

# Lymphangioma circumscriptum of the vulva treated with radiofrequency ablation: case report

## Radyofrekans ablasyonla tedavi edilen vulvar lenfanjioma sirkumskriptum: Olgu sunumu

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### Abstract

Lymphangioma circumscriptum (LC) is a rare benign skin disease involving hamartomatous lymphatic malformation of the deep dermal and subcutaneous lymphatic ducts. Upon occurring in uncommon areas such as axilla, shoulder, groin, and vulva, it is a therapeutic challenge for the dermatologist. Various methods such as surgical excision, lasers, and sclerotherapy have been used in the past to treat this unsightly skin condition. In this article we report the efficacy of a radiofrequency ablation in a female patient with vulvar LC. The treatment efficiency of radiofrequency was satisfactory in our patient without recurrence during the 6-month follow-up period. Based on previous studies, radiofrequency technique is a safe and cost-effective treatment for LC management, as in our case.

**Key words:** radiofrequency, lymphangioma, hamartomatous malformation

### Özet

Lenfanjiyoma sirkumskriptum (LS), derin dermal ve subkutan lenfatik kanalların hamartomatöz lenfatik malformasyonu ile giden nadir benign bir deri hastalığıdır. Aksilla, omuz, kasık ve vulva gibi farklı alanlarda meydana geldiğinde, dermatologlar için terapötik bir zorluk yaratır. Geçmişte hoş olmayan bu cilt görünümünü tedavi etmek için cerrahi eksizyon, lazerler ve skleroterapi gibi çeşitli yöntemler kullanılmıştır. Bu makalede vulvar LS'li bir kadın hastada radyofrekans ablasyonunun etkinliğini bildiriyoruz. Radyofrekansın tedavi etkinliği, hastamızda 6 aylık takip süresi boyunca nüks olmaksızın tatmin ediciydi. Önceki çalışmalara dayanarak, radyofrekans tekniği, bizim vakamızda olduğu gibi, LS yönetimi için güvenli ve uygun maliyetli bir tedavidir.

**Anahtar kelimeler:** radyofrekans, lenfanjiom, hamartomatöz malformasyon

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## Introduction

Lymphangioma circumscriptum (LC) is a hamartomatous formation consisting of enlarged lymph channels surrounded by lymphatic endothelium. It is considered a malformation, not like a neoplasia.<sup>1</sup> The lesions are usually seen as typical fluid-filled vesicles containing lymphatic fluid. While it can be seen anywhere on the body, it usually occurs in the proximal limb, shoulder, and neck, genital involvement is very rare.<sup>2</sup> The most common type of LC usually occurs at birth and in childhood, rarely in adulthood.<sup>3</sup> Clinical manifestations consist of many vesicles that may be pink to copper colored, red or black if secondary bleeding is present. These vesicles contain lymph and are often likened as “frog’s spawn”.<sup>4</sup> LC is not life-threatening but may cause psychosexual and cosmetic problems in patients due to swelling, bleeding, pain, xeroderma, eczema and infection. In differential diagnosis, it may be confused with herpes zoster, molluscum contagiosum, genital verruca, leiomyoma, cellular angiofibroma, angioblastoma and aggressive angiofibroma.<sup>5</sup> Various medical and ablative treatment modalities are available. Since deeper subcutaneous cysternal component, treatment is also difficult because of the persistent nature of the disease. Here we report a female patient, who presented with vulvar swelling which was clinically misinterpreted as a malignant disorder.

After this misdiagnosis was excluded histopathologically, the patient was diagnosed as LC of vulva and successfully applied radiofrequency ablation.

