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MODERN APPROACHES IN TREATMENT OF LYMPHATIC MALFORMATIONS IN CHILDREN

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Summary.

Lymphatic malformations (LMs) represent a prevalent subtype of vascular anomalies associated with significant morbidity in pediatric patients. Clinical manifestations, disease progression, and prognosis correlate with cyst size and anatomical location. However, optimal therapeutic strategies remain incompletely defined.

***The aim of this study** was to analyze the clinical presentation, complications, and treatment outcomes of LMs in children.*

***Methods.** This retrospective, non-randomized case series included 259 patients with LMs who underwent hospital treatment at a single center between 2014 and 2024.*

***Results.** The majority of patients presented with LMs during the first two years of life (81,5%), and a significant number were diagnosed prenatally (15,83%). Cervicofacial LMs were the most common type (52,51%), with mediastinal involvement observed in 11.9% of cases. Airway obstruction necessitated tracheostomy in 4.2% of patients. Abdominal and retroperitoneal LMs were diagnosed in 43 (19,1%) while external genitalia involvement was noted in 17 (6,56%) patients. Sclerotherapy was the primary treatment for head and neck LMs (117/136 (86,03%)), whereas surgical resection was more frequently employed for abdominal LMs (32/43 (74,42%)) and external genitalia LMs (10/26 (38,46%)). Extremity, axillary, and truncal LMs were predominantly managed with sclerotherapy. Systemic sirolimus therapy was administered in 3.1% of cases (n=8) with complicated LMs, resulting in symptomatic improvement in 78.5% of these patients, though no complete resolutions were achieved.*

***Conclusion.** Common LM complications included inflammation and intralesional bleeding with mass effect. Surgical resection proved effective for abdominal and external genitalia LMs but carried potential risks for cervicofacial and limb LMs. mTOR inhibitor therapy demonstrated efficacy in complex cases. A multidisciplinary approach remains essential for LM management, with further refinement of treatment strategies required to optimize outcomes.*

***Keywords:** Vascular Anomalies; Lymphatic Malformations; Children; Cclerotherapy; Curgical Treatment; mTOR Inhibitors.*

Introduction

Lymphatic malformations (LMs) encompass a spectrum of disorders with heterogeneous clinical presentations, disease courses, and prognoses. Cystic LMs represent the most prevalent subtype. Classification by cyst size has clinical relevance, as macrocystic LMs demonstrate superior treatment responsiveness to both surgical intervention and sclerotherapy compared to microcystic LMs, whose infiltrative growth pattern often precludes effective therapy [1].

This article presents our institutional experience in managing cystic LMs, with particular emphasis on complications and therapeutic approaches. We demonstrate that clinical variability stems not only from anatomical location and cyst size, but also from complication status and relationship to major lymphatic collectors. While sclerotherapy remains the gold standard for cystic LMs [2], surgical resection maintains clinical utility, particularly for abdominal lesions. Emerging evidence supports mTOR inhibitors as targeted therapy for complex cases [3].

The introduction of mTOR inhibitors may herald a new paradigm in managing severe, complicated LMs, given their promising therapeutic profile [4].

Materials and methods

The Committee on Clinical Investigation at Bogomolets National Medical University approved this study, which was conducted in compliance with Good Clinical Practice (ICH-GCP) guidelines and the Declaration of Helsinki [5]. Written informed consent was obtained from all participants' parents/legal guardians, with the approval of the study protocol granted by the Local Ethics Committee.

Statistical analysis utilized R software (version 4.2.2, 2022-10-31 Continuous variables with normal distribution are presented as arithmetic mean (M) ± standard error (m) (M±m); other data are reported with 95% confidence intervals.

Study design: retrospective nonrandomized case series.

In total, 259 patients with cystic LMs who underwent hospital treatment within a single center between December 2014-December 2024 were included in the study.

Inclusion criteria comprised: age less than 18 years old, ≥6-month follow-up, centralized diagnostic/therapeutic management, and absence of malignant neoplasms.

