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Surgical and non-surgical treatment modalities for lymphangioma circumscriptum

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To the Editor:

We have read, with great interest, an article published in the *Dermatology Online Journal* by Heller et al. entitled "Lymphangioma circumscriptum" [1]. In this paper, the authors report on a 54-year-old-man with lymphangioma circumscriptum (LC). Although this paper sheds light on clinical and histological nuance, it is also important for physicians to be aware of the literature regarding various treatment modalities for LC to educate patients on the decision-making process. This letter is written to discuss the various surgical and non-surgical treatment options for LC.

The literature has described surgical excision as the primary treatment modality for LC [2]. The excision should extend to subcutaneous tissues and the deep fascia to ensure complete removal of abnormal hypertrophied lymphatic cisterns which serve as conduits of recurrence. A cohort of 158 patients with LC treated with surgical excision by Flanagan et al. demonstrated a cure rate of 75% with re-excision curing an additional 12% [3]. In situations where complete excision would be too disfiguring, there has been some evidence that lipectomy is a less-invasive surgical approach with therapeutic benefit [4]. Shrestha et al. report on two patients who received therapeutic lipectomy; however, one of these patients had a recurrence of her LC within 6 months [4].

Although excision has empirically been the primary treatment modality for LC, preliminary data demonstrated that non-surgical procedures may be efficacious in cases of LC not amenable to surgical resection. Niti et al. reported 10 cases of LC treated with radiofrequency ablation followed by sclerotherapy and demonstrated that 9 of 10 patients exhibited near-complete clearance [5]. In contrast, Okazaki et al. reported that in their cohort of 128 LC patients that sclerotherapy was not as effective as surgery and deemed it only useful in single cystic and macrocystic subtypes of LC [6]. Eliezri et al. reported three patients in whom CO_2 laser was effective in resolving symptoms of LC with excellent cosmesis [7]. In contrast, Huilgol et al. described two patients with vulvar LC, both of whom had recurrence despite treatment with CO_2 laser [8]. Lastly, electrocoagulation therapy has also demonstrated therapeutic promise as Yang and colleagues demonstrated near-complete clearance in 12 patients in their cohort who were treated with this modality [9]. While promising, it is clear that there is much discrepancy regarding the efficacy of sclerotherapy and laser ablation due to the small sample sizes in each study. Although further study is needed, it is prudent to consider these modalities in situations where the patient is surgery-averse or not a surgical candidate.

Despite the promise of newer procedural technology, it is also important for treating physicians to be aware of medical management of LC for patients who are not suitable candidates for invasive therapy or decline these interventions. In a case series of two patients, Wang et al. successfully

used topical imiquimod to treat LC [10]. Imiquimod induces the production of interferons and interleukins which may inhibit vessel formation while inducing endothelial cell apoptosis. There is also data to support the use of propranolol for the treatment of LC. Ozeki and colleagues published their experience using propranolol to treat a patient with diffuse lymphangiomatosis which resulted in significantly reduced drainage volume [11]. Furthermore, Leboulanger et al. published a case report in which propranolol was used to minimize hemorrhage of a lymphangioma on a patient's tongue [12]. Lastly, bleomycin has also been shown to be effective in treating LC. Chakravarti and colleagues reported two patients with LC of the tongue who were successfully treated with bleomycin [13]. Medical management of LC has the least supporting data as it is derived from limited case series. Nevertheless, these options may be the only recourse for select patients and warrant further research.

The management of LC should be evaluated on a case-by-case basis. Although surgery has been shown to be the most definitive treatment option, there are situations in which less invasive treatment modalities may prove to be efficacious, especially on cosmetically sensitive areas or large surface areas not amenable to resection. As of now, the data for less invasive procedures and medical management are limited to case series and small cohorts. However, there seems to be therapeutic benefit with these modalities. We hope that this letter will help physicians discuss the various treatment options with their patients. Awareness of all treatment modalities will aid in clinical decision-making and will result in better care for our patients.

Potential conflicts of interest

The authors declare no conflicts of interest

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