Ulcus Vulvae Acutum

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An acute vulvar ulcer was diagnosed based on a case history of a 17-year-old girl with genital ulcers. No relevant causative factors could be established. The gangrenous form of the disease, cutaneous anergy, and the cultured anaerobic pathogens suggested the possibility of an infectious pathogenesis.

In 1913 an Austrian dermatologist named Lipschütz[1] was the first to identify an acute disease with fever, ulceration of the external genital organs, and lymphadenomegaly that occurs in young women. The syndrome has been termed acute vulvar ulcer. Despite the characteristic symptoms, the disease is very rare and is usually misdiagnosed by specialists. The following typical case history is a good illustration of this interesting clinical entity.

Case Report

A 17-year-old girl had had, as an infant, frequent respiratory infections with fever and complications. At school the patient was observed to have optical atrophy, transient hemiparesis, Raynaud symptoms, articular complaints, splenomegaly, and lymphadenomegaly. The diagnosis based on observation was of nondifferentiated autoimmune disease.

At admission she complained of a high fever (39°C) with sudden onset and malaise, which persisted for 4 days. Later, tenderness and swelling of the external genital organs was followed by the appearance of very painful nodules and "wounds" that impeded walking and urination. The patient had never had sexual contact. She did not complain of any kind of respiratory infection or other internal disease.

At admission her vulva was slightly swollen and erythematous. An ulcer about 10 mm in diameter on the right labium majora and three ulcers 15 mm in diameter each, spreading at both labia minora at the vestibulum, were noted. 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 to 2 cm in diameter (Figures 1 and 2).

Laboratory Data

Results of laboratory tests showed a red blood cell sedimentation rate of 47 mm/hour, a white blood cell count of 5000, and sustained anemia. Urine showed

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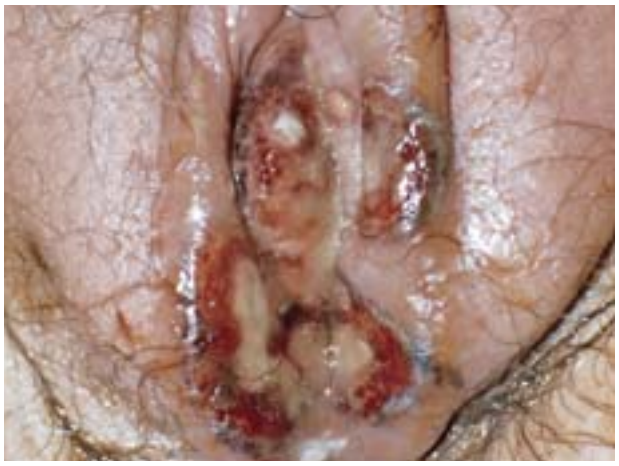

FIGURE 1. Crusty ulcers affecting the vestibulum and labia majora.

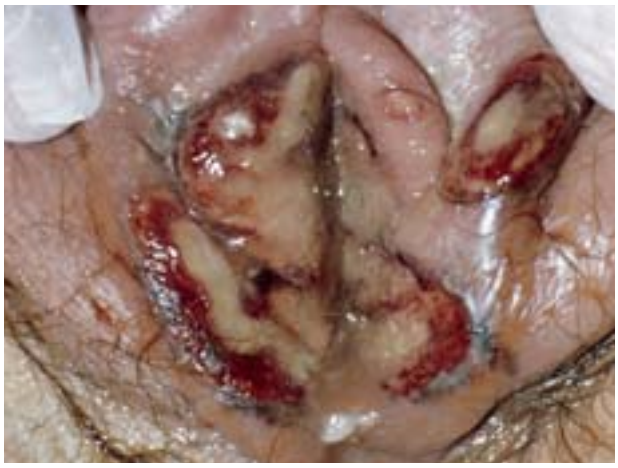

FIGURE 2. Crusty ulcers affecting the vestibulum and labia majora.

visible pus. Complete anergy was documented by results of the Mantoux test and Multitest. A pathergic test gave negative results. Serologic tests for reactive plasma reagin, Monostikon, Epstein-Barr virus, herpes simplex, and varicella zoster virus gave negative results. Direct identification of pathogens from the genital ulcers showed no Neisseria gonorrhea (methylene blue and Gram's staining). Bacterial culture identified *Bacteroides fragilis*, *Porhyromonas asaccharolyticus*, and *Peptostreptococcus anerobius*. Native study for *Trichomonas* gave negative results. Results of tests for fungi, Mycoplasma, yeasts *Ureaplasma urealyticum*, and Chlamydia (enzyme-linked immunosorbent assay) were negative.

Treatment was begun with oral doxycycline, potassium permanganate sitz baths, and povidone–iodine (Betadine®) dressings and ointment. After the second day of treatment the ulcers started to colliquate, and by the third day sloughed leaving behind loci of tissue lesions. Pain disappeared in a week with dysuria remaining the most persistent complaint. By the end of the third week the ulcers healed leaving scarry retracted areas (Figure 3).