## Case report

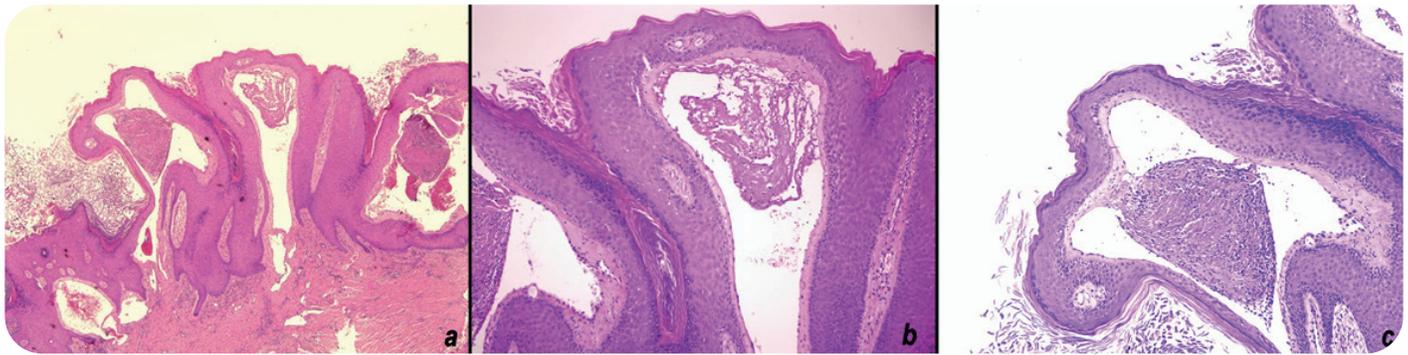
A 55-year-old female was presented to our clinic with the complaint of extremely itchy vulvar lesions. The lesions had developed when the patient was 36 years old. She was suspected as vulvar carcinoma and referred to the gynecology department for surgical therapy. On the other hand, she refused surgery, because she hoped that her disease could be improved by topical or systemic drugs and consulted in our clinic for biopsy. Dermatological examination revealed oedema and multiple, grouped, oozing vesicles on the mons pubis involving both labia majora and minora (Fig. 1a-b). The lesions had slowly grown in number and size over the years, and there was a lymphedema in the lower both limbs that appeared three years before the vulval lesions. She had no history of sexually transmitted disease, surgery, and radiotherapy. General physical examination and gynecologic examination were normal. She had normal Pap smear, normal laboratory work-up including blood biochemistry and serology for sexually transmitted diseases. A biopsy specimen was obtained from the lesions and histopathological examination revealed epidermal orthokeratosis and acanthosis, numerous dilated lymphatics and angiomatous



**Fig. 1a-b.** Oedema and multiple, grouped, oozing vesicles on the mons pubis involving both labia majora and minora, **1c.** Application radiofrequency ablation

spaces with proliferation of capillaries (Fig. 2a-c). A clinical diagnosis of LC was made and the patient was given a systemic antibiotics clindamycin 600 mg and ciprofloxacin 750 mg twice daily for seven days. Radiofrequency ablation was done a week later in cut and coagulate mode with wire loop electrode, under local anesthesia. The patient was advised to clean the area with antiseptic lotion and apply topical antibiotic fusidic acid on the lesions. All the lesions were successfully

des”, later Morris first used the term “lymphangioma circumscriptum” in 1889. The exact etiology of this malformation is unknown.<sup>7</sup> Lymphangiogenic growth factors, vascular endothelial growth factor-C (VEGF-C) and VEGF-D and their receptors on lymphatic endothelial cells, VEGF-3, may play a role in mechanisms controlling the development of LC. LC is clinically characterized by thin-walled, translucent vesicles, most commonly seen in the axilla, chest, mouth and tongue.<sup>8</sup>



**Fig. 2a-c.** Epidermal orthokeratosis and acanthosis, numerous dilated lymphatics and angiomatous spaces with proliferation of capillaries (H&E,x100)



**Fig. 3a.** After 30 days, **3b.** After five months of treatment with radiofrequency ablation

ablated at first session (Fig. 1c). A month later, the lesions completely healed and swelling subsided. There were no side effects of the treatment. Patient was called for monthly follow up. No recurrence has occurred in the last five months and the patient is still under surveillance (Fig. 3a-b).

