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Clinical features of mesenteric lymphatic malformation in children

Soo-Hong Kim [a], Hyun-Young Kim [b,*], Cheol Lee [c], Hye Sook Min [c], Sung-Eun Jung [b]

[a] Department of Pediatric Surgery, Pusan National University Yangsan Hospital, Yangsan, Korea
[b] Department of Pediatric Surgery, Seoul National University Children's Hospital, Seoul, Korea
[c] Department of Pathology, Seoul National University Hospital, Seoul, Korea

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ABSTRACT

Background: Mesenteric lymphatic malformations (MLs) are a heterogeneous group of benign diseases of the lymphatic system that present with cystic dilated lymphatics of the mesentery. MLs are rare and represent less than 5% of all lymphatic malformations. The aims of this study were to analyze the characteristics of MLs in children and to suggest a modified classification.

Patients and methods: We investigated 25 patients who underwent ML surgery. The clinical data and pathological findings were reviewed retrospectively. We divided the patients into 4 groups according to the operative findings. Group 1 included patients with MLs involving the intestinal walls. Group 2 included patients with pedicle-type MLs with no relationship to the mesenteric vessels. Group 3 patients presented with MLs located in the mesenteric boundaries near the mesenteric vessels. Group 4 patients had multicentric and diffusely infiltrated MLs.

Result: The male-to-female ratio was 11:14, and the median age at diagnosis was 5 years of age. The most common symptom was abdominal pain. The jejunal mesentery was the most frequently involved site in this study. Five patients showed the macrocystic type and 20 patients showed the mixed cystic type. With the exception of one patient with a large mixed cystic-type ML who underwent incomplete mass excision, 24 patients underwent complete mass excision. The group 1 patients ($n = 14$) underwent mass excision performed with segmental resection of the bowel. The group 2 patients ($n = 3$) only underwent mass excision surgery. The patients in group 3 ($n = 7$) underwent mass excision with segmental resection of the intestine because ML excision altered the blood supply of the adjacent intestines. The group 4 patients ($n = 1$) presented with MLs involving the entire mesentery and underwent incomplete excision.

Conclusion: The relationships between MLs and the neighboring organs determine the surgical strategy, and the size and location of MLs affect the operative methods. The modified classification based on these findings can facilitate effective treatment planning.

Mesenteric lymphatic malformations (ML), previously termed mesenteric lymphangiomas, are a heterogeneous group of benign malformations of the lymphatic system composed of cystic dilated lymphatics of the mesentery. Most common cystic lymphatic malformations present as cervical and axillary lesions, whereas mesenteric locations are rare and represent only 5% of lymphatic malformations [1,2]. MLs present in 1 per 20,000 admissions in the pediatric age group, with a male-to-female ratio of 1.5:1–3:1 [2–4].

Depending on the site and size of the ML, various symptoms, including abdominal pain, abdominal distension, and abdominal masses, might occur, which could be accompanied by complications, such as rupture, hemorrhage and volvulus [4,5]. Because of the diversity of symptoms, an ML might not be diagnosed preoperatively, and in some cases, an ML is found incidentally.

* Corresponding author at: Department of Pediatric Surgery, Seoul National University Children's Hospital, 101, Daehang-ro, Yeongeon-dong, Jongro-gu, Seoul, Korea, 110-744. Tel.: +82 2072 2478; fax: +82 747 5134.
E-mail address: spkhy02@snu.ac.kr (H.-Y. Kim).

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The treatment of choice is complete excision. The site, size and relationship of the ML with neighboring organs could influence its resectability [6]. Recently, sclerotherapy and medications, such as propranolol and siroliumus, have been used to treat lymphatic malformations instead of surgical treatment, achieving good outcomes [7–9].

The aim of this study was to analyze the clinicopathological characteristics of MLs in children. To more clearly describe the lesions, we modified the classification previously reported by Losanoff et al. [6] according to our operative findings.

1. Patients and methods

We investigated 25 patients who underwent ML surgery in the Department of Paediatric Surgery, Seoul National University Children's Hospital from 1991 to 2011. The clinicopathological data, including gender, age, clinical signs and symptoms, preoperative diagnosis, radiological findings, histology, operative findings, operative methods and outcomes, were reviewed retrospectively. To investigate recurrence, we conducted

