Percutaneous Sclerotherapy for Lymphatic Malformations: A Retrospective Analysis of Patient-evaluated Improvement

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PURPOSE: To evaluate the midterm outcomes of percutaneous sclerotherapy of lymphatic malformations (LMs) as judged by patients.

MATERIALS AND METHODS: A 13-item survey questionnaire was sent to 74 patients who had undergone at least one sclerotherapy procedure in our hospital from January 1997 through January 2003. Information regarding the anatomic location, specific symptoms reported, history, treatment satisfaction, postprocedural complications, and number of treatment sessions was elicited. Four sclerosing agents (as single agents or in combination with other agents) were used: ethanol, sodium tetradecyl sulfate 3% (STS), OK-432, and doxycycline.

RESULTS: Fifty-five patients or their caregivers completed the survey. The patients' ages ranged from 6 months to 48 years at the time of the first procedure (mean, 12 y; median, 4 y). A majority of LMs were located in the cervicofacial region. The size and location of the lesion, recurrent infection, and pain were the most frequent indications for treatment. Fifty-one percent of these patients received sclerotherapy alone or in conjunction with surgery as primary treatment. Ethanol was the most common sclerosing agent used, followed by doxycycline, STS, and OK-432. Response varied with the type of LM, with 100%, 86%, and 43% of the patients reporting good to complete response for macrocystic, microcystic, and combined-type LMs, respectively. Skin blistering and ulcers were the most common complications. Permanent complications were uncommon and were largely related to ethanol use.

CONCLUSIONS: Percutaneous sclerotherapy provides effective midterm primary treatment for LMs. Treatment outcomes appear to vary according to the morphology of the malformation.

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Abbreviations: LM = lymphatic malformation, STS = sodium tetradecyl sulfate 3%

LYMPHATIC malformations (LMs) are uncommon congenital vascular dysplasias of the developing lymphatic system. Although surgical resection of such malformations is the conventional treatment, a surgical approach can be complicated by a high incidence of recurrence, infection, lymphatic leak, and local nerve damage (1). Alternative modes of treatment are therefore needed.

Sclerotherapy has been documented in the literature as an effective primary treatment modality of LMs (2–12). However, most of the published studies are flawed by relatively short-term follow-up and are largely based on the treatment of macrocystic lesions with single sclerosing agents (2–12). In these published series or case reports, macrocystic lesions appear to respond favorably compared with microcystic or combined lesions. However, we believe that the notion that sclerotherapy is an option for only large cystic spaces (13) is a misconception. In our experience, technically successful treatment can be feasible in all patients regardless of the morphology of the LM. Although percutaneous access of the large cysts in macrocystic LMs is relatively easy, especially with sonographic guidance, microcysts and even ectatic lymphatic channels are amenable to percutaneous access and sclerotherapy.

To verify and document the success of percutaneous sclerotherapy in various types of LMs, we conducted a retrospective analysis of such treatments at our tertiary care facility. Because interference with quality of life and issues such as pain and leakage are often precipitating reasons for treat-

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Table 1
Survey Respondents' Profile

Patient Group	No. of Patients (%)
All patients	55 (100)
Sex (M/F)	27/28
Age <5 years	19 (34.5)
Sex (M/F)	7/12
Age 5–20 years	27 (49.1)
Sex (M/F)	16/11
Age >20 years	9 (16.4)
Sex (M/F)	3/6

ment, this study was designed to use patient or caregiver evaluation of symptom improvement at midterm follow-up as a measure of safety and efficacy of percutaneous sclerotherapy in the treatment of LMs.

MATERIALS AND METHODS

Patient Demographics

Overall, 74 patients with LM in various anatomic locations presented during the study period. Of these, 55 patients (74%; 27 male patients and 28 female patients) completed the survey. The ages of the patients ranged from 6 months to 48 years (mean, 12 y) (**Table 1**). Eighty-four percent of respondents were aged 20 years or younger at the time they had their first procedure; one third of the patients were aged 5 years or younger. Patients were followed up for an average of 2.6 years, with a range of 2 months to 7.6 years after the last sclerotherapy session (median, 4 y).

