Acquired lymphangioma circumscriptum of the genitals in an individual with chronic hidradenitis suppurativa

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INTRODUCTION

Acquired lymphangioma circumscriptum (LC), also known as *localized LC*, is characterized by small, clear, fluid-filled, clustered vesicles and papules that resemble a gelatinous mass of "frog spawn" on the surface of the skin. We present a case of genital LC associated with hidradenitis suppurativa (HS), which is significant because it represents a rare causal relationship.

Abbreviations used:

HS:	hidradenitis suppurativa
LC:	lymphangioma circumscriptum
PP:	pseudoverrucous papules
SCC:	squamous cell carcinoma

CASE

A 48-year-old man presented with a greater than 10-year history of hidradenitis suppurativa of the buttocks, gluteal cleft, and perineal area treated with antibacterial soaps, topical and oral antibiotics, deroofing procedures, and cryoinsufflation. Over the last 5 years, the patient noted asymptomatic nondraining papules diffusely on the scrotum, penis, and perineal area. The lesions were asymptomatic except when the drainage from his hidradenitis caused irritation and itching in the area.

Physical examination found multiple semitranslucent skin-colored papules coalescing into plaques on the penile shaft, scrotum, and perineum (Figs 1 and 2). The patient had fibrosis and scarring of the perineum, gluteal cleft, and bilateral buttocks, greater on the left than the right, with multiple interconnected draining sinus tracts. Shave biopsy specimens obtained from a smaller plaque on the penile shaft (Figs 3 and 4) showed dilated vascular spaces lined by a thin layer of endothelial cells and surrounded by hyperplastic epidermis. Chronic inflammation was seen within the stroma. These findings were consistent with lymphangioma circumscriptum, and because the lesions are asymptomatic, no further treatment has been initiated. Treatment of his hidradenitis suppurativa is being pursued with adalimumab, topical clindamycin solution, and benzoyl peroxide and staged excision of sinus tracts.

DISCUSSION

The differential diagnosis of genital papules is wide, and LC is not often considered when treating patients that present with this chief complaint. The vesicles and papules of LC may have verrucous alterations and associated hyperkeratosis, giving them a warty appearance, often leading to a prebiopsy diagnosis of human papillomavirus infection.[1] Histologic examination of LC shows superficial dermal multiloculated cystic vascular spaces filled with hypocellular fluid and lined by endothelium.[2] Although LC was acquired in our patient, the condition may also be congenital.

In most cases of localized LC, there is a predisposing tissue alteration such as Crohn's disease, radiation exposure, or cancer.[3] A pre-existing

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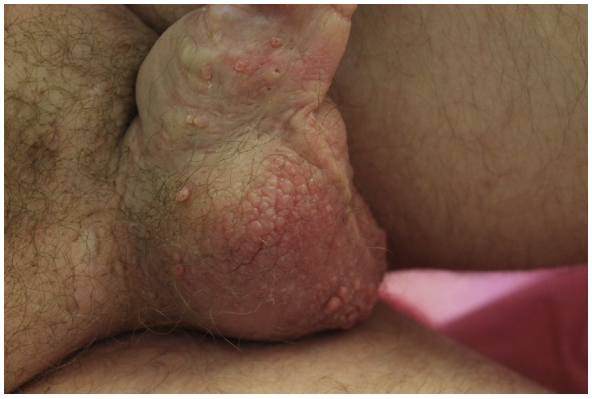

Fig 1. Lymphangioma circumscriptum on the penile shaft.

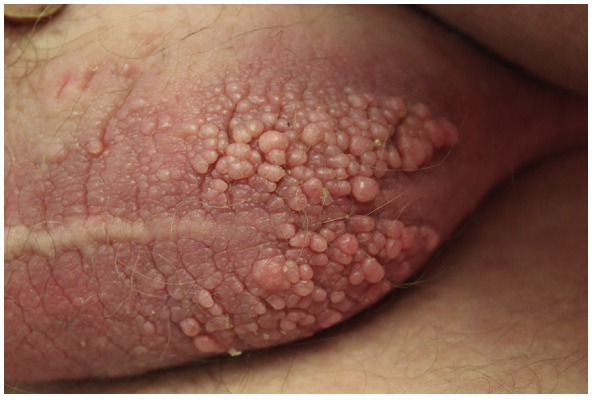

Fig 2. Lymphangioma circumscriptum coalescing vesicles and papules on the scrotum.

