Acquired Lymphangioma Circumscriptum Of The Vulva And Chronic Hidradenitis Suppurativa: A Case Report

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Citation

R Leiria Gomes, A Calhau, R Salgueiro Neto, T Freitas, H Gaspar, I Oliveira, L Farinha, M Cardoso. *Acquired Lymphangioma Circumscriptum Of The Vulva And Chronic Hidradenitis Suppurativa: A Case Report*. The Internet Journal of Gynecology and Obstetrics. 2023 Volume 27 Number 1.

DOI: 10.5580/IJGO.56526

Abstract

Lymphangioma circumscriptum is a form of lymphangioma characterized by benign dilation of lymphatic channels, which affects the skin and subcutaneous tissues. Lesions usually are grouped translucid, thin-walled vesicles filled with a clear liquid, resembling a “frog spawn”. The condition is usually congenital but can be acquired. Vulvar involvement is uncommon and only a few cases have been reported. It represents a diagnostic and management challenge because it can be confused with condyloma acuminata, molluscum contagiosum, or other vulvar disorders. Treatment options include observation, surgical excision, laser ablation, or sclerosing therapy. We present a case of acquired vulvar lymphangioma circumscriptum in a woman with hidradenitis suppurativa, since this association is rare.

INTRODUCTION

Lymphangioma circumscriptum (LC) is a benign lymphatic malformation characterized by dilation of lymphatic vessels in the skin and subcutaneous tissue. These abnormal lymphatic malformations do not communicate with the normal lymphatics [1,2]. The exact cause of LC is unknown. It can be congenital or acquired due to damage of lymphatic vessels of various etiologies [1,2]. The chest, mouth, axilla, and tongue are the commonly affected sites while the vulva is an infrequently encountered site for LC [1,2]. Physical examination typically demonstrates vesicles in the pilosebaceous and suprapubic regions of the vulva, which rarely exceed 2 cm in diameter [3]. These vesicles contain lymph, so usually are filled with clear colorless fluid but occasionally can be red or black if secondary bleeding is present. The physical appearance resembles “frog spawn” or “cobblestone” [3, 4]. Compared to those arising in nongenital areas, lymphangiomas of the vulva and genital areas are more hyperplasic, possibly due to the loose connective tissue, with a warty appearance and may easily be misdiagnosed as genital warts or molluscum contagiosum [5]. LC is not life-threatening and the clinical presentation is highly variable, ranging from being asymptomatic to a highly disabling condition. Discomfort, itching, burning, pain, and lymph oozing are the most frequent symptoms [6]. The diagnosis is usually made by recognizing the characteristic clinical appearance and confirmed by biopsy [3]. Various methods of treatment of LC have been described, including observation, surgical excision, laser therapy, and sclerosing therapy; however, local recurrence may occur [3].

Till date, only a few cases of vulvar LC have been reported in the literature. We intend to report a case of vulvar LC in a 54-year-old female with hidradenitis suppurativa (HS), since this is an uncommon association.

CASE PRESENTATION

A 54-year-old woman was referred to vulvar pathology’s appointment in the gynecology department of our hospital after being observed in the primary healthcare system. The major complains were follicular lesions involving the vulvar and inguinal regions that were compatible with HS, enlargement of the right labia minora, pruritus, vulvar pain, and the appearance of verrucous lesions in the past year. The patient was treated with topical clindamycin solution for HS. From her personal and obstetrical history, she had psychiatric disorder treated with antidepressants and two cesarean deliveries. She had no history of sexually

transmitted disease, radiotherapy, malignancies, or other predisposing conditions of LC. She was obese (BMI 31,8) and had smoking habits of 7 cigarettes per day. Menopause was at the age of 50-year-old. On physical examination, there were visible multiple lesions of HS on the mons pubis and inner tights, a 4-centimeter polypoid lesion of the right labia minora, and multiple grouped hyperkeratotic papules with semitranslucent vesicles in the lower third of the right labia majora and the anterior commissure of the labia (Figure 1 and 2).

