Acquired vulva lymphangioma circumscriptum from Crohn's disease: A case report

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ABSTRACT:

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Lymphangioma circumscriptum (LC) is a rare benign disorder involving lymphatic channels. It can occur in congenital and acquired form. Vulva presentation of LC is rare. Acquired vulva LC may arise from infections, Crohn's disease and pelvic radiation. We herein reported a case of vulva LC that developed over 20 years after Crohn's disease.

Key words: Lymphangioma circumscriptum, Crohn's disease, vulva

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Introduction

Lymphangioma circumscriptum (LC) is a benign vascular tumor involving the lymphatic channels in deep dermal and subcutaneous layers[1]. It typically presents as multiple frogspawn-like vesicles. The common locations are the neck, axilla, chest wall and groin. Vulva presentation is rarely seen. LC can occur in congenital and acquired forms. Acquired LC may arise from architectural disruption of the lymphatic channels such as after surgery and pelvic radiation[1,2].

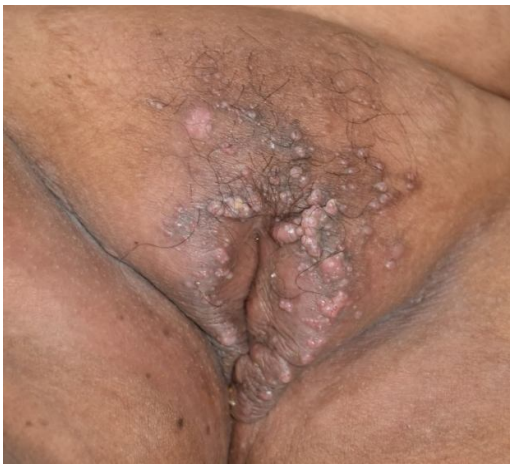

Figure 1 Multiple clusters of semi-translucent frogspawn-like vesicles on labia majora

Cutaneous manifestations of Crohn's disease (CD) are classified into specific manifestations such as edema, erythema, fissures or ulcer of the labia, penis or anus and non-specific manifestations such as aphthous stomatitis, erythema nodosum, pyogenic granuloma and sweet's syndrome[3]. We herein report a case of acquired vulva LC associated with CD.

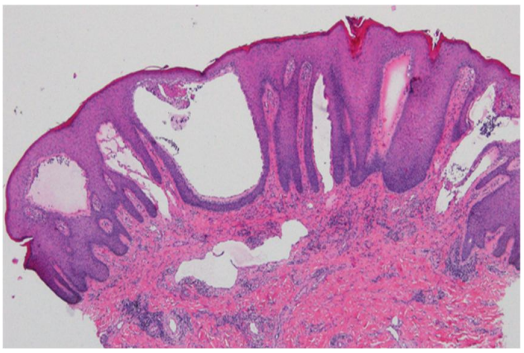

Figure 2 The sections show dilated vascular spaces lined by flattened endothelial cell. Valves were noted at the dilated vascular spaces. Overlying acanthotic epidermis is also noted. (H&E, X40)

Case report

A 55-year-old female presented to Dermatology out-patient clinic with multiple labial papules for 6 months. The lesions were mild pruritic and gradually increased in size. The patient was diagnosed with CD for 20 years based on the clinical, colonoscopy and histopathology findings. She developed rectovaginal fistula for 10 years. Her disease was controlled by medications. She had no history of pelvic malignancy or previous pelvic surgery or radiation. Physical examination revealed multiple clusters of semi-translucent frogspawn-like

vesicles on labia majora (Figure1). No inguinal lymphadenopathy was identified, and her vulva fistula was inactive.

Skin biopsy was performed. The sections show dilated vascular spaces lined by flattened endothelial cell. Valves were noted at the dilated vascular spaces. Overlying acanthotic epidermis was also noted (Figure 2).

Magnetic resonance imaging (MRI) of the lower abdomen showed large transsphincteric fistula tract with the opening at the upper anus with multiple complex fistula tracts at upper perineum, lower of the rectum and lower vagina.

Treatment options for LC were offered. However, the patient preferred a watchful observation due to very mild symptoms.

Table 1 Summary of patients with vulva LC and Crohn's disease (CD)

Case	Age	Duration of CD	Fistula	History of previous surgery and radiation	Exam	Treatment
Mu[2]	44	20	Yes	No	Flesh colored papules	Excision
Handfield[3]						
Patient 1	38	17	Yes	No	Weeping vesicular lesion	Resection
Patient 2	40	19	Yes	No	Papulovesicular lesion	CO2 laser
Papalas[7]						
Patient 1	74	56	Yes	No	Vesicles	Excision
Patient 2	48	21	Yes	No	Inflamed, thickened skin	Excision
Patient 3	43	18	Yes	No	Esthiomene	Excision
Patient 4	53	20	Yes	No	Exophytic verrucous	Excision
Chang[6]						
Patient 1	44	N/A	No	N/A	N/A	None
Patient 2	44	N/A	Yes	N/A	N/A	None
Patient 3	19	N/A	No	N/A	N/A	Excision
Our case	55	20	Yes	No	Semi-translucent frogspawn-like vesicles	Punch biopsy

N/A: not available

Discussion

The vulva presentation is an uncommon site for the development of LC. Congenital LC is often observed at birth until the age of two. Acquired LC is associated with surgery, radiation, recurrent infections and CD. The etiology is not clear. Blockage and disruption of lymphatic from inflammatory, neoplastic or surgical are proposed to cause this disease[2,4].

From the literature review, acquired LC in CD patients are noted in a few case reports/series. [2,5,6] (Table1) Fistula, longstanding CD and no history of previous pelvic surgery and radiation are noted in most cases. The duration from the onset of CD to the development of LC varies between 18 to 56 years. In our patient, acquired LC also developed after longstanding CD and rectovaginal fistula was identified. LC secondary to CD when compared to radiation; was larger in size, later in onset, lower in incidences of associated comorbidity, lower risk of disease progression and usually do not required additional surgical intervention[7]. In patients of vulva LC who have no underlying diseases, no history of surgery and radiation, computed tomography (CT), MRI or abdominal and pelvic ultrasound are recommended to assess the causes of obstruction[6].

Abnormal of lymphatic drainage plays an important role in the pathogenesis of CD. The proposed mechanism of vulva LC in patient with CD is direct blockage of lymphatic channels caused by chronic inflammation associated with fistula tract formation[8,9].

Although frogspawn-like vesicles are a clue for diagnosis LC, vulva LC can mimic condyloma accuminata, vulva intraepithelial neoplasia, molluscum contagiosum or bullous dermatosis due to reactive epidermal changes. A high index of suspicion is important especially in cases with atypical presentations. Skin biopsy should be performed to make a definite diagnosis[2].

The treatment of vulva LC is not standardized. Many different treatments were proposed with various results. The options include surgical excision, electrocautery, cryotherapy and ablative laser. The recurrence was frequently occurred. Vigilant observation was acceptable treatment. Multiple lesions and previous radiotherapy had been identified as a factor of poor prognosis due to comorbidity and high rate of recurrence[2,6,7].

Conclusion

Acquired LC is a rare benign malformation of lymphatic channels. We reported a case of vulva LC which developed over 20 years after CD. Blockage and disruption of lymphatic channels from chronic inflammatory in CD is proposed to cause the disease. The clinical of frogspawn-like vesicles on vulva is the clue to make the diagnosis.

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