Study Design

The study protocol for this retrospective analysis was reviewed and accepted by an institutional review board. Medical records and imaging studies of percutaneous sclerotherapy performed in all patients with LMs from January 1997 through January 2003 were reviewed.

Imaging studies before sclerotherapy were reviewed by two pediatric radiologists with experience in the diagnosis of vascular anomalies for confirmation of the diagnosis and classification by content, location, and extent of LMs. Data obtained from the medical records and imaging studies included age, sex, type, and anatomic location of the LM, number of sclerotherapy treatments, and sclerosing agents used. Subjective data related to indication, response, and complications were obtained from the survey, which was completed by the patients or their caregivers.

Morphologic Types

LMs can be classified into three relatively distinct morphologic types: macrocystic, microcystic, and combined (1). The macrocystic form is primarily composed of clinically or radiologically identifiable cysts filled with lymphatic fluid (**Fig 1**).

The microcystic type of LM is composed of dysplastic lymphatic tissue with a variable fibrous/fatty component, tiny cysts, or ectatic channels (**Fig 2**). Extensive microcystic LMs can be associated with local osseous overgrowth and deformity, airway obstruction, muscular atrophy, cutaneous vesicles, and disturbance of central lymphatic flow.

The combined form of LM is a combination of macrocystic and microcystic types. In this type, a solid soft-tissue mass is accompanied by a cystic component (**Fig 3**). Sclerotherapy may change one type of LM into another (eg, combined into microcystic).

Sclerotherapy Technique

Sclerotherapy was performed by two pediatric interventional radiologists according to previously reported techniques (13). Most procedures were carried out with the patient under general anesthesia. In general, fluid-filled spaces were cannulated with 21-gauge or 20-gauge needles with use of ultrasound (US) guidance, and lymphatic fluid was aspirated completely. A small amount of contrast medium was injected into the LM to confirm the position of the needle, then aspirated (**Fig 4**). The lesion was then filled with the same volume of the sclerosing agent under US or fluoroscopic guidance. Sclerosant volume injected was determined by estimating the volume of the cysts or observing the filling of the cysts sonographically. The sclerosing agent was generally not removed after injection. Light manual compression was applied at the puncture sites after needle removal to prevent leak of the sclerosing agent. When possible, very large cysts were drained with use of a pigtail catheter. Sclerosant was injected, allowed to dwell for 4 hours, then drained. This process was repeated daily or every other day until there was minimal or no drainage from the catheter (<10% of the original fluid volume).

For microcystic-type LMs, ectatic lymphatic channels and tiny cysts were accessed percutaneously with a small-gauge needle under US guidance. Lymphatic/chylous fluid return confirmed the position of the needle. Contrast medium injection assessed the nature of the malformation and determined how much sclerosant was necessary to inject. Aspiration of the fluid was performed carefully so as not to lose access. Under US guidance, the appropriate sclerosant was then injected while any changes in the overlying skin were monitored. The injection should be stopped immediately if abnormal change in the skin color, such as pallor or cyanosis, occurs. Lymphatic vesicles were injected directly with unopacified ethanol with use of small-gauge needles (25–30 gauge).

Dehydrated ethanol was opacified with the addition of oily contrast medium (eg, Ethiodol at a 10:2 ratio; Savage Laboratories, Melville, NY). The mixture was injected under digital subtraction fluoroscopic guidance only, with use of the roadmap technique. Contrast agent opacification of the LM was used to confirm appropriate placement of the needle and avoid nontarget injection. The use of ethanol was avoided in some special anatomic locations, such as the orbit, hand, and foot, as a result of the potentially higher risk of complication.

Sodium tetradecyl sulfate 3% (STS) may be used in foam or liquid forms. STS opacified with Ethiodol (10:2 ratio) added to the same volume of air can be "foamed" by exchanging the mixture between the syringes through a three-way stopcock. As with ethanol, this agent was injected under digital subtraction fluoroscopic or US guidance. The foam form requires less agent volume and can be clearly visualized with US because of the artifact effect of the added air.

In our facility, we commonly add 10 mL of half-strength contrast medium to 100 mg of lyophilized doxycycline powder (Doxy 100 and 200;