Vulval elephantiasis

Abstract

Vulval elephantiasis is one of the rarest conditions affecting female genitalia. The disease can lead to a significant psychological impact caused by functional impotence in cases of large tumors. The diagnosis is mainly based on clinical history and the surgical treatment remains the only relevant intervention for improving the quality of life. We present a rare case of vulval elephantiasis associated with hidradenitis suppurativa that underwent surgery with complete recovery.

Keywords: *vulva, elephantiasis, surgery, hidradenitis suppurativa*

Rezumat

Elefantiazisul vulvar este una din cele mai rare afecțiuni genitale ale femeii. Boala poate avea un impact psihologic semnificativ, prin impotența funcțională, în cazul tumorilor voluminoase. Diagnosticul este bazat mai ales pe istoricul clinic, iar tratamentul chirurgical rămâne singura intervenție care poate îmbunătăți calitatea vietii. Prezentăm un caz rar de elefantiazis vulvar, asociat cu hidradenitis suppurativa, care a beneficiat de intervenție chirurgicală, cu recuperare completă.

Cuvinte-cheie: *vulvă, elefantiazis, chirurgie, hidradenitis suppurativa*

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Funding sources: None

Submission date: 24.11.2021
Acceptance date: 30.11.2021

Elefantiazisul vulvar

Suggested citation for this article: Peltecu G, Botezatu R, Gică N, Chirculescu R, Panaitescu AM. Vulval elephantiasis. Ginecologia.ro. 2021;34(4):43-45.

Introduction

Vulva can be affected by a wide variety of lesions. One of the rarest conditions is vulval elephantiasis, which is a disease secondary to the obstruction of lymphatic channels.

Genital elephantiasis significantly affects the physical, mental and social well-being of the patient. The majority of cases in Africa are due to filariasis caused by *Wuchereria bancrofti* o *Brugia malayi*[1]. In the Northern hemisphere, it is due to sexually transmitted infections (STI), tuberculosis (TB), dermatologic diseases or iatrogenic causes.

The pathology and laboratory investigations are essential for diagnosis.

The differential diagnosis can be difficult due to multiple clinical features associated with the disease. Filariasis should be suspected when there is a relevant epidemiological exposure associated with an acute clinical picture (fever, eosinophilia, lymphangitis). The diagnosis is confirmed with microfilariae Giemsa stains from blood smear, polymerase chain reaction[2] or antifilarial antibody tests[3]. Ultrasound Doppler can be used for diagnosis to detect the presence of adult worms ("filarial dance" sign)[4].

Lymphogranuloma venereum, a genital ulcer caused by *Chlamydia trachomatis*[5], can be suspected when there is a recent history of travel in tropical or subtropical areas[6]. However, the diagnosis is often seen in immunocompromised patients and needs to be confirmed by serological testing[7].

Genital hidradenitis suppurativa (HS) is a chronic inflammatory disease that primarily involves the genital sweat glands. The clinical manifestations vary from malodor and pain to skin abscesses and disfigurement[8]. The mechanism is considered to be most likely the follicular occlusion causing plugging[9]. However, there is a strong association between HS and smoking[10]. Hidradenitis suppurativa mainly affects axillar and inguinal areas, but can also be a widespread disease[11].

Case presentation

A 56-year-old woman presented for a large, pediculated, right labial tumor (25/15 cm), evolving over the last six months, with a rapid growth in the last three months (Figure 1). She was obese (BMI 34.9 kg/m^2), had type 2 diabetes and had a total abdominal hysterectomy for uterus myoma 10 years before. The genital examination revealed a non-ulcerated vulval tumor, with a smooth surface, with soft consistency, painless, covered at the bottom by a thin layer of clear fluid. There was no sign of infection to the skin (redness, raised local temperature) covering the tumor. The superficial inguinal lymph nodes were not palpable.

Blood tests were within normal range. Right groin was unremarkable at the ultrasonography. Magnetic resonance imaging (MRI) of the pelvis revealed an edematous benign tumor well delimited from pubic bone and muscles. Special tests for TB and STI were negative. Filaria was excluded because the patient never left the country. The patient had significant difficulties to walk and her quality of life was poor.

She had surgery under spinal anesthesia. The cosmetic result was good (Figure 2). The pathology report revealed dilated lymphatic channels containing proteinaceous material and lymphocytic infiltrate within the papillary dermis (Figure 3). The bacterial cultures examination of the samples taken from the sectioned tumor were negative.

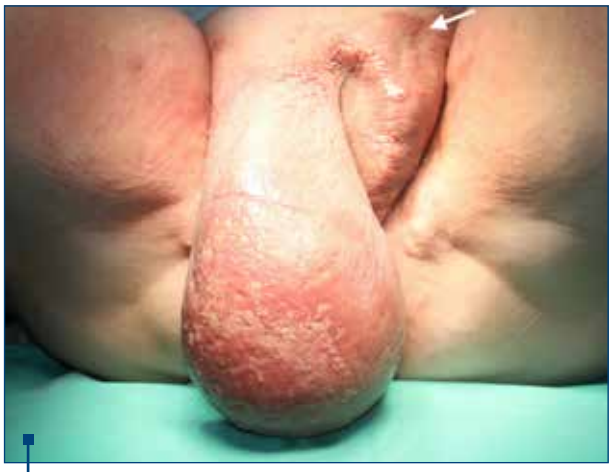

Figure 1A. *Non-ulcerated right labial tumor, with a smooth surface, with soft consistency, painless, covered at the bottom by a thin layer of clear fluid. There was no sign of skin infection*

