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Sclerotherapy for intramuscular vascular malformations: A single-center experience

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Abbreviations:
DIC, Disseminated Intravascular Coagulopaty
IVM, Intramuscular Venous Malformation
LIC, Localized Intravascular Coagulopathy
LMWH, Low Molecular Weight Heparin
STS, Sodium Tetradecyl Sulfate
VM, Venous Malformation

ABSTRACT

Background: Vascular malformations isolated to skeletal muscles are rare and often debilitating due to pain and very challenging to treat. Multi-modal management options include compression garments, medical therapy, sclerotherapy, and surgical resection.
Methods: A retrospective review of patients who underwent sclerotherapy for intramuscular venous malformations (IVM) between 2008 and 2016 was performed. Demographics, indications, and clinical follow-up were analyzed.
Results: Twenty patients underwent sclerotherapy for IVM. Six males and 14 females underwent 58 procedures. All patients presented with pain and were treated initially with compression garments. Median age at first treatment was 13 years (+/− 5.06 years). Initial protocol consisted of 2 sclerotherapy procedures with sodium tetradecyl sulfate (STS) within a 2–3 month interval. Median volume of the lesion was 40 cm^3 (+/− 28.7), mostly located in the lower extremities (15/20). Median number of treatments was 2 (+/− 1.95). Treatment prior to puberty resulted in a median symptom-free time of 4 years (+/− 2.18), while after puberty resulted in a symptom-free time of 2 years (+/− 2.28). Two patients had an underlying coagulopathy and were admitted for observation and peri-procedural Lovenox. No procedure related complications were noted with a median follow-up of 4 years (+/− 2.27).
Conclusion: IVMs are rare but can be incapacitating secondary to pain. Sclerotherapy is a useful minimally invasive procedure generally requiring at least two consecutive treatments. Treatment of patients prior to puberty appears to provide a more durable result, and surgical resection may be avoided.
Type of study: retrospective.
Level of evidence: IV

Venous malformations (VM) are the most common vascular malformation consisting of abnormal veins which can be present within the superficial dermis and subcutaneous tissue or infiltrating muscle, bones and joints [1]. Although already present at birth, deeper VMs are not always apparent in the first years of life, but generally become symptomatic later, presenting typically with pain and swelling [2]. Intramuscular VM (IVM) are difficult to manage due to the severity of symptoms and poor overall response to therapy in general compared to other VMs.

Multiple treatment options have been described, including medical management generally with compression and medication, percutaneous embolization and sclerotherapy and surgical resection. Sclerotherapy has emerged as the first line interventional treatment for symptomatic venous malformation. Several different sclerosing agents have been described in literature with different mechanisms of action and similar results [3]. In our institution, sodium tetradecyl sulfate (STS) is used as the agent of choice.

1. Material and methods

After institution review board (IRB: 2015–7096) approval, a single-center, retrospective review of all isolated intramuscular VM treated with sclerotherapy between 2008 and 2016 was performed. VMs with extension in the subcutaneous tissue, joint or other structure outside the muscle were excluded. All patients were evaluated at our Hemangioma and Vascular Malformation Center by a multidisciplinary team

Abbreviations: DIC, Disseminated Intravascular Coagulopaty; IVM, Intramuscular Venous Malformation; LIC, Localized Intravascular Coagulopathy; LMWH, Low Molecular Weight Heparin; STS, Sodium Tetradecyl Sulfate; VM, Venous Malformation.

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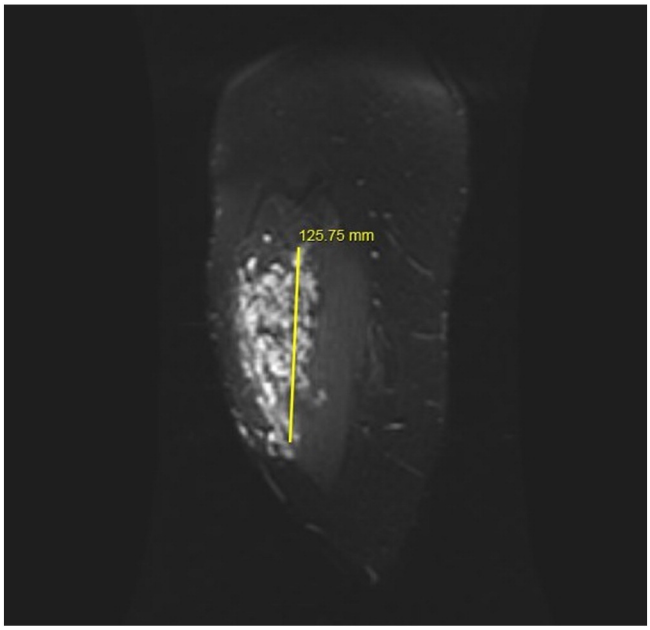

Fig. 1. MRI of an intramuscular venous malformation in the right vastus lateralis.

