Successful Treatment of Acquired Vulvar Lymphangiectasia with 1% Polidocanol Sclerotherapy

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Acquired lymphangiectasia, also known as lymphangioma circumscriptum, of the vulva is a rare malformation characterized by dilated superficial lymphatic channels within the papillary dermis. Clinical presentation consists of clusters of thin-walled vesicles containing clear to serosanguinous fluid. Occasionally, warty hyperkeratosis may be present, leading to a verrucous appearance. Acquired lymphangiectasia of the vulva may arise in inflammatory conditions, such as Crohn's disease, in the setting of intralymphatic metastases, or following surgery, lymphadenectomy or radiotherapy. Symptoms, including pain, frequent drainage and recurrent infections, may motivate patients to seek treatment. Numerous therapeutic modalities have been utilized, including surgery, electrocautery, cryosurgery, and laser resurfacing. However, there is a lack of consensus regarding the optimal treatment approach. We report here a case of acquired lymphangiectasia of the vulva that was treated successfully with 1% polidocanol sclerotherapy.

CASE REPORT

A 70-year-old woman presented for evaluation of vesiculopapular lesions on the left vulva, of approximately 3 years' duration. She reported that the lesions would fluctuate in size and occasionally bleed. Her past medical history was remarkable for cervical squamous cell carcinoma treated with radical vulvectomy, lymph node dissection, chemotherapy and radiotherapy 10 years previously. Physical examination revealed clusters of translucent to haemorrhagic papulovesicles along her left vulva and inguinal fold (**Fig. 1**a). Punch biopsy showed dilated vascular spaces containing lymphatic fluid within the papillary dermis, confirming the diagnosis of lymphangiectasia (**Fig. 2**). A total of 3 ml intralesional 1% polidocanol solution was administered to the affected areas during a single session. Approximately 20 sites were injected with 0.1–0.2 ml polidocanol per injection site. Four weeks later the patient reported improvement in her symptoms, and physical examination revealed near-complete resolution in the treated areas. Six months later the patient returned for treatment of newly appearing lesions across the bilateral vulva and 3 ml intralesional 1% polidocanol solution was administered to the affected areas with subsequent improvement (Fig. 1b).

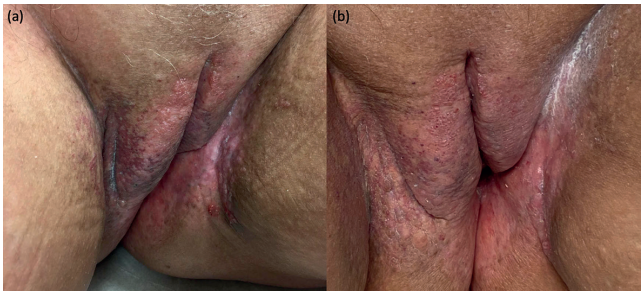

Fig. 1. (a) Clusters of translucent to haemorrhagic papulovesicles along the left inguinal fold prior to treatment with polidocanol sclerotherapy. (b) Significant improvement with few residual papulovesicles following 2 sessions of polidocanol sclerotherapy.

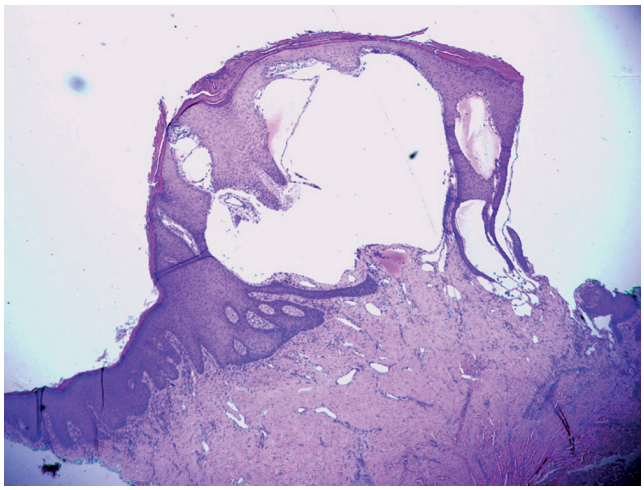

Fig. 2. Dilated, thin-walled vascular spaces containing lymphatic fluid within the papillary dermis (haematoxylin & eosin ×40).

