

VULVAR NON-FILARIAL ELEPHANTIASIS ASSOCIATED WITH SCABIES

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ABSTRACT Vulvar elephantiasis is a very rare condition often caused by filariasis infection. Other causes like general inflammatory disease, cancer treatment, traumatic causes or other infections can also explain the occurrence of this condition. In some cases, it is due to unknown causes. In this study, we present the clinical case of a Moroccan youth female patient with no significant history, who presented vulvar elephantiasis associated with scabies with no evidence of filariasis infection, general inflammatory disease, malignancy, or other infection. This clinical case is unusual because of the large volume of lymphedema, its atypical localisation, its association with generalised scabies, and the absence of a well-defined aetiology, which may explain its occurrence.

KEYWORDS Elephantiasis, vulvar, scabies, lymphedema.

Introduction

Elephantiasis describes a massive enlargement of a limb or the external genitalia because of chronic lymphedema [1]. The elephantiasis of the vulva is a rare condition often caused by filariasis in developing nations [3]. In this study, we present the clinical case of a 27-year-old patient with no significant medical history, who presented elephantiasis of the vulva evolved one year ago, associated with scabies. The patient did not show any evidence of filariasis infection or any other known cause which can explain the occurrence of this condition.

Case report:

A female patient of 27-years-old, with no previous medical history, was admitted to our department for vulvar elephantiasis of one-year evolution associated with generalised itchy skin lesions. The general clinical examination found: BMI=26, normal body temperature, generalised, smooth and slightly elevated



Figure 1: Anterior view of the vulva.

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Figure 2: Posterior view of the vulva.



Figure 3: view of the left and right labia majora with the two masses.

erythematous papules, and scratch lesions. The vulvar examination found a voluminous pendulous swelling mass in the left labia majora with minimal skin inflammation, and moderate lymphedema in the right labia majora, (figure 1, 2). The clinical examination also found bilateral inguinal adenopathy, and lower limb varices. However, the clinical and radiological examinations did not reveal the presence of an abdominal or gynaecological mass. The biological testing showed an increasing number of circulating eosinophilic granulocytes. C-reactive protein (CRP) was also elevated (84 mg/L), and the routine biochemistry, glucose, serological testing for sexually transmitted infection (hepatitis B, HIV, syphilis) didn't show any particularity. We also performed a Midnight peripheral blood smear for filarial worms, but the result was negative. The patient received an antiscabies treatment for her skin lesions, and she underwent into a complete excision of her vulvar masses, which carried out a mass of 2 kg from the left labia majora and a mass of 500 g from the right labia majora (Figures 3 and 4). Simple cutaneous su-



Figure 4: the two masses after excision.



Figure 5: Vulvoplasty.

tures were realised to close the residual substance loss (figure 5). The pieces were sent to the anatomopathological study which returned for vulvar lymphedema, without any sign of malignancy. The surgical follow-up was simple, but the patient presented again after two months with a new recurrence of lymphedema in the left labia majora (figure 6), without any associated general skin lesions.

Discussion:

Lymphedema can be defined as swelling of soft tissues which are the result of the accumulation of protein-rich interstitial fluids caused by a low output failure of lymph [3].

The Impairment of lymphatic drainage can be due to congenital abnormal vessel development (primary lymphedema) or damaged lymphatic vessels from infection, trauma, surgery, ra-



Figure 6: Recurrence of lymphedema after one month.

diation, persistent inflammation, obstruction due to metastases or parasite infestation (secondary lymphedema) [1].

Lymphedema is frequently encountered in the limbs, less commonly in the face and the genital organs [3]. In its earliest stages, lymphedema is soft and pitting; however, with persistence, lymphedema has a brawny, non-pitting appearance likened to peau d'orange because of accentuation of skin folds and follicular ostia. Most of the swelling occurs in the subcutaneous tissue, but the skin exhibits the most changes, which, when severe, is termed elephantiasis [1]. Elephantiasis described a gross enlargement of the part of the body that has been involved. Review of the world's literature reveals that chronic vulvar elephantiasis is a very rare condition [3].

The majority of cases of vulvar elephantiasis found in the literature are due to filariasis [3] [4]. Other causes include bacterial sexually transmitted infections (STI's), especially lymphogranulomavenerum (LGV) and donovanosis; tuberculosis, haematological malignancies, and dermatological diseases and in some cases from unknown cause [4, 1, 2].

Lu et al. [1] presented a clinical series of 24 cases of localised lymphedema; he made a comparison between those patients and the patients with diffuse lymphedema. Lu found that the most frequent localisation in these patients is anogenital, and overall, women were more frequently affected by lymphedema, particularly in the anogenital region and trunk, than men.

The patients in Lu et al. series did not have any form of cancer treatment, they were significantly more likely to have history of trauma to the site (vaginal delivery), and less likely to have a coexisting, chronic inflammatory process (rosacea and Crohn's disease). No etiology or associated disease was found in about one-third of all cases. Lu and al. Also, show in his study that 85% of patients were obese or overweight which can promote lymphedema by obstruction of the lymphatic canals by fat cells.

Antonio et al. [2] presented two clinical cases of unilateral vulvar lymphedema similar to the Lu and Al series, but unlike the Lu et al. series, they did not find any factors that may explain the onset of Lymphedema except obesity in one patient.

In our study the patient did not have any previous medical history or an apparent cause that can explain the vulvar elephantiasis, the research of filariasis was negative and also no sign of sexually transmitted infection, neither a sign of malignancy or inflammatory disease. Histological examination of the excised mass showed Acanthosis papillomatous epidermis surmounted by hyperkeratosis and orthokeratosis without cytonuclear atypia; in the dermis, we found many dilated lymphatic vessels of variable size. Important interstitial oedema without any sign of malignancy. The only abnormality she presented beside the vulvar elephantiasis was the skin lesions which were resolved after antiscabies treatment and, the slightly elevated level of circulating granulocyte and the CRP which may correlate with the previous scabies infestation.

Our patient presented a recurrence of lymphedema after excision. In Lu and al series the majority of lymphedematous tumours were cured with excision, and those that recurred or progressed were associated with factors known to aggravate lymphedema: obesity, cellulitis, signs of persistent inflammation. In our patient, we believe that persistent of inflammation can explain recurrence. However we planned to realise more investigation to explain the recurrence, but the patient refuse to go under supplementary examination.

Conclusion

This clinical case is unusual because of the large volume of lymphedema, its atypical localisation, its association with generalised scabies, and the absence of a well-defined aetiology, which may explain its occurrence.

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Disclosure Statement

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Competing Interests

Written informed consent obtained from the patient for publication of this case report and any accompanying images.

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