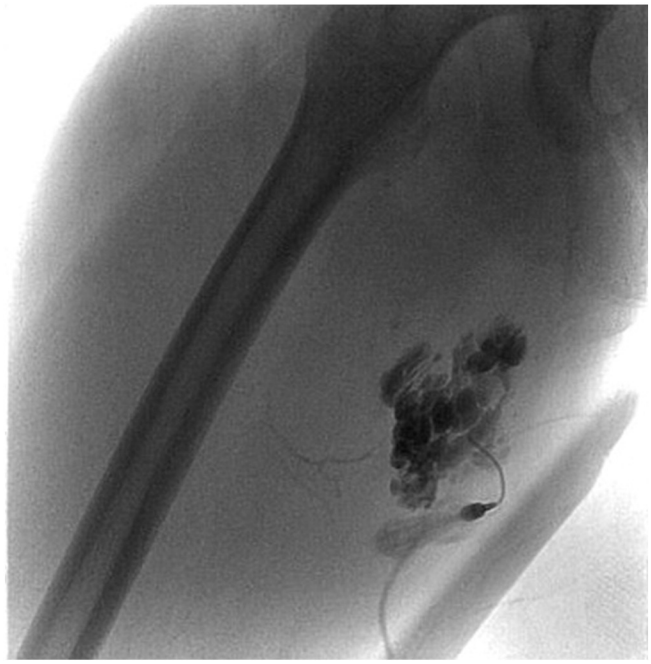

Fig. 2. Venogram of an intramuscular venous malformation in the right posterior thigh before injection of sclerosing agent.

including pediatric surgery, interventional radiology and hematology-oncology. All lesions were confirmed with a combination of both clinical examination and magnetic resonance imaging (MRI) (Fig. 1). Medical charts were reviewed for demographic variables, VM characteristics, patients' symptoms, treatment course and follow up information were gathered. When available, comparison of size was made on 2 consecutive images.

Sclerotherapy was performed by a single interventional radiologist under general anesthesia. The VM was accessed with a needle under US and fluoroscopic guidance and contrast was then injected to assess size and determine if there was significant venous outflow (Fig. 2). 3% STS was injected into the lesion using imaging guidance. The median amount of STS administered was 3 ml (+/− 2.47) and it was mixed 1:1 with air and 1:0.5 with contrast agent. Patients that did not have a coagulopathy were discharged post procedure on oral pain medication as needed. Patients with elevated D-dimers (>2) were treated with low molecular weight heparin (LMWH) in the peri-operative period (2 weeks prior and 2 weeks post procedure) and were admitted overnight for observation. All patients were instructed to continue to wear compression garments after the procedure.

Initial treatment protocol consisted of 2 sclerotherapy procedures within a 2–3 month interval. The efficacy of the treatment was determined as resolution or significant improvement of symptoms after the protocol completion.

For patients with symptomatic recurrence, the interval time was noted; if no further procedure were required, the interval elapsed from the last procedure was considered as the symptoms-free interval. For this analysis, the patients were also divided between pre-pubertal (13yo and younger) and pubertal/post-pubertal (14yo and older) based on the age at the treatment.

Imaging was generally not repeated unless further procedures were considered.

2. Results

A total of 300 patients underwent sclerotherapy for vascular malformations between 2008 and 2016 at a single tertiary care hospital. Of these, 20 patients presented with isolated IVM, 6 males and 14 females. 13 patients had venous malformation while 7 patients had mixed venous-lymphatic malformation based on radiologic imaging. These 20 patients underwent a total of 58 procedures. All patients were treated initially with compression garments and only underwent sclerotherapy when consistently worn compression did not improve symptoms. The median age at first treatment was 13 years (+/− 5.06 years) (Table 1).

The most common location was the lower extremity (15/20, 8 in the thigh musculature and 7 were below the knee), 4 were located in the upper extremities and 1 in the chest wall. The most common presenting symptoms were pain (20/20), swelling (5/20) or mass (5/20). The malformation significantly affected activities of daily living in 7 out of 20 patients. The median volume of the lesions prior to the treatment was 40 cm3 (+/− 28.7). Median number of treatments for each patient was 2 (+/− 1.95). When the treatment was performed before puberty, it resulted in a median symptom free time of 4 years (+/− 2.18) while treatment after puberty resulted in a symptom free time of 2 years (+/− 2.28). 2 patients had coagulopathy with baseline elevated D-dimers: as per protocol were admitted for observation and were treated with peri-procedural LMWH. No procedure related complications were recorded with a follow-up of median 4 years (+/− 2.27). No patients went on to require surgical resection due to failure of sclerotherapy.

3. Discussion

Intramuscular venous malformations pose both a diagnostic and treatment dilemma. Although present at birth, it is not unusual to diagnose an IVM only when it causes pain which often occurs during puberty. Growth pattern of these lesions is not linear and is influenced by environmental factors. Trauma, infection and hormonal stimulation, such as puberty, pregnancy or oral contraceptive pills, can accelerate the growth and development of symptoms [4].

Trauma and infection are noted to cause a localized inflammatory reaction within the venous malformation and can often incite symptoms. Several hormone receptors have been noted within the endothelium of vascular anomalies. Maclellan et al. demonstrated an increased expression of follicle-stimulating hormone receptors [5]. Duyka et al. also confirmed the presence of progesterone receptor in vascular malformations suggesting that progesterone may play a role in