Ulcus vulvae acutum

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SUMMARY

A case of a 17 year-old girl with genital ulcers diagnosed as acute vulvar ulcer is reported. No relevant etiologic factors could be established. The gangrenous form of the disease, cutaneous anergy, and the cultured anaerobic pathogens suggest the possibility of infectious pathogenesis.

Introduction

In 1913 an Austrian dermatologist named Lipschütz first identified an acute disease with fever, ulceration of the external genital organs, and lymphadenomegaly, which occurs in young women. The syndrome has been termed acute vulvar ulcer. Despite the characteristic symptoms, the disease is very rare and often misdiagnosed. The following typical case history is a good illustration of this interesting clinical entity.

Case history

The patient was a 17 year old female. As an infant, she had frequently had respiratory infections with fever and complications. At school, the patient exhibited symptoms of optical atrophy, transient hemiparesis, Raynaud symptoms, articular complaints, splenomegaly, and lymphadenomegaly. Diagnosis at observation: not further defined autoimmune disease.

Complaints at admission: high fever (39°C) of sudden onset and malaise, which persisted for four days. Later, tenderness and swelling of the external genital organs occured, followed by development of very painful nodules and "wounds" impeding walk and urination. The patient never had sexual contacts. She did not complain of any kind of respiratory infection or other internal disease.

Status at admission: slightly swollen and erythematous vulva. An ulcer of about 10 mm in diameter on the right labium majus and three ulcers of 15 mm in diameter each, distributed over both labia minora at the vestibulum. The pressure-sensitive, round or elliptical ulcers had sharp edges and were covered with grayish-yellowish crusts. The hymen was intact. The firm inguinal lymph nodes were 1,5-2 cm in diameter.

KEY WORDS

genital ulceration, acute vulvar ulcer

Laboratory data

Erythrocyte sedimentation rate: 47 mm/h; WBC 5000; mild anemia. Urine: ++++ leukocytes. Immune status: Mantoux test and Multitest: complete anergy. Pathergic test negative. Serologic tests: RPR negative, Monostikon test: negative. *Epstein Barr virus, Herpes simplex, Varicella zoster virus*: negative. Microscopy of pathogens from the genital ulcers: *Neisseria gonorrhoeae* (methylene blue and Gram staining) negative. Bacterial culture: *Bacteroides fragilis, Porhyromonas asaccharolyticus and Peptostreptococcus anerobius positive. Trichomonas* (native study): negative. Fungi: negative; *Mycoplasma, Ureaplasma urealyticum, Chlamydia* (ELISA): negative. *Herpes virus,* EBV: negative.

Course of the disease and therapy

Antibiotic treatment with Doxycyclin 100 mg b.i.d. was introduced. External treatment (sitz bath) with potassium permanganate; Betadine dressings and Betadine ointment. By the second day of treatment the ulcers began to colliquate, and by the third day sloughed, leaving behind defects. Pain disappeared after a week, dysuria remaining the most persisting complaint. By the end of the third week the ulcers healed leaving scarred retracted areas.

Discussion

The clinical entity of hyperacute genital ulceration generally occurs in young women, predominantly virgins (1), although cases of ulceration developing after sexual intercourse (defloration) have also been reported. Similar symptoms have been observed in wives of healthy military staff, who had sexual contacts with their husbands following long periods of absence (2).

The cause and pathogenesis of the disease still remains unknown, and only some hypotheses are discussed in the literature. Lipschütz assumed that the disease is caused by autoinoculation with *Bacillus crassuss (Döderlein's lactobacillus)*, while other physicians of his generation ascribed the disease to poor hygiene of the young women (3, 4). In some cases, *Epstein-Barr virus* and *Ureaplasma* were identified. Recently, genital ulceration very similar to acute vulvar ulcer has been found in HIV-positive women. These ulcers healed after zidovudin therapy (5, 6, 7).

The clinical picture starts with sudden onset of fever, chills and malaise without any prodrome. 3-4 days later, painful ulcers develop on the internal surface (vestibulum) of the labia minora, but they can affect the labia majora as well. The symptoms are accompanied by dysuria. Based on clinical course and macromorphologic findings, three main forms of the course of the disease can be identified:

1. Gangrenous form: characteristic hyperacute onset, ulcers covered with grayish-yellowish crusts, extreme pain. Crusts slough after several days with scarring. This is is the most frequent form.

