Vol. 24(3), pp. 34-38, August 2024
DOI: 10.5897/AJMHS2024.0270
Article Number: D62678072470
ISSN 2384-5589
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African Journal of Medical and Health Sciences

Case report

Acquired vulvar *Lymphangioma circumscriptum* managed by simple partial vulvectomy: A case report

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Received 30 March, 2024; Accepted 11 July, 2024

Acquired *Lymphangioma circumscriptum* is among the rarest and most benign conditions of the lymphatic system. The etiology of *L. circumscriptum* is unclear. A case is presented involving a 42-year-old Para 4 women diagnosed with acquired *L. circumscriptum* after presenting with vulvar swelling, for which a simple partial vulvectomy was performed. The clinical presentation of a patient with acquired vulvar *L. circumscriptum* may resemble that of genital warts and malignancies. Therefore, vulvar lymphangioma should be considered as a differential diagnosis for vulvar swelling. Simple partial vulvectomy is one management option if sclerotherapy and vaporization with CO_2 are not available.

Key words: Vulvar swelling, acquired vulvar *Lymphangioma circumscriptum*, simple partial vulvectomy, case report.

INTRODUCTION

Acquired *Lymphangioma circumscriptum* is among the rarest and benign condition of the lymphatic system that results from secondary obstruction of pelvic lymphatic channels. The etiology of *L. circumscriptum* is unclear; however, it may be due to abnormality of congenital lymphatic system or secondary damage of the previously normal lymphatic system which is called acquired. It can have an effect on any parts of the body including the vulva (Amouri et al., 2007; Vlastos et al., 2003; Ikeda et al., 2011). Acquired *L. circumscriptum* can be resulted from surgical procedures like radical hysterectomy, with or without adjuvant radiation therapy for cervical cancer, chronic lymph edema of the lower limbs, or infectious causes including tuberculosis, Lymphogranuloma venereum, Filariasis (Ikeda et al., 2011; LaPolla et al., 1985). The clinical presentation of *L. circumscriptum* could start from being asymptomatic to highly disabling condition. Some of the most common symptoms observed from patients include vulvar discomfort, itching, and oozing. Physical examination findings include multiple nodular lesions, Verruciform or Polypoid, edematous and shiny skins with different size and having normal skin in between (Heyi et al., 2022; LaPolla et al., 1985). *L. circumscriptum* creates diagnosis dilemmas because it resemble genital warts or Condyloma acuminate (Sinha et al., 2015). This diagnosis is made utilizing clinical assessment and tissue histopathology (Ikeda et al., 2011; Toshyan et al., 2016). Management options include observation for asymptomatic cases to laser ablative therapy and surgery (Vlastos et al., 2003). In all management cases, there is a variable risk of recurrence (LaPolla et al., 1985). A case was presented of a 42 year-old Para 4 women diagnosed to have acquired *L. circumscriptum* following presentation with vulvar swelling for which simple partial vulvectomy was done.

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Figure 1. Acquired vulvar lymphangioma circumscriptum pre-operative image.

CASE PRESENTATION

A 42-year-old Para IV woman, with a history of all deliveries being vaginal and amenorrheic for the past 2 years, presented with progressively increasing vulvar swelling of 16 years' duration. She reported that the swelling had worsened significantly over the past 7 years, accompanied by itching and oozing. Her marriage ended 6 years ago due to discomfort during sexual intercourse. She had been a known retroviral infection (RVI) patient on highly active antiretroviral therapy (HAART) for the past 13 years. Her recent CD4 count was 283, and she had an undetectable viral load, stating that she had been adherent to her medication and follow-ups. She had a history of treatment for pulmonary tuberculosis (TB) 13 years ago, diagnosed after presenting with a productive cough, weight loss, and loss of appetite, confirmed through sputum acid-fast bacilli (AFB) testing and chest X-ray. Despite repeated treatments for sexually transmitted infections (STIs) following the development of vulvar swelling, no symptom improvement was observed. She was referred from a tertiary hospital with an initial diagnosis of genital warts to exclude vulvar cancer. Colposcopy raised suspicions for cancer, leading to a presumptive diagnosis of vulvar elephantiasis and a referral to the Urogynaecology clinic. At the current presentation, she had no cough, chest pain, weight loss, loss of appetite, or any other symptoms suggestive of tuberculosis. Physical examination revealed stable vital signs, with no significant lymphadenopathy in the lymphoglandular system. The respiratory examination was normal. Genital examination showed a grossly swollen vulva with clustered cauliflower-like nodular swellings involving the upper part of the vulva bilaterally, with the largest nodule measuring 2x3 cm. The clitoris was buried under the swelling, and there was blood oozing from the swelling, visible at the posterior fourchette (Figure 1). Additionally, satellite warty-like lesions were observed at the mons pubis. The swelling appeared shiny, and the cervix was smooth with no masses.

