Lipshütz Ulcer: An Undiagnosed Entity

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Abstract
Lipshütz ulcer is a rare cause of vulvar ulcerous diseases and it is often underdiagnosed, probably because of the lack of knowledge about this diagnosis. The aim of this article is to review the pathogenesis, clinical presentation, differential diagnosis and management of this entity.

Keywords: Lipshütz ulcer; Epstein-Barr virus; Pain

Introduction
Lipshütz ulcer is an entity that occurs in sexually inactive young women. The typical presentation in the emergency department is a healthy teenage girl with a first episode of painful vulvar lesion preceded, generally, by a flu-like syndrome a few days before.

Discussion
Lipshütz ulcer was firstly described in 1912 by Benjamin Lipschütz, and it is also known by “ulcers vulvae acutum”. It mainly affects non-sexually active adolescents and young adults, but there are even reported cases in children under two [1].

The precise incidence is unknown, and it seems to be a rare condition, probably because it is greatly under-reported and many cases are misdiagnosed with other vulvar ulcerous diseases.

The exact pathogenesis is still unclear. One hypothesis suggests that the ulcer is a clinical manifestation of a hypersensitivity reaction to a distant viral or bacterial infection, such as the Epstein-Barr virus infection [2]. The causative agent might reach the vaginal mucosa and disturb the microbiota, turning the Döderlein lactobacilli into pathogenic species [3].

Although the ulcers are not infectious or sexually transmitted, a case of Lipshütz ulcer in twin sisters with a probable airborne infectious transmission has been reported [4].

The diagnosis is usually made by exclusion of other common causes of genital ulcerations. The first step is to be aware of the existence of this entity. The physical examination shows a single or multiple ulcers, with an overlying yellowish-grey exudate and raised borders on labia minor or major, very painful to the touch. Bilateral ‘kissing lesions’ are characteristic [5]. A very detailed anamnesis must be carried out to find out a possible recent history of upper respiratory tract infections, among other important aspects. The differential diagnosis to consider includes sexually and non sexually transmitted infections, autoimmune conditions, drug reactions and local manifestations of systemic illness [6]. It can also be confused with sexual abuse, leading to unnecessary investigations and anxiety to the family [7].

Proposed diagnostic criteria include the following [2,7]:

a. First episode of acute genital ulceration.
b. Being younger than 20 years.
c. Presence of one or multiple deep, well delimited, painful ulcers with a necrotic base on the labia minor or major.
d. Bilateral “kissing” pattern.
e. Absence of sexual contact during the past 3 months.
f. Absence of immunodeficiency.
g. Acute course of the genital ulcer (abrupt beginning and healing without scarring within 4 to 6 weeks).

The complementary tests should include a viral culture or PCR for HSV and a serological test for EBV. Other tests will be performed upon clinical suspicion. The biopsy is not necessary, but if the ulcer is persistent, it may be helpful (Table 1).
Table 1: Vulvar ulcerous diseases [6].

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infectious Venereal</td>
<td>Herpes simplex virus, Syphilis, acute HIV infection, Granuloma inguinale, Lymphogranuloma venereum, Chancroid</td>
</tr>
<tr>
<td>Nonvenereal</td>
<td>Epstein-Barr virus, Candida, Bacteria, Parasites, Mycobacteria</td>
</tr>
<tr>
<td>Traumatic</td>
<td>Mechanical, Thermal, Chemical, Factual</td>
</tr>
<tr>
<td>Tumors</td>
<td>Basal cell carcinoma, Squamous cell carcinoma, extramammary Paget’s disease, Leukemia/lymphoma</td>
</tr>
</tbody>
</table>

In most cases, the ulcer heals spontaneously within four to six weeks. The treatment is mainly supportive and should include pain relief and topical treatment of the lesion. Empiric treatment will include analgesics and topical anesthetics, as well as sitz baths. In case of severe pain, hospitalization is indicated. If urination is very painful permanent catheterization should be considered [8]. Use of systemic corticoids and broad-spectrum antibiotics can be indicated in severe cases.

Conclusion

Lipschütz ulcer is often under diagnosed and mistaken with other vulvar ulcerous diseases. It must be considered in young virgin patients with painful necrotic ulcers and a history of a recent viral or bacterial infection. Keeping in mind this diagnosis should avoid unnecessary treatments to the patients, since it is a self-limiting disease.

References

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