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Primary Classic Kaposi's Sarcoma Confined to the Vulva in an HIV-Negative Patient

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Dear Editor:

Kaposi's sarcoma (KS) is a vascular neoplasm associated with human herpesvirus-8 (HHV-8) infection that is primarily categorized into 4 different types: endemic (African), iatrogenic (associated with immunosuppressive therapy), AIDS-associated (rapidly progressive form in HIV-infected patients), and classic. The classic type is frequently observed in elderly male patients of Mediterranean origin and Ashkenazi Jews, presenting as an indolent disease with bluish-red (hematoma-like) cutaneous macules that tend to progress only slowly both horizontally and vertically and develop into firm angiomatous plaques and nodules. The most frequently affected sites are the lower limbs and feet; extracutaneous involvement is uncommon[1]. The lesions of KS are rarely localized exclusively in unusual mucocutaneous areas, thus leading to frequent diagnostic errors[2]. To our knowledge, there are no reports of KS confined to female external genitalia not associated with HIV infection. Here, we report the first case of classic KS restricted to the vulva in an HIV-negative patient.

An 85-year-old Caucasian woman presented to our clinic for evaluation of asymptomatic vulvar lesions that appeared over the last year and progressed. Physical examination showed several brownish-violaceous angiomatous nodules and slightly raised plaques on both labia majora and minora (Fig. 1). No other significant mucocutaneous

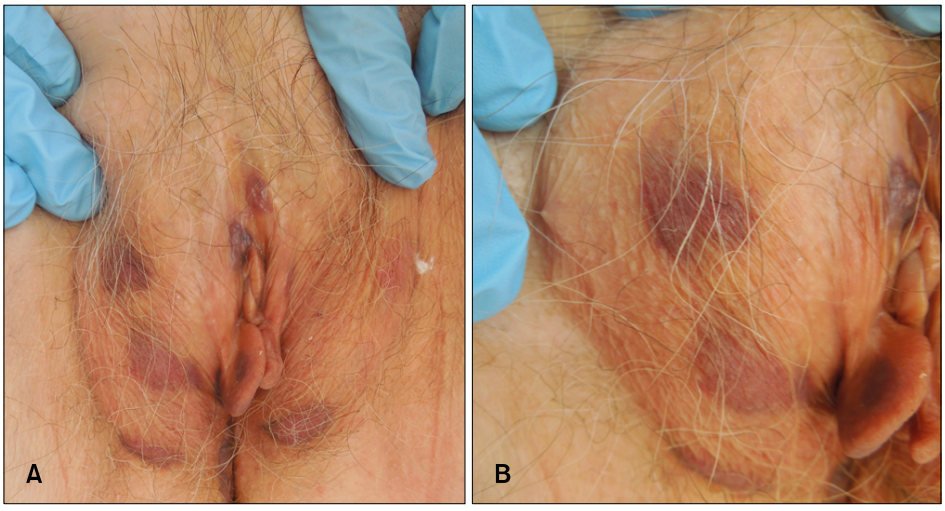

Fig. 1. (A) Several brownish-violaceous angiomatous nodules and slightly raised plaques on both the labia majora and minora, (B) magnified image of the lesions on the right labia majora.

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