

Ulcus vulvae acutum

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S U M M A R Y

A 29-year-old female presented with a history of sudden onset of genital ulcers with fever and malaise. The clinical picture and course of the disease corresponded to the diagnosis of acute vulvar ulcer. No relevant etiologic factors could be ascertained. Treatment with doxycycline and systemic corticosteroids led to the resolution of the ulcerations after three weeks. Although the disease is rare, it is important to include it in the differential diagnosis of genital ulcers.

Introduction

Ulcus vulvae acutum is a rare but not under-recognized (unreported?) disease characterized by painful genital ulceration occurring in young women without venereal infections. Lipschütz initially described it in 1913. The ulcers are often multiple and develop in association with malaise, fever, and inguinal lymphadenopathy. Healing is spontaneous but may take several weeks and leaves some scarring. The etiology is still unknown. Histopathologic examination is of no diagnostic value.

Case report

A 29-year-old female presented at our department with a sudden onset of fever and two papules on the external genitalia, followed by the development of pain-

ful ulcers and burning on urination. The pain was so severe that the patient had great difficulty walking. There was no history of a previous sexually transmitted disease. The patient had a healthy long-term sexual partner. She had been taking birth control pills for several years and did not complain of any kind of respiratory infection or other internal disease.

Clinical examination revealed an ulcer 15 mm in diameter on the right labium minus at the vestibulum and a shallow ulcer of 10 mm in diameter on the left labium minus (Fig. 1). Both were sharply demarcated and covered with necrotic adherent crusts. No lymphadenopathy was noted. The laboratory tests revealed a slightly elevated C-reactive protein whereas the complete blood count, blood chemistry profile, and urine analysis were normal. The results of laboratory tests – including herpes simplex virus culture, gonorrhea, di-

K E Y W O R D S

**ulcus vulvae,
acutum,
29-year-old
female,
doxycycline
treatment**



Figure 1. Ulcer vulvae acutum: an ulcer on the right labium at the vestibulum.

rect immunofluorescence test for Chlamydia trachomatis, screening tests for syphilis, serological tests for antibodies to hepatitis A, B, and C viruses as well as for HIV 1 and 2, and Epstein-Barr virus – were all negative. Bacterial culture of the smear identified *Escherichia coli* and *Morganella morganii*, and microscopic examination and culture for fungi were negative.

The patient received oral treatment with doxycycline 100 mg b.i.d., and topical with fusidic acid cream. After exclusion of infectious disease, a short course of dexamethasone was started for symptomatic relief.

Therapy led to rapid improvement and epithelization of ulcers. After three weeks the ulcers healed with an atrophic scars.

Discussion

Etiology and pathogenesis of acute vulvar ulcers are still unknown. Lipschütz blamed these lesions on *Bacillus grassus*, which was later identified as Döderlein's bacillus (1). Ulcers can be associated with systemic infections such as Mycoplasma infection (2) and infectious mononucleosis (3–13), in which genital ulcerations can be an initial manifestation of Epstein-Barr virus infection in adolescents that are neither sexually active nor immunocompromised. Genital ulcerations similar to acute vulvar ulcers have been found in HIV-positive women (14). Some authors believe that anaerobic bacteria can cause ulcerations (15).

Several types of genital ulcerations have been described. The gangrenous form is the most frequent, and is still occasionally called Lipschütz disease. It has a characteristic hyperacute onset with fever and malaise. This type usually affects adolescent girls. The ulcerations are very painful, with thick adherent slough and a sur-

rounding red areola. They are most often solitary, on one side of the vulva, usually the inner aspect of the labium minus. As in our case, however, the lesions may occur bilaterally. Healing is spontaneous, though somewhat slow, and leaves some scarring. Recurrence is unlikely.

In the chronic form, small yellow ulcers with undermined edges and red surrounding areola arise on the introitus, labia minora, and labia majora. They tend to be associated with relatively few symptoms other than local pain and marked edema, and tend to recur.

In the miliary form, the purulent, fibrinous ulcerations are 2 to 3 mm in size with inflammatory edges and are more likely to involve the labia majora and minora as well as the perineal region. The general symptoms are mild and healing is rapid. In light of present knowledge, the last two forms would be assigned to either aphthous ulcerations or Behçet's syndrome. Only the gangrenous form is still considered a separate entity.

Histopathology is of no diagnostic value (14). The upper part of the edematous corium reveals dilated capillaries with polymorphous infiltrate composed of lymphocytes, histiocytes, plasmocytes, and fibroblasts. Proliferation and local thickening of the vessel wall is also observed. In advanced cases, the infiltrate contains mainly neutrophils with subsequent formation of a pseudo-abscess and ulceration.