Figure 1

Evidence of lesions of hidradenitis suppurativa mainly in pubic area, polypoid lesion of the right labia minora and hyperkeratotic papules with semi-translucent vesicles in the lower third of right labia majora and the anterior commissure of the labia.

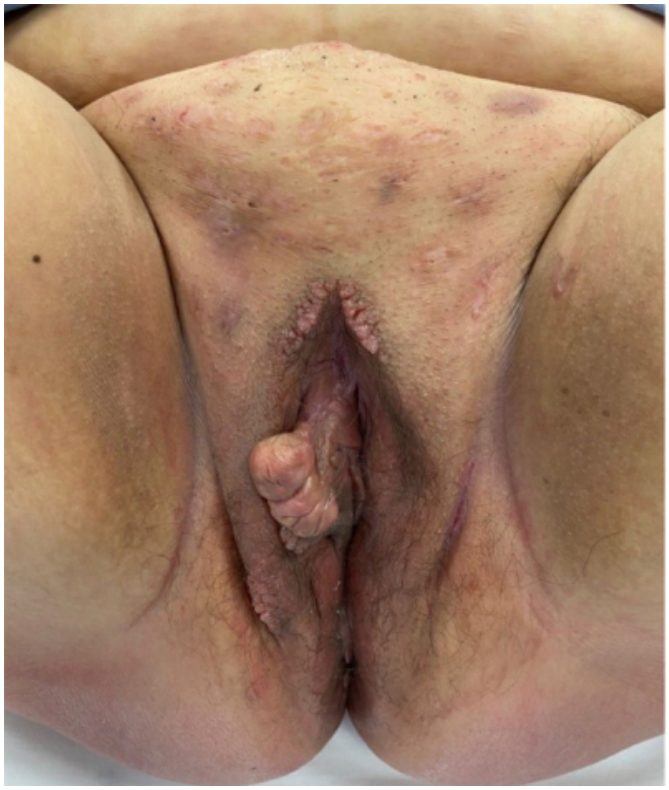
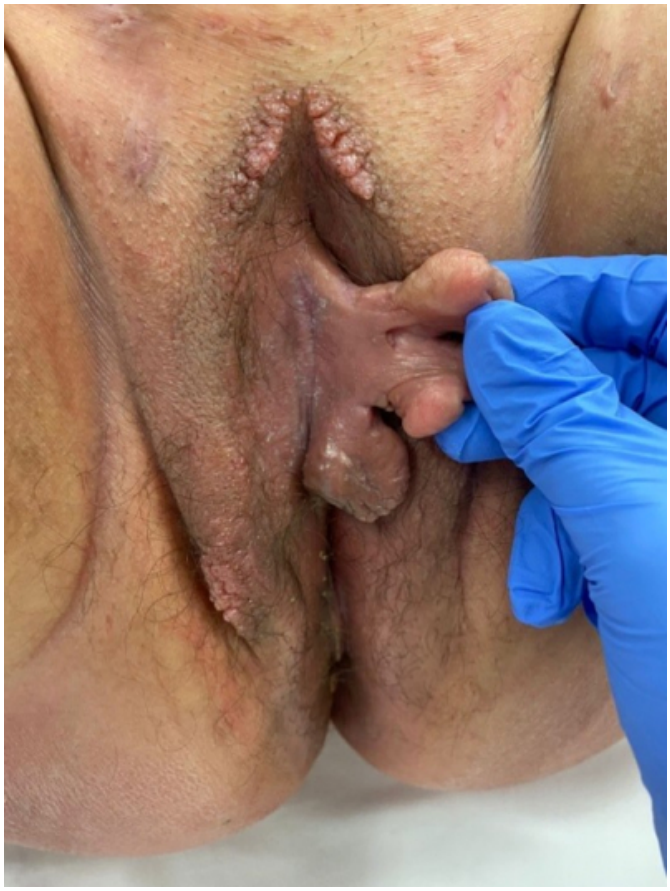


