Zoon Vulvitis as a Differential Diagnosis of Ulcerative Lesions on the Vulva: Case Report

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Abstract
Vulvar ulcers are a frequent reason for doubt and difficulty of etiological diagnosis. Among the causes of genital ulcers, we can mention sexually transmitted infections such as herpetic simplex virus infection, syphilis, chancroid, granuloma inguinale or Donovanosis, and lymphogranuloma venereum, as well as non-infectious etiologies including psoriasis, sexual trauma, Behçet’s syndrome, Wegener’s granulomatosis, and Zoon Vulvitis. In the present study, a case of a seven months evolution genital ulcer patient (who did not respond to the various proposed therapies) was referred to the Service of Vulvar Pathology of the Hospital Universitário Antônio Pedro (Universidade Federal Fluminense). A biopsy of the lesion was performed, and the histopathological report was compatible with Zoon Vulvitis. The treatment with corticosteroids was initiated. Patient presented a successful evolution. An extensive review of the literature on differential diagnoses of vulvar ulcers was carried out in order to guide a clinical reasoning and a therapeutic approach to the different etiologies.

Keywords: ulcer; vulvitis; sexually transmitted diseases.

Introduction
Vulvar ulcers are a frequent cause of doubt and difficulty of etiological diagnosis. It may be associated with sexually transmitted diseases (STDs), but also with autoimmune diseases (Behçet Disease), cancer, trauma, or be related to the use of pharmaceuticals (hormonal non-steroidal anti-inflammatory drugs), among others.

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DISCUSSION

Sexually transmitted infections should be considered among the causes of genital ulcers, including: infection by herpes simplex virus (HSV), syphilis (Treponema pallidum), soft chancre (Haemophilus ducreyi), inguinal granuloma or Donovanosis (Klebsiella granulomatis), and lymphogranuloma venereum (Chlamydia trachomatis serotypes L1, L2 and L3). Non-infectious etiologies include sexual trauma, Lipschütz ulcer, Behçet syndrome, drug source ulcers and ZoonVulvitis(4).

The herpetic lesions may originate from both the HSV-1 and HSV-2, which are most commonly associated with subclinical conditions. Genital herpes begins with pruritus and erythema, followed by the development of vesicular lesions on the external genitalia, and may evolve into ulcerating lesions of clean fundus(5).

Primary syphilis is characterized by a localized cutaneous lesion called chancre. The lesion begins as a papule, which is typically (but not always) painless, appearing at the site of inoculation and evolving into an ulcer with a high and hardened margin, with non-exudative basis and associated with a moderate, often bilateral, regional lymphadenopathy. Soft chancre evolves into spontaneous healing within 3 to 6 weeks, even if not treated(6).

The soft chancre arises as painful ulcerated lesions with dirty fundus with purulent exudate and may progress in suppurative lymphadenopathy by a drainage of a single orifice(7).

The granuloma inguinale or Donovanosis begins with the uprising of a subcutaneous nodulation, generally unilateral and painless, whose erosion produces well defined flat edge or hypertrophic ulcerations, causing tissue destruction and subsequent urethral and vesical fistulas(8).

Drug source ulcers occur 30 minutes to 8 hours, approximately, after administration of drug associated with itching and burning. These lesions tend to disappear in 1-2 weeks, followed by cicatrization with hyperpigmentation, which can persist for months. This drug reaction is mainly associated with the use of non-steroidal anti-inflammatory drugs, although medications like clotrimazole, tetracyclines and ampicillins can also initiate this type of ulcer. Intercourse with partners using these drugs can also cause the reaction(9).

It is also worth mentioning the possibility of acute aphthous ulcers triggered by immune reaction by Epstein Barr virus, Mycoplasma pneumoniae, Varicella infection, viral gastroenteritis and respiratory infections, which develop multiple painful ulcers that last from 1 to 3 weeks(10).

As already mentioned, the differential diagnosis of genital ulcers involves a number of infectious and non-infectious etiologies. Evaluation of patients during the initial approach should always take place in order to verify the presence of STDs, since these infections may be transmitted and increase the risk of contracting HIV.

Faced with the possibility of co-infection by multiple organisms, the determination of the etiology of a genital ulcer based on a classic sign can lead to misdiagnosis and inappropriate treatment.(12) In addition, immunocompromised patients may have atypical clinical symptoms, including more widespread and serious diseases(13).

Once the diagnostic tests results may not be available in the initial medical appointment and the appropriate laboratory analyses does not identify the pathogen in 25% of genital ulcers patients, the empirical treatment of these patients can be performed(14).

The empirical treatment should take into consideration the most likely diagnosis based on the patient’s epidemiological and clinical history, as well as on careful physical examination. The disadvantage of an empirical treatment approach is the administration of inadequate therapy, affecting the ability to confirm the diagnosis and causing an impact on the contactants trace. This is a particular

The lymphogranuloma venereum is characterized by inoculation lesion, papule, pustule or painless ulcerations, which disappear without sequelae, followed by generally one-sided painful inguinal adenopathy, with suppurration and fistultization of multiple holes(15).

Lipschütz ulcer, or ulcus vulvae acutum — a rare entity, but probably under-diagnosed disease —, is an acute painful vulvar ulcers in young women. Etiology and pathogenesis are unknown although there are reports associating the vulvar acute ulcers with infection by Mycoplasma, influenza A, and especially the primary infection by the Epstein-Barr virus (EBV) and Cytomegalovirus (CMV). This disease can be preceded with feverish symptoms and is characterized as influenza prodromal symptoms, showing lesions that affect labia majora, perineum and vaginal introitus, well-defined ulcers, and may show discreet gray exudate with adhered pseudomembranous lesions in mirror(16).

Behçet’s disease is an autoimmune disease manifested by recurrent oral ulcers and may be associated or not with genital well-defined ulcers. These are usually deeper lesions than the oral ones with necrotic material in their base and an erythematous halo. The lesions in labia minora usually heal without scar evidence, however the most common ulcerations occur in the vagina and may evolve into deep ulcerations, causing tissue destruction and subsequent urethral and vesical fistulas(17).

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Figure 1 – Patient’s genital ulcer at first appointment in Hospital Universitário Antônio Pedro.
concern when treating supposed syphilis patients. However, this situation is a compensation for the risk of unattendance of an untreated patient for monitoring or infection transmission while waiting for the diagnostic tests results(2).

The Zoom Vulvitis, the subject of our study, was first described in 1954. It is a chronic inflammation of the vulvar mucosa characterized by itching, burning, dysuria and dyspareunia affecting primarily the labia minora, clitoris, vaginal and urethral meatus. The symptoms are usually erythematous, bright, limited and symmetrical plaques affecting the patient’s life quality. The lesions tend to chronicity, but the progression to malignancy was not described. The definitive diagnosis is histopathological, showing plasmocytic and vascular changes — vascular hyperplasias and extravasation of red blood cells (Figure 2 and 3). There are many proposed therapeutic schemes, but their results are inconsistent, including estrogens, steroids, antibiotics, antifungals, cryotherapy, laser ablation and surgical resection(13).

The conclusion is that it is important to refer those genital ulcer patients who do not respond to the proposed treatment to a service of reference so that the histopathological diagnosis can be confirmed and the monitoring done properly.

Conflict of interests

The authors declare no conflict of interests.

REFERENCES


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