Bacillary angiomatosis is a pseudoneoplastic infectious vascular proliferative disorder first described by Stoler et al.1 The disorder primarily affects patients with AIDS, but also occurs in other immunocompromised patients, organ-transplant recipients, and, rarely, in immunocompetent patients.2,3 Bacillary angiomatosis presents most commonly as cutaneous papules or subcutaneous nodular lesions, but also occurs in other organs and soft tissues.2,3 Although bacillary angiomatosis mimics the lesions of Kaposi sarcoma, it is readily treated with antibiotics. We know of one other instance in which bacillary angiomatosis involved the vulva,2 but no previous examples of bacillary angiomatosis are known in the cervix. We report the case of a woman with AIDS who had lesions of bacillary angiomatosis on both the vulva and cervix.

Case Report

The patient was a 32-year-old woman with a history of intravenous drug use and prostitution. She had been diagnosed as seropositive for the human immunodeficiency virus (HIV) 2 years earlier, and had a CD4 count of 105 at presentation. Her medical history included herpes simplex virus and trichomonal infections. Before her first admission, she had a 1-month history of diarrhea, weight loss, fatigue, fevers, chills, and shortness of breath. Staphylococcus aureus was detected in blood cultures. Asymptomatic lesions on the vulva and cervix were identified and excised during a routine gynecologic exam. The vulvar lesion was a red-purple nodule located at the junction of the right labium minus and majus. The lesion was firm, well-demarcated, and measured 1.5 cm in maximum diameter. The cervical lesion was at one o'clock position, 2 cm from the cervical os. The nodule was raised, well-demarcated, red-purple, and dome-shaped, and it measured 0.5 cm in diameter (Figure 1). A 1.5-cm purple nodular mass on the hard palate was also identified and biopsied. Following appropriate antibiotic therapy for staphylococcal septicemia, the patient’s fevers resolved and she was discharged on acyclovir, trimethoprim-sulfamethoxazole, and acetaminophen. The biopsies were reviewed after discharge. The hard palate biopsy showed Kaposi sarcoma, and those of the vulva and cervix exhibited features of bacillary angiomatosis. The patient failed to keep her follow-up appointments.

She was readmitted 1 month later with persistent fevers, severe watery diarrhea, weakness, and dizziness. She was febrile, tachycardic, and pale, with cervical and inguinal lymphadenopathy and an enlarged and tender liver and spleen measuring 7 and 4 cm below the costal margins, respectively. The vulvar lesion had recurred and multiple tiny papules were identified around the introitus. An additional 0.8 × 0.3 × 0.8-cm purple nodule was identified on the skin of the thigh. Blood cultures were positive for Staphylococcus aureus, Mycobacterium avium intracellulare, and Bartonella henselae. Cryptosporidia were identified from a duodenal biopsy. An abdominal computed tomography scan revealed a 2.6-cm mass in the liver, which was compatible with an abscess, infarct, hematoma, neoplasm, hemangiomata, or bacillary peliosis. The spleen contained multiple focal lesions, some of which enhanced centrally, compatible with microabscesses, peliosis, Mycobacterium avium intracellulare, tuberculosis, or lymphoma. The patient was first treated with erythromycin 500 mg four times a day, then switched to clarithromycin, ethambutol, clofazimine, and trimethoprim-sulfamethoxazole. She responded well to treatment, and her vulvar, cervical, and thigh lesions resolved. However, the hard palate lesion remained unchanged. She was transferred to a long-care hospice and, after her decision to cease antibiotic therapy, the lesions recurred, and she died of unspecified causes 4 months after her second admission.

On pathologic examination, the vulvar specimen consisted of a single triangular piece of firm red-tan tissue measuring 0.8 × 0.6 × 0.5 cm. One aspect was covered by intact red-purple mucosa. The cervical biopsy was a small irregularly shaped fragment of red-tan-purple tissue measuring 0.2 × 0.2 × 0.2 cm.

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The vulvar and cervical lesions were microscopically similar. The surface squamous epithelium of the vulva showed parakeratosis, marked ballooning degeneration, and disruption, while the surface squamous epithelium of the cervix contained a mild acute and chronic inflammatory infiltrate and was mildly spongiotic. The most striking histologic finding was the presence of circumscribed, lobular subepithelial capillary proliferations (Figure 2).

