Physical examination reveals an anxious child, crying with pain. Her temperature is 38.7°C, heart rate is 109 beats/min, blood pressure is 114/73 mm Hg, respiratory rate is 20 breaths/min, and oxygen saturation is 99% on room air. She has swelling and redness of clitoris, labia majora and labia minora with clear vaginal discharge observed on examination (Figure 1). There are bilateral ruptured vesicles on labia minora with dark discoloration of <1 cm on the right side. She also has moderate tenderness to palpation over the genital area, inner thighs, and bilateral enlarged inguinal lymph nodes. Her laboratory results are as follows: WBC count, 2.56×10^9/mcL (2.56×10^3/L) (52% neutrophils, 38% lymphocytes, 8% monocytes, 0.5% eosinophils); Hgb, 13.0 g/dL (133 g/L); Hct, 39% (0.39); and platelet count, 189×10^9/ mcL (189×10^3/L). Her serum electrolytes and liver enzymes are normal. Her urinalysis obtained through a catheter shows clear yellow urine with negative leukocyte esterase and nitrite tests, 0 WBC/high power field (hpf), and 0 red blood cell/hpf. Viral PCR from genital lesions is negative for HSV 1 and 2. Serology for Cytomegalovirus (CMV) is negative for IgG and IgM. Epstein - Barr virus (EBV) mononuclear spot test and serology panel are negative for acute or past infection. ANA Screen Multiplex, ANCA Screen, and HCG Beta Subunit are all negative. A CT scan of the pelvis with contrast shows swelling and protrusion of the clitoral hood without evidence of any collection (Figure 2). A blood test reveals the diagnosis.

Clinical presentation

A 12-year-old Caucasian girl presents with vaginal pain, redness and swelling of her vulva for 3 days. The onset was associated with fever, chills, and dysuria. She denies any sexual activity or trauma to the area. She has no history of sore throat, skin rash, arthralgia or arthritis, diarrhea, bloody stools, weight loss, abdominal or flank pain, frequency or urgency of urination. She denies ingestion of any prescribed or over the counter medications. The patient has no identified sick contacts. Her past medical history is unremarkable except for a previous episode of vaginal yeast infection 6 months ago.

Physical examination reveals an anxious child, crying with pain. Her temperature is 38.7°C, heart rate is 109 beats/min, blood pressure is 114/73 mm Hg, respiratory rate is 20 breaths/min, and oxygen saturation is 99% on room air. She has swelling and redness of clitoris, labia majora and labia minora with clear vaginal discharge observed on examination (Figure 1). There are bilateral ruptured vesicles on labia minora with dark discoloration of <1 cm on the right side. She also has moderate tenderness to palpation over the genital area, inner thighs, and bilateral enlarged inguinal lymph nodes. Her laboratory results are as follows: WBC count, 2.56×10^9/mcL (2.56×10^3/L) (52% neutrophils, 38% lymphocytes, 8% monocytes, 0.5% eosinophils); Hgb, 13.0 g/dL (133 g/L); Hct, 39% (0.39); and platelet count, 189×10^9/ mcL (189×10^3/L). Her serum electrolytes and liver enzymes are normal. Her urinalysis obtained through a catheter shows clear yellow urine with negative leukocyte esterase and nitrite tests, 0 WBC/high power field (hpf), and 0 red blood cell/hpf. Viral PCR from genital lesions is negative for HSV 1 and 2. Serology for Cytomegalovirus (CMV) is negative for IgG and IgM. Epstein - Barr virus (EBV) mononuclear spot test and serology panel are negative for acute or past infection. ANA Screen Multiplex, ANCA Screen, and HCG Beta Subunit are all negative. A CT scan of the pelvis with contrast shows swelling and protrusion of the clitoral hood without evidence of any collection (Figure 2). A blood test reveals the diagnosis.

Clinical progression

The patient had Foley’s catheter placement due to pain and difficulty to urinate. She was started empirically on ampicillin-sulbactam and vancomycin for possible cellulitis, and awaiting bacterial cultures and laboratory test results. Vaginal swabs for bacterial cultures and nuclear acid amplification (NAA) for Chlamydia trachomatis, Neisseria gonorrhoeae, Trichomonas vaginalis, and Candida albicans were sent and were all negative. Both HSV 1&2 IgG antibodies in serum and Mycoplasma hominis PCR in urine tested negative. Antibiotics were discontinued after bacterial culture results reported no growth. The fever persisted for 1 week and the patient started to have transaminitis with ALT value of 219 U/L (normal, 0 to 55 U/L) and AST value of 268 U/L (normal, 5-34 U/L). The spleen was palpable 3 cm below the costal margin in the second week of illness. Quantitative EBV polymerase chain reaction (PCR) was sent in the blood and the patient had 12,800 copies/ml. EBV serology was repeated with a reported value of EBV VCA IgM antibody >4.0 (positive, >=1.1).

