

# FAMILY MEDICINE GRAND ROUNDS

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## Vaginal pain and fever in a premenarchal girl How would you treat?

**A 13-year-old girl is brought to your office by her father** because she has had vaginal discomfort for 2 days. She also has had a fever up to 104°F and a small perineal rash. She was seen the previous day in the emergency department (ED) for the same symptoms. The ED evaluation included urinalysis, urine pregnancy test, and complete blood count with differential. Results were reportedly normal, with the exception of a left shift without leukocytosis. Slightly indurated and “pebble-like” lesions were noted on the perineal exam. DNA probes for gonorrhea and chlamydia were obtained. The patient was given acyclovir for presumed new-onset herpes simplex and was instructed to continue acetaminophen and ibuprofen for fever. The patient and her father are now seeking follow-up care.

**Q:** What are some causes of vaginal pain with fever? How would you proceed with the evaluation?

**A:** \_\_\_\_\_

### Other medical history

- Enuresis, recurrent perioral rash around age 7 with annual recurrences
- No chronic infections or illness
- Premenarchal
- Negative trauma history, including abuse
- Bike riding 2 days earlier but no falls
- Taking acyclovir as prescribed by ED; acetaminophen/ibuprofen for fever
- No known allergies

### Family and social history

- Parents alive and well; no siblings
- Straight-A student
- Involved in JROTC (was on a retreat 2 weeks ago)
- No use of tobacco, alcohol, street drugs, or suspicious substances
- Denies any sexual activity

### Physical examination

- Review of systems: Positive for chills, fever, dysuria, and perineal pain; otherwise negative
- Temperature 101.6°F, pulse 112, respiratory rate 22, blood pressure 110/70 mm Hg, weight 94 lb
- Alert female in no distress, normal neurologic exam
- HEENT: normal exam with clear oropharynx without lesions
- Skin/integument: no rashes, including arms, legs, hands, feet, and trunk
- Heart, lungs, and abdomen: tachycardia, otherwise normal
- Reproductive examination (chaperone present): Tanner stage III; perineal edema with marked purple ecchymoses located bilaterally at posterior introitus; multiple lacerations also

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noted as well as tenderness; some scabbed areas noted; no periurethral or perianal lesions or bruising; no

vaginal discharge; speculum and bimanual exam were deferred due to pain.

**Q: What is your presumptive diagnosis and plan?**

**A:**

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You are concerned about abuse. You interview the patient alone and she again denies any type of sexual encounter. She has never used tampons. She does report that she is a heavy sleeper and that she recently went on a retreat where she and a girlfriend slept in the same room. As far as she knows, there was no intruder. She tearfully says, “If anything happened, I can’t remember.”

You also interview the father alone. He says he and his wife “keep a tight rein” on their daughter. She has been out of their supervision only for the recent retreat. He is concerned that his daughter may have been given a “date-rape” drug and requests testing. His demeanor seems appropriate during the conversations both in the presence and absence of his daughter.

According to state law, you notify the Department of Social Services of unexplained perineal trauma. You also contact the local sexual abuse/rape experts in your area, who have arranged for an evaluation the following day. The father assists in making the arrangements for the evaluation while he is still in your office.

■ **Department of Social Services forensic interview and medical examination**

The next day, the patient and her family undergo a comprehensive evaluation by local medical and investigative professionals from the Department of Social Services.

The differential diagnosis for genital ulcers in a sexually inexperienced female

includes sexual abuse, herpes simplex virus, Behçet’s disease, Epstein-Barr infection, pilonidal disease, Crohn’s disease, and hidradenitis suppurativa. Definitive diagnosis can be difficult (TABLE 1).

One retrospective series reviewed the case of 9 adolescent females with vulvar ulcers and found that 6 had no formal diagnosis. Most important, the initial presentation should prompt healthcare professionals to take steps to ensure a patient’s safety.

After performing a physical examination, the abuse experts report that the patient’s presentation is consistent with Behçet’s disease. A genital culture was obtained, and the patient was given pain medication and azithromycin.

The expert also stated that the father’s reaction to the situation was appropriate and not that of an abusive father. Most perpetrators are very hostile and defensive, whereas this father was extremely concerned and cooperative. Still, a report to Child Protective Services had to be made because of the unexplained physical findings, especially in the genital area.

