CASE REPORT

Lymphangioma circumscriptum of vulva successfully cured with sclerotherapy

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ABSTRACT

Sclerotherapy is used in venous malformations and hemangiomas, as well as chronic venous insufficiency and hemorrhoids.

Objective: To report on the usefulness of sclerotherapy in the treatment of unusual vascular lesions, such as lymphangioma circumscriptum.

Methods: A 42-year-old female clinically and histopathologically proved as lymphangioma circumscriptum on the vulva treated with five sessions of an intralesional injection of 1% sodium tetradecyl sulfate (sclerotherapy).

Results: After this treatment, there was no more discharge and a reduction in the lesion's size by 90%.The patient's lesions were almost cleared with the sclerotherapy treatment.

Conclusion: Sclerotherapy can be recommended as an effective treatment method for unusual vascular lesions, such as lymphangioma circumscriptum.

KEY WORDS: Lymphangioma circumscriptum (LC), vulva, sclerotherapy, sodium tetradecyl sulfate

INTRODUCTION

Lymphangioma circumscriptum (LC) is a rare benign skin disorder involving hamartomatous lymphatic malformation of deep dermal and subcutaneous lymphatic channels. Fluid-filled vesicles that contain lymphatic fluid are typically seen in patients with LC. Vulval LC is very rare and may present as a congenital condition or, rarely, it might develop secondary to radiotherapy, infection, or surgery. Several medical and abrasive treatment modalities are available including laser therapy. Surgical resection is the most commonly used method to treat vulval LC but there are high rates of recurrence. Other treatment modalities with success are intra lesional sclerotherapy using doxycycline or picibanil (OK-432) and vaporization by carbon dioxide laser. Here we report the case of a patient who presented with multiple papular grouped vulval swelling which was clinically misinterpreted as an infectious disorder. This clinical misdiagnosis points to the fact that histopathology is necessary even if clinically the gynaecologist suspects as an inflammatory disorder. The patient had no significant past medical or surgical history and the cause of her rare vulval LC remained unidentified. This unusual case was treated with a sclerosant, sodium tetradecyl sulfate. The patient was almost cleared with several treatments of sclerotherapy. She is still being followed-up on a regular basis.

CASE REPORT

A 42-year-old woman attended our Medical College Hospital with multiple skin coloured papular

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lesions on the vulva. The lesions had developed when the patient was 35 years old. There was no preceding history of local trauma. She had no other organ complaints. The lesions were increasing gradually with the time. On examination, there were multiple verrucous, coalescent papules on the vulva (Fig. 1). There was some drainage of fluid from the lesions. No significant excoriation, crusting was observed. The remaining genital and physical examination was normal. Routine laboratory tests including complete blood count and differential WBC counts were within normal limits. Erythrocyte sedimentation rate was 20 mm/hr. Mantoux test was negative and X-ray chest was normal. The differential diagnoses suggested, based on clinical presentation included genital warts, lymphangioma circumscriptum and cystic hygroma. A biopsy specimen was obtained from the lesions and histopathological examination revealed focal parakeratosis, focal acanthosis and elongation of the rete ridges. In the papillary dermis, there were cystic proteinaceous fluid-filled spaces. Also noted in the sections was superficial perivascular lymphohistiocytic infiltrate (Fig. 2). The diagnosis of LC was entertained. The patient was treated initially with topical and systemic antibiotic which led to flattening of the surface of the lesions. Later on, we treated the patient with five sessions of sclerotherapy, with which the patient showed good improvement. (Fig. 3).

Fig. 1 Multiple verrucous, coalescent papules on the vulva.

