Acquired lymphangiectasia: a rare mimic of genital warts

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Abstract

Acquired lymphangiectasia of the vulva is very uncommon. Owing to the non-specific papillomatous manifestation and the vast array of possible differential diagnoses, lymphangioma circumscriptum (LC) still presents a diagnostic challenge. In this report, we present a very rare form of acquired vulvar LC in a 71-year-old patient with a longstanding history of asymptomatic lesions over the labia majora that had been previously treated as genital warts. On examination, the patient had multiple clustered translucent papules up to 15mm in diameter, morphologically reminiscent of vesicles, that oozed clear fluid throughout her groin and swollen labia majora. The patient also suffered concomitant bilateral lower-extremity lymphedema. A skin biopsy showed multiple, irregular-shaped lumina containing eosinophilic material in the upper dermis. Dilated lymphatic channels were lined by a single layer of flattened endothelial cells and the overlying epidermis showed acanthosis, hyperkeratosis, focal mild pseudoepitheliomatous hyperplasia. There is still no consensus on the optimal management of LC. Our patient was referred to a plastic surgeon for further evaluation and treatment. Although there are a variety of therapeutic modalities for LC, positive results are few and relapses are observed.

Keywords: acquired lymphangiectasia, lymphangioma circumscriptum, vulva

Introduction

Acquired lymphangiectasia, also known as acquired lymphangioma circumscriptum (LC), is a rare entity characterized by dilated superficial lymphatics that occur as a consequence of damage to previously normal lymphatic channels [1]. The most prominent clinical appearance of acquired lymphangiectasia are numerous small elevated spawn-like papules with clear discharge [1,2]. This variety is most often localized to the axillary, inguinal, and genital areas and there is often coexisting lymphedema [2]. Owing to the non-specific manifestation and the vast array of possible differential diagnoses, LC still presents a diagnostic challenge. In this report, we present a very rare form of acquired vulvar LC.

Case Synopsis

A 71-year-old woman presented with a history of multiple asymptomatic lesions over the vulva that initially developed on her right inguinal fold and subsequently slowly spread to the right labia majora. She previously failed a course of cryotherapy and podophyllotoxin for possible external genital warts. The patient had some improvement, but later presented with recurrence and noted increasing size and number.

She had a medical history of cervical carcinoma surgery, lymphadenectomy, and post-operative

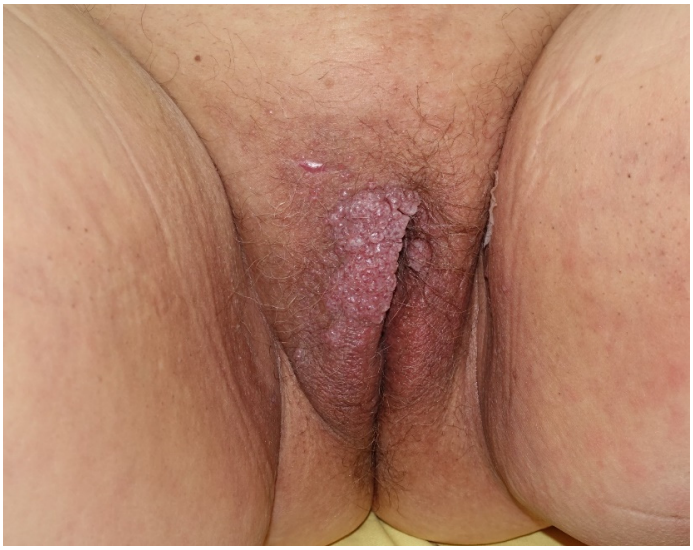

Figure 1. *Vulva with extensive involvement of the right labia majora by a lesion consists of multiple clusters of translucent vesicles that contain clear lymph fluid.*

radiotherapy more than 25 years prior. Following her cancer treatment she also developed a persistent, bilateral lower-extremity lymphedema.

On examination, the patient had multiple clustered translucent papules that oozed clear fluid throughout her groin and labia majora, morphologically reminiscent of vesicles. Lesions measured up to 15 mm in diameter. The labia minora were swollen and tender (**Figure 1**). The inguinal lymph nodes were not enlarged. Diffuse thickening of both feet and legs was observed along with profound, non-pitting edema.