**Further consideration**

Additional information on Behçet’s disease is found in an article in the *New England Journal of Medicine* from 1999: “Behçet’s disease is an inflammatory disorder of unknown cause, characterized by recurrent oral aphthous ulcers, genital ulcers, uveitis, and skin lesions.”

You ponder this information and note that your patient has no oral lesions currently but apparently has a history of

**FAST TRACK**

**Initial presentation of vulvar ulcers should prompt steps to ensure a patient’s safety**

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**TABLE 1**

### Differential diagnosis of vulvar ulcers in sexually inexperienced females

Sexual abuse
Herpes simplex virus infection
Behçet's disease
Epstein-Barr viral infection
Crohn's disease
Pilonidal disease
Hidradenitis suppurativa

some oral lesions. You review your records indicating a history of recurrent perioral rash but do not find a history of oral lesions. However, the consultant remains constant in his diagnosis.

You review the results of the ED laboratory evaluation:

- Gonorrhea/chlamydia DNA probe negative
- Herpes culture and herpes immunoglobulin M (IgM) negative

- Rapid plasma reagin (RPR) nonreactive
- Serum pregnancy negative
- Urine drug screen negative for PCP, benzodiazepines, amphetamines, THC, opiates, barbiturates, methadone, tricyclic antidepressants
- Complete blood count normal; white blood count 9600 with a normal differential

As the family physician, you interpret these results and support the clinical plan for this patient, pending further changes in the clinical picture.

### ■ Next contact— ED visit 2 days later

The patient's vaginal pain is worsening and she cannot void. Large ulcers with adhered pus are on the left labia; a smaller lesion is on the right labia. There are also questionable lacerations of the posterior fourchette. She is catheterized and 300 mL urine is emptied from the bladder. A gynecology consultation is obtained by the ED physician with your concurrence.

### FAST TRACK

**In Behçet's, oral ulcers appear in 70% of cases; genital lesions are often present as well**

**Q: This patient has had an extensive work-up. Is there any other diagnostic testing you would consider?**

**A:**

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**Q: Is this Behçet's disease?**

**A:**

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### ■ Details of Behçet's disease

Behçet's disease is most common in the third or fourth decade of life. It has an association with the human leukocyte antigen HLA-B51 and HLA-B5 allele. Vascular injuries, hyperfunction of neutrophils, and autoimmune responses lead to the clinical findings. Behçet's disease is presumed to be an autoimmune disease, with the primary lesion being related to the vasculitis.

### Clinical criteria of Behçet's

Diagnosis of Behçet's disease is based on clinical criteria. No single test can determine if a patient has Behçet's.

An International Study Group of physicians was convened to develop a set of guidelines to diagnose Behçet's disease. These criteria include recurrent, painful oral ulceration with at least 2 other symptoms (recurrent genital ulcers,

eye lesions, skin lesions, and positive pathergy test) (TABLE 2). These diagnostic criteria may be accessed at the website of the American Behçet's Disease Association, www.behcets.com.

### Prevalence of signs

Oral ulcerations are the presenting signs in about 70% of cases. These ulcers may remain for as long as 3 weeks. In women, the genital lesions are often present in the vulvo-vaginal region; in men, they often appear in the scrotal area. Ocular complaints occur in about one-half of patients, sometimes with photophobia, watering, and blurred vision. The most common skin lesions are pseudofolliculitis and erythema nodosum.

The pathergy test uses a sterile needle to make a skin prick. The test result is positive if an aseptic erythematous nodule or pustule (>2 mm in diameter) occurs within 24 to 48 hours, indicating neutrophil hyperfunction.

### Differential diagnosis

Skin, joint, gastrointestinal, vascular, and central nervous systems may also be involved. The differential diagnosis includes chronic oral aphthosis, herpes simplex virus infection, Sweet's syndrome, ankylosing spondylitis, inflammatory bowel disease, and multiple sclerosis (TABLE 3).

### Laboratory findings and treatment

Examination of the blood can reveal nonspecific findings consistent with inflammation, including elevated C-reactive protein, erythrocyte sedimentation rate, and positive markers for autoimmune diseases.

**Treatment** depends upon the particular symptoms and clinical findings and must be coordinated with the various specialists involved.

Topical steroids are used for oral and genital ulcers as well as ocular lesions.

Oral steroids are useful for gastrointestinal and neurological symptoms but may also help with skin, joint, vascular, and ocular symptoms.