## Discussion

Lymphangioma circumscriptum was first described in 1878 by Fox and Fox, who called it “lymphangiocy-

Vulvar LC is usually asymptomatic, but sometimes itchy, burning is accompanied by pain. Swelling and itching were the most important symptoms for our patient.

Histopathological findings classically shows enlarged lymphatics in the epidermis and papillary dermis. Generally they are lined by smooth endothelial cells and contain red and white blood cells.<sup>9</sup> Vulva is a rare region for lymphangioma placement. Vulvar LC can be congenital or acquired. Congenital forms of this tumor occur at birth or before the age of 5. Acquired forms usually occur in older women secondary to radical hysterectomy, pelvic lymphadenectomy, or post-cervical cancer radiotherapy that can damage the lymphatic system.<sup>10</sup> In our case, the lymphedema of the lower limbs may have played a pathogenic role in the appearance of the vulval lesions, but we did not find any cause of the lymphatic obstruction.

There is no standard treatment for the management of LC. Among the most common treatments are abrasive

therapy, sclerotherapy, electrocoagulation, and surgical resection.

In addition, surgical excision may not always be possible due to its disruptive effect to normal anatomy of the vulva, especially when large areas of the vulva are involved. Other treatment methods include electrocautery, CO<sub>2</sub> and Er:YAG laser and sclerotherapy.<sup>11</sup> Even with best treatment option, recurrence is common. In order to treat LC, we applied radiofrequency ablation, which provides almost complete clinical ablation with coagulation of lesional and perilesional skin causing fibrosis of perivesicular lymphatics. Although the large cluster of vesicles dissolved to a light pink atrophic scar, the result was satisfactory for the patient. We followed up our patient for five months, which showed no recurrence. This successful experience in our patient demonstrates the versatility and effectiveness of radiofrequency surgery.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

#### References

1. Erdogan AG, Balaban D, Koc K. Vulvar lymphangioma circumscriptum: a case report. *Turkderm* 2005;39:54-56.
2. Haroon S, Hasan SH. Lymphangioma circumscriptum in the scrotum: a case report. *J Med Case Rep* 2012;6:233.
3. Lapidoth M, Ackerman L, Amitai DB, Raveh E, Kalish E, David M. Treatment of lymphangioma circumscriptum with combined radiofrequency current and 900 nm diode laser. *Dermatol Surg* 2006;32:790-4.
4. Bikowski JB, Dumont AM. Lymphangioma circumscriptum: treatment with hypertonic saline sclerotherapy. *J Am Acad Dermatol* 2005;53:442-4.
5. Harwood CA, Mortimer PS. Acquired vulval lymphangioma mimicking genital warts. *Br J Dermatol* 1993;129:334-6.
6. Sachdeva S. Lymphangioma circumscriptum treated with radiofrequency ablation. *Indian J Dermatol* 2011;56:77-8.
7. Kudur MH, Hulmani M. Extensive and invasive lymphangioma circumscriptum in a young female: a rare case report and review of the literature. *Indian Dermatol Online J* 2013;4:199-201.
8. Sinha A, Phukan JP, Jalan S, Pal S. Lymphangioma circumscriptum of the vulva: report of a rare case. *J Midlife Health* 2015;6:91-3.
9. Whimster IW. The pathology of lymphangioma circumscriptum. *Br J Dermatol* 1976;94:473-86.
10. Jappe U, Zimmermann T, Kahle B, Petzoldt D. Lymphangioma circumscriptum of the vulva following surgical and radiological therapy of cervical cancer. *Sex Transm Dis* 2002;29:533-5.
11. Khunger N. Combination technique of radiofrequency ablation with sclerotherapy in acquired lymphangiectasis of the vulva. *J Cutan Aesthet Surg* 2009;2:33-5.