2. Chronic form: (pseudo-veneric Lipschütz ulcer, Scherber's pseudo-tuberculotic form). A relapsing form with marked edema, circular or partially undercut superficial ulcers. The ulcers usually heal in 4-6 weeks.

3. Miliary form: Purulent, fibrinous ulcers with inflammatory edges of pinhead size. The ulcers typically affect the margins of the labia majora and minora, as well as the perineal region. The general symptoms are mild and healing is rapid.

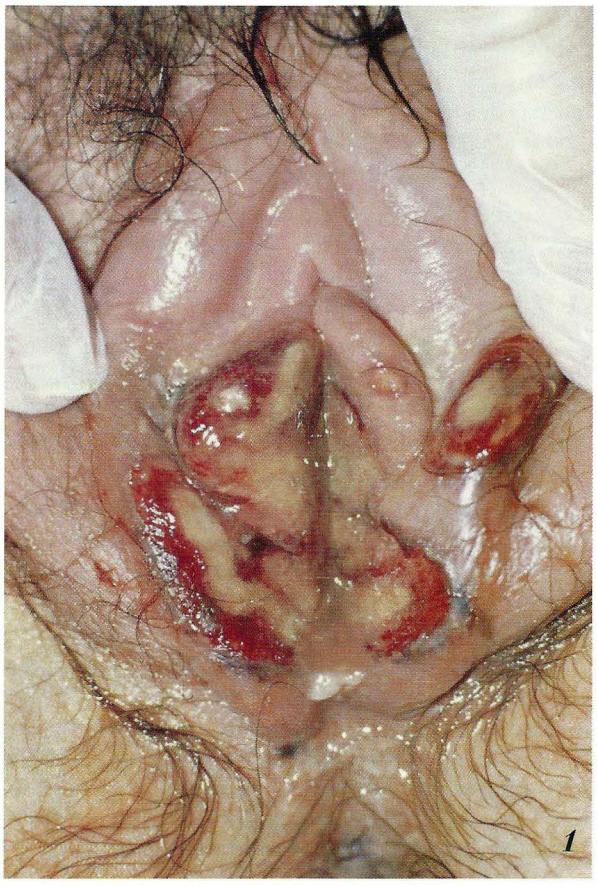
Apart from the characteristic labial localization, the involvement of the vagina and the urethra have also been reported. Aphthous ulcers of the buccal mucosa developing together with polymorphic and nodose erythema can be regarded as an extragenital form of this disease (8) and may possibly be included into Behcet's syndrome or a primary herpes infection.

Histolopathology is of no diagnostic value (7). The upper part of the edematous corium reveals dilated capillaries with a lymphocyte, histiocyte, plasmocyte and fibroblast infiltrate. Proliferation and local thickening of the vessel wall is also observed. In advanced cases, the infiltrate contains mainly polymorphic neutrophils with subsequent formation of a miliary pseudoabscess and ulceration.

The disease should be differentiated from venereal (syphilis, ulcus molle) and non-venereal infections (herpes simplex, herpes zoster, miliar ulcerative tuberculosis). Of the non-infectious diseases, Behçet's syndrome, various aphtous ulcers, ano-vaginal fistula, Mb. Reiter, and myeloproliferative diseases should be primarily considered (8). Evaluation of the entire skin and mucosal surface may help in the differential diagnosis.

Therapy: Because the etiopathogenesis remains unclear, therapy is mainly symptomatic. Bed rest is important. In acute gangrenous forms, wide-spectrum antibiotic therapy has proved to be beneficial. It should however be emphasized that due to the hyperacute character of the disease, its course can scarcely be affected after onset. In severe Behçet syndrome-like forms steroids and colchicin therapy should be considered. The use of externally applied desinfectants and ointments stimulating epithelization is worthwhile.

