Original Article

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Role of surgical excision in the treatment of large macrocystic lymphatic malformations in children

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ABSTRACT

Background: Lymphatic malformations are rare benign congenital cystic lesions that can develop anywhere in the body. Therapeutic options for these lesions include intralesional sclerotherapy, surgical excision, and other modalities. **Aim**: This study aims to assess the safety and efficacy of complete surgical excision as a therapeutic option in the treatment of large macrocystic lymphatic malformations in children, as well as to introduce a novel modification to the surgical procedure. **Methods**: A prospective study was conducted on 14 patients with large macrocystic lymphatic malformations in different body locations, treated between October 2018 and October 2021 at our institution. Complete surgical excision using a continuous traction technique was the principle therapeutic approach, with no alternative modalities applied. Patients were followed up to assess postoperative cosmetic and functional outcomes, as well as recurrence. **Results**: Most enrolled patients, comprising 12 (85.7%), presented before the age of 2 years, with eight (57.1%) having lesions in the neck. Clinical diagnosis was possible in 11 (78.6%) cases. Complete surgical excision was successfully achieved in 12 (85.7%) patients, with no significant injury to vital structures. No postoperative functional problems were reported, and esthetic results were satisfactory in the vast majority (92.8%) of patients. Recurrence was not reported in any of the patients enrolled. **Conclusion**: Complete surgical excision proves to be a safe and effective initial therapeutic modality in the management of large macrocystic lymphatic malformations in children, yielding excellent postoperative results in terms of functional outcomes and recurrence. Utilizing a continuous traction technique during surgical excision has been instrumental in achieving these favorable outcomes.

Keywords: children, lymphatic malformations, surgical excision

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INTRODUCTION

Lymphatic malformations (LMs), previously known as cystic hygromas and lymphangiomas, are rare benign congenital cystic lesions that result from localized disordered embryologic development of the lymphatic vessels in the soft tissues.[1,2] These malformations can manifest anywhere in the body, with the head and neck being the most commonly affected anatomical regions (75%), followed by the axilla and mediastinum.[3,4] LMs are broadly classified into the macrocystic type, which consists of cysts greater than 1 cm in diameter, and the microcystic type, which consists of cysts smaller than 1 cm in diameter. However, the two types can coexist in a

mixed variety.[5] These lesions can be solitary or multifocal, and present a wide spectrum of clinical presentations depending on their size and anatomical location.[1] Typically, LMs present as asymptomatic, soft, partially compressible swellings in the affected anatomical regions, with parental esthetic concern being the primary consideration. They usually progress and expand slowly over time. Sudden enlargement may be the result of infection or intracystic hemorrhage, leading to respiratory distress, swallowing difficulties, and speech problems.[1,6,7] Approximately 90% of LMs are diagnosed by history and physical examination.[6] Ultrasonography (U/S), computed tomography (CT), and magnetic resonance imaging (MRI) are commonly used in assessing the size and extent of these lesions.

Therapeutic options for these lesions include intralesional sclerotherapy, surgical excision, or a combination of both. Historically, surgical excision has been the first-line therapy for LMs and is still favored by many surgeons.[1,3] Total surgical resection offers the advantage of a complete removal of the disease and its coexisting morbidity in a single stage, with low recurrence rates. However, surgical excision can be a tedious and challenging procedure and may be associated with a risk of neurovascular injury, especially for infiltrative lesions involving the cervicofacial regions, for which intralesional sclerotherapy has emerged as an alternative or adjuvant therapeutic modality yielding encouraging and excellent results. Various other therapeutic modalities, including radiation, laser excision, radiofrequency ablation, and embolization, have been attempted for the management of pediatric LMs, returning variable results. Maximal surgical excision that preserves important neurovascular structures and cosmesis is considered the treatment of choice for large macrocystic lymphatic malformations by many authors.[2,8]

This study was designed to assess the safety and efficacy of surgical excision as an initial therapeutic option in the treatment of large macrocystic LMs in children.

MATERIALS AND METHODS

A prospective descriptive study was conducted on 14 children with LMs who were managed in the Pediatric Surgery Department of the Basrah Children's Specialty Teaching Hospital from October 2018 to October 2021. Inclusion criteria consisted of patients who were clinically and radiologically diagnosed with large focal macrocystic LMs in different body locations. Large is the term used to describe lesions equal to or greater than 5 cm in diameter, while focal is used to describe LMs that develop in a single body location. Additionally, macrocystic refers to an LM that is greater than 1 cm in diameter. Patients with small, multifocal, and predominantly microcystic lesions were excluded from the study.

