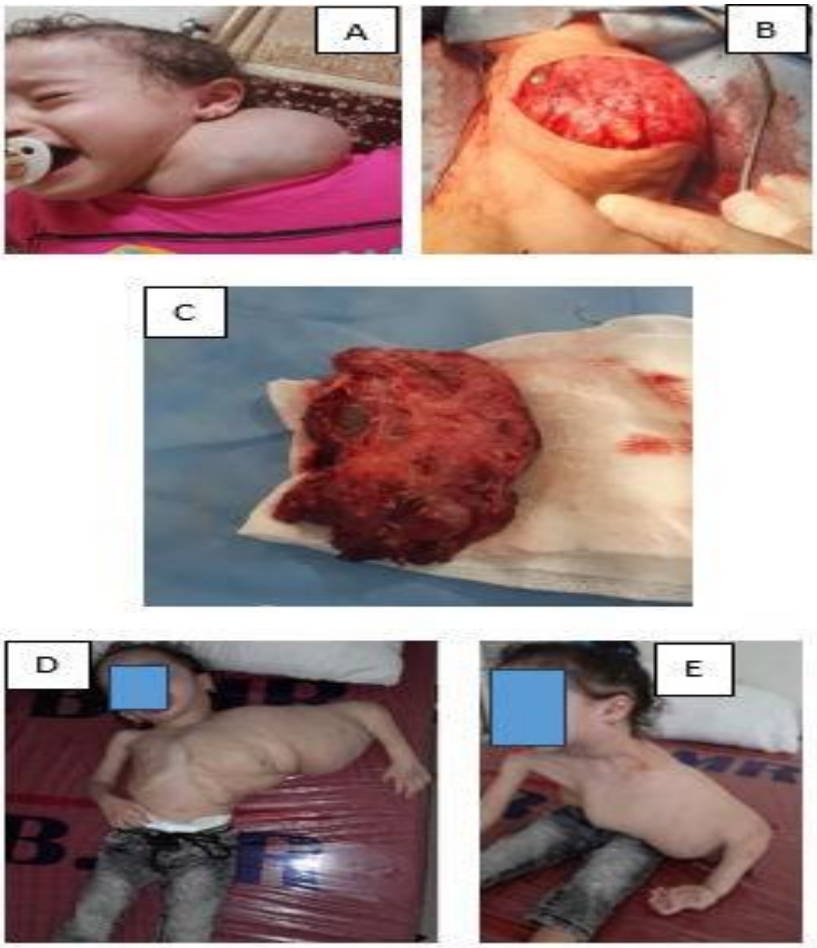

Figure 5: A) The cervical mass located on the left lateral side of the neck; B) left lateral transverse cervicotomy; C) resection of the tumor entirely in its compressive cervical portion on the upper airways; D,E) the patient post-operatively (after 6 months).

Histopathological examination concluded it was a cystic lymphangioma. Various classifications exist in the literature. Some authors prefer to classify lymphangiomas as capillary, cavernous, and cystic. However, the most widely used classification currently divides CLMs based on cystic formation size: macrocystic (greater than 1 cm) and microcystic (less than 1 cm) [20], or mixed (comprising small and large cysts). There is no uniform consensus on the definition of macrocystic and microcystic [10] lymphatic malformations. Cervical and axillary CLMs are more often macrocystic or mixed [4]. Macrocystic lymphatic malformations present as subcutaneous, round, or lobulated masses, usually several centimeters in diameter, under normal-colored skin, with a firm and elastic, or soft and depressible consistency. The quality of life of affected individuals and their families can be significantly impacted [1,4].

The treatment is essentially surgical, allowing for the complete removal of the tumor, which is essential for a full recovery. Other therapeutic methods have been tried without success, such as radiotherapy, drainage through mediastinoscopy, and chemical sclerosis with intravenous cyclophosphamide. These are primarily reserved for tumors that are not resectable due to their size, location, or the general condition of the patient.

Date of Intervention? for the Giant Brachial Lymphangioma

The patient is currently 22 months old. We postponed the surgery until this age, considering the possibility of regression. The decision to operate was made based on the following factors **(Figure 6F, 6G, 6H)**:

- Combatting pulmonary atelectasis and promoting the expansion of the left lung
- Preventing complications, especially inflammatory flare-ups
- Addressing aesthetic concerns and parental pressure
- Freeing the upper limb (from the axilla to the elbow) and restoring its mobility
- The postoperative course was uncomplicated, with no sensory-motor sequelae.

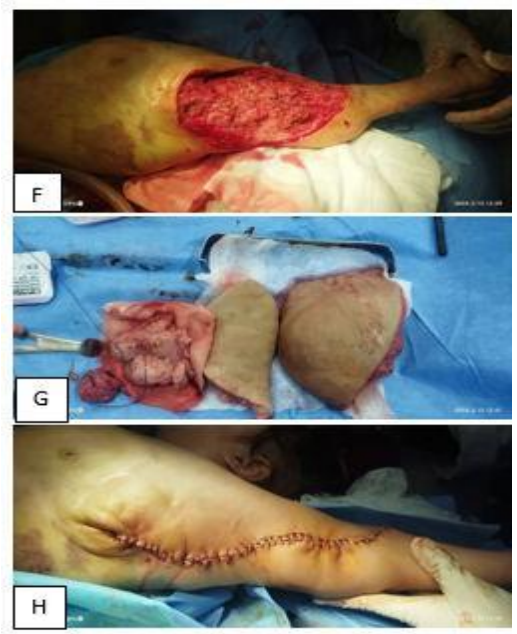

Figure 6: F) longitudinal incision taking the excess skin and centered on the mass; G) The resected mass with excess skin;H) The appearance of the arm after resection of the mass

Conclusion

Cervicothoracic-brachial cystic lymphangiomas are rare. They typically present as a giant mass. Ultrasound and MRI are the two imaging modalities indicated to confirm the diagnosis and assess the extent of a cystic lymphatic malformation (evaluating the intra-thoracic, mediastinal extension and its relationship with vascular structures). The definitive diagnosis is based on histology. Surgical treatment remains the gold standard in managing these CLMs, along with sclerotherapy. However, this treatment can be challenging in cases of extensive lymphangioma ramifications, with a risk of residuals and, therefore, recurrence.

Conflicts of Interest

The authors declare no conflict of interest.

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