Standardized evaluation included: demographic characteristics, disease onset age, anatomical localization, clinical manifestations, complications, prior treatments, therapeutic outcomes, current treatments and procedure-related adverse events. Diagnostic protocols incorporated grayscale ultrasonography with color/spectral Doppler for initial screening and surveillance. MRI was performed at the beginning of treatment (n=239) and at following treatment stages (n=155). Contrast-enhanced CT (n=23) was reserved for acute airway compromise or osseous involvement assessment. Histopathological confirmation (n=59) required positivity for podoplanin (lymphatic endothelial marker recognized by antibody D2-40) in resected specimens.

OK-432 (Picibanil), bleomycin, or their combination were utilized for sclerotherapy. Under ultrasound guidance, the procedure involved cystic cavity puncture, fluid aspiration (with subsequent cytological confirmation), and sclerosing agent administration. Laparoscopic guidance was employed for patient's safety during sclerotherapy of extensive intraabdominal/retroperitoneal cystic LMs. Dosing regimens were: OK-432 at 0.5-1 KE per session (12-24 week intervals); bleomycin at 0.5-1.0 mg/kg per session (8-12 week intervals), not exceeding 5.0 mg/kg annually.

The mTOR inhibitor sirolimus was administered both systemically (initial dose 0.8 mg/m^2 twice daily, target serum level 5-15 ng/mL [6]) and topically (0.2% gel twice daily).

All systemic therapy recipients received trimethoprim-sulfamethoxazole for Pneumocystis pneumonia prophylaxis, with biweekly laboratory monitoring

Treatment efficacy was quantified by percentage LM volume reduction ≥6 months post-treatment, categorized as: excellent (90-100%), good (60-89%), satisfactory (20-59%), or absent (0-19%)

Results

Patient demographics and other characteristics are presented in Table 1. A slight male predominance can be observed. Although presentation ages varied, 81.1% (n=210) manifested within the first two years of life, including 15.8% (n=38) with prenatal diagnosis via routine ultrasonography. Among prenatal cases, 5.3% (n=2) required fetal MRI for suspected airway compromise, with one EXIT procedure performed. Cervicofacial localization predominated (52.51%), without left-sided predominance despite thoracic duct anatomy (Figure 1).

Table 1

Demographic and clinical characteristics of patients with lymphatic malformations (n=259)

Total group n=259
Gender
Male 142 (54,83%)
Female 117 (44.17%)
Age at Disease Onset
Prenatally 41 (15,83%)
Immediately postnatally 78 (30,12%)
between 1-5 years 91 (35,14%)
between 6-12 years 27 (10,42%)
> 12 years 22 (8,49%)
Anatomical Distribution
Head and neck 136 (52,51%)
Abdominal/retroperitoneal locations 43 (16,60%)
Axillar 11(4,25%)
Extremities 24 (9,27%)
Thoracic/abdominal wall 9 (3,47%)
Perineum 9 (3,47%)
Multiple lesions 10 (3,86%)
External genitalia 17 (6,56%)
Clinical Variants
Cystic (common) form 244 (94,21%)
Generalized lymphatic anomaly (GLA) 5 (1,93%)
Kaposiform lymphangiomatosis (KLA) 3 (1,16%)
LM in GSD 3 (1,16%)
Congenital telangiectatic LM (CTLM) 4 (1.54%)
Primary Patients 212 (82.2%)
Treatment History
ß-blocker therapy 3 (1,16%)
Systemic steroids 1 (0,39%)
Surgical management 36 (13,9%)
Lesion-related Complications
Airways compression 17 (12,5%)
Proptosis 4 (1,54%)
Acute exophthalmos 2 (0,77%)
Mucosal lymphorrhea 8 (3,09%)
Cutaneous lymphorrhea 18 (6,95%)
Coagulation disorders 5 (1,93%)
Chyloperitoneum 1 (0,39%)
Chylopericardium 3 (1,16%)
Chylothorax 5 (1,93%)
Osseous involvement 11 (4,25%)

Most cases (82.2%, n=212) represented initial presentations, while 18.2% (n=47) had prior interventions: surgical (17.0%, n=44) or medical (1.2%, n=3; β-blockers n=3, steroids n=1). Previous treatment complications included: facial nerve palsy (n=2), phrenic nerve injury (n=1), lymphorrhea-associated vesicles (n=18), and deforming scars (n=22).

