Sudden onset of painful genital ulcers

VICTORIA HARRIS MB BS, LLB; **GAYLE FISCHER** MB BS, MD, FACD

Test your diagnostic skills in our regular dermatology quiz. What has caused this vulval ulceration?

Case presentation

A 23-year-old woman presents with painful vulval ulcers and severe dysuria (Figure). She has not been sexually active for the past three months and is otherwise well. She has no history of gastrointestinal disease.

On examination, several superficial ulcers are visible that have a red areola and a sloughy yellow-green base measuring 3 to 5 mm. The lesions are located mainly on the labia minora, which are oedematous. She does not have any oral ulceration.

The patient reports similar episodes over the past few years that have resolved spontaneously. On this occasion, however, the ulcers are so painful that she is not able to urinate and she requires admission to hospital for catheterisation.

Differential diagnoses

Conditions to consider in the differential diagnoses include the following.

- **Herpes simplex infection.** Genital herpes is common and should be ruled out in any case of painful acute and recurrent genital ulceration. In immunocompetent patients, small clear vesicles form that rapidly rupture to form groups of shallow erosions. In immunocompromised patients, ulcers and persistent crusted lesions are common. The patient in this case did not describe typical prodrome symptoms (burning, itching, stinging at the site) or any vesicles, and her ulcers are deeper and larger than would be expected in a woman who is otherwise well.
- **Primary syphilis.** The ulcer of primary syphilis (chancre) is typically painless with a clean base and little or no pus or crust. On palpation, the base of the ulcer feels firm and indurated. Painless regional lymphadenopathy may or may not be present. The ulcer is usually solitary and not recurrent but there is a red surrounding ring and often oedema.
- **Crohn's disease.** Aphthous ulcers that occur in association with Crohn's disease have the same appearance as those that occur in an idiopathic setting. Crohn's disease can affect the skin with or without bowel involvement, and vulval oedema may accompany ulceration in addition to typical sinuses and fistulae.[1] Crohn's disease is unlikely in this patient, who is otherwise well and has had about one episode of genital ulceration per year.
- **Behçet's syndrome.** This multisystem disease can present with aphthous ulceration. However, Behçet's syndrome would be highly unlikely in this young woman, who has no ocular or systemic complaints.
- **Trauma.** Excoriations can cause accidental trauma, usually in the setting of any severely itchy dermatitis. The surrounding skin is abnormal and the onset is rarely acute. Ulceration due to trauma is associated with pain as well as itch and is unlikely to be severe enough to cause urinary retention. Traumatic ulceration may occur in paraplegic patients and in patients in wheelchairs as a result of chronic friction from catheters and other medical equipment.
- **Squamous cell carcinoma and vulval intraepithelial neoplasia**. Malignancy must be considered in any patient with a persistent ulcer but is unlikely to be acute. It may cause pain, but this is usually not severe. In this patient, the acute nature of the ulceration makes malignancy very unlikely.
- **Nonsexually acquired genital ulceration.** This is the correct diagnosis. Nonsexually acquired genital ulceration (NSAGU) almost always occurs in girls and women aged between 8 and 25 years. It is a benign condition of unknown aetiology. Episodes may be triggered by viral infections, particularly when there is a viral prodrome. However, no cause has been clearly identified in patients with recurrent ulcers who do not have an underlying condition such as Crohn's disease or Behçet's syndrome. The ulcers of NSAGU have the same characteristics as oral aphthous ulcers (canker sores), with the ulcers at both

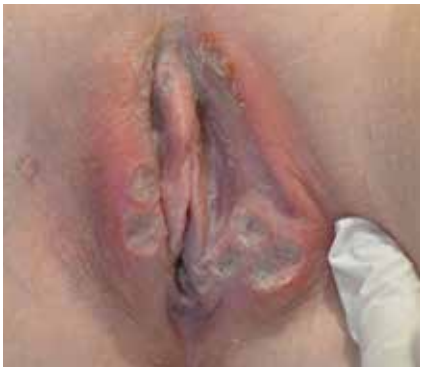

Figure. Superficial vulval ulcers at presentation.

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Dr Harris is a Dermatology Research Fellow and Associate Professor Fischer is Associate Professor of Dermatology at Sydney Medical School – Northern, University of Sydney, Royal North Shore Hospital, Sydney, NSW.

sites being painful and sharply marginated (punched out). The colour of the lesions is typically yellow-green on the base with a red areola. When they occur on the labia minora there may be severe accompanying oedema that makes them difficult to visualise. In the acute situation there is often a febrile prodrome. Approximately one-third of affected patients with NSAGU have had, or will develop, recurrent vulval ulceration. Approximately half of patients have had, or will develop, oral aphthous ulcers. NSAGU is also known as Lipshutz ulcer, Sutton's ulcer, vulval aphthae and complex aphthosis.[2]

Investigations

The diagnosis of NSAGU is based on clinical findings. Skin biopsy can be traumatic for a patient who is already in severe pain, particularly an adolescent, and the results are nonspecific. Investigation for primary herpes simplex virus and secondary bacterial infection is essential and should include skin swabs for polymerase chain reaction (PCR) testing and culture.

Identification of aphthous ulceration is very important in the emergency setting. Minimal investigations are required and biopsy is traumatic and nonspecific. Many young women are over-investigated and interrogated about sexual encounters, which they and their families find traumatic. If investigations for herpes simplex virus are negative then it is not necessary to perform a screen for sexually transmitted infections.

Management

NSAGU can range in severity and there are no formal treatment guidelines. For mild cases of genital ulceration, the mainstays of treatment are avoidance of irritating factors (tight clothing, perfumed soaps, pads and liners) and use of analgesia and topical treatment. The anti-inflammatory properties of corticosteroids can be useful. Potent topical corticosteroids are generally safe to use,[3] and when applied in an ointment base for two weeks or less to settle a minor flare they do not cause side effects.

In the situation where a patient is unable to walk or urinate, a short course of oral prednisone (25 mg/day) results in rapid relief of pain and hastens healing, with prednisone being rapidly withdrawn as soon as ulcers are healed. For severe cases, hospitalisation for pain management including catheterisation may be required. Prophylactic doxycycline (50 to 100 mg/day) may be effective to control flare-ups and prevent recurrence. MT

References

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COMPETING INTERESTS: None.