Figure 2

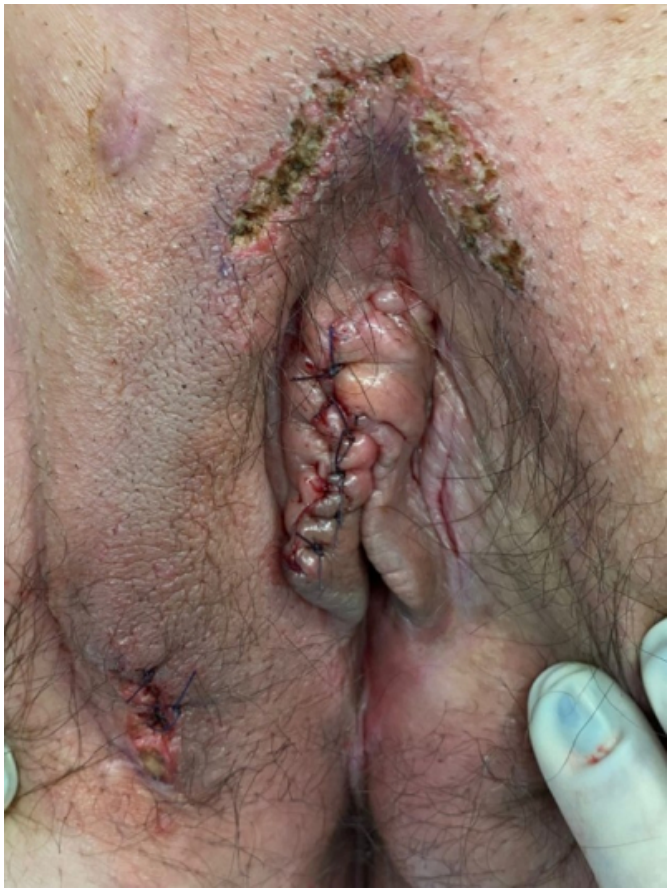
Evidence of lesions of hidradenitis suppurativa mainly in pubic area, polypoid lesion of the right labia minora and hyperkeratotic papules with semi-translucent vesicles in the lower third of right labia majora and the anterior commissure of the labia.



The diagnosis of LC was suspected by recognizing the characteristic clinical appearance and the patient was proposed for surgical excision of the polypoid lesion and excisional biopsy of the presumptive LC lesions. Cervical cytology and laboratory analysis performed before surgery had no alterations. During surgery, oozing of clear fluid from the vesicles was evident, which corroborates the diagnosis of LC. The lesions were superficially removed, fulgurated, and sent for pathology evaluation (Figure 3).

Figure 3

After surgical removal of the polypoid lesion and excision of the presumptive LC lesions.



Histopathological examination demonstrated a fibroepithelial polyp of the right labia minora and biopsy of the lesions revealed acanthosis and hyperkeratosis of the epidermis with numerous dilated lymphatic channels containing eosinophilic proteinous material in the epidermis and papillary dermis (Figure 4). The immunohistochemical study confirmed the lymphatic origin (poloplanin (D2-40) +, CD34 -) (Figure 5 and 6).

Figure 4

Histopathological section of the lesion showing large vascular channels in the epidermis and papillary dermis, containing eosinophilic proteinous material (H&E, 100x).