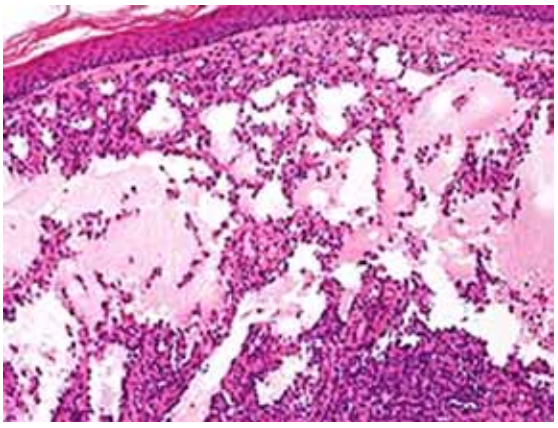

Fig. 2 Cystic protienaceous fluid filled spaces of Lymphangioma circumscriptum

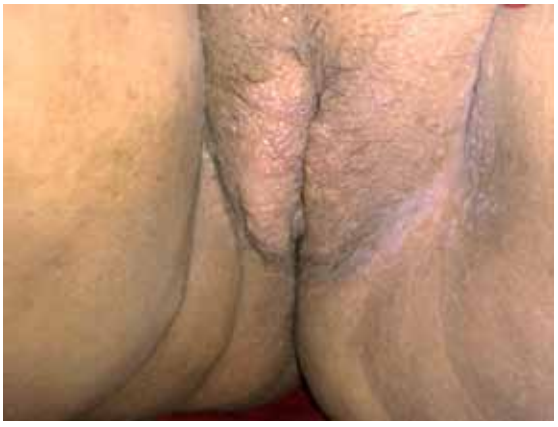

Fig. 3 Significant improvement after sclerotherapy treatment.

DISCUSSION

Natural resolution of LC is rarely seen, and surgical excision has long been the preferred treatment option.[1-3] However, the complications encountered with surgical excision, include infection, nerve and vital organ injury, episodic bleeding and significant functional and cosmetic deformity, as well as the need for hospital admission and the difficulty of complete excision with a relatively high recurrence rate of up to 25%, have led many physicians to consider other less invasive procedures for the treatment of LC.[4]

CO_2 laser ablation has been used with good results but with significant scarring.[2] The use of pulsed dye lasers (PDL) has shown efficacy; however, it is costly and cannot be done in every case because it is dependent on the presence of a chromophore, hemoglobin.[2] Thus, patients with LC with clear fluid-filled vesicles might not benefit from PDL.

Surgical excision is the definitive treatment for lymphangioma circumscriptum.[4] Magnetic Resonance Imaging may be used to define the extent of the lesion and minimize recurrence rate.[5] Other destructive techniques used with variable success include hypertonic saline sclerotherapy, carbon dioxide laser, pulse dye laser/intense pulse light system and radiotherapy.[6-10] Propranolol has been used to treat a patient with diffuse lymphangiomatosis associated with chylothorax and significantly reduced the drainage volume.[11] In another report, propranolol was used to successfully minimize hemorrhage of a lymphangioma that was located on the tongue.[12] However, determination of propranolol's efficacy in the treatment of cutaenous lymphangioma will require evaluation through clinical trials.

The idea of treating lymphangiomas with sclerosing substances is not new. This type of treatment involves the injection of sclerosing substances to eliminate abnormal blood vessels by inducing local endothelial destruction extending to the entire adventia with minimal thrombus formation, which can induce an inflammatory reaction leading to fibrosis and eventually to obliteration of the vessel lumen.[13] The procedure of sclerotherapy is simple, quick, cheap, does not require general anesthesia or a hospital admission, and is not associated with scarring. Sclerosing agents are classified into three groups on the basis of chemical structures and the mechanism of injury to the endothelium: hyperosmotic agents (e.g., hypertonic saline), detergent sclerosants (e.g., polidocanol and STS), and chemical irritants (e.g., polyiodinated iodine).[13] The side effects of sclerosing agents are minimal and include hyperpigmentation, telangiectatic matting, cutaneous ulcerations, necrosis, superficial thrombophlebitis, and arterial injection.[14] Hypertonic saline sclerotherapy has shown good results in treating LM.[14] OK-432 (picibanil) sclerotherapy has been reported by many investigators to be an effective mode of treating microcystic lymphangiomas with equally good initial and long-term results.[15] Bleomycin, an antitumor agent discovered in 1966, has been used as a sclerosant agent in the treatment of LM and induced the complete disappearance of treated lesions in 45-60% of the cases in most series; the success rate reached 80% on including partial responders.[16-18] A literature search revealed a common recurrence of LC after surgical excision and failed to find any detailed information about recurrence after treatment with the other modalities including sclerotherapy.[19]

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