**TABLE 2**

### Diagnostic criteria of Behçet's disease

#### Recurrent oral ulceration

- Aphthous ulcers or herpetiform ulcers
- Three times in 12-month period

#### AND AT LEAST 2 OTHER SYMPTOMS:

#### Recurrent genital ulcers

#### Eye lesions

- Uveitis, vitreous cells on slit-lamp exam, retinal vasculitis

#### Skin lesions

- Erythema nodosum, pseudofolliculitis, papulopustular lesions, or acneiform nodules

#### Positive pathergy test

- Determined at 24 to 48 hours

Adapted from Sakane et al, *N Engl J Med* 1999.<sup>1</sup>

**TABLE 3**

### Differential diagnosis of Behçet's disease

Chronic oral aphthosis

Herpes simplex virus infection

Sweet's syndrome

Ankylosing spondylitis

Inflammatory bowel disease (Crohn's disease, ulcerative colitis)

Multiple sclerosis

Adapted from Sakane et al, *N Engl J Med* 1999.<sup>1</sup>

Colchicine may help alleviate oral and genital ulcers as well as skin, ocular, and joint problems.

Topical tetracycline may be used to treat oral ulcers.

Anticoagulants may be indicated for vascular disturbances and progressive central nervous system lesions.

Cytotoxic and antirheumatic agents may treat vascular, ocular, neurological, and joint manifestations.

### FAST TRACK

**Treatment of Behçet's depends on symptoms: oral and topical steroids, colchicine, topical tetracycline, anticoagulants, cytotoxic and antirheumatic agents**

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**Q: If the patient does not meet the criteria for Behçet's disease, what are the next steps?**

**A:** \_\_\_\_\_  
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### ■ Gynecology consultation

The gynecologist recommends that the patient be admitted to the hospital for examination under anesthesia. This examination found an ulcerated area of perineum, cellulitis, and edema, without a palpable abscess. Necrotic tissue is debrided.

Biopsies are obtained for herpes, aerobic, and anaerobic cultures. Repeat gonorrhea and chlamydia polymerase chain reaction (PCR) probes are obtained as well. The patient is started on ampicillin/sulbactam (Unasyn) after the debridement.

Further laboratory testing is also done at admission, including a throat culture, cryoglobulin, hepatitis screen, antinuclear antibody (ANA), HIV, lupus anticoagulant, cardiolipin, herpes IgG, antithrombin III level, immune complex detection panel for C1q, and tissue transglutinin AB4 IgA. All results are negative. A possible rheumatology consultation is also discussed.

Culture results from the previous sexual abuse evaluation show strep and gram-negative rods. Initial cultures from exam under anesthesia reveal gram-positive cocci, gram-positive rods, and gram-negative rods on the Gram's stain with final report pending. An infectious disease consultation is recommended. The family physician agreed.

### ■ Infectious disease consultation

The consultant believes streptococcal infection is the most likely culprit and

recommends changing the IV antibiotic from ampicillin/sulbactam to piperacillin/tazobactam (Zosyn) with the addition of clindamycin for broad-spectrum coverage. Later, the final culture results from the sexual abuse evaluation show heavy *Enterococcus* colonization. Surgical culture results reveal moderate *Escherichia coli*, moderate *Enterococcus*, light *Staphylococcus epidermidis*, heavy bacteroides, and moderate *Lactobacillus*. All are susceptible to the piperacillin/tazobactam.

The patient does well and is successfully switched to oral amoxicillin/clavulanate (Augmentin) with no recurrent fever and continued healing of vaginal ulcers. She is sent home to continue to heal.

One week later, the patient is seen in follow-up with continued healing and no further lesions. A few months later, the patient has healed completely and no other lesions have occurred. Incidental discussions included recent menarche. The family physician, patient, and patient's family do a final review of all clinical and pathological findings.

The pathology report shows inflammation and necrosis. In addition, the vulvar biopsy reveals "multiple hair follicles adjacent to the inflammation and necrosis, raising the possibility of infected hair follicles and pilonidal disease."

Unlike the implications with the presumed Behçet's, the patient's localized disease process has no long-term health consequences. She is discharged with a diagnosis of labial ulcers with cellulitis, resolved urinary retention, and a pathological diagnosis of pilonidal disease.