Individual capillaries were round to oval and variably sized. They were lined by plump, epithelioid endothelial cells, which often protruded into the vascular lumen. The interstitium was mildly edematous, and contained foci of granular amphophilic material, abundant extravasated red cells, nuclear fragments, and a mixed inflammatory infiltrate dominated by neutrophils.

A modified Warthin-Starry stain, optimized for identification of the cat-scratch bacillus, revealed multiple clumps and tangles of darkly staining bacillary organisms (Figure 3). Bartonella henselae was cultured from blood using the techniques and procedures described by Koehler et al.

Discussion

Bacillary angiomatosis is a well-documented clinicopathologic entity that most commonly involves the skin, but it also occurs in other body sites. We are aware of only one previous report of bacillary angiomatosis in the obstetrics and gynecology literature. That report documented the presence of bacillary angiomatosis on the skin of a pregnant woman with AIDS. Our case is the first reported example of cervical bacillary angiomatosis and only the second that we know of with vulvar involvement. Bacillary angiomatosis can mimic several other disease entities. The condition can be treated with antibiotics, but if misdiagnosed or untreated, it can result in disseminated infection or even death.

Lesions of bacillary angiomatosis are typically elevated, dome-shaped or acuminate, rubbery to firm, and tan to bright red to red-purple. They can be friable or ulcerated and bleed profusely when punctured. Individual papules measure from 1 mm to several centimeters in diameter, and tend to become smaller and more widespread with time. Microscopically, our case exhibited the typical histopathologic features of bacillary angiomatosis. Lobular aggregates of well-formed capillaries were lined by enlarged protuberant endothelial cells. The stroma varied from edematous to mucinous or fibrotic, and contained leukocytoclastic debris, an inflammatory infiltrate dominated by neutrophils, and foci of granular amphophilic or purple material. A Warthin-Starry stain revealed darkly staining bacillary organisms within these granular zones. The bacteria were present individually and, more commonly, in clumps and tangles.

Bacillary angiomatosis is most often diagnosed in tissue biopsies, but the diagnosis can also be made by culture or polymerase chain reaction (PCR). Relman
et al. using a PCR technique, first identified Bartonella henselae (formerly known as Rochalimaea henselae) as a causative agent of bacillary angiomatosis. Both B henselae and Bartonella (Rochalimaea) quintana have been isolated from histologically confirmed lesions of bacillary angiomatosis by Koehler et al. In our case, B henselae organisms were isolated from blood cultures.

The differential diagnosis of bacillary angiomatosis includes pyogenic granuloma, endothelioid hemangioma, dermatofibroma, Kaposi sarcoma, angiosarcoma, granulation tissue, leukocytoclastic vasculitis, and verruga peruana. In the immunocompromised patient, differentiating the lesions of bacillary angiomatosis from those of Kaposi sarcoma is the most common and important challenge. The absence of macules, patches, or plaques can help distinguish bacillary angiomatosis from Kaposi sarcoma. However, plaque-like lesions have been described in bacillary angiomatosis. Moreover, bacillary angiomatosis and Kaposi sarcoma can be present together in the same individual, as was seen in our patient, who had histologically proven Kaposi sarcoma in her palate. In contrast to bacillary angiomatosis, Kaposi sarcoma is composed of fascicles of spindle cells that contain hyaline globules, bizarrely shaped vascular lumina, and a predominantly lymphoplasmacytic infiltrate. Kaposi sarcoma does not exhibit leukocytoclasia, a predominantly neutrophilic infiltrate, or amphophilic interstitial granular aggregates, and no bacillary organisms are seen in Warthin-Starry–stained sections.

The drug of choice for treatment of bacillary angiomatosis is erythromycin 500 mg four times a day. However, doxycycline has proven to be effective. Usually, skin lesions show significant clinical improvement after 4–7 days of therapy, with complete resolution within 3–4 weeks. Patients with extensive skin or mucosal lesions, lytic bone lesions, or visceral disease may require prolonged treatment.

In summary, this case documents examples of bacillary angiomatosis involving the female genital tract. Women represent one of the fastest growing groups with AIDS, and, consequently, gynecologists can expect to encounter this lesion with increasing frequency in coming years. Clinicians and pathologists must be aware of this entity and recognize it when present because 1) it can mimic several other disease processes, 2) it is effectively treated with antibiotics, and 3) if untreated, it may progress to disseminated infection and death.

References


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