Clinical data, including gender, age at presentation, lesion site and size, mode of presentation, diagnostic approach, and perioperative complications, were reported. The diagnosis was confirmed in all patients through history and physical examination, U/S and MRI, and histopathology.

This study's principal objective was complete surgical excision, with a specific focus on avoiding injury to vital neurovascular structures using the continuous traction technique. This approach was attempted as a therapeutic option in all patients. No other alternative or adjuvant therapeutic modality was employed.

Patients underwent a period of follow up for 16 months, with two-month interval visits to assess postoperative recurrence as well as cosmetic and functional outcomes. Cosmetic results were assessed based on a family questionnaire regarding esthetic satisfaction, whereas functional outcomes were evaluated based on the presence or absence of issues with breathing, phonation, and swallowing.

RESULTS

The study enrolled 14 patients, of whom eight (57.1%) were males and six (42.9%) patients were females, accounting for a male to female ratio of 4:3. The age at presentation ranged from 10 months to 8 years with a median of 12.5 months. Twelve (85.7%) patients presented before the age of 2 years, while two (14.3%) presented after the age of 2 years.

Among the patients, eight (57.1%) had lesions in the neck, with two having bilateral neck involvement. Two (14.3%) patients had their LMs in the axilla, two in the chest wall, one in the upper back, and one in the gluteal region.

The majority (78.6%) of the patients presented with asymptomatic mass lesions, with special parental concerns regarding future cosmetic and functional outcomes. Two (14.3%) patients reported a sudden increase in lesion size owing to infection, confirmed by symptoms such as redness and tenderness of the overlying skin, fever, and elevated white blood cell count. Respiratory distress was evident in only one

(7.1%) patient with a large bilateral cervical lesion causing compression of the upper airway. The demographic and clinical characteristics of patients are summarized in Table1.

The clinical diagnosis of LM depending on history and physical examination was possible in 11 (78.6%) patients and was confirmed by U/S and MRI. MRI was necessary to achieve the diagnosis in two patients, one with a lesion involving the left chest wall and another with a lesion involving the left upper back. The diagnosis was only possible through histopathology in one patient with LM involving the right gluteal region.

Complete surgical excision with the lesion's capsule intact was achieved in 12 (85.7%) patients (Fig. 1). Two patients had minimal residual disease: one with bilateral cervical LM and the other with a lesion involving the left axilla.

Fortunately, no significant injury to neurovascular structures was reported during surgical excision. Wound healing was good in 12 (85.7%) patients, although partial wound dehiscence occurred in one patient and wound infection in another.

All patients underwent a period of postoperative follow up that ranged from 8 to 16 months, with an average of 12.07 months. None of the patients with cervical lesions had reported problems regarding breathing, phonation, and swallowing. Patients with LMs in other body locations did not experience any functional problems. Esthetic results were satisfactory in the vast majority (92.8%) of patients. There were no recurrences in patients with complete lesion excision, and no regrowth of residual lesions was observed in patients with incomplete lesion excision during follow up.

Table 1: Demographic and clinical characteristics of patients.

No.	Sex	Age	Lesion's site	Lesion's size	Mode of presentation	Excision
1	Female	10 months	Neck (right side)	9 x 7 cm	Asymptomatic cyst	Total
2	Male	10 months	Chest wall (left side)	11 x 8 cm	Asymptomatic cyst	Total
3	Female	11 months	Neck (right side)	10 x 8 cm	Asymptomatic cyst	Total
4	Male	11 months	Neck (right side)	8 x 6 cm	Asymptomatic cyst	Total
5	Female	12 months	Axilla (left side)	10 x 7 cm	Infected cyst	Near total
6	Male	12 months	Neck (left side)	9 x 6 cm	Asymptomatic cyst	Total
7	Male	12 months	Neck (bilateral)	10 x 8 cm	Asymptomatic cyst	Total
8	Male	13 months	Upper back (left side)	6 x 5 cm	Asymptomatic cyst	Total
9	Female	13 months	Neck (right side)	8 x 6 cm	Asymptomatic cyst	Total
10	Female	14 months	Gluteal region (right side)	5 x 5 cm	Asymptomatic cyst	Total
11	Male	16 months	Axilla (right side)	9 x 7 cm	Asymptomatic cyst	Total
12	Female	18 months	Neck (bilateral)	12 x 8 cm	Respiratory distress	Near total
13	Male	2.5 years	Chest wall (left side)	12 x 9 cm	Infected cyst	Total
14	Male	8 years	Neck (left side)	10 x 6 cm	Asymptomatic cyst	Total