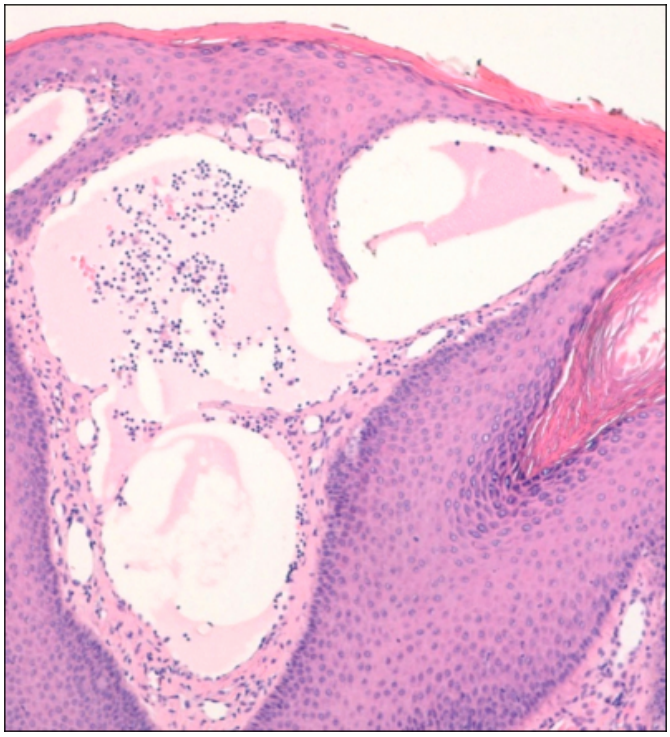


Figure 5

Immunohistochemistry confirms the lymphatic origin of the vascular channels (D2-40+, CD34-).

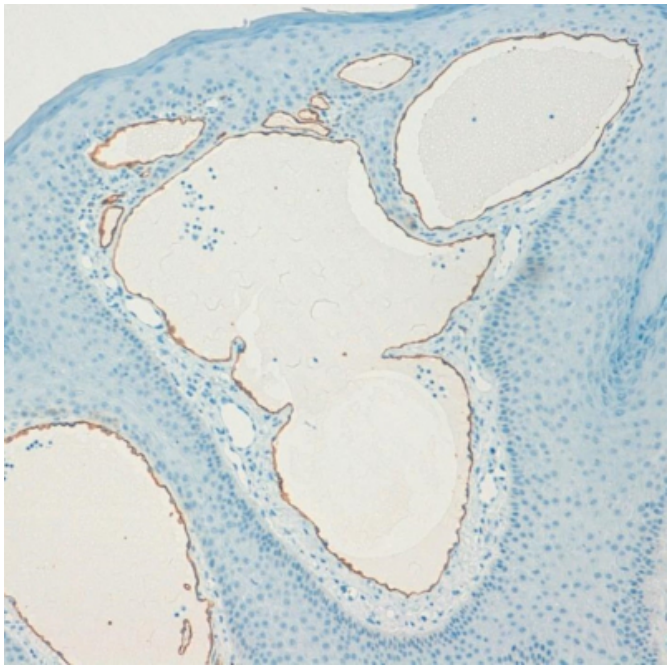
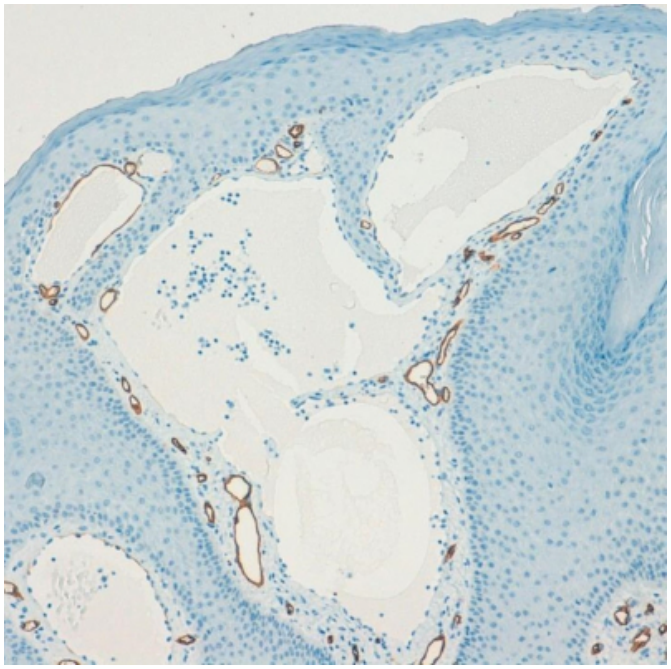


Figure 6

Immunohistochemistry confirms the lymphatic origin of the vascular channels (D2-40+, CD34-).



Therefore, a diagnosis of lymphangioma circumscriptum was done. Because the patient is now asymptomatic, no further treatment has been initiated and the patient remains in follow-up.