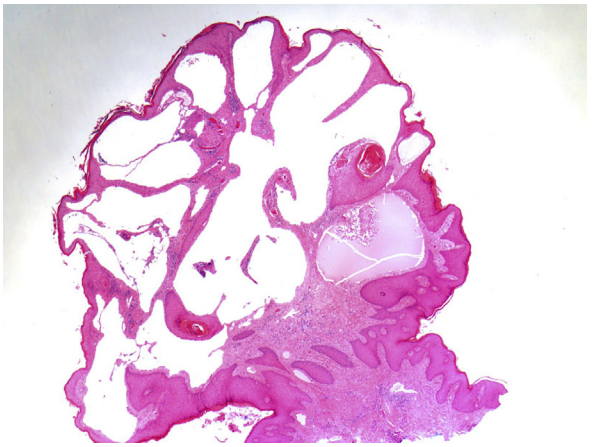

Fig 3. Histology of LC shows dilated vascular spaces. (Hematoxylin-eosin stain.)

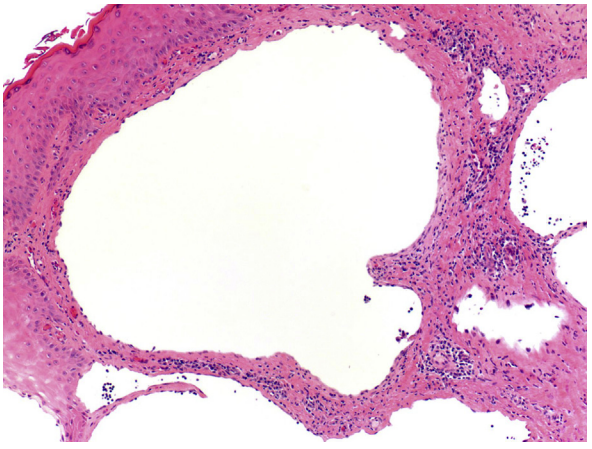

Fig 4. Thin layer of endothelial cells line a vascular space of an LC papule. Note the chronic inflammation in the dermis. (Hematoxylin-eosin stain.)

pathologic process likely limits the subcutaneous lymphatic cisterns from completely draining into the general lymphatic system. The accumulation of lymphatic fluid and the contraction of smooth muscle surrounding these cisterns leads to lymphatic fluid being forced into surface vesicles.[1] In our patient, HS is the only pre-existing condition near the LC lesions. His long-standing and severe HS resulted in extensive scarring in the groin and buttocks. This case is an interesting presentation because HS has rarely been associated with LC of the genitals in men.

Depending on location, the differential diagnosis of localized LC is diverse. On the genitals, the vesicles may be misdiagnosed as molluscum contagiosum, pseudoverrucous papules (PP), condyloma acuminata, or squamous cell carcinoma (SCC), leading to painful and fruitless treatments or apprehension by the patient in participating in sexual activity.[2-4] The presentation of SCC is diverse, but it may mimic LC when warty in appearance. Almost 95% of penile cancers are SCCs, of which 6% are the verrucous subtype.[5] Furthermore, SCC has been associated with chronic HS in the anogenital region; therefore, a high index of suspicion is indicated.[6] Pseudoverrucous papules on the genitals present as flat, skin-colored papules and nodules and typically appear in areas with chronic irritation because of urine or stool incontinence. The papules appear moister than condyloma and resolve when the underlying condition improves.[7,8] Histologic examination of PP finds irregular epidermal acanthosis, hyperkeratosis with intense parakeratosis, and papillomatosis. A dermal infiltrate of lymphocytes and plasma cells may be present.[8] Neither condyloma acuminata, SCC, nor PP display the dilated dermal lymphatics or subcutaneous cisterns seen in LC.

Palliative treatment, rather than definitive, may be used in sensitive areas like the scrotum to reduce morbidity. Such treatments include sclerotherapy, electrocautery, and radiofrequency ablation. Definitive treatment of symptomatic LC can include carbon dioxide laser ablation of

superficial lymphatics with destruction of vesicles. Surgical management requires removal of deep communicating lymphatic channels with success dependent on the extent of disease, location of the lesions, and depth of lymphatic cisterns within the dermis.[2,9] If left untreated, LC of the genitals may lead to pain, pruritus with excoriation, and secondary infection.[10] It is also important to address the underlying cause of LC when possible. Our patient is undergoing surgical treatment of his sinus tracts with the goal of reducing the clinical symptoms associated with HS. It should be noted that this treatment could potentially contribute to worsening of the LC because of increasingly aberrant lymphatic drainage.

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