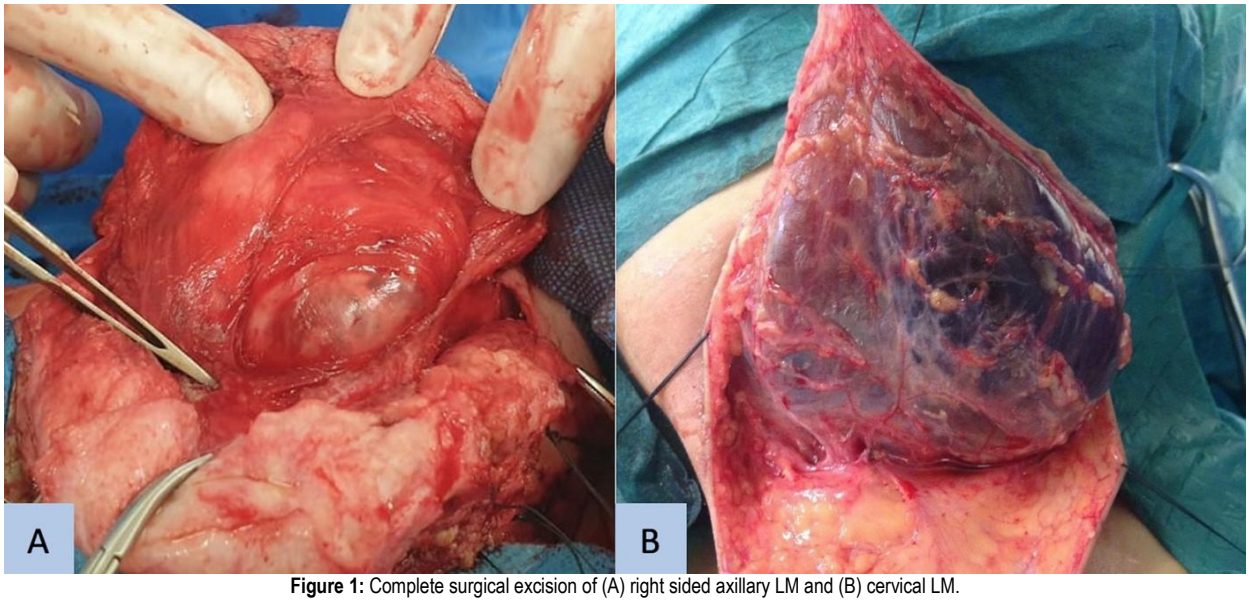

Figure 1: Complete surgical excision of (A) right sided axillary LM and (B) cervical LM.

DISCUSSION

The majority (85.7%) of patients in this study presented and were treated before the age of 2 years, consistent with findings from previous studies by Hassanein and Eliasson et al.[9,16] Our study reported a male to female ratio of 4:3, while Ma et al. reported a ratio of 1:0.94[1] and Khanwalker et al. a ratio of 0.84:1.[10] Most (57.1%) patients in this study had their lesions in the neck, aligning with reports by Gallego Herrero et al. and Lee et al.[11,12]

Most patients (78.6%) in this study presented with asymptomatic mass lesions, a trend also noted by Ma et al. and Hassanein who observed that 82.3% and 76.7%, respectively, had presented with asymptomatic mass lesions.[1,2] Our standard practice is to excise LMs between six and 12 months of age, unless there is an urgent indication like respiratory distress during neonatal and early infancy periods, which was not observed in our study.

LMs can become infected due to the abnormal lymphatic vessels' limited ability to clear foreign substances. Additionally, the high proteinaceous content of these lesions invites bacterial overgrowth.[6] Infected lesions were evident in 14.3% of the patients in the present study. Previous studies by Hassanein and Chen et al. reported infection rates of 11.5% and 17% in their patients, respectively.[2,13]

Respiratory distress is a serious complication of cervical LMs, often caused by a sudden increase in lesion size following infection or intralesional hemorrhage, or by mass compression of the upper airway. This life-threatening condition was reported in only one (7.1%) patient in this study. A comparable observation was reported by Bouwman et al.[14]

About 90% of LMs can be diagnosed by history and physical examination.[6] In the present study, clinical diagnosis was achieved in 11 (78.6%) patients. It was not possible to confirm the diagnosis on clinical grounds alone in three lesions located in anatomical regions where LMs usually do not predominate, one in the chest wall, one in the upper back, and another in the gluteal region.