DISCUSSION

Treatment of vulvar lymphangiectasia is challenging, and several therapeutic approaches have been proposed. Surgical excision has historically been the treatment of choice, but may require extensive resection, leading to undesirable functional and cosmetic outcomes. In addition, failure to adequately address deeper lymphatic cisterns may result in recurrence (1, 2). Reported rates of post-surgical recurrence range from 23.1% to 25% (2). Alternative approaches, including laser therapy, have been utilized with some success. Fully ablative carbon dioxide laser resurfacing has been shown to be effective by vaporizing superficial tissue and sealing lymphatic channels. However, use is limited by the need for prolonged recovery and risks, including infection and scarring. Ablative fractional erbium:yttrium aluminium garnet laser has been proposed recently as a more tolerable, yet effective, alternative (3). Available data regarding prevalence and outcomes of various therapeutic approaches are limited. In a retrospective study of 34 patients with acquired vulvar lymphangiectasia, 26.5% were treated with surgery and electrocoagulation, 20.6% were treated with surgery alone, 2.9% were

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treated with electrocoagulation alone, and 50% were untreated. Recurrence developed in 52.9% of treated patients (1).

Sclerotherapy is a minimally invasive, well-tolerated, cost-effective modality that may be underutilized in the treatment of acquired lymphangiectasia. A variety of sclerosing agents are available and can be divided into 3 classes based on their mechanism of action. Hyperosmotic agents (hypertonic saline or hypertonic saline with dextrose) cause endothelial damage through dehydration. Chemical irritant detergents (chromated glycerin, polyiodinated iodine) function as corrosives. Detergent sclerosants (polidocanol, sodium tetradecyl sulphate, and sodium morrhuate) cause vascular injury by altering the surface tension around endothelial cells. Some of these agents, including hypertonic saline, sodium tetradecyl sulphate and bleomycin, have been used for treatment of non-vulvar lymphangiomas with good outcomes (4). In addition, OK-432 (picibanil), a sclerosant, containing group A *Streptococcus pyogenes* combined with benzylpenicillin, was used to treat vulvar lymphangioma circumscriptum in a paediatric patient. A total of 5 sessions were administered at monthly intervals, resulting in near-complete clearance (5). In the current case, polidocanol was selected due to favourable tolerability and safety profiles. However, other sclerosing agents may have similar efficacy.

Of note, some reports have described good outcomes with a combination approach involving sclerotherapy and radiofrequency ablation (6). These modalities may work synergistically to address superficial and deep lymphatic channels resulting in reduced risk of recurrence. In a case report, a patient with acquired vulvar lymphangiectasia was treated with a single session of radiofrequency ablation and 3% polidocanol sclerotherapy, followed by 3 sessions of sclerotherapy alone. The patient remained free of recurrence at a 2-year follow-up visit (6). This multimodal approach may be useful in extensive or refractory cases; however, sclerotherapy alone may achieve desired outcomes in patients with focal areas of involvement. Finally, as demonstrated in the current case, 1% polidocanol can achieve favourable outcomes, while theoretically carrying less risk for skin necrosis compared with a 3% concentration.

In summary, we propose that 1% polidocanol sclerotherapy be considered for treatment of acquired lymphangiectasia of the vulva. This approach may be particularly well-suited for patients who are not surgical candidates or wish to avoid the need for prolonged recovery.

The authors have no conflicts of interest to declare

REFERENCES

1. Simon L, Trévidic P, Denis P, Vignes S. Vulvar lymphangioma circumscriptum: comparison of primary and acquired forms in a cohort of 57 patients. J Eur Acad Dermatol Venereol 2018; 32: e56–58.
2. Yoon G, Kim H-S, Lee Y-Y, Kim T-J, Choi C-H, Kim B-G, et al. Clinical outcomes of primary surgical treatment for acquired vulvar lymphangioma circumscriptum. Arch Gynecol Obstet 2016; 293: 157–162.
3. Makdisi J, de Feraudy S, Zachary CB. Vulvar lymphangioma circumscriptum treated with fractional ablative erbium:yttrium aluminium garnet laser. Dermatol Surg 2018; 44: 1149–1151.
4. AlGhamdi KM, Mubki TF. Treatment of lymphangioma circumscriptum with sclerotherapy: an ignored effective remedy. J Cosmet Dermatol 2011; 10: 156–158.
5. Ahn S-J, Chang S-E, Choi J-H, Moon K-C, Koh J-K, Kim D-Y. A case of unresectable lymphangioma circumscriptum of the vulva successfully treated with OK-432 in childhood. J Am Acad Dermatol 2006; 55: S106–S107.
6. Khunger N. Combination technique of radiofrequency ablation with sclerotherapy in acquired lymphangiectasis of the vulva. J Cutan Aesthet Surg 2009; 2: 33–35.