Laboratory tests including CBC, renal function, and liver enzymes were within normal limits. Management options included conservative management with manual lymph drainage, perineal care, tight clothing and surgical management with vulvectomy. Per patients' request, a simple partial vulvectomy was performed, removing affected skin and subcutaneous tissue (Figure 2). The procedure was performed by Urogynaecology and pelvic reconstructive surgery fellows and skin approximated with interrupted vicryl 2/0 suturing material (Figure 3). Histopathology confirmed *L. circumscriptum* (Figures 4 and 5). The post-operative course in the hospital was smooth with stable vital sign and no sign of surgical side

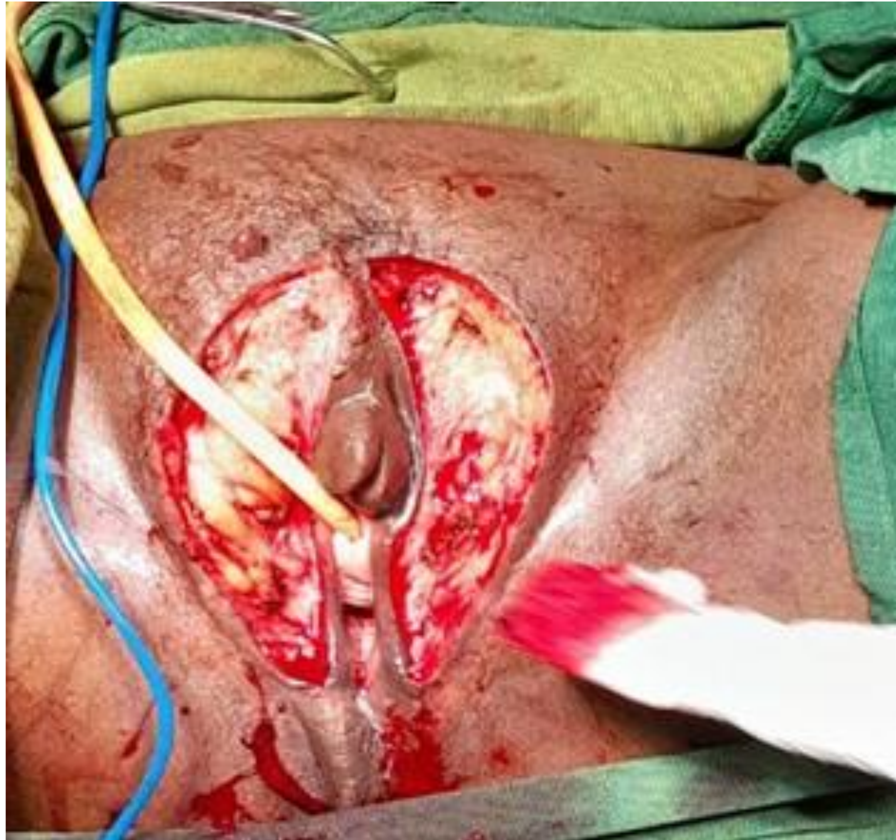

Figure 2. Image taken intra operatively after superficial vulvectomy but before closure.

Figure 3. Image showing superficial vulvectomy after skin was closed with interrupted stitches.

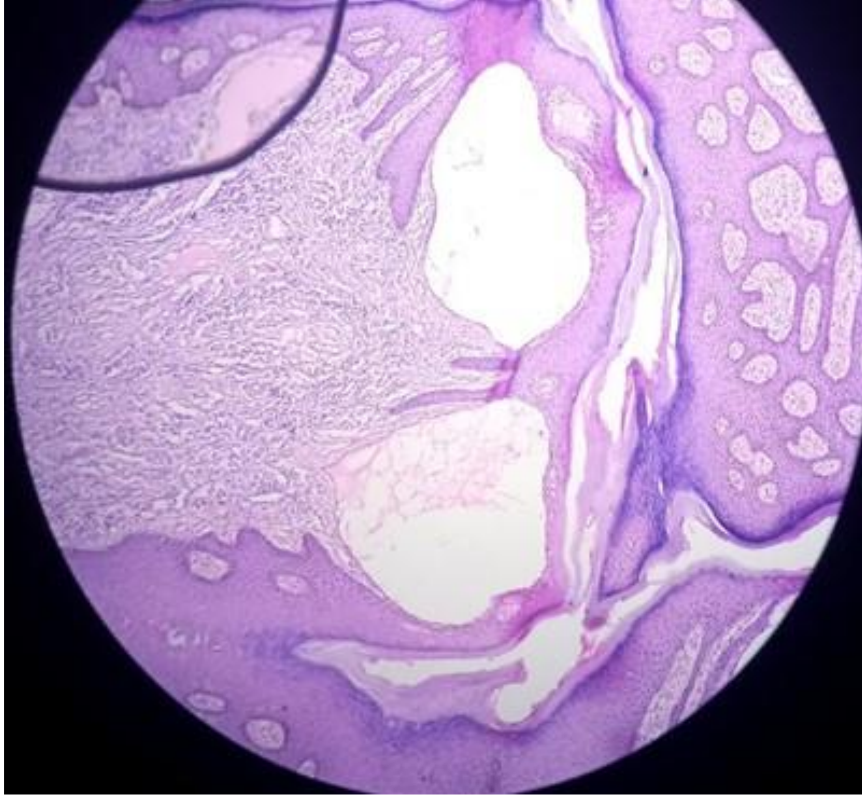

Figure 4. Image showing histopathology.