U/S served as the initial imaging modality in this study, revealing classical characteristics of LMs as reported by Jiaoling et al. with a U/S sensitivity rate of about 90%.[15] However, in this study, the diagnosis of LMs was successfully achieved by U/S in 78.6% of the cases. In three cases, the provisional U/S diagnosis was lipoma for a patient with a left sided chest wall lesion, and teratoma in a patient with an upper back lesion and another with a gluteal lesion. The variability in diagnoses may be attributed to the operator-dependent nature of this study and the lack of clinical correlation for lesions in less common anatomical locations.

MRI was performed for all patients to confirm the diagnosis, identify the full extent of the lesion, its interface with surrounding normal tissue, and its relation with vital neurovascular structures. This information served as a key measure in planning complete surgical excision. MRI confirmed the diagnosis of LM in 13 (92.8%) patients. However, one patient with a lesion in

the right gluteal region was diagnosed with a dermoid cyst. The correct diagnosis of LM was only possible through histopathology in this case.

This study aimed to evaluate the safety and efficacy of surgical excision as the initial and sole therapeutic option in the treatment of large macrocystic LMs in children, using a novel modification in the surgical procedure, and compare the results with those reported by previous similar studies in the English literature.

Historically, surgical excision has long been considered the initial therapeutic option for the treatment of LMs by many surgeons.[16,17] This is especially true for large and focal macrocystic lesions, which are usually characterized by clear boundaries and more limited invasion of nearby muscles and neurovascular structures, rendering them suitable to surgical excision. In contrast, predominantly microcystic LMs have a tendency to interdigitate into tissue planes and encase adjacent neurovascular structures, making complete resection difficult and hazardous.[2,16]

LMs have thin, delicate walls composed of endothelium. During surgical excision, the fundamental principle is to keep dissection close to the walls, in the interface between the lesions and normal surrounding tissues, while avoiding cyst rupture to prevent complication. Thin-walled empty cysts are difficult to identify, making careful dissection crucial.

The novelty behind our technique of continuous traction is that instead of beginning with a simple transverse incision, we start with an elliptical skin incision made over the maximum diameter of the mass, keep this skin ellipse in place, and then insert several stay sutures along its axis. Making this generous elliptical incision will create a more tension-free space that allows easier dissection and delivery of the mass, and the stay sutures will be used to exert continuous uniform and indirect traction on the mass in all directions to facilitate dissection and delivery throughout the whole procedure without violating the integrity of its capsule, thereby aiding in its complete excision (Fig. 2). To the best of our knowledge, this technique has not been reported in previous similar studies.

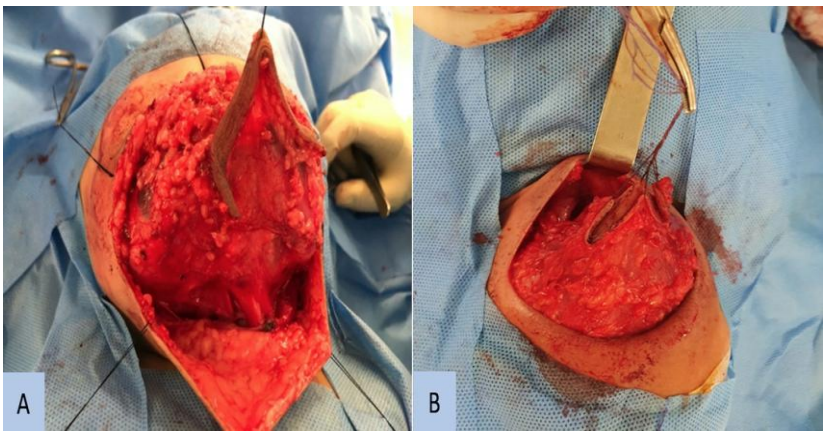

Figure 2: Continuous traction technique in (A) patient with bilateral cervical LM and (B) patient with left sided cervical LM.

LMs have a proclivity to grow along fascial planes and around neurovascular structures, making complete surgical excision of these lesions—particularly those involving the cervicofacial region—a critical procedure. Understanding anatomical details and conducting dissection close to the cyst wall will minimize the risk of neurovascular injury. In our study, we successfully dissected these lesions safely off vital structures, including the carotid sheath, jugular vein, and the trachea (Fig. 3).

