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Lymphangioma Circumscriptum: Treatment Modalities for this Unyielding Condition

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Abstract

Lymphangioma circumscriptum is a superficial lymphatic malformation, presenting as lymph-filled micro-to-macroscopic vesicles. It is oftentimes disfiguring and can negatively affect one's quality of life due to complications such as infection, pain and lymphorrhea, making it of high value to elucidate current treatment options for the condition. High recurrence rates and a risk of scarring make lymphangioma circumscriptum difficult to treat. A broad literature search was performed using PubMed in January 2017 to compile all available published articles that studied treatments for lymphangioma circumscriptum. Incidentally, the treatment options that have been described for the condition are primarily restricted to case reports and limited case series, rendering need for further large, randomized-controlled studies to accurately assess their efficacy and safety. Our correspondence consolidates information about the most commonly used treatment options for LC, many of which are noninvasive, minimize complications, and result in improved aesthetic outcomes. More specifically, we describe of surgical excision, laser therapy, sclerotherapy, cryotherapy, radiotherapy, electrocautery, electrodessication and more recently, imiquimod in treating lymphangioma circumscriptum. The majority of the described treatments provide palliative care, with only resection being used as a definitive treatment for lymphangioma circumscriptum. Nevertheless, most of the therapeutic options facilitated clearance of lymphangioma circumscriptum with minimal adverse effects.

Lymphangioma circumscriptum (LC), or microcystic lymphatic malformation, is a rare superficial lymphatic malformation involving the skin and, at times, subcutaneous tissue and muscle. The condition presents as grouped micro-to-macroscopic vesicles filled with lymph and oftentimes blood [1,2]. Treatments improve cosmetic outcomes, but also control complications including refractory rupture, lymphorrhea, hemorrhage, infection, and pain [3,4]. Herein, we describe treatments that have been implicated for LC, albeit with varying outcomes, including surgical excision, lasers, sclerotherapy, cryotherapy, radiotherapy, electrocautery, electrodessication, and imiquimod. These treatments have been described in case reports and case series, indicating need for further large and controlled studies proving efficacy and safety. Surgical excision remains the mainstay of treatment for LC, for it removes sequestrated lymphatic cisterns in the subcutaneous plane and has the lowest reported rate of recurrence (17%) [5,6]. Nevertheless, since certain LC lesions may be unresectable, and surgery may result in disfiguring scars, hematomas, infections, and nerve injuries, there is need for alternative treatments.

Lasers, mainly the pulse dye laser (PDL) and CO_2 laser, have been implicated in the treatment of LC with varying results. The high-energy of PDL targets the chromophore hemoglobin (585-595 nm) within blood vessels and minimizes collateral damage to surrounding tissues. Effectiveness for LC may be limited due to the minimal hemoglobin content in lymph vessels. Lai et al. described significant improvement of LC with minimal scarring and no oozing or bleeding after treatment with a flash lamp-excited PDL. Unfortunately, recurrence was noted three years after treatment [7]. Railan et al. reported the successful treatment of two cases of LC with PDL, one of which also recurred after three years [8]. Karadag et al. described regression of LC after five sessions at four-week intervals of PDL treatment combined with five sessions of double freeze-thaw cycles of cryotherapy at three-week intervals with no recurrence after two years. Cryotherapy uses very low temperatures to cause vasoconstriction immediately followed by reactive vasodilation resulting in cellular necrosis. This, in combination with laser therapy, may have a synergistic effect for LC [6]. Emer et al. failed to observe clinical change in an LC lesion after two treatments with PDL delivered two weeks apart [6].

CO_2ablative lasers have a much longer wavelength than PDL, effectively vaporizing water within tissue and fusing lymphatic channels [6]. Although many case reports and series have achieved successful results with the CO_2 laser, especially in large, deeply-involved lesions, patients may require anesthesia, and the laser carries risk of scarring and post-inflammatory hyperpigmentation [8].

Sclerotherapy is a simple, safe, quick, and inexpensive method of managing LC over the long-term. Ideal sclerosing agents should produce panendothelial destruction and lack systemic toxicity. Further, they should produce an inflammatory reaction causing fibrosis and ultimately vessel lumen obliteration. Al Ghamdi et al. achieved a 70% reduction in the size of an LC lesion after two sessions of intralesional 1% sodium tetradecyl sulfate (STS) [2]. No side effects were reported at a one-year follow-up [2]. Similarly, Chang et al. injected an LC lesion with 0.5-1% STS seven times monthly until all lesions cleared. Adverse effects were mild but included post inflammatory hyperpigmentation and atrophic scars [9]. Lastly, Bikowski et al. attained complete resolution of LC with minimal postinflammatory erythema and hyperpigmentation after a single injection of 23.4% hypertonic saline solution [3].

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For large or unresectable lesions with limited treatment options, radiotherapy is an alternative treatment. Denton et al. described the effective management of two cases of LC which were unsuitable for excision with radiotherapy [10]. Nevertheless, the role of radiotherapy remains controversial as cases have been reported in which LC developed as a complication of previous radiotherapy sessions.

Emer et al. successfully used electrodessication, a treatment which causes protein denaturation through electrical pulses, for LC. Three sessions at one-month intervals showed smoothing and a decrease in the number of superficial vesicles in addition to a reduction in bleeding, oozing and pain [6]. Additionally, Yang et al. reported the use of electro electrocoagulation in the treatment of LC with a special isolated needle that is able to penetrate the skin to treat the superficial and deep components of LC. Only the needle tip is electrically conductive; the isolated portion of the needle protects the epidermis from electrical damage. Twelve patients were treated, all of which achieved at least a 90% reduction in lesion size with minimum side effects. Only one case was complicated by ulceration, which ultimately resulted in a large scar [4].

Imiquimod is another potential noninvasive treatment for LC. Wang et al. successfully treated LC in two patients with marked lesion regression without recurrence after two-to-three year follow-up. Imiquimod stimulates cell production of endogenous interferons and interleukins. While the interferons can inhibit vascular motility and invasion, the interleukins inhibit endothelial proliferation and formation. Thus, imiquimod appears to treat LC by inhibiting vessel formation and inducing endothelial cell apoptosis [1]. Bikowski and colleagues, however, did not successfully treat LC with imiquimod and found that lesions had actually become more prominent following treatment [3].

Laser therapy, sclerotherapy, cryotherapy, radiotherapy, electrocautery, electrodessication and imiquimod have been used as palliative therapies for lymphangioma circumpscriptum, although only surgical is deemed a definitive treatment. Nevertheless, local recurrences tend to occur with surgery, and adequate excision of the lymphangioma may prove difficult. Further, certain lymphangiomas, especially those that are large, are not suitable candidates for resection. Lasers such as the PDL and CO_2 laser have demonstrated efficacy, but also run the risk of recurrence and adverse effects. Further, sclerotherapy is a simple, quick, and cheap method of clearing LC with only mild adverse effects. Cryotherapy may prove beneficial for treating LC when combined with the PDL laser, and radiotherapy should be preceded with caution, for several cases actually noticed the novel development of LC following radiotherapy sessions. Both electrocoagulation and electrodessication may be valuable treatments for LC, although electrocoagulation has been associated with adverse effects. Lastly, imiquimod may prove promising; the case reports that used the treatment, however, are not in accordance with its efficacy. As stated previously, case reports and limited case series primarily described the mentioned treatments, rendering need for additional large, randomized-controlled clinical studies to accurately establish a safety and efficacy profile. Nevertheless, the noted studies provide a valuable basis for such prospective studies, and introduce various therapeutic options that may successfully treat LC in clinical practice.

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