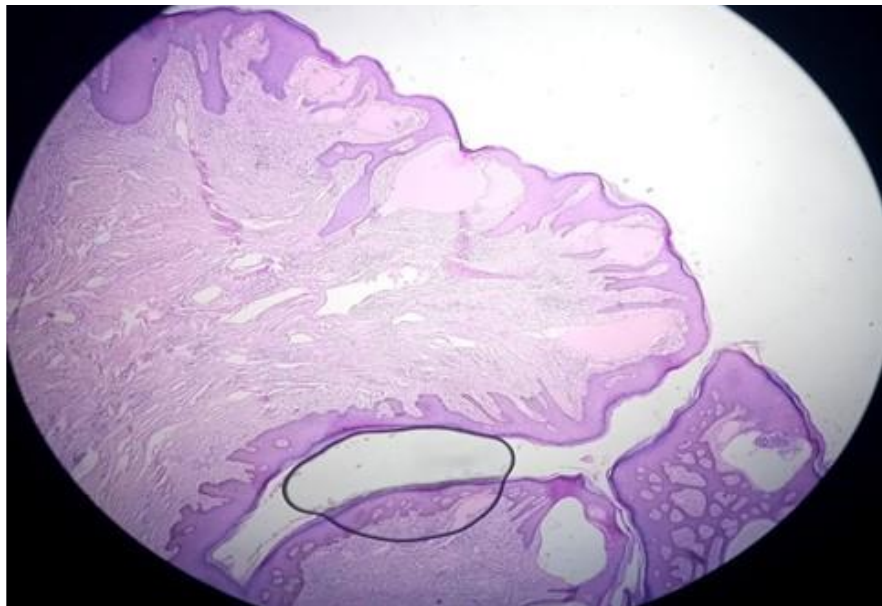

Figure 5. Image of histopathology.

infection. She was on broad spectrum antibiotics and discharged on the 5th post-operative day with post-operative advice and scheduled appointment. Follow up appointment after 1 month revealed well-healed wound with no subjective complaint. On her subsequent 6-month post-operative follow up, a well healed wound was

observed, however the clitoral head was slightly swollen. She was advised for perineal care and to wear tight clothing. Follow-up scheduled for one year later.

DISCUSSION

Acquired vulvar *L. circumscriptum* can occur as a result of secondary lymphatic channel obstruction in previously normal lymphatic channels (Ikeda et al., 2011). The causes can be classified into three broad categories: malignancy-associated, Crohn's disease-associated, and tuberculosis-associated (Ikeda et al., 2011; LaPolla et al., 1985). This case is likely tuberculosis-associated *L. circumscriptum*, causing secondary pelvic lymphatic channel obstruction, as the patient had a history of treatment for pulmonary tuberculosis. Several classifications for lymphangioma have been proposed, with the most accepted being localized and classic. The clinical appearance of both forms is similar, characterized by clustered or diffuse thin-walled, translucent vesicles filled with clear lymphatic fluid (Ikeda et al., 2011). In this case, the *L. circumscriptum* was of the localized type, involving only the vulva.

The clinical presentation of acquired *L. circumscriptum* may resemble genital warts or condyloma acuminatum, creating diagnostic dilemmas both clinically and histopathologically (Vlastos et al., 2003). Histopathology is paramount to confirm the diagnosis and outline the potential causes of lymphatic obstruction. The histopathology results in this case were consistent with *L. circumscriptum*. Initially, the patient was diagnosed with genital warts to rule out vulvar cancer. Management of acquired *L. circumscriptum* depends on the clinical condition and includes conservative or surgical options. Conservative management may involve manual lymph drainage, exercise, compression, abrasive therapy, sclerotherapy, electrocoagulation, and laser therapy with carbon dioxide (CO_2) (36).

Although recurrence is a possibility with treatment, surgical excision carries the lowest recurrence risk, and vaporization with CO_2 is recommended for its acceptable cosmetic results (Amouri et al., 2007; Vlastos et al., 2003). In this case, a simple partial vulvectomy was performed because laser ablation and CO_2 vaporization were not available in the current setup, and follow-up over 6 months showed no recurrence. Complications of *L. circumscriptum* include cellulitis and psychosexual dysfunction. This case involved sexual dysfunction, and the patient had been divorced 6 years prior.

CONCLUSION

The clinical presentation of a patient with acquired vulvar *L. circumscriptum* can vary and may resemble genital warts and malignancies. In cases of vulvar swelling, vulvar lymphangioma should be considered as a differential diagnosis. Simple partial vulvectomy is one management option if sclerotherapy and vaporization with CO_2 are not available.

CONFLICT OF INTERESTS

The authors have not declared any conflict of interests.

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