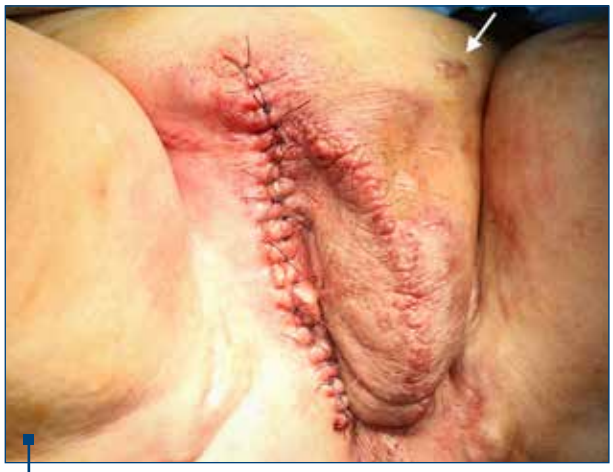

Figure 1B. *Hidradenitis suppurativa (white arrow). Local aspect after right labial tumor excision*

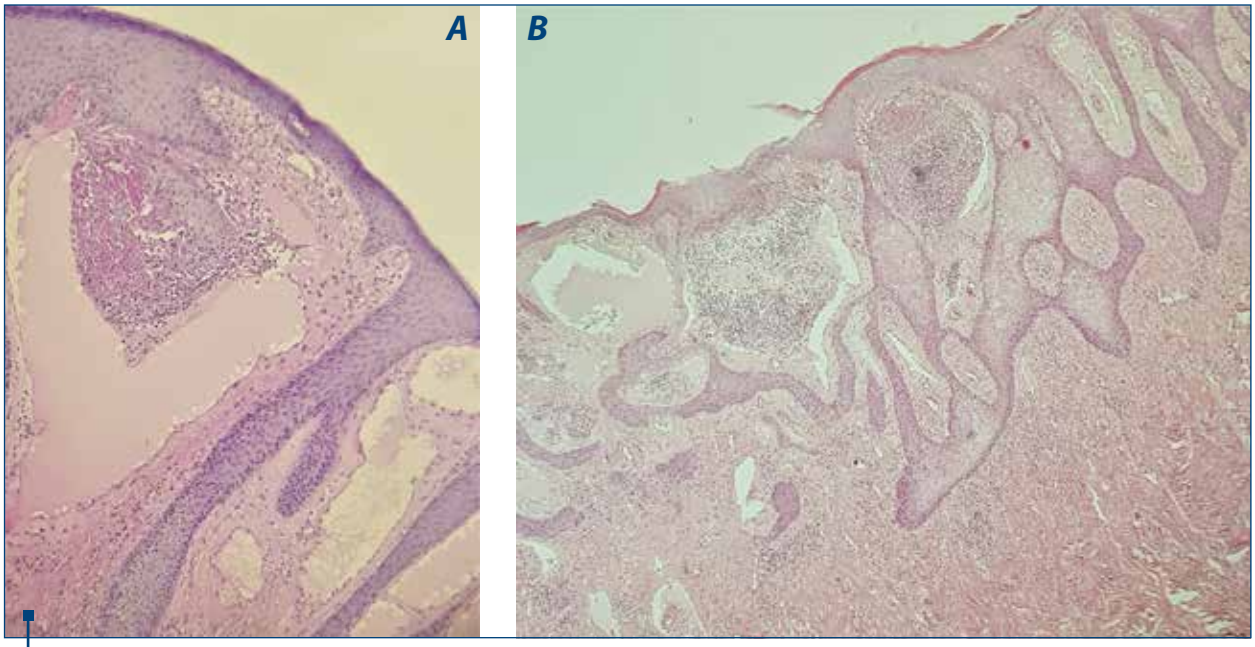

Figure 2. A, B. *Epidermal acanthosis and hyperkeratosis; dilated lymphatic channels containing proteinaceous material and lymphocytic infiltrate within the papillary dermis [(HE staining, 4x (A); HE staining 10x (B)]*

Discussion

Elephantiasis of the vulva was first described by J. Halliday Croom in 1893[12] and was considered with an unknown etiology.

Lymphedema is a swelling of the soft tissue secondary to the accumulation of interstitial fluid, rich in protein, due to the difficult lymphatic drainage of the involved area. In case of long-term lymphatic obstruction, it could result an exaggerated form of lymphedema, known as elephantiasis[13]. The incidence of vulval elephantiasis is the lowest of all cases of elephantiasis, representing only 1-2%.

In the past, the patient had recurrent skin infections of the right axilla, clinically highly suggestive for hidradenitis suppurativa, treated empirically with antibiotics by her family doctor. The same kind of skin infection was described in both groins, predominantly on the right groin, one year before, treated in the same way. Hidradenitis suppurativa (HD) is a chronic, recurrent inflammatory disease presenting as painful subcutaneous nodules. It is characterized by double comedones, deep sinus tracts and abscesses, followed by scars formation[14]. HS is a clinical diagnosis, without using laboratory tests. It is frequently associated

with diabetes mellitus and typically occurs in the axilla, inguinal, perianal, perineal and inframammary regions[15]. In this case report, HS is the only etiological correlation.

Conclusions

Vulvar elephantiasis that is not associated with filaria is an extremely rare condition, associated with an important psychological impact on social life. These cases are often diagnosed late due to delayed seeking of medical advice. The surgical treatment improves the overall condition, but it does not treat the cause, which needs further investigations for understanding the underlying lymphatic obstruction mechanism. ■

Abbreviations used: STI *– sexually transmitted infections;* ***MRI*** *– magnetic resonance imaging;* ***HS*** *– hidradenitis suppurativa;* ***TB*** *– tuberculosis*

Conflicts of interests: The authors declare no conflict of interests.

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