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Original Article

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Lymphangioma: A 2019 Survey Conducted by the Korean Association of Pediatric Surgeons

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Conflict of Interest
No potential conflicts of interest relevant to this article are reported.

ABSTRACT

Purpose: Report of a nationwide survey on lymphangioma conducted by the Korean Association of Pediatric Surgeons (KAPS) in 2019.
Methods: The authors reviewed and analyzed the clinical data of pediatric patients who started treatment for lymphangioma in hospitals of KAPS members from 2011 to 2013. Their follow-up data is also included in the study.
Results: A total of 532 patients with lymphangioma from 18 institutes were registered for the study. The results were discussed at the 35th annual meeting of KAPS, which was held in Gyeongju on June 13–14, 2019.
Conclusion: This study provides general information on lymphangioma and comprehensive treatment outcomes for this disease. The study is expected to be an important reference for improving pediatric surgeons' understanding and treatment of lymphangioma.

Keywords: Lymphangioma; Pediatrics; Surveys and questionnaires

INTRODUCTION

Lymphangioma or lymphatic malformation includes a wide spectrum of abnormalities because it can be found in any region of the body and has a variety of appearances [1-4]. Although pediatric surgeons occasionally encounter this disease, the incidence of lymphangioma is relatively low. This means that it takes a long time for surgeons to accumulate experience in the treatment of lymphangioma. Therefore, a nationwide study is helpful for surgeons to understand this disease and choose optimal treatment methods. The 2019 annual nationwide survey of the Korean Association of Pediatric Surgeons (KAPS) was on the subject of lymphangioma, and the results were first discussed at the annual meeting of KAPS in Gyeongju on June 13–14, 2019.

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METHODS

The authors reviewed and analyzed the clinical data of patients who started treatment for lymphangioma in KAPS members' hospitals from 2011 to 2013. Their follow-up data were also included in the study. We used Microsoft Access 2016® (Microsoft, Redmond, WA, USA) for patient registry and data collection. All the data were analyzed using the IBM SPSS version 23 statistical software package (IBM Co., Armonk, NY, USA).

RESULTS

1. Demographics

A total of 532 patients with lymphangioma from 18 institutes were registered for the study (**Fig. 1**). Although 550 patients were reported, 18 patients were reported by 2 institutes. Therefore, we unified the data of these patients. The patients' demographics and clinical characteristics are summarized in **Tables 1** and **2**. Age at diagnosis varied; 39.3% of patients (209/532) were diagnosed in the infant period, and 25% (133/532) were diagnosed after the age of 5. Only 12.8% of patients (68/532) were diagnosed postnatally. The majority of patients (81.0%) were treated by pediatric surgeons. Most patients (94.9%) had a lesion at a single site, but 27 patients had multiple lesions at more than one site. The most common symptom was mass, followed by pain. The most common imaging study was ultrasound, followed by magnetic resonance imaging. We classified lymphangioma with a diameter of more than 1 cm as macrocystic. Macrocystic lymphangioma was the most common type of lymphangioma, and the median diameter size was 4.8 cm.

2. Treatment

We grouped treatment methods into 5 categories: observation only; operation; sclerotherapy; operation and sclerotherapy; and other treatments (**Fig. 2**). Operation with observation and sclerotherapy with observation were classified as operation and sclerotherapy, respectively.

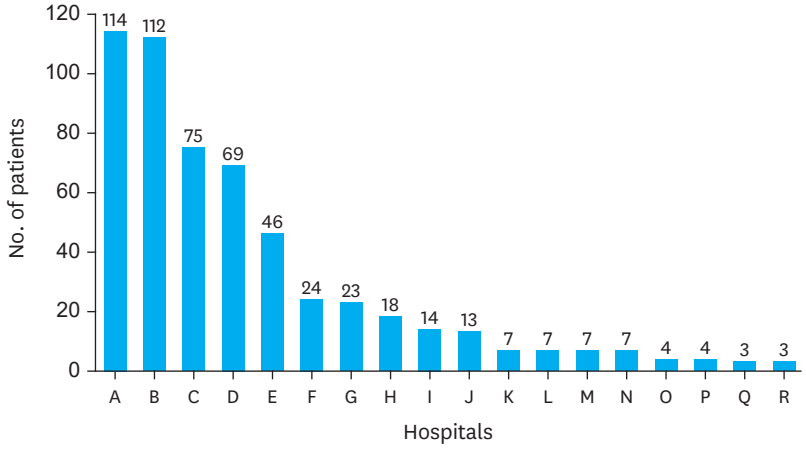

Fig. 1. Hospital distribution of patients with lymphangioma.