Differential diagnosis

Acute genital ulcers (AGU) in nonsexually active young adolescents have etiologies other than infections. Women with progesterone autoimmune dermatitis develop AGU that recur in the late luteal phase of their menstrual cycle when hormone levels are high. AGU were also reported in some pubertal premenarchal females or in the first year after menarche, which might be explained by high
Clitoromegaly and vulvar ulcers in a young adolescent girl

10.15406/jpnc.2018.08.00365

Citation:

culture for HSV has a limited sensitivity. Diagnostic tests for syphilis, PCR 1&2 of the lesion to exclude sexually transmitted infection. Viral were reported to have recurrent episodes of AGU.

Around 71% of reported patients in the literature presented with

uncommon. Size of ulcers is variable but lesions can measure greater

than 1 cm in diameter. Most of ulcers resolve with complete healing.

Lipschutz ulcers present as single or multiple ulcers with raised
demarcated borders. Many lesions will develop gray pseudomembrane
or adherent brown eschar with secondary erythema, edema, and
inguinal lymphadenopathy. Our patient had significant clitoral
enlargement which was not reported previously in the literature. Usually, ulcers are distributed on the medial aspects of the labia
minora, but they might be found on the labia majora, perineum, and in
the lower vagina. “Kissing” ulcers on opposing labial surfaces are not
uncommon. Size of ulcers is variable but lesions can measure greater
than 1 cm in diameter. Most of ulcers resolve with complete healing.
Around 71% of reported patients in the literature presented with
systemic symptoms in the form of fever, myalgia, and fatigability. In
addition, 47% of cases had a history of oral ulcers, and around 30%
were reported to have recurrent episodes of AGU.

The laboratory evaluation of AGU should include a viral HSV
PCR 1&2 of the lesion to exclude sexually transmitted infection. Viral
culture for HSV has a limited sensitivity. Diagnostic tests for syphilis,
chancroid, and lymphogranuloma venereum might be considered in
the initial work up depending on the prevalence and epidemiology
of these infections. A complete blood count will help in detecting,
anemia, thrombocytopenia, and neutropenia. Serologic testing for
EBV is highly indicated. Negative serology for EBV with AGU does
not exclude a primary infection with EBV since genital ulcers might
proceed the onset of other systemic manifestations. Detection of EBV
DNA by PCR in serum or directly from the ulcer or vulval swabs
is diagnostic. Serology for CMV IgM and IgG is recommended.
Iron, folate, Vitamin B12 levels, and a biopsy of the vulvar lesion
are indicated for patients with recurrent AGU without identifiable
etiology.

Treatment of non-venereal AGU is mainly supportive. Sitz baths
and a mixture of lidocaine, epinephrine and tetracaine might be helpful
in controlling local pain in AGU. Alternating topical corticosteroid
ointment or amlexanox paste with a local anesthetic are reported to
be helpful. Antibiotics are not indicated except if secondary cellultis
develops during the acute onset of genital ulcers which is reported in
30% of the cases.

Most of non-venereal AGU heal within 16-21 days. EBV or CMV
serology can be repeated especially if the initial test was negative.
Evidence of seroconversion supports the diagnosis of an acute
primary infection and carries a lower risk of recurrence of AGU. It
is important for patients to follow up yearly with their primary care
providers for ruling out progression to other systemic manifestations
that will change the diagnosis.

The patient’s vulvar swelling improved significantly throughout the
admission, and Foley’s catheter was removed 2 days prior to discharge.
The pain medications were tapered off to oral acetaminophen. She
was instructed to use topical corticosteroid ointment and zinc oxide
barrier cream. She was discharged from the hospital on day 14.
The patient’s ulcers healed completely and the liver enzymes dropped
to normal levels within four weeks of the onset of illness.

Lessons for the clinician

• Lipschutz ulcers can be an early manifestation of acute EBV in
young adolescent girls that are not sexually active. AGU might
precede the onset of infectious mononucleosis.

• Negative serology for EBV in the first week of illness does not
exclude the diagnosis.

• Evidence of seroconversion of serology or detection of EBV
DNA PCR in blood or vulvar swabs is diagnostic.

• Lipschutz ulcers secondary to EBV primary infection heal
completely and the treatment is mainly supportive.

• Genital HSV infection should be excluded before diagnosing
patients with Lipschutz ulcers.

• Other laboratory work up for sexually transmitted infections
should be considered on a case-by-case basis.

Funding source

none

Financial disclosure

I have no financial disclosures related to this article to disclose.
Conflict of interest
I have no conflicts of interest to disclose.

References