### FAST TRACK

**Further testing includes biopsy for herpes, PCR, throat culture, ANA, cardiolipin, and tests for hepatitis, HIV, and lupus**

**Q: What considerations exist in the diagnosis of pilonidal disease?**

**A:**

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### ■ Pilonidal disease

Pilonidal disease, this patient's final diagnosis, is more common than Behçet's disease. Men are affected more often than women, and ages range from puberty to the third decade of life with decreased frequency after the age of 45.

Pilonidal disease becomes a problem when "nests of hair" become infected. It is mostly found in the natal cleft but can present in almost any hairy area, mostly in the midline. Other examples include the umbilicus and scalp. There are also reports of acquired pilonidal disease from trauma in the hands of barbers, hair dressers, sheep shearers, and animal groomers. It is important to distinguish pilonidal disease

from other disorders in the same region, such as anal fistulas, perirectal abscesses, and hidradenitis suppurativa.

Recurrences of pilonidal disease are quite common, with painful inflamed masses, abscesses, or sinus tracts. An acute abscess must be incised and drained. Hair removal by laser may prevent the growth of hair temporarily. For recurring infections, the definitive treatment is surgical. Procedures range from simple incision and curettage to excision of the area with plastic flap reconstruction. Antibiotics are not routinely needed, but they may be appropriate when cultures indicate specific microbes, when significant cellulitis is present, or for perioperative prophylaxis.

**Q: What are the comparative likelihoods of pilonidal disease and hidradenitis suppurativa?**

**A:**

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### Could it be hidradenitis suppurativa?

Hidradenitis suppurativa is a possibility, but the clinical and pathological findings make pilonidal disease more likely. Hidradenitis suppurativa similar to pilonidal disease, but it usually is located in the axillary, perineal, or inguinal areas. Incidence is as high as 1 in 300, and it frequently begins with puberty.

Hidradenitis suppurativa differs from pilonidal disease in that the hair follicle becomes occluded due to a defect in the epithelium of the follicle's terminal ends.

This defect leads to trapping of bacteria and formation of firm, pea-sized nodules or cysts. Some associations between hidradenitis suppurativa and sex hormones have been proposed, but hormonal states of pregnancy, menstruation, and menopause have not shown clear association with hidradenitis suppurativa. Initial symptoms of hidradenitis suppurativa may be pruritus, erythema, and hyperhidrosis. Recurrences are common. Treatment includes medications such as antibiotics, antiandrogens, and retinoids, as well as surgical excision. ■

### FAST TRACK

**Pilonidal disease can be found in the natal cleft, umbilicus, and scalp**

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### Family physician commentary

**A**lthough this case is unusual, several key points are worth noting.

- The family physician as case coordinator allows patients access to specialized medical care, particularly if problems are potentially difficult medically or socially. In this instance, multispecialty care with family physician support is what enabled the final diagnosis.
- The question remains: Why is a pre-menarchal female affected by pilonidal disease? The most reasonable explanation is that the patient began to experience other changes associated with puberty including secondary hair growth. The “nests of hair” in the deep intergluteal regions may also have been stimulated, leading to the inflammation, necrosis, and infection demonstrated by the pathology and culture findings.
- Fortunately, in follow up, the patient has done well and has continued to be followed by the family practitioner, who provided ongoing care and interpretation for this family.

**Discussing sexual abuse.** The approach to discussing possible sexual abuse is a difficult one. Many physicians do not feel comfortable

dealing with such a sensitive situation. The physician cannot take away his or her human side when presented with this kind of situation. The key is clear, unbiased communication with both the parent and the child.

The child can initially be interviewed with the parent present. Then the child should be interviewed alone in a nonthreatening environment. When the family physician takes the time to sit and talk with the child, the child may reveal things otherwise not discussed. Any questions must be age-appropriate and open-ended, being careful not to lead the child. The parent should also be interviewed alone in a nonconfrontational manner, stating what is known at the present and what is the plan, including further evaluation and reports to appropriate agencies. Again, a calm demeanor is important. In all of these interviews, clear documentation is paramount.

Referral to child advocacy centers as this patient experienced can also be appropriate. Referral may allow the physician to serve as the case coordinator as done in this instance. The physician can also remain neutral, giving a unique perspective of the family as the situation is investigated.

### FAST TRACK

**When discussing possible sexual abuse, talk to the parents alone in a calm and nonconfrontational manner**

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#### CONFLICT OF INTEREST

The author has no conflicts of interest to declare.

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