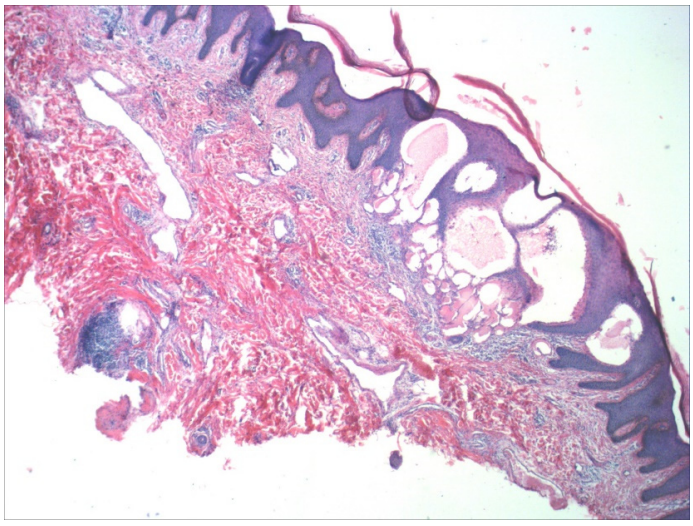

Figure 2. *Histology: multiple, irregular-shaped lumina containing eosinophilic material in the upper dermis. Dilated lymphatic channels were lined by a single layer of flattened endothelial cells. The overlying epidermis showed acanthosis, hyperkeratosis, and areas of mild pseudoepitheliomatous hyperplasia. H&E, 100×.*

A skin biopsy from a papulovesicle on the vulva was performed. The histopathologic findings showed multiple, irregularly-shaped lumina containing eosinophilic material in the upper dermis. Dilated lymphatic channels were lined by a single layer of flattened endothelial cells. The overlying epidermis showed acanthosis, hyperkeratosis, and areas of mild pseudoepitheliomatous hyperplasia (**Figure 2**). Based on the correlation of the history, lesion morphology, and histopathologic findings, a diagnosis of acquired lymphangiectasia was established.

After a thorough education of the patient about the nature of her disease she was referred for a gynecological and oncological consultation to rule out underlying malignancy. She was then referred to a plastic surgeon for further evaluation and treatment. Nevertheless, she refused any surgical intervention and unfortunately did not return for follow-up.

Case Discussion

Acquired lymphangiectasia of the vulva is very uncommon. It is usually caused by a disruption of the lymphatic vessels related to surgery, trauma, radiation, or infection. The mean time of initial presentation of lesions after cancer therapy of the genitalia and lymph node dissection is around 7-15 years [1,2]. Obesity is the most dominant risk factor for development of lymphangiectasia, following lymphadenectomy and radiation therapy [3].

Owing to its papillomatous nature, the primary conditions in the differential diagnosis includes condyloma accuminata, squamous cell carcinoma, and molluscum contagiosum, as was the case with our patient. The clinical appearance could also lead to the misdiagnosis of other conditions like infected Bartholin cyst, epidermoid cyst, herpesvirus infection, or extramammary Paget disease. Hence, biopsy is often warranted [4,5]. A notable condition in the differential diagnosis is angiokeratoma [6].

Reports of vulvar LC include asymptomatic [7] pruritic [8], burning [9] or painful [8] variations. Similarly, other than cosmetic defects, our patient was asymptomatic for the whole course of her consultation and therapy.

There is still no consensus on the optimal management of LC. If symptomatic, excision of the affected lymphatic channels may be considered. Other therapeutical modalities include vaporization with CO_2 laser, cryotherapy, and superficial radiotherapy [10-12]. Owing to the rarity of the vulvar cases, there is no reported recurrence rate after vulvectomy, quite a radical solution that would rarely be considered. Yet, there is a 23.1% recurrence rate reported after surgery for all cases of cutaneous LC, ranging from 6 to 81 months follow up period [13]. Cellulitis, lymphorrhea and lymphedema are common complications. Moreover, dyspareunia and psychosexual difficulties can also occur. Additionally, in long-standing cases, squamous cell carcinoma can rarely occur [14].

Conclusion

Owing to its course of progression, time of development, and papillomatous appearance, acquired vulvar lymphangiectasia often bestows a diagnostic challenge. A lesional biopsy is the golden standard for diagnosing LC. Although there are an array of therapeutic modalities for LC, results are often poor and relapses are commonly observed. Hence patient education and compliance is prudent for reasonable therapeutic outcomes.

Potential conflicts of interest

The authors declare no conflicts of interests.

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