The incidence and severity of LM complications correlated strongly with anatomical localization. Among 136 patients with cervicofacial and mediastinal cystic LMs, airway compression developed in 12.5% (n=17). Clinical manifestations ranged from transient stridor (5.1%, n=7) to complete airway obstruction requiring tracheostomy (4.4%, n=6), with 2.9% (n=4) necessitating emergency surgical intervention for acute respiratory failure. All tracheostomies were performed prior to definitive treatment. Bilateral involvement and mediastinal extension were identified as primary risk factors for clinically significant airway compromise.

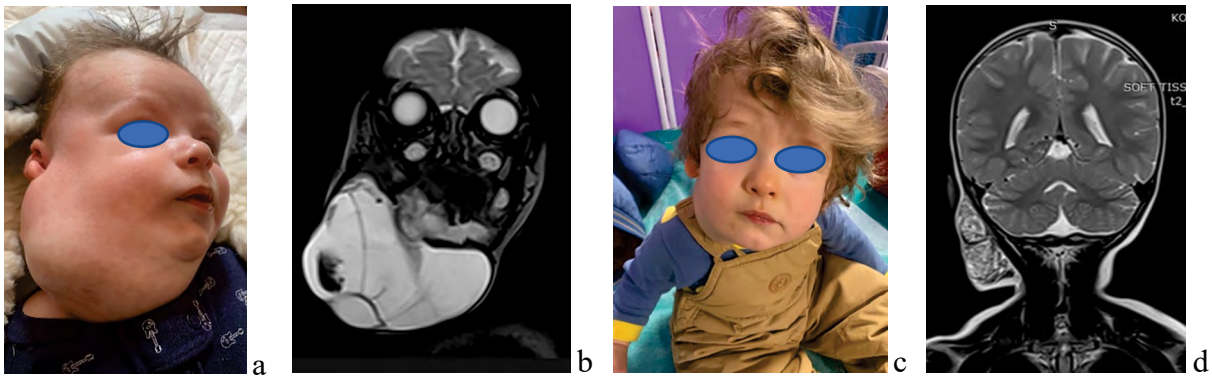

Figure 1. Cervicofacial LM before (A, B) and after (C, D) sclerotherapy.

Ophthalmic complications included proptosis secondary to upper eyelid infiltration (n=4), carrying substantial risk of amblyopia development without prompt intervention. Bleomycin sclerotherapy demonstrated particular efficacy in these cases. Two patients developed acute exophthalmos from retrocanal orbital lesions.

Oral cavity involvement manifested as mucosal vesicles with lymphorrhea in 5.9% (n=8) of cervicofacial cases, with associated macroglossia and open-bite deformity in five instances.

Cutaneous lymphorrhea affected 7.0% (n=18) of the total cohort, predominantly (77.8%, n=14) following incomplete surgical resection. Anatomical distribution of cutaneous involvement included extremities (n=7), perineum (n=3), and facial regions (n=3).

Coagulopathy, characterized by marked D-dimer elevation (>5×ULN) with mild hypofibrinogenemia, was identified in 1.9% (n=5) of patients with extensive LMs.

The most severe complications arose from lymphatic leakage, intralesional hemorrhage, and inflammatory-mediated mass effect, particularly in mediastinal LMs.

Chylothorax developed in two patients with massive thoracoabdominal LMs (one postoperatively). Surgical exploration revealed cisterna chyli injury as the etiology of postoperative chylous leakage. Mediastinal involvement was documented in 7.0% (n=18) of cases, comprising isolated lesions (11.1%, n=2), combined cervicofacial-mediastinal disease (61.1%, n=11), and concurrent truncal involvement (11.1%, n=2). Acute hemorrhage within mediastinal LMs precipitated respiratory compromise in 16.7% (n=3), requiring emergent surgical management (complete excision in one case, debulking procedures in two) with successful outcomes.

Therapeutic approaches for LMs included sclerotherapy (n=149, 57.5%), surgical excision (n=49, 18.9%), combined sclerotherapy and surgery (n=25, 9.7%), and sirolimus therapy (n=4, 1.5%), with systemic sirolimus administered to 8 patients and topical sirolimus gel applied in 2 cases.