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Table 1. Patient demographics

Characteristics	Value (n=532)
Sex	M:F=1.18:1 (288:244)
Mode of delivery	
Normal spontaneous vaginal delivery	203
C-section	128
Unknown	201
Age at diagnosis	
≤28 days	106
28 days<Age≤6 months	53
6 months<Age≤1 year	50
1 year<Age≤3 years	115
3 year<Age≤5 years	74
>5 year	133
Unknown	1
Prenatal diagnosis	68 (12.8)
Department of treatment	
Pediatric surgery	431
Other department	101
Pediatrics	13
Plastic surgery	17
Radiology	5
Otolaryngology	30
Dermatology	14
Cardiothoracic surgery	5
Orthopedic surgery	9
Ophthalmology	2
General surgery	2
Neurosurgery	2
Oral and maxillofacial surgery	2

Values are presented as number (%).

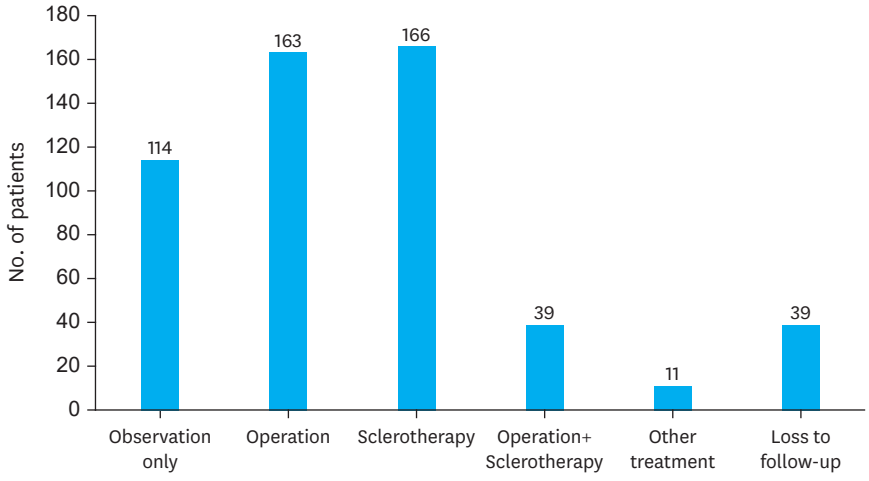

Fig. 2. Treatment methods.

Observation only

Patients who underwent observation only are summarized in **Table 3**. A total of 114 patients (21.4%) did not undergo any specific treatment, and the median observation period was 316 days. Among the patients in this group, 67.9% (88/114) showed grossly normal or partially improved appearance.

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Table 2. Clinical characteristics

Characteristics	Total	Observation (n=114)	Operation (n=163)	Sclerotherapy (n=166)	Operation+Sclerotherapy (n=39)	Others (n=50)
Location						
Single lesion	505 (94.9)					
Head and neck	228	53	39	97	18	21
Chest and mediastinum	52	12	19	12	4	5
Abdominal wall and flank	56	10	37	6	2	1
Axilla	40	11	13	10	2	4
Back and extremities	103	22	23	34	8	16
Intra-abdominal and pelvic cavity	24	1	22			1
Other	2	1				1
Multiple lesions	27 (5.1)	4	10	7	5	1
Chest+xilla	6	2	2	2		
Chest+Extremities	2		2			
Chest+Abdominal wall	4		2	1	1	
Abdominal wall+Extremities	3	1	1	1		
Neck+Chest	4		1	1	2	
Neck+Extremities	2			1	1	
Generalized	2		1		1	
Other	4	1	1	1		1
Symptoms[a]						
Mass	476 (89.5)	100	147	147	36	46
Pain	56	6	31	11	2	6
Infection	11	2	7	1	1	
Bleeding	16	7	4	2	1	2
Swelling	12	4	2	6		
Other	22	1	12	4	2	3
Imaging studies[a]						
US	355 (66.7)	79	105	121	28	22
MRI	239 (44.9)	44	63	90	24	18
CT	153 (28.8)	25	73	37	9	9
Other	11	5	3	1	0	2
Type of appearance[b]						
Macrocystic	380 (71.4)	75	120	132	29	24
Microcystic	67	21	18	16	6	6
Cavernous	25	8	11	2	1	3
Unclassified	60	10	14	16	3	17
Size (cm)						
Measurement	492	103	153	162	37	37
Median (range)[c]	4.8 (0.6–26.0)	3.7 (0.7–10.0)	6.0 (0.6–26.0)	5.0 (1.2–15.0)	6.5 (1.6–20.0)	4.1 (0.7–14.0)

Values are presented as number (%).
[a]Including multiple selection; [b]Data of imaging study or histopathology; [c]Measurement of the largest diameter.

