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OLGU SUNUMU - CASE REPORT

# AN UNUSUAL CASE OF LYMPHANGIOMA CIRCUMSCRIPTUM OF THE VULVA

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**ABSTRACT:**

Lymphangioma circumscriptum (LC) is a benign disorder of the lymphatic channels that rarely occurs in the vulva. Here, we report a case of LC of the vulva that occurred 11 years after surgery and radiotherapy for endometrial stromal sarcoma. The case was managed surgically.

**Key Words:** Lymphangioma Circumscriptum, Vulva, Postoperative Radiotherapy

**VULVASINDA LENFANJİOMA SİRKUMSKRİPTUMU OLAN NADİR BİR OLGU**

**ÖZ:**

Lenfanjioma sirkumskriptum (LS) vulvada nadir olarak görülen, lenfatik kanalların benign hastalığıdır. Burada, endometriyal stromal sarkom nedeni ile cerrahi tedavi ve radyoterapi alan ve 11 yıl sonra vulvasında LS ortaya çıkan bir olguyu rapor ettik. Olgu cerrahi olarak tedavi edildi.

**Anahtar Kelimeler:** Lenfanjioma Sirkumskriptum, Vulva, Postoperatif Radyoterapi

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## INTRODUCTION

Lymphangioma circumscriptum (LC) is a benign disorder of the lymphatic channels that rarely occurs in the vulva. It is clinically characterized by thin-walled, translucent vesicles concentrated in groups most commonly located on the axilla, chest, mouth, and tongue. It may be congenital or acquired. The complications include a clear exudate, secondary infections, and minor hemorrhages, and a case of squamous cell carcinoma on lymphangioma lesions has been described[1].

The English language medical literature review since 1960 revealed that there have been 17 cases of congenital and 26 cases of acquired LC, including the one reported in this article[2-9]. In this report we present an acquired case of LC following surgery and radiotherapy for endometrial carcinoma.

## CASE REPORT

A 54-year-old woman presented to our centre in January 2005 with the complaint of profuse vulvar itching of three months' duration. She had been treated with radical gynecologic surgery and had received postoperative radiotherapy for endometrial stromal sarcoma 11 years before. On clinical examination there was thick subcutaneous tissue containing gray, coalescent, hyperkeratotic 1 to 5 mm vesicular papules (Figure1), marks of excoriation were detected, and no oozing was observed. The other findings on the genital and physical examination were unremarkable. Biopsy of the lesion revealed epidermal acanthosis, hyperkeratosis, and dilatation of lymphatic channels in the papillary dermis (Figure 2). She was diagnosed with acquired LC. No residual malignant gynecological disease was found and an ultrasonographic evaluation of the abdomen and pelvis revealed no abnormalities. The lesions were surgically excised by simple vulvectomy with wide excision. After 3 days of follow-up in the hospital she was discharged without any complications.

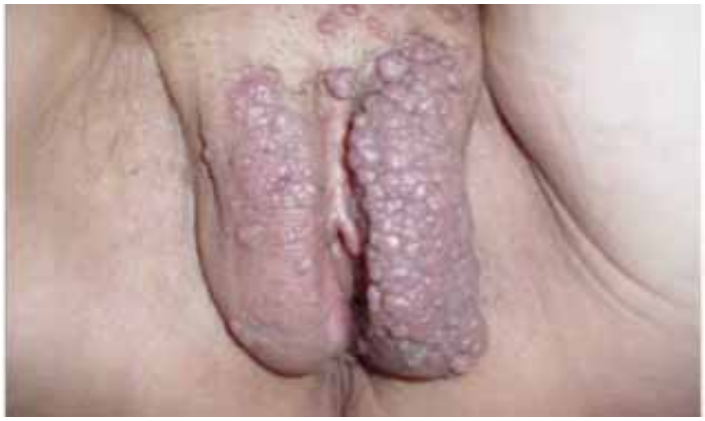

**Figure 1.** Gross view of the lesion.

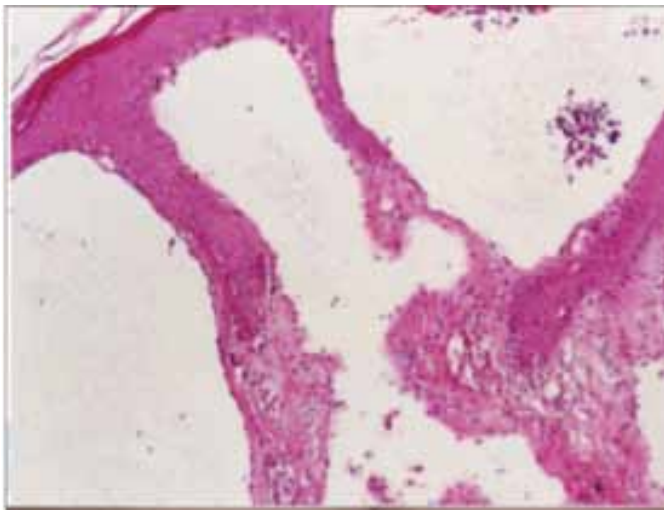

**Figure 2.** Subepidermal multiloculated cystic spaces (H&E, X200)

## DISCUSSION

The specific etiology of LC is not known; however, the underlying pathology is thought to be the altered lymphatics that form cisterns, which communicate with the skin and form vesicles[3]. It seems likely that the surgical procedure and radiotherapy cause the occlusion of the lymph vessels in the pelvis and, due to increased hydrostatic pressure within vessels proximal to the lymphatic obstruction, LC develops.

The vulva is a rare location for LC and this uncommon pathology poses a diagnostic challenge, especially in the presence of hyperkeratosis, which makes it difficult to distinguish between genital warts and some infectious processes as molluscum and leads to improper treatment. The presenting symptoms may be pain, lymphorrhea, itching, and recurrent cellulitis caused by excoriations accompanying the lesions, which are typically multiple persistent translucent vesicles. However, the morphologic characteristics may range from papules and nodules to much firmer hyperkeratotic lesions and it may be difficult to distinguish it from cutaneous metastasis of cervical carcinoma. For this reason histopathologic confirmation is mandatory.

Among the many treatment modalities, like observation, electrocoagulation, radiotherapy, cryosurgery, argon laser surgery, and carbon dioxide laser, surgical extirpation appears to be the best option with the lowest recurrence rates[2]. Vlastos et al. recommended the creation of a database of these cases so that different treatment modalities may be compared[2]. Recently intralesional sclerotherapy with OK-432 (Picibanil; Chugai Pharmaceutical, Tokyo, Japan) became a new therapeutic approach for macrocystic lesions[5]. In our case we surgically removed the involved vulvar parts by simple vulvectomy with wide excision. On the other hand, Ghaemmaghami et al. recently reported that major labiaectomy seems to be a more successful method than the others[4].

In conclusion, the differential diagnosis of vulvar lesions that occur after surgical and radiologic cancer therapy should include LC. Surgical excision is still the preferred mode of treatment for LC, and recurrence is a common problem.

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