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Management of pediatric intramuscular venous malformations

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ABSTRACT

Background: Intramuscular venous malformations (VMs) are rare, but can be highly symptomatic. There are few reports on outcomes, particularly pain, functional limitations, and muscle contractures. We aimed to compare results of medical management, sclerotherapy, and surgical resection.
Methods: We retrospectively reviewed 45 patients with an extremity or truncal intramuscular VM between June 2005 and June 2015 at a single institution. Outcomes were compared between treatment modalities with ANOVA and $\chi 2$ tests.
Results: Six patients (13%) were treated with medical management, 4 (9%) with surgical resection, 23 (51%) with sclerotherapy, and 12 (27%) with both surgery and sclerotherapy. Sclerotherapy alone decreased pain in 72%. Only 20% of patients presented with muscle contracture. For these patients, 33% resolved with sclerotherapy, physical therapy, and aspirin; 22% resolved with surgery, and 45% had persistent contracture. 40% of patients treated with sclerotherapy then surgery developed new muscle contractures, compared to 4% of sclerotherapy only patients and 0% of surgery only patients ($p = 0.04$).
Conclusions: Medical management, surgery and sclerotherapy are effective treatments for intramuscular VMs. Observation and supportive care can be a primary treatment for patients with minimal symptomatology and no functional limitations. Sclerotherapy is more effective for treating pain than contractures and when used alone, rarely causes a new muscle contracture.

Venous malformations (VMs) are congenital, dysplastic vascular malformations and are the most common vascular malformation seen at vascular anomalies centers. [1] Up to 40% are intramuscular, usually in the head and neck and extremities. [2] These VMs typically present with pain and swelling [2–4], but intramuscular VM in particular can also cause significant functional limitations because of pain, mass effect, and neural involvement. [2,5–7] Furthermore, extremity and truncal VMs are more prone to enlarge and become more symptomatic as children age, particularly during puberty. [8] Consequently, although rare, these intramuscular VMs can be highly symptomatic requiring treatment, but no clear management algorithm has been defined.

Spontaneous resolution of VMs has not been described and symptomatic enlargement is common as the child ages. Multiple treatment options have been described with varying results based on lesion size, location and extent of tissue involvement. These include medical management, direct percutaneous embolization, radiofrequency ablation, sclerotherapy with various liquid and foam sclerosants, and surgical resection. [3,4,9–11] Of these, medical management, sclerotherapy and surgical resection are the mainstays of therapy, but few recent case series have been published comparing outcomes of each treatment modality. Thus, the purpose of this study was to review to our experience with multimodality treatment of venous malformations in the pediatric patients. In particular, we further sought to focus on outcomes and complications related to muscle dysfunction, which is often the most debilitating symptom of intramuscular VM.

1. Methods

With the approval of the institutional review board (CHLA-15-00279), we performed a single-center, retrospective review of all children diagnosed with an intramuscular VM of the trunk, back, or extremity between June 2005 and June 2015. VMs located on the head, neck, or hands, and subcutaneous VM were excluded. All VM were confirmed by imaging with ultrasound, CT scan, or MRI. All patients were evaluated at our Vascular Anomalies Center (VAC) by a multidisciplinary team including pediatric surgery, plastic surgery, interventional radiology, dermatology, orthopedics and hematology-oncology. To fully assess the impact of various treatment modalities, patients without any follow up were excluded.

Medical charts were reviewed for information regarding demographics, VM location, symptomatology, treatment course, clinical

Abbreviations: VM, vascular malformation; STS, sodium tetradecyl sulfate.

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improvements, postoperative complications, and length of follow up. Since VM can be highly infiltrative and diffuse, size measurements were inconsistent and not evaluated. The primary outcome measure was resolution of symptoms. Secondary outcome measures were number of treatments required, recurrence rate, and rate of muscle contracture.

Treatment plans were developed by the entire VAC team. Treatment options included medical management, sclerotherapy and surgery. Medical management included physical therapy, compression garments, braces, daily low-dose aspirin, nonsteroidal anti-inflammatory drugs and low molecular heparin or a combination thereof. Surgical treatment included a complete resection and reconstruction as needed. For sclerotherapy, Sotradecol® (sodium tetradecyl sulfate) was injected into the lesion using imaging guidance.

Complications included recurrence and development of new muscle contracture. Contractures were either managed nonoperatively with physical therapy and braces, or through surgical muscle lengthening procedures.

Patients were grouped into cohorts based on their treatment modality. Outcomes were compared between these groups with the ANOVA and chi-squared tests using GraphPad 6.0 (GraphPad Software, Inc., La Jolla, CA) software. A p-value <0.05 was considered significant.

2. Results

A total of 121 patients with extremity or truncal VM were identified. Of these, 61 were classified as intramuscular. Fifteen were excluded because of lack of follow-up and 1 was excluded after being treated with laser ablation. This resulted in inclusion of 46 patients, aged 3 months to 19 years old. Follow up ranged from 2 months to 9 years. Comparison of age, weight, gender, and length of follow up based on treatment group did not reveal any significant differences between the treatment groups (Table 1).