The presented case corresponded to the most frequently occurring gangrenous form and may have been



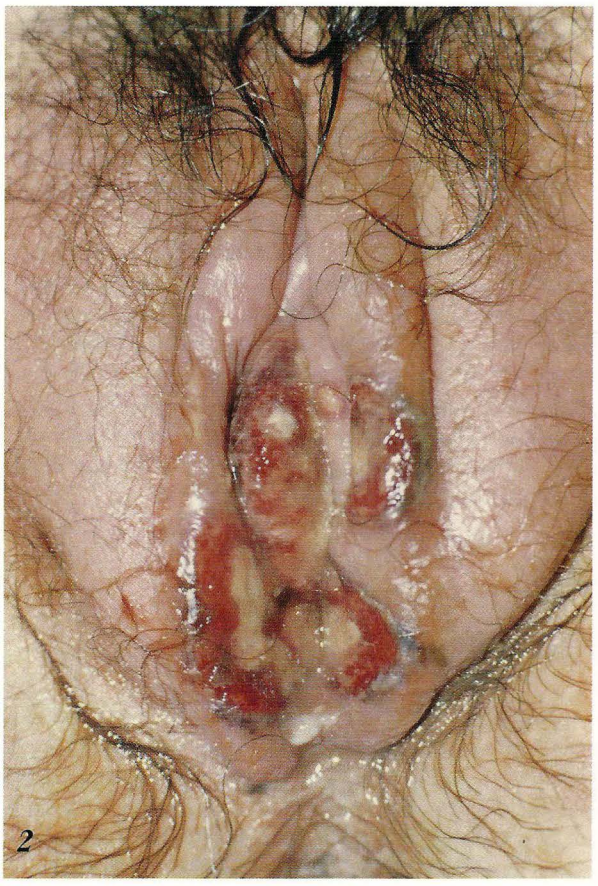


Figure 1-2: crusty ulcers affecting the vestibulum and the labia majora

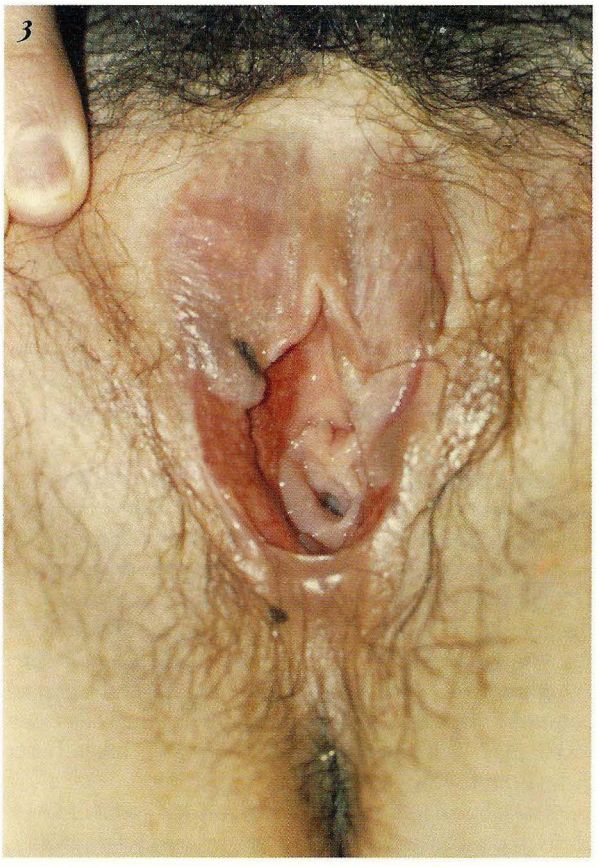


Figure 3: tissue lesions left after sloughing of the necrotic tissue

induced by a background infection. The latter suggestion is supported by the presence of anaerobic pathogens detected by bacteriological study; these microorganisms, however, can not be unambiguously considered as pathogens, since they can occur in normal vaginal flora. We failed to identify any other pathogens (*Herpes simplex, Herpes zoster, Epstein-Barr viruses, Mycoplasma* or *Chlamydia*) as mentioned by other authors. Together with the lack of other symptoms, the negative pathergy test excludes Behçet's syndrome. Cutaneous anergy of the patient may have contributed to the infection by facultative anaerobic pathogens.

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