A 14-year-old previously healthy, postmenarcheal adolescent girl with a family history of thyroid disease and rheumatoid arthritis presented with vulvar pain and swelling. Vulvar pruritus was noted 6 days prior, which worsened and became associated with vulvar swelling, yellow vaginal discharge, difficulty walking, and a fever (temperature, 39.3°C). Her condition did not improve after a course of cephalexin and trimethoprim-sulfamethoxazole. She denied being sexually active or exposing foreign objects or chemicals to the vaginal area.

WHAT’S THE DIAGNOSIS?

a. Behçet disease  
b. candida  
c. chlamydia  
d. Epstein-Barr virus  
e. herpes simplex virus
PHOTO CHALLENGE DISCUSSION

THE DIAGNOSIS:
Epstein-Barr Virus

Physical examination revealed bilateral 1-cm ulcerated lesions on the labia minora with vulvar edema (Figure). She had a palpable liver edge but no splenomegaly, oral ulcers or lesions, conjunctivitis or scleral icterus, or cervical or inguinal lymphadenopathy. A detailed genitourinary examination was performed under anesthesia, but the hymen was not commented on. Inflammatory markers were elevated with a C-reactive protein level of 16.4 mg/L (reference range, 0.08–3.1 mg/mL), erythrocyte sedimentation rate of 39 mm/h (reference range, 0–20 mm/h), white blood cell count of 7.1×10^3/L (reference range, 4.5–11.0×10^3/L) with 57% neutrophils and 30% lymphocytes, an alanine aminotransferase level of 41 U/L (reference range, 10–40 U/L), and an aspartate aminotransferase level of 126 U/L (reference range, 10–30 U/L).

Bacterial and fungal cultures of vulvar tissue were negative as well as blood and urine cultures. Serological tests for herpes simplex virus (HSV), syphilis, and cytomegalovirus were negative, and urine testing for gonorrhea and chlamydia were negative. Serologies for Epstein-Barr virus (EBV) all were strongly positive with an EBV viral capsid antigen (VCA) IgM greater than 160 U/mL, early antigen IgG of 68 U/mL, and EBV VCA IgG of 456 U/mL. Two years after the initial presentation, repeat EBV serologies were obtained, showing a strongly positive EBV VCA IgG (>8.0 antibody index; reference range, 0–0.8), and a negative EBV VCA IgM.

Infectious etiologies of genital ulcers in a sexually active female include HSV, syphilis, lymphogranuloma venereum, and chancroid. Herpes simplex virus often is the assumed etiology of genital ulcers, especially in sexually active patients, and misdiagnosis in the setting of negative HSV testing may be high. Less common infectious causes such as mumps and cytomegalovirus also have been reported.1,2 Lichen planus and lichen sclerosus are noninfectious inflammatory causes, both of which may involve and be limited to the genitals. Autoimmune disorders include Crohn disease and Behçet disease, and vulvar ulcers with an eschar, consistent with aphthous major or complex aphtous, has been used to describe patients with severe recurrent oral and genital ulcerations without other systemic manifestations of Behçet disease.3

Genital ulcers are an uncommon manifestation of EBV infection. The formation of genital ulcers in EBV infection has been hypothesized to be due to immune complex formation during the acute phase that becomes activated in the vasculature, leading to microthrombosis and eventually necrosis of the tissue.4 The mode of transmission for EBV-related acute genital ulcers has been postulated to be hematogenous spread in lymphocytes or EBV shedding in the urine with subsequent transfer to the genital mucosa.5

Epstein-Barr virus–related acute genital ulcers are self-limiting. The average healing time for the ulcers is 14 to 18 days.6,7 Antivirals are ineffective in treating this condition; however, supportive treatment with systemic glucocorticoids for associated swelling and pain medications could be considered. Our patient was treated symptomatically. Two weeks after debridement, granulation tissue was noted at the site and her pain and discomfort had resolved. This case illustrates an uncommon manifestation of EBV in a sexually inactive adolescent and is a reminder for the dermatologist of the diverse spectrum of illness caused by this common virus.

REFERENCES