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Symptom resolution and volumetric reduction of abdominal lymphatic malformations with sclerotherapy

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ABSTRACT

Background: Lymphatic malformations (LMs) are congenital and arise from errors in vascular embryogenesis. LMs are categorized by cyst size as microcystic, macrocystic, or combined. Abdominal LMs are rare. Surgical resection of abdominal LMs has been the mainstay of therapy, but recurrence and morbidity are high. We sought to determine the effectiveness of sclerotherapy treatment for abdominal LM.

Methods: A single-center, retrospective review from 2014 to 2018 was conducted evaluating pediatric patients with abdominal LM.

Results: Ten patients were included, $n = 9$ had macrocystic LM and one patient had combined disease. The average age at first treatment was 6.8 y. The most common presenting symptoms were abdominal distention, pain, infection, and anemia. Preprocedural imaging was performed for all patients; median pretreatment volume was 1572.9 cm^3 (range, 67.2-13,226.4). LMs were accessed using ultrasound guidance and injected with opacified doxycycline. Patients received a mean of 7.1 sclerotherapy injections. Complications included intraperitoneal doxycycline extravasation ($n = 1$), managed conservatively, and LM infection ($n = 1$), treated with intravenous antibiotics and drainage. One patient went on to surgical resection due to inability gain stable intracystic access; follow-up ultrasonography showed no recurrence. Postprocedural imaging was available in $n = 8$. Volume decreased by 96.7% after sclerotherapy. The median remaining volume was 0 cm^3 (range, 0-599.7) ($P = 0.016$). Postsclerotherapy magnetic resonance imaging was obtained in $n = 6$, with complete resolution in 83.3%. All patients had resolution of presenting symptoms. Follow-up duration was 12.3 mo.

Conclusions: Initial results demonstrate that sclerotherapy is an effective and durable treatment for symptom resolution and volume reduction of abdominal LM.

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Introduction

Pediatric lymphatic malformations (LMs) are congenital vascular anomalies that arise due to errors during vascular embryogenesis.[1-4] LMs are rare with a prevalence of 1 in 20,000 children.[5] The most common location for an LM is the head and neck, with only 10% occurring in the abdomen.[2,5] Cystic LMs are categorized by the size of their lymphatic channels as either macrocystic, microcystic, or combined. In general, macrocystic LMs are those which can be accessed by a hypodermic needle and undergo visible decompression; however, the microcystic form comprises numerous small cysts that are difficult to enter with a needle. Combined LMs contain both macrocysts and microcysts.[2] LMs can spontaneously regress, but this is the exception as most persist and can enlarge over time.[1,2] Abdominal LMs typically present with vague abdominal complaints of pain and distention. They can also present acutely with vomiting, peritonitis, intestinal obstruction, or intestinal necrosis.[6,7] Additional complications of intra-abdominal LMs include anemia due to intracystic bleeding, sepsis due to bacterial translocation, and/or inability to empty dilated lymphatic channels, and chylous ascites.[2]

Historically, the first-line treatment for problematic LMs has been surgical resection.[3,6,8-12] Regardless of location, LMs tend to be infiltrative, involve vital structures, and are poorly demarcated.[3,13] Complete excision is difficult and can require staged interventions. Surgical principals for LM include limiting the resection to a defined anatomic area, performing as complete a resection as possible, preserving vital structures, and minimizing blood loss. Prolonged closed suction drainage is often needed for large resections involving soft tissues. Resection of intra-abdominal or mesenteric LM can require segmental bowel resection; consideration of post-surgical enteral length is essential.[8,14,15]

Expectant management of an abdominal LM is a viable option in some instances in an effort to avoid anesthesia in very young infants; however, treatment is preferred over observation to avoid the aforementioned complications.[1,5] Sclerotherapy has become an important mainstay and alternative to surgical resection for the treatment of macrocystic lymphatic malformation.[2,13,16-21] Sclerotherapy involves aspiration and subsequent injection of a sclerosant that causes scarring of cyst walls to one another. We sought to add to the collective experience for the treatment of abdominal lymphatic malformation with doxycycline, as the body of literature supporting this treatment remains sparse.[11,15,18,19,22,23]

Methods

Study population

After approval by the institutional review board with a waiver of consent, a retrospective review was conducted evaluating patients with macrocystic lymphatic malformation treated with doxycycline sclerotherapy from 2012 to 2018. The clinical records and imaging for presclerotherapy and post-sclerotherapy treatment were reviewed for age, gender, radiographic imaging, sclerotherapy intervention, hospital admissions, and follow-up information. Diagnosis of LM and classification of cyst size was based on magnetic resonance imaging (MRI) characteristics. Dimensions were calculated using width, height, and length at time of diagnosis. Follow-up data included imaging to determine presence and size of the LMs. Subjective data regarding symptom resolution was obtained from patients and families.

Technique

Preprocedural MRI was used to plan the sclerotherapy intervention. Sclerotherapy was performed by an experienced interventional radiologist. General anesthesia was administered in all cases. For the initial treatment, ultrasound guidance was used to access the macrocysts with a needle. Under fluoroscopic guidance the needle was exchanged for a pigtail catheter. Cysts were aspirated and opacified doxycycline was injected via the pigtail catheters at a concentration of 10 mg/mL. Volume injected was equal to the aspirated volume of each macrocyst, up to a typical maximum of 1000 mg per treatment. Each catheter was clamped for 8 h and left to gravity drainage after this 8 h period. The patients returned to interventional radiology for subsequent injections of sclerosant through the existing catheter during the same cycle. After a median of three sclerotherapy injections, the catheter was removed. The next cycle was initiated after 4-8 wk if imaging demonstrated the presence of continued macrocysts, or if the patient experienced a complication or increase in symptoms.

Statistical analysis

Wilcoxon matched pairs signed-rank test was used to determine if the difference between pre- and post-treatment volume was significant. The difference was considered significant when $P < 0.05$. Calculations were performed in GraphPad Prism 7.0 (GraphPad Software Inc, La Jolla, California).

Results

Ten patients underwent sclerotherapy as the primary treatment of their abdominal LM. Diagnosis for all patients was based on pretreatment imaging, MRI was used in $n = 9$ and CT was used in $n = 1$. One patient was diagnosed prenatally with ultrasonography and confirmed with postnatal MRI. Average age at diagnosis was 6.8 y (range, 0-17 y). Nine patients were male. Common presenting symptoms were abdominal distention ($n = 5$), abdominal pain ($n = 5$), infection ($n = 2$), and anemia ($n = 1$). Macrcocystic LM was identified in nine patients; one patient had a combined LM. Patient characteristics, LM type, and symptoms are presented in (Table 1).

Nine patients were treated solely with sclerotherapy. Sclerotherapy was unable to be performed in one patient due to inability to achieve a stable intracystic location after two distinct, failed attempts. This patient underwent resection of the mesenteric LM with a 5 cm segmental small intestinal