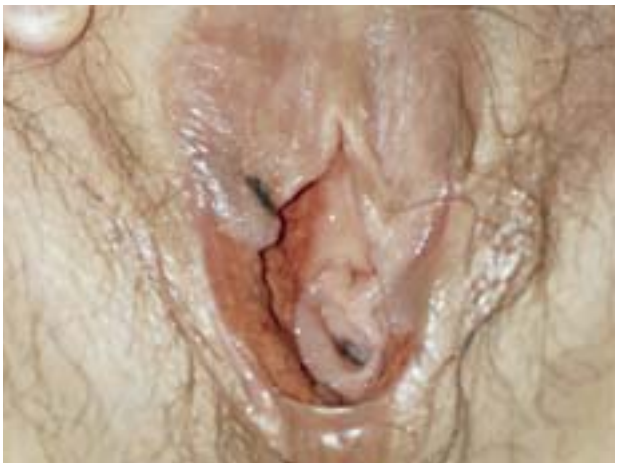

FIGURE 3. Tissue lesions left after sloughing of necrotic tissue.

Comments

Hyperacute genital ulceration generally occurs in young women, predominantly virgins,[1] although cases of ulceration occurring after defloration have also been reported. Similar symptoms have been observed in wives of otherwise healthy military staff who had sexual contact with their husbands after the latter were absent from home for a long time.[2]

The cause and pathogenesis of the disease remain unknown, and only some hypotheses have been reported in the literature. Lipschütz[1] 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 hygienic conditions.[3,4] In some cases Epstein-Barr virus and Ureaplasma could be identified. Recently, genital ulceration very similar to acute vulvar ulcer was found in women infected with human immunodeficiency virus. The ulcers healed after zidovudine therapy.[5-7]

The clinical appearance starts with a sudden onset of fever, chills, and malaise without any precedent. Three to 4 days later, painful ulcers appear 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 macromorphologic findings, three main forms of the course of the disease can be identified. The gangrenous form has a characteristic hyperacute onset, ulcers covered with grayish-yellowish encrustation, and extreme pain. Crusts slough in several days with scarring. This form is the most frequent. The chronic form (pseudoveneric Lipschütz ulcer, Scherber's pseudotuberculotic form) is a relapsing form with marked edema and circular or partial undercut superficial ulcers. The ulcers usually heal in 4 to 6 weeks. The miliar form involves purulent, fibrinous ulcers with inflammatory edges of a 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 fast.

As well as the characteristic labial localization, involvement of the vagina and urethra are also reported. Aphthous buccal mucosa occurring in parallel with polymorphic and nodose erythema can be regarded as an extragenital form of the disease.[8] The latter forms might probably be ascribed to Behçet's syndrome and primary herpes infection.

The histologic picture is of no diagnostic value.[8] The upper part of the edematous corium reveals dilated capillaries with lymphocyte,[1] histiocyte, plasmocyte, and fibroblast infiltration. Proliferation and local thickening of the vessel wall were also observed. In advanced cases infiltration mainly contains polymorphic neutrophils, with subsequent formation of a miliar pseudoabscess and ulceration.

The disease should be differentiated from venereal (syphilis, ulcus molle) and nonvenereal infections (herpes simplex, herpes zoster, miliar ulcerative tuberculosis). Of the noninfectious diseases, Behçet's syndrome, different aphthae, Reiter's syndrome, anovaginal fistula, and myeloproliferative diseases should be primarily considered.[8,9] Evaluation of the whole skin surface and the mucosa might help in the differential diagnosis.

Because of this condition's unclear etiopathogenesis, the therapy is mainly symptomatic. Bedrest is of importance. In the acute gangrenous forms, wide-

spectrum antibiotic therapy proved to be beneficial. It should, however, be emphasized that due to the hyperacute character of the disease, its course can be affected only in a limited way after the onset. In severe forms resembling Behçet's syndrome, steroids and colchicin therapy should be considered. The use of externally applied disinfectants and ointments stimulating epithelialization is worthwhile.

Our patient's condition was the most frequently occurring gangrenous form and was probably caused by an underlying infection. The latter suggestion was supported by the presence of anaerobic pathogens on bacteriologic study. These microorganisms, however, cannot be unambiguously considered as pathogens, since they can normally occur in the vaginal flora. We failed to identify any other pathogens (herpes simplex, herpes zoster, Epstein-Barr virus, Mycoplasma, Chlamydia) mentioned by other authors. Together with other lacking symptoms, the negative results of a pathergic test preclude Behçet's syndrome. The patient's cutaneous anergy might have contributed to infection caused by facultative anaerobic pathogens.

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