DISCUSSION

Lymphangioma circumscriptum is a rare, benign disorder of lymphatics located in the deep dermal and subcutaneous tissues, which can be of primary or secondary origin [3, 5]. The exact aetiology of LC is still not clear. However, various growth factors such as vascular endothelial growth factor-C (VEGF-C) and VEGF-D and their receptors on lymphatic endothelial cells may have a role in the mechanisms that control the development of LC [1, 4]. Acquired vulvar LC is induced by obstruction of lymph flow from many causes, including radiotherapy, malignancies (most common pelvic carcinomas), fistulous Crohn disease, infections such as tuberculosis, pelvic surgery and HS sinus tract removal [1, 5, 7]. The injury of the deep collecting channels in the connective tissue causes dilation of superficial dermal lymphatic channels and subsequent development of typical thin-walled translucent vesicles that range from 1 to 4mm in size [3-6, 9]. The vesicles and papules of LC may have verrucous alterations and associated hyperkeratosis, giving them a warty appearance [1, 9]. Clinical presentation is often asymptomatic. However, when symptoms are present, they usually are described as vulvar itching, burning, pain, or exudative discharge. Vesicles can rupture and cause secondary bacterial infection. Because of its clinical appearance and size, LC often results in aesthetic and psychosexual disorders [5, 8]. The most common differential diagnosis includes genital warts or condyloma accuminata, herpes zoster, molluscum contagiosum, pseudoverrucous papules, or squamous cell carcinoma [1, 6, 9].

Biopsy and histopathology provide the correct diagnosis [1]. Microscopic findings demonstrate superficial dermal dilated endothelial-lined and lymph-containing cystic spaces that are typically located in the papillary dermis but may also be found in the reticular dermis. These endothelial-lined cystic spaces may be multiloculated, and the overlying epithelium may be eroded or hyperkeratotic. The vascular spaces in the papillary dermis may nearly abut the basal layer of the overlying epithelium. The vascular channels contain lymph that is largely acellular and eosinophilic [3, 5].

Treatment for LC is intended in the presence of recurrent infection, persistence of exudative lymphatic fluid or blood, and for cosmetic purposes. There is no standard treatment for the management of LC [1, 5, 8]. Various methods have been described, including surgical excision, radiotherapy, laser therapy, sclerosing therapy, cryotherapy, electrocautery, and radiofrequency ablation [1, 3-6, 8, 9]. High degree of recurrence is seen, especially in lesions with deeper involvement [5]. Surgical management requires removal of deep communicating lymphatic channels with success dependent on the extent of disease, location of the lesions, and the depth of lymphatic cisterns within the dermis [9]. Accordingly to the literature, in most cases observation was chosen as the principal management, followed by surgical excision [7]. Rare cases of squamous cell carcinoma arising in vulvar LC have been reported, and patients should be followed clinically if resection is not performed [7].

It is also important to address the underlying cause of LC when possible. In our case, hidradenitis suppurativa was the only preexisting condition near the LC lesions. This chronic disease is characterized by the presence of recurrent, painful, deep-seated, rounded nodules that develop into abscesses and sinus tracts, with suppuration and hypertrophic scarring of apocrine gland-bearing skin, most commonly, the axillary, inguinal, and anogenital regions [10]. Smoking and overweight are the two main factors associated with HS [10]. Lymphatic obstruction and lymphedema may complicate

longstanding inflammation, which could be the cause of LC [10]. Medical treatments include topical therapy, such as soaps and antibiotics, and systemic therapy, including oral antibiotics and newer therapies like tumor necrosis factor alpha inhibitors or interleukin-23 inhibitors. Surgical removal of sinus tracts in severe refractory HS is limited to patients who have failed medical therapy. There are some reported cases of LC in patients submitted to this surgery, therefore we should be aware that this treatment could potentially cause or contribute to worsening of the LC [7].

Lastly, we have reported this case because it´s a rare association between vulvar lymphangioma circumscriptum and hidradenitis suppurativa. It is possible to misdiagnose this condition and therefore our goal is to highlight this diagnosis by recognizing the clinic and confirming it with a biopsy, to achieve an adequate management.

Conflict of interest statement: The authors declare that they have no conflict of interest regarding the publication of this case report.

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