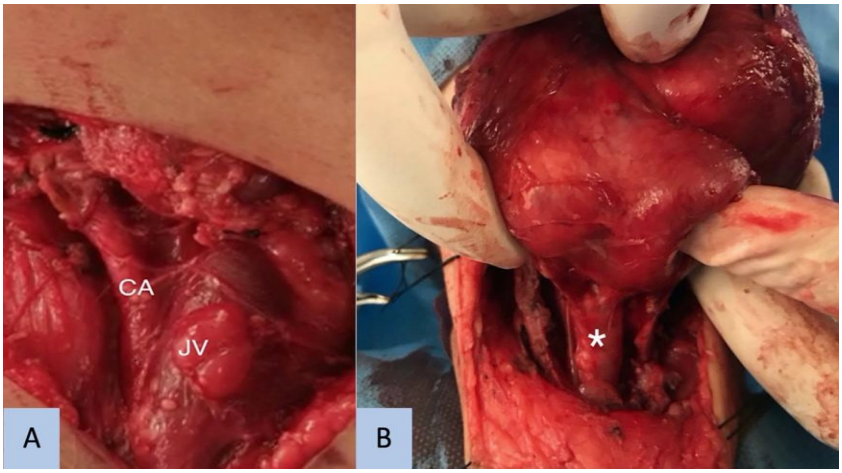

Figure 3: Avoiding injury to vital structures while excision: (A) Dissecting lesion's off the carotid artery "CA," and internal jugular vein "JV." (B) Dissecting lesion's off the trachea "✶".

Out of the 14 patients enrolled in this study, complete surgical excision was achieved in 12 (85.7%) patients. This is comparable to the results reported by previous studies.[1,13,16]

Near total resection was achieved in two patients, while minimal residual lesions were left in one patient with a huge cervical LM involving both sides of the neck. In this case, after completing near total release, it was necessary to leave a very small part of the lesion due to its deep infiltration between the left carotid artery and the internal jugular vein in anticipation of any possible vascular injury. Another patient with a left sided axillary LM had a small residual lesion that was adherent to the skin and subcutaneous tissue, following a bout of infection in anticipation of uncontrolled rupture of the cyst capsule.

The LMs in this study were located in various parts of the body, with sizes ranging from 5 x 5 cm to 12 x 9 cm (average 9.2 x 6.8 cm). Complete surgical resection was possible regardless of the lesion site and size.

During surgical excision, there were no significant neurovascular injuries reported. A similar observation has been reported by Ma et al.[1] Wound healing was good in the majority (85.7%) of patients, with comparable results reported by previous studies.[2,13] Partial wound dehiscence was reported in one patient, and wound infection was observed in another. Both responded well to conservative therapy.

All patients underwent a period of postoperative follow up that ranged from eight to 16 months, accounting for an average of 12.07 months. Esthetic results were satisfactory in the vast majority (92.8%) of patients. Moreover, no recurrence (0%) was reported in any of the patients studied. Ma et al. reported a recurrence rate of 2.9% in their series of 68 patients with macrocystic LMs.[1] This may be attributed to the larger population sample in their study in comparison to ours. Hassanein et al. reported an even a higher recurrence rate of 8.3% in their study.[16] This is because they included the more infiltrative LMs of microcystic and mixed types that usually interdigitate more among tissue planes, making their complete resection more difficult, hence leaving more lesions with subtotal resection.

Over the last few decades, intralesional sclerotherapy has emerged as an alternative or adjuvant therapeutic modality, especially for the management of infiltrative lesions that pose a high risk of injury to vital structures or have a potential for recurrence, yielding encouraging and excellent results. Some authors observed that using sclerotherapy is usually associated with leaving a residual lesion.[18] Others reported that intralesional sclerotherapy is not suitable for lesions surrounding the trachea due to upper airway obstruction that results from post-injection edema.[19] Several studies have raised concerns about the lengthy treatment period associated with sclerotherapy, as well as the uncertainty surrounding the possibility of recurrence and long-term toxicity.[20] In 2014, Balakrishnan et al. were the first to make a comparison between primary surgery and primary sclerotherapy in the management of head and neck LMs, reporting equal effectiveness.[21]

One major limitation in our study is the small sample size owing to the rarity of the pathology itself. This has resulted in our inability to compare our results with those of other therapeutic options.

Future studies and more extensive reviews will be required to thoroughly investigate the safety and

effectiveness of surgical excision in the management of large macrocystic LMs in children.

CONCLUSIONS

Complete surgical excision has proven to be a safe and effective initial therapeutic modality in the management of large macrocystic LMs in children, yielding excellent postoperative results in terms of functional outcomes and recurrence. The continuous traction technique adopted during surgical excision in this study has been crucial in achieving these results.

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