Determining the cause of vulvar ulcers can be challenging. The differential diagnosis is extensive and includes venereal infections: herpes simplex, syphilis, chancroid, granuloma inguinale, lymphogranuloma venereum, HIV, bowenoid papulosis with papules that can easily erode and ulcerate, and nonvenereal infections by candida, Epstein-Barr virus, bacteria, parasites, and mycobacteria. As for noninfectious diseases, numerous inflammatory diseases such as nonbullous lichen planus, lichen sclerosus, inflammatory bowel disease, idiopathic and secondary aphthae, Sweet's syndrome, Behçet's syndrome, drug reaction, Reiter syndrome, and bullous diseases (pemphigus vulgaris, bullous pemphigoid, cicatricial pemphigoid, and paraneoplastic pemphigoid) should be considered (16). Traumatic causes such as mechanical, thermal, chemical, and factitial should be kept in mind, as well as malignant tumors (squamous cell carcinoma, basal cell carcinoma, extramammary Paget's disease, and leukemia/lymphoma). Careful surface examination of the entire skin and mucosa surface may be helpful in establishing a differential diagnosis.

The therapy is mainly symptomatic. Topical measures may include disinfectants, topical antibiotics, and topical anesthetics with or without topical corticosteroids. Systemic therapy includes analgesics with or without broad-spectrum antibiotic therapy and, in severe and painful forms, a short course of oral corticosteroids may be required.

Conclusion

Ulcus vulvae acutum is a rare diagnosis most often seen in adolescent girls, but should be considered if the patient's history and clinical manifestations do not fit the diagnoses of herpes simplex, syphilitic chancre, bacterial infection, Behçet's syndrome, or other causes of ulceration.

Because the ulcerations can be very painful, it is important that they be diagnosed and treated. The ul-

cers in ulcus vulvae acutum heal spontaneously, but because of subjective symptoms it is preferable to treat them systemically with corticosteroids, to which they respond well. Broad-spectrum antibiotic therapy has also proved to be beneficial.

It is important to keep this diagnosis in mind, in addition to syphilis or Behçet's syndrome, because the most frequent cause of ulceration is herpes simplex and patients risk being treated for herpes simplex without the physician even considering other causes of ulceration in the genital area.

REFERENCES

1. Lipschutz B. Über eine eigenartige Geschwürsform des weiblichen Genitales (Ulcus vulvae acutum). Archiv für Dermatologie und Syphilis (Berlin) 1913;114:363–95.
2. Korting GW, Hinterberger G. Ulcus vulvae acutum with cold-agglutinin-positive, Mycoplasma-caused atypical pneumonia. Hautarzt. 1979 Oct;30(10):550–2.
3. Brown ZA, Stenchever MA. Genital ulceration and infectious mononucleosis. Am J Obstet Gynecol. 1977;127:673–4.
4. Lampert A, Assier-Bonnet, Chevallier B et al. Lipschutz's genital ulceration: a manifestation of Epstein-Barr virus primary infection. Br J Dermatol. 1996;135:663–5.
5. Portinoy J, Aronheim GA, Chibu F et al. Recovery of Epstein-Barr virus from genital ulcers. New Engl J Med. 1984;311:966–8.
6. Lampert A, Assier-Bonnet H, Chevallier B, Clerici T, Salag P. Lipschutz's genital ulceration: a manifestation of Epstein-Barr virus primary infection (letter). Br J Dermatol. 1996;135:663–5.
7. Lawee D, Shafir MS. Solitary penile ulcer associated with infectious mononucleosis. Can Med Assoc J. 1983 Jul 15;129(2):146–7.
8. Sisson BA, Glick L. Genital ulceration as a presenting manifestation of infectious mononucleosis. J Pediatr Adolesc Gynecol. 1998 Nov;11(4):185–7.
9. Hudson LB, Periman SE. Necrotizing genital ulcerations in premenarcheal female with mononucleosis. Obstet Gynecol. 1998 Oct;92(4 Pt 2):642–4.
10. Cheng SX, Chapman MS, Margesson LJ, Birenbaum D. Genital ulcers caused by Epstein-Barr virus. J Am Acad Dermatol. 2004 Nov;51(5):824–6.
11. Halvorsen JA, Brevig T, Aas T, Skar AG, Slevolden EM, Moi H. Genital ulcers as initial manifestation of Epstein-Barr virus infection: Two new cases and review of the literature. Acta Derm Venereol. 2006;86(5):439–42.
12. Huppert JS, Gerber MA, Deitch HR, Mortensen JE, Staat MA, Adams Hillard PJ. Vulvar ulcers in young females: a manifestation of aphthosis. J Pediatr Adolesc Gynecol. 2006 Jun;19(3):195–204.
13. Barnes CJ, Alio AB, Cuningham BB, Friedlander SF. Epstein-Barr virus-associated genital ulcers: an under-recognized disorder. Pediatr Dermatol. 2007 Mar–Apr;24(2):130–4.
14. Covino JM, McCormack WM. Vulvar ulcer of unknown etiology in a human immunodeficiency virus-infected woman, response to treatment with zidovudine. Am J Obstet Gynecol. 1990;163:115–8.
15. Török L, Domjan K, Farago E. Ulcus vulvae acutum. Acta Dermatovenereol APA. 2000;9(1):30–3.
16. Daunt SO, Kotowski KE, O'Reilly AP et al. Ulcerative vulvitis in Reiter's syndrome. Br J Vener Dis. 1982;58:405–7.

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