



Case report

Lymphangioma circumscriptum of the vulva: A case report and review of literature

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ABSTRACT

Lymphangioma circumscriptum (LC) is a rare skin disorder of lymphatic channels in deep dermal and subcutaneous layers. Typically fluid filled vesicles that contain lymphatic fluid with frogspawn-like appearance are seen in any part of body. Vulvar LC is seen as congenital condition or might develop secondary to radiotherapy or surgery. Variety of medical and abrasive treatment modalities are present including carbon dioxide laser. Surgical treatment is the most commonly used method to treat vulvar LC. In this report we present surgically treated vulvar LC in a 50-year-old women with three-year follow-up.

Key words: *Lymphangioma circumscriptum, vulva, labium majus, surgical treatment.*

Introduction

Lymphangioma circumscriptum (LC) is a rare benign disorder involving the deep dermal and subcutaneous lymphatic channels [1]. Typical clinical appearance is fluid filled septated vesicles that contain lymphatic fluid. LC may be seen in any part of the body. Most common locations are proximal part of limbs that contains extensive lymphatic network (axilla, shoulders, groins, buttocks, perineum) [2].

In this case we report a case of vulvar LC treated with surgical excision.

Case

A fifty-year-old woman was admitted to our hospital with the complaint of extremely itchy vulvar lesions. On physical examination, there were multiple vesicles grouped into textures resembling frogspawn particularly on the left side of the vulva and broad swelling of the vulva involving both labia majora and minora from the mons pubis to the perineal raphe (**Figure 1A**). These fluid-filled vesicles had been first appeared 15 years before she admitted to our hospital and these lesions had progressed in size very slowly till last year. Especially itching complaint had been increased last year. She has been taking oral antidiabetic drugs for last five years because of type 2 diabetes mellitus. 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 workup including blood biochemistry and serology for sexually transmitted diseases.

A diagnostic punch biopsy was performed. The histopathological examination revealed the diagnosis of vulvar LC. Wide local excision to take out all affected areas was performed. Histopathological examination confirmed the punch biopsy result (**Figure 1B**) typically showing thin walled fluid filled vessels. After three years patient is free of complaints (**Figure 1C**).

Discussion

Lymphangioma circumscriptum (LC) is term used for a lymphatic malformation which can be found anywhere in the body; on the skin surface, intraperitoneal or extraperitoneal region or in bone but the commonest sites are shoulders, axillary folds, proximal parts of the extremities, flanks and perineum [3, 4] and can present at any age. "Capillary lymphangioma", "lymphangiectasia", and "dermal lymphangioma" are other terms sometimes used to denote the same entity [4].

Vulvar LC might be congenital or acquired [5]. Youngest age of presentation was 3 years for congenital form [6].



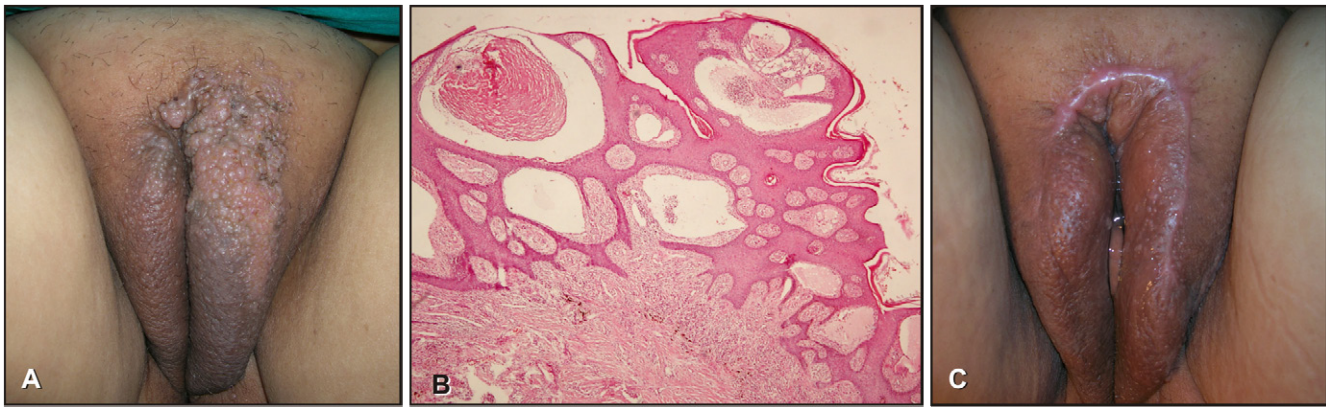


Figure 1. **A.** Before surgical treatment. **B.** Low magnification view shows focal elevations of epidermis due to underlying thin walled dilated lymphatic vascular channels that cause clinical polypoid appearance. Dermis shows mononuclear inflammatory cell infiltration with fibrosis (HE) **C.** Three years after surgical diagnosis.

Most common cause of acquired form is surgery and radiotherapy especially for cervical carcinoma [7-9]. Interestingly, radiotherapy was also used for treatment [10].

Correct diagnosis of LC is important due to similar clinical presentation with infectious processes to prevent inappropriate treatment [11-14]. If associated with significant hyperkeratosis clinical presentation may mimic condyloma acuminata [12, 13]. The management of LC is not standardized and involves only observation, surgical excision (local excision, simple or extended vulvectomy), and abrasive modalities (carbon dioxide laser, liquid nitrogen, 5-fluorouracil, electrocoagulation, or sclerosing therapy) [4, 15-20].

There is no approved medical treatment for vulvar LC and primary alternative to surgery is carbon dioxide laser ablation [21, 22] which is efficient and patient compliance is high, but recurrences are seen with this therapy as occurred with surgery. However, carbon dioxide laser ablation may lead to pain, aggravation of number of vesicles and keloid formation [8]. Furthermore, this procedure is expensive, necessitate specialty and not easily available in developing countries neither did our hospital [23]. Sclerotherapy with OK-432 (Picibanil; Chugai Pharmaceutical, Tokyo, Japan) is a new medical treatment especially effective in macrocystic lesions [24].

Currently, surgery is an effective and well-tolerated treatment for vulvar LC in most women [25]. It is efficient, well tolerated, and easily applicable. Lesion recurrence is major problem but surgical resection can be repeated with confidence [25]. Incomplete excision is the most common reason for recurrence after surgical treatment. In order to prevent recurrence complete excision of involved areas is important. Involvement of tissues as shown by magnetic resonance examinations is generally wider and deeper compared to clinical assessment [26]. Therefore, excision should include all subcutaneous tissues down to fascia [15].

Our patient, after three years, has no signs or symptoms of recurrence.

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