Table 3. Observation only

Characteristics	Value (n=114)
Length of observation	(n=88)
Median days (range)	316 (7–4,490)
Results	
Grossly normal	43 (37.7)
Partial improvement (residual lymphangioma)	45
No improvement	2
Loss to follow-up	24

Values are presented as number (%).

Operation

A total of 163 patients (30.6%) underwent an operation. The predominant operation site was the head and neck, followed by the abdominal wall and flank. Characteristically, lesions in the intra-abdominal and pelvic cavity were more common in this group compared to

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APS Advances in Pediatric Surgery

the other treatment method groups. The most common histopathological diagnosis was lymphangioma, followed by hemangiolymphangioma. Most patients underwent complete excision in a single operation. Only 8% (13/163) of patients required more than 1 operation. Three patients underwent sclerotherapy during operation. Postoperative complications were not common; only 7.4% of patients experienced complications, and complications were of various kinds. After operation, 84.5% (138/163) of patients showed grossly normal results or no residual lesions (**Table 4**).

Sclerotherapy

A total of 166 patients (31.2%) underwent sclerotherapy, which is similar to the number of patients who underwent operation. The most common sclerosing agent was OK-432, followed by bleomycin and doxycycline. General anesthesia was required in the majority of patients, and more than one-third of patients required multiple sessions of sclerotherapy. Complications after sclerotherapy were extremely rare; only 3 patients experienced complications. After sclerotherapy, 57.2% (95/166) of patients showed grossly normal results (**Table 5**).

Table 4. Operation[a)]

Characteristics	Value (n=163)
Age at time of surgery (median, yr)	3.4 (0.0–19.7)
Duration from diagnosis to operation (median, days)	11 (0–2112)
Body weight at time of surgery (mean, kg)	21.0±17.9
Operation duration (mean, min)	113.5±82.0
Co-operation with other department	6
Emergency operation	8[b)]
Pathology	
Lymphangioma	141 (86.5)
Hemangiolymphangioma	12
Hematoma or Pseudocyst	5
Others	5
Type of operation	
Complete excision	140 (85.9)
Incomplete excision	22
Unknown	1
Sclerotherapy during operation	3
No. of operations	
1	150 (92.0)
2	11
≥3	2
Postoperative complications	12 (7.4)
Skin necrosis	1
Hematoma	1
Postoperative intestinal obstruction	2
Lymphatic fluid collection	2
Proptosis (orbital lymphangioma)	1
Wound dehiscence	1
Secondary lymphedema	1
Postoperative ileus	1
Drain site infection	1
Ipsilateral ptosis	1
Result of operation	
Grossly normal	138 (84.5)
Residual lymphangioma	21
Loss to follow-up	4

Values are presented as number (%).
[a)]In cases of multiple operation, the data were based on the first operation; [b)]Eight cases of intestinal obstruction.

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APS Advances in Pediatric Surgery

Table 5. Sclerotherapy

Characteristics	Value (n=166)
Age at time of sclerotherapy (median, yr)	1.4 (0.0–18.9)
Sclerosing agents	
OK-432	131 (78.9)
Bleomycin	16
Doxycycline	13
Ethanol	6
Anesthesia[a]	
General	103 (62.0)
Local	32
None	31
Radiologic intervention	
Yes	23 (13.9)
No	143
Complication	3 (1.8)
Skin necrosis	2
Ecchymosis	1
No. of sclerotherapy sessions	
1	102 (61.4)
2	38
3	10
4	4
≥5	12
Results	
Grossly normal	95 (57.2)
Residual lymphangioma	48
Loss to follow-up	23

Values are presented as number (%).
[a]Multiple injections with general anesthesia or other methods were classified as general anesthesia.