The most common locations were on the lower leg (45%), upper leg (28%), and upper arm (23%). Only 19% presented with a VM on the trunk. The most common presenting symptoms were pain (80%), swelling or mass (42%), and functional limitations, which were often because of pain or the location of the mass (38%). Muscle contraction was only present in 9 patients (20%) at presentation. 5 patients (11%) had an asymptomatic mass at presentation and none presented with bleeding. Duration of symptoms ranged from 1 month to 17 years, often starting from birth. There was no significant difference in duration of symptoms between treatment groups. Forty patients (87%) were initially evaluated with an MRI. Patients who had only an ultrasound or CT scan were typically referred with completed imaging and no further imaging was required.

Six patients were treated with medical management alone, including the five patients who were asymptomatic. The sixth patient actually presented with muscle dysfunction but had a type IV lesion with involvement of the deep venous system that was not amenable to any procedural intervention. All of these patients remained asymptomatic and none required any procedural intervention. Two patients were initially observed but after 1.5 and 5 years, they started to develop symptoms which prompted sclerotherapy and surgical resection, respectively.

Four patients were treated with a single surgical resection that resulted in complete resolution and no recurrence in any of these patients. Two of these lesions involved the abdominal and thoracic wall, one involved the gluteus, and one involved the gastrocnemius. For these patients, surgery was the initial treatment because the lesion's location facilitated easy excision or made sclerotherapy less likely to be successful, or for diagnostic uncertainty about the type of vascular malformation.

Twenty-three patients were treated with sclerotherapy alone. At the completion of treatment, 91% of these patients had clinical improvement after an average of 3.7 ± 3.6 procedures. Sclerotherapy was notably effective for patients presenting with pain alone, resulting in resolution of pain in 72%. The only complication noted was bleeding from the sclerotherapy site after the patient fell on the affected area. Thus, both surgery and sclerotherapy had similar rates of clinical improvement, but sclerotherapy required significantly more procedures (Table 2).

Twelve patients required both surgery and sclerotherapy. One patient had a forearm VM that was successfully resected, but then had a recurrence which was treated with 2 sclerotherapy sessions. This lesion was first resected without an attempt at sclerotherapy because its lesion made it easily excisable. The other 11 patients all had sclerotherapy first followed by surgery because of persistent symptoms. On average, 3 attempts at sclerotherapy (range 1–6) were performed before the multidisciplinary VAC team decided to pursue surgical resection. All surgeries were radical resections or excisions; no debulkings were performed. The most commonly resected muscles were the gastrocnemius (3 patients), sartorius (3 patients), and semimembranosus (2 patients). These patients experienced a similarly high rate of clinical improvement (83%) but required the highest average number of procedures (4.3 ± 1.7).

Since muscle contracture is a unique feature of intramuscular VMs, we further sought to review our experience with patients who either presented with muscle contracture or later developed postop procedural muscle contracture. Of the nine patients who presented with muscle contracture, all but one was treated with some combination of cast or brace, physical therapy, aspirin, and compression garment. Compliance with medical management was highly variable, however. Five patients were also treated with sclerotherapy alone and 4 were treated with sclerotherapy followed by surgery. Following treatment, only 2 treated with sclerotherapy and 2 treated with both sclerotherapy and surgery had resolution of their muscle dysfunction.

Post procedural muscle contracture occurred in 5 patients (11%). One patient with a VM of the lower leg developed new muscle contracture after 2 sclerotherapy sessions approximately 1 year after the first sclerotherapy session. The remaining four developed new muscle contractures only after both sclerotherapy and surgery. Thus, treatment with both sclerotherapy and surgery had a significantly higher incidence of muscle contracture (33%) compared to treatment with only one modality (4% sclerotherapy only, 0% surgery only, $p = 0.04$). Only one patient of the five patients with muscle contracture had been treated with muscle release so far with resolution of the muscle contracture. The other 4 were not candidates for contracture release and are currently being treated with medical management.

3. Discussion

Venous malformations remain a challenging clinical entity to diagnose and treat. Because of their rarity and potential for high morbidity, optimal treatment is best managed by multidisciplinary specialists. [1] At our vascular anomalies center, 88% of patients with intramuscular

Table 1
Demographics.

	Medical (n = 6)	Surgery only (n = 4)	Sclerotherapy only (n = 23)	Surgery + sclerotherapy (n = 12)	p-value
Age (y)	13.2 ± 4.0	9.5 ± 2.6	9.9 ± 5.2	10.0 ± 4.9	0.31
Weight (kg)	47.9 ± 32.4	38.4 ± 19.0	44.1 ± 26.0	46.4 ± 24.5	0.94
Gender (F:M)	2:4	1:3	13:10	6:6	0.23
Length of follow up (mo)	17.1 ± 19.4	21.6 ± 33.5	32.4 ± 22.4	42.2 ± 30.5	0.28
Duration of symptoms (mo)	102.8 ± 85.6	59.8 ± 38.4	53.7 ± 52.8	53.9 ± 49.5	0.26

Data are described as mean ± SD.