Sclerosing agents comprised OK-432 (49.1%, n=80), bleomycin (19.3%, n=50), and their combination (12.7%, n=33). Both agents demonstrated good tolerability, though OK-432 was associated with allergic skin rash (1.3%, n=1/80) and edema requiring decompression (5.0%, n=4/80). Bleomycin-related adverse events included local infection (1.2%, n=1/83), predominantly with oral mucosa applications, and hyperpigmentation (2.4%, n=2/83).

Cervicofacial LMs were managed primarily with sclerotherapy (86.0%, n=117/136), utilizing OK-432 (55.4%, n=62), bleomycin (25.0%, n=28), or combination therapy (19.6%, n=22). Surgical intervention was required in 2.9% (n=4) for atypical histopathology (n=2), recurrent inflammation (n=1), or OK-432 allergy (n=1). Combined approaches (9.6%, n=13) involved sclerotherapy with adjunctive procedures: tracheostomy with mass resection (n=3), partial glossectomy for macroglossia (n=3), intraorbital LM resection for exophthalmos (n=1), and debulking of residual masses (n=6). Spontaneous regression occurred in 2.2% (n=3). Mediastinal-extending cervicofacial LMs were treated with sclerotherapy alone (54.5%, n=6/11), sclerotherapy with tracheostomy (27.3%, n=3), or combined sclerotherapy and mediastinal resection (18.2%, n=2).

Treatment outcomes for cervicofacial LMs were excellent (50.7%, n=69), good (34.6%, n=47), or satisfactory (14.7%, n=20), with no mortality. Macrocystic LMs demonstrated superior outcomes, particularly unilateral cases showing 100% excellent/good results, including 81.2% (n=52/64) requiring single sclerotherapy sessions. Microcystic variants, especially upper lip localization, showed poorer response (10-30% regression). Prior incomplete resection predicted worse outcomes, with satisfactory results in 31.3% (n=5/16) of such cases.

Three (81.8%) of 11 patients with axillary LMs underwent sclerotherapy, with OK-432 used in 9 (60.0%) cases and OK-432 combined with bleomycin in 2 cases. Excellent results were achieved in 8 (72.7%) patients and good results in 2 (18.2%) patients. In 2 patients who had undergone prior debulking procedures, sclerotherapy proved insufficiently effective, but topical application of 0.2% sirolimus gel successfully controlled skin lesion progression.

Among 24 patients with extremity LMs, 10 presented as primary cases while 8 had previous surgical interventions. Notably, all patients with prior procedures developed complications, including incision scar vesicles (n=7), lymphedema (n=2), and prolonged lymphorrhea (>30 days, n=3). Treatment approaches included sclerotherapy (54.2%, n=13) and surgical excision (4.2%, n=1).

Abdominal and retroperitoneal LMs were diagnosed in 39 (15.1%) children, with isolated lesions in 92.3% (n=36) and combined mediastinal involvement in 7.7% (n=3). Surgical management included: complete laparoscopic resection (41.0%, n=16), video-assisted transumbilical bowel resection (16.4%, n=6), and primary laparotomy (n=5). Five cases required conversion to laparotomy due to previous surgeries or severe inflammatory processes. Sclerotherapy was employed for 4 retroperitoneal LMs,

including one case with thoracic duct connection via cisterna chyli managed with laparoscopic-guided sclerotherapy.

Splenic lesions required splenectomy in one patient for refractory pain, while two patients were managed conservatively. Treatment outcomes for abdominal LMs included excellent (76.9%, n=30), good (17.9%, n=7), satisfactory (5.1%, n=2), and unsatisfactory (2.3%, n=1) results.

Systemic sirolimus therapy was administered to 8 (3.1%) patients with complicated cases: cervicofacial and mediastinal cystic LMs with upper airway obstruction (n=2), thoracoabdominal LMs complicated by chylothorax (n=2), extensive chest wall lesions with mediastinal extension (n=2), and external genitalia/perineal LMs (n=2). Clinical improvement (symptom reduction, decreased LM size, and cosmetic enhancement) was observed in 78.5% (n=7/8) of cases, though no complete resolutions were achieved. Treatment duration ranged from 6 to 24 months (median 7±2.4 months). One 2-year-old patient with extensive abdominal/retroperitoneal LMs and postsurgical chylothorax succumbed to secondary immunodeficiency and sepsis.