Operation and sclerotherapy

Thirty-nine patients (7.3%) underwent operation and sclerotherapy separately, not simultaneously. The majority of these patients underwent sclerotherapy either before or after operation, but 3 patients underwent sclerotherapy both before and after operation. Multiple sclerotherapy sessions were required in 51.3% (20/39) of patients. After treatment, 53.8% (21/39) showed grossly normal results (**Table 6**).

Other treatments

A total of 11 patients underwent other treatments. Of these, 4 had drug treatment (2 cases of steroid medication and 2 cases of antibiotic medication), 4 underwent ultrasound-guided aspiration, and 3 had CO_2 laser abrasions. Only 2 patients showed grossly normal results at follow-up.

DISCUSSION

Although lymphatic malformations can occur in any anatomic region, most clinical studies of lymphangioma, or lymphatic malformation, focus on lymphatic-rich areas, such as the head and neck, the axilla, and the mediastinum [3,5,6]. However, understanding the overall clinical characteristics of lymphangioma is important because the basic concepts in the treatment of lymphangioma are similar, regardless of the lesion site [2,4]. The significance of this study lies not only in the fact that it is the first nationwide survey on lymphangioma in Korea but also in the fact that it integrates clinical characteristics and treatment results of patients with lymphangioma.

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Table 6. Operation and sclerotherapy

Characteristics (n=39)	Sclerotherapy		
	Before operation	After operation	Before and after operation
No. of patients	17	19	3
Age at time of sclerotherapy (median, yr)	0.5 (0.0–8.3)	2.4 (2.1–19.2)	0.4 (0.0–0.6)
No. of sclerotherapy sessions			
1	9	10	
2	3	2	
3	3	2	2
4		1	
≥5	2	4	1
Sclerosing agents			
OK-432	17	16	3
Bleomycin		2	
Doxycycline		1	
Anesthesia			
General	10	11	3
Local	4	3	
None	3	5	
Radiologic intervention			
Yes	4	6	3
No	13	13	0
Complications			
Fever		1	
Bleeding	1		
Pain	1		
Age at time of surgery (median, yr)	2.0 (0.2–10.1)	1.4 (0.1–18.5)	1.1 (0.6–1.6)
Duration from diagnosis to operation (median, days)	375 (14–1917)	25 (5–399)	403 (210.0–555)
Body weight at time of surgery (mean, kg)	13.4±6.3	17.9±18.3	9.5±1.7
Operation duration (mean, min)	152.8±168.1	229.3±173.8	81.3±31.5
Co-operation with other department	3	7	0
Emergency operation	0	0	0
Pathology			
Lymphangioma	12	17	3
Hemangiolymphangioma	3	2	
Unknown	2		
Type of operation			
Complete excision	10	7	1
Incomplete excision	7	12	2
Sclerotherapy during operation	1	1	1
No. of operation			
1	16	14	1
2	1	3	1
≥3		2	1
Postoperative complications		1 (swelling)	
Result			
Grossly normal (21/39, 53.8%)	11	9	1
Residual lymphangioma	5	9	2
Loss to follow-up	1	1	

The results of this study are consistent with those of previous studies with respect to the most common locations of lesions, the major symptoms, the kinds of imaging studies, the types of appearances [1,7], and favoring of OK-432 as a sclerosing agent [8-10]. However, in contrast with other studies, we included observation without other treatments as a method of treatment [6]. We believe that the benign characteristic of lymphangioma and the possibility of spontaneous regression makes observation a viable treatment option [11].

Among the treatment methods considered, the best results were obtained from operation, followed by sclerotherapy, co-treatment with operation and sclerotherapy, and observation.

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However, the correlation between the choice of treatment method and the difficulty of treatment should be considered when interpreting these results. The difficulty of treating lymphangioma generally varies significantly depending on whether it takes a simple or complex form [5,12]. We think it was possible that we avoid the operation in complex form of lymphangioma, and non-operative treatment methods were selected as the preferred treatment method over operation. In this study, we did not distinguish between simple and complex forms of lymphangioma, which is one of the limitations of this study.

Other limitations are that we did not include long-term follow-up results, and we did not compare results according to the area where the lesion occurred. These issues should be addressed in future studies. Despite its limitations, this study provides general information on lymphangioma and comprehensive treatment outcomes. Therefore, we expect it will be an important reference for pediatric surgeons seeking to better understand lymphangioma and treatment options for this disease and that it will ultimately lead to better outcomes.

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