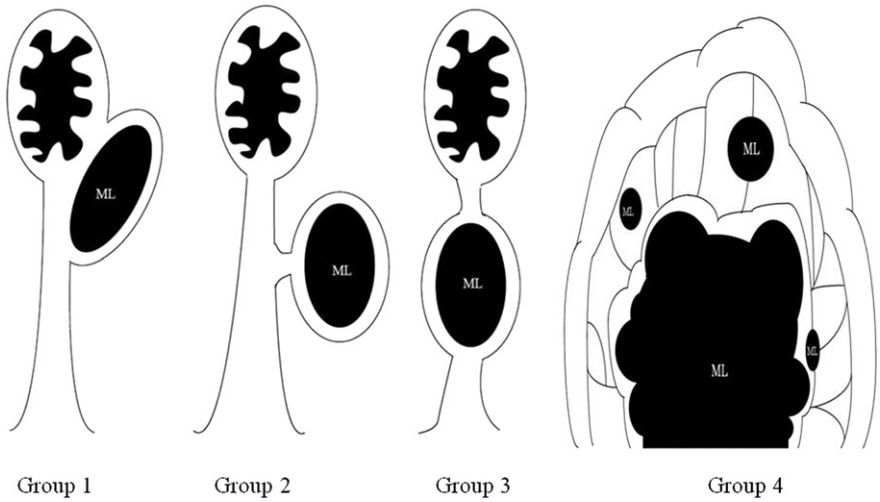

Fig. 1. Group 1 included the patients with MLs involving the intestinal walls. The group 2 patients had pedicled-type MLs with no relationship with mesenteric vessels. Group 3 included the patients with MLs located in the mesenteric boundaries near the mesenteric vessels. The group 4 patients presented with multicentric and diffusely infiltrated MLs.

telephone interviews. The median follow-up period was 10.9 ± 6.9 months (2 months–10 years).

Upon evaluation of the histological type and the extent of the involved mesentery and intestine, all of the cases were reviewed by two pathologists. Histologic reviews were conducted according to the classification system of the International Society for the Study of Vascular Anomalies (ISSVA) [10]. There are no definite criteria to distinguish morphologic types. Macrocystic types are large, smooth and multilocular structures consisting of numerous cysts that vary in size, and microcystic types exhibit a complex of many tiny cysts. Mixed cystic types are mixture of micro- and macrocystic types [11]. In this study, we defined macrocystic types as MLs composed of cysts bigger than 0.5 cm. To confirm the diagnosis of ML, the expression of D2-40, a marker of lymphatic epithelium [12], was checked in all patients, and the results were all positive for D2-40.

We divided the patients into 4 groups according to the modified classification of the operative findings. Group 1 included patients with MLs involving the intestinal walls. Group 2 patients had pedicle-type MLs with no relationship to the mesenteric vessels. Group 3 patients had MLs located in the mesenteric boundaries near the mesenteric vessels. Group 4 patients had multicentric and diffusely infiltrated MLs (Figs. 1 and 2).

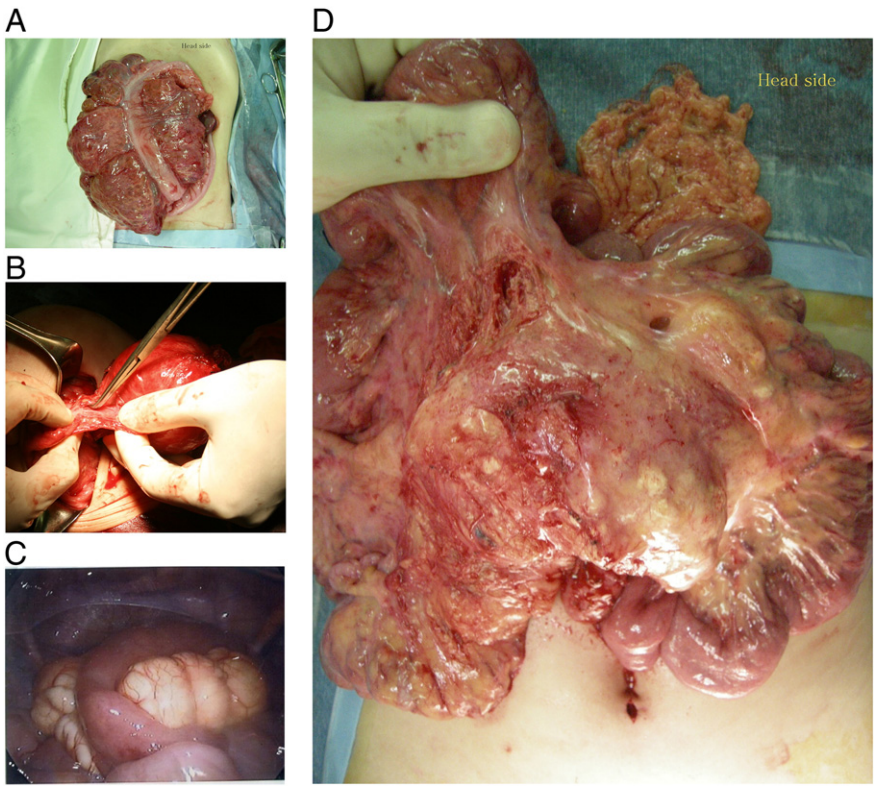

Fig. 2. Operative findings of each group: A. Group 1, B. Group 2, C. Group 3, D, Group 4.