Sirolimus-related adverse events included oral mucositis (n=2), dyslipidemia (n=2), and leukopenia (n=1). Mucositis responded to topical antiseptics, while dyslipidemia was managed with dietary modification. The leukopenic episode (occurring in an infant) resolved after temporary sirolimus discontinuation, with subsequent uneventful therapy continuation.

All 26 patients with external genitalia/perineal LMs received individualized treatment. Sclerotherapy (inducing obliteration of pathological lymphatic vessels) was performed in 4 cases. Surgical intervention (partial/complete resection for functional and aesthetic improvement) was undertaken in 9 patients. Two patients received systemic sirolimus (mTOR inhibitor therapy targeting abnormal lymphatic proliferation). This multimodal approach achieved pathological regression while minimizing recurrence risk.

The assessment of treatment efficacy demonstrated positive outcomes in most cases. Complete resolution of clinical manifestations or only minimal residual symptoms with no functional impairment was observed in 7 patients (26.9%), indicating an excellent treatment outcome. A significant improvement with minimal residual symptoms, classified as a good outcome, was achieved in 12 patients (46.2%). A satisfactory outcome, characterized by partial regression of the pathological process, was recorded in 7 cases (26.9%).

The choice of treatment modality for LMs should be guided by their anatomical localization, as highlighted in Table 2.

Table 2

Treatment of cystic (common) LMs according to anatomical localization

Anatomical localization	Sclerotherapy	Surgery	Systemic Sirolimus	Combined	Spontaneous regression, observation
Head and neck Total 136	117 (86,03%)	4(2,94%)		13 (9,56%) (sclerotherapy and surgery)	2 (1,47%)
Abdominal and retroperitoneal Total 43	5 (11,63%)	32 (74,42%)	2 (4,65%)	4 (9,30%) (sclerotherapy and surgery)	-
Axillar Total 11	9 (81,82%)			2 (18,18%) (sclerotherapy and topical Sirolimus)	
Extremities Total 24	13 (54,17%)	1 (4,17%)		1 (4,17%)	9 (37,5%
External genitalia and perineum Total 26	5 (19,23%)	10 (38,46%)	2 (7,69%)	5 (19,23%) (sclerotherapy and surgery)	4 (15,38%)

Multivariate analysis revealed that treatment outcomes for cystic LMs were primarily determined by LM subtype and prior treatment history. Patients with macrocystic LMs demonstrated superior treatment outcomes ($p<0.005$). Previous therapeutic interventions were associated with reduced treatment efficacy ($p<0.005$), while lymphatic leakage and mediastinal involvement correlated with poorer prognosis.

The overall results of cystic LM treatment are summarized in Table 3.

Table 3

Results of treatment

Anatomical localizations	Excellent	Good	Satisfactory	Pure
Head and neck Total 136	69 (50,74%)	47 (34,56%)	20 (14,71%)	0
Abdominal and retroperitoneal Total 39	30 (76,92%)	7 (17,95%)	2 (5,13%)	0
Axillar Total 11	8 (72,73%)	2 (18,18%)	1 (9,09%)	0
Extremities Total 24	3 (12,5%)	18 (75%)	3 (12,5%)	0
External genitalia and perineum Total 26	7 (26,92%)	12 (46,15%)	7 (26,92%)	0
Other, including multiple lesions Total 23	6 (26,09%)	9 (39,13%)	7 (30,43%)	1 (4,35%)
Total 259	123 (47,49%)	95(36,68%)	40 (15,44%)	1 (0,39%)

Discussion

When characterizing cystic LMs, their principal clinical features include: an incidence of 1 per 4000-16000 live births [7,8], predominant localization in anatomical regions containing major lymphatic collectors – specifically the head and neck region (45-53%) [1,9] (corresponding to the thoracic duct venous angle junction) and abdominal cavity (cisterna chyli area where abdominal lymphatic tributaries converge) [10]. While disease onset typically occurs within the first two years of life, clinical manifestations may emerge at any age [1,8]. Our findings align with published data [11]: 62.5% of cases involved cervicofacial localization, 17.9% demonstrated abdominal/retroperitoneal involvement, 70.7% manifested within the first two years of life, while 6.3% presented after age 12. Prenatal diagnosis was established in 15.8% of cases [12].

Historically, surgical excision represented the sole therapeutic option for cystic LMs, though outcomes were frequently suboptimal. Cervicofacial LM resection has been associated with complication rates of 12-33% and recurrence rates of 15-53% [9,13]. Recurrences and complications typically result from inadequate preoperative evaluation or suboptimal treatment planning. Surgeons must remain vigilant for unexpected anatomical findings [14]. In our cohort, 80% of patients experiencing residual masses or postoperative complications had undergone procedures without preoperative CT or MRI.

The efficacy of surgery in achieving satisfactory cosmetic outcomes for cervicofacial LMs remains questionable. Procedural planning must account for several pathological characteristics: fascial plane adherence, tissue infiltration, and anatomical distortion by LMs. Iatrogenic injury to neurovascular structures during resection of benign lesions represents an unacceptable risk [15]. Furthermore, postoperative progression of facial asymmetry occurred in 31.3% of cases in our series, with lymphatic vesicle adhesion to surrounding tissues and scar formation representing additional concerns [16].

While no standardized treatment protocol exists for cystic LMs, sclerotherapy has emerged as a predominant therapeutic approach in recent decades [2,9,17]. Current literature consistently reports superior outcomes for macrocystic LMs regardless of sclerosing agent selection [9,17,18]. In our clinical practice, both OK-432 and bleomycin demonstrated favorable efficacy profiles with minimal adverse effects.

Most LMs present as isolated lesions that do not threaten pediatric viability, particularly small localized variants demonstrating spontaneous regression in 2.8% of cases. This observation supports conservative management strategies, as therapeutic interventions in young children carry inherent risks given the frequent proximity of LMs to vital structures governing respiration, deglutition, auditory, and visual functions. Clinicians must recognize that complete LM eradication may not always be achievable. In many instances, vigilant observation («watch and wait» strategy) proves preferable to premature intervention without comprehensive pathological understanding, which may precipitate detrimental outcomes [19]. Our data indicate a mean treatment initiation age of 3.9±3.6 years (95% CI), consistent with published norms of 3.4 years [15].

Traditional pathophysiological models attributed LM development to lymphatic network maturation defects resulting in dilated, dysfunctional channels lacking smooth muscle components. Contemporary molecular research has identified somatic genetic abnormalities within malformation tissue as the fundamental etiology [7,13].

Specifically, somatic mutations in PIK3CA (encoding phosphatidylinositol-4,5-bisphosphate 3-kinase catalytic subunit alpha) frequently occur in isolated lymphatic malformations and certain vascular malformations with lymphatic components [20]. These discoveries substantiate the pivotal role of PI3K/mTOR pathway dysregulation in vascular morphogenesis, rationalizing mTOR inhibitor therapy. Since the inaugural 2011 report of sirolimus application for LMs [21], subsequent case series [9,22-23] and a prospective multicenter trial [20] have corroborated its therapeutic potential. However, methodological limitations persist, including heterogeneous patient populations compromising statistical rigor. Critical questions remain regarding optimal treatment duration, toxicity mitigation, and long-term oncological risks (particularly cutaneous malignancies and lymphoproliferative disorders) [24]. Dosing protocols exhibit minor variability across studies, though most employ an initial regimen of 1.6 mg/m^2 divided twice daily [4,21], which we have adopted. Therapeutic drug monitoring targets 5-15 ng/mL plasma concentrations, supported by 76% of reviewed literature (19/25 sources) [13]. Our protocol mandated 10% dose escalation for subtherapeutic levels (<5 ng/mL), while maintaining concentrations below the 20 ng/mL toxicity threshold [13]. Therefore, deviations in sirolimus clearance must be carefully monitored in pediatric patients as they reflect drug metabolism capacity. The most comprehensive sirolimus dosing guidelines originate from pharmacologists at Cincinnati Children's Hospital Medical Center [3]. Target plasma concentrations of 5-15 ng/mL were achieved within 2-3 months in 94% of patients aged 3 months to 18 years. For children >2 years, the average dose required to maintain ~10 ng/mL was 1.8 mg/m^2 twice daily (range 0.8-2.9 mg/m^2), while infants (3 weeks-2 years) required 0.7-1.6 mg/m^2 twice daily. Both literature evidence and our data indicate heightened sirolimus toxicity risks in younger populations. In our cohort, leukopenia developed in one 12-month-old patient, with no marrow suppression observed in others. Notably, the first reported sirolimus-related fatalities occurred in 2018: two infants with kaposiform hemangioendothelioma developed fatal Pneumocystis pneumonia [25], emphasizing the critical importance of prophylaxis.

Our experience demonstrates sirolimus efficacy in giant LMs, consistent with recent publications. However, prognosis significantly worsens with concurrent chylothorax, mediastinal involvement, or airway compromise [3].

Conclusions

LMs exhibit remarkable clinical heterogeneity, from incidental findings to life-threatening masses.

Common complications include inflammatory episodes, intralesional hemorrhage with mass effect, sepsis, and hemorrhagic shock.

Current therapeutic options include surgical resection, sclerotherapy, and emerging targeted therapies. While surgery remains preferred for abdominal and genital LMs, its application

for cervicofacial and extremity lesions carries substantial risks of recurrence, complications, and suboptimal cosmesis. mTOR inhibitors show particular promise for complex cases.

These findings validate multidisciplinary management while highlighting the need for optimized treatment protocols to improve long-term outcomes.

Keywords: Vascular Anomalies; Lymphatic Malformations; Children; Sclerotherapy; Surgical Treatment; mTOR Inhibitors.

Conflict of Interest. The authors declare no conflict of interest.

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НЕОНАТОЛОГІЯ, ХІРУРГІЯ ТА ПЕРИНАТАЛЬНА МЕДИЦИНА
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СУЧАСНІ ПІДХОДИ В ЛІКУВАННІ ЛІМФАТИЧНИХ МАЛЬФОРМАЦІЙ У ДІТЕЙ

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Резюме.

Лімфатичні мальформації (ЛМ) є поширеним типом лімфатичних мальформацій, які можуть спричинити значну захворюваність у дітей. Клінічна картина, перебіг і прогноз ЛМ варіюють залежно від розміру і локалізації кіст. Однак оптимальна стратегія лікування ЛМ ще не до кінця встановлена.

Метою цього дослідження був аналіз клінічної картини, ускладнень та результатів лікування ЛМ у дітей.

Методи. Ця ретроспективна не рандомізована серія випадків включала 259 пацієнтів з ЛМ, які проходили стаціонарне лікування в одному центрі в період з 2014 по 2024 роки.

Результати. У більшості пацієнтів ЛМ були виявлені протягом перших двох років життя (81,5%), а значна кількість була діагностована пренатально (15,83%). ЛМ голови та шиї були найбільш поширеним типом (52,51%), зі значною часткою середостіння (11,9%). З них 4,2% потребували трахеостомії з приводу обструкції дихальних шляхів. Також відносно поширеними були абдомінальні та заочеревинні ЛМ, діагностовані у 43 (19,1%) дітей, зовнішні статеві органи були залучені у 17 (6,56%). Склеротерапія була найбільш поширеним методом лікування ЛМ голови та шиї (117/136 (86,03%)), тоді як хірургічна резекція частіше застосовувалася для ЛМ черевної порожнини (32/43 (74,42%)) та ЛМ зовнішніх статевих органів (10/26 (38,46%)). Склеротерапію переважно застосовували у хворих з ЛМ кінцівок, пахвових та тулупчастих ЛМ. Системну терапію сиролімусом застосовували у невеликої кількості пацієнтів (n=8 (3,09%)) з ускладненими ЛМ. З них у 7/8 (78,5%) пацієнтів спостерігалося поліпшення симптомів, але в жодного не спостерігалося повне зникнення захворювання.

Висновок. Поширеними ускладненнями ЛМ є запалення і внутрішньосеансові кровотечі з мас-ефектом. Хірургічне втручання є методом вибору при ЛМ черевної та зовнішніх статевих органів, але потенційно шкідливому при ЛМ шийно-лицевих органів та кінцівок. У складних випадках системна терапія інгібіторами mTOR була ефективною. Мультидисциплінарний підхід є ефективним у лікуванні ЛМ, вимагаючи подальшого удосконалення стратегій для оптимізації результатів.

Ключові слова: судинні аномалії; лімфатичні мальформації; діти; склеротерапія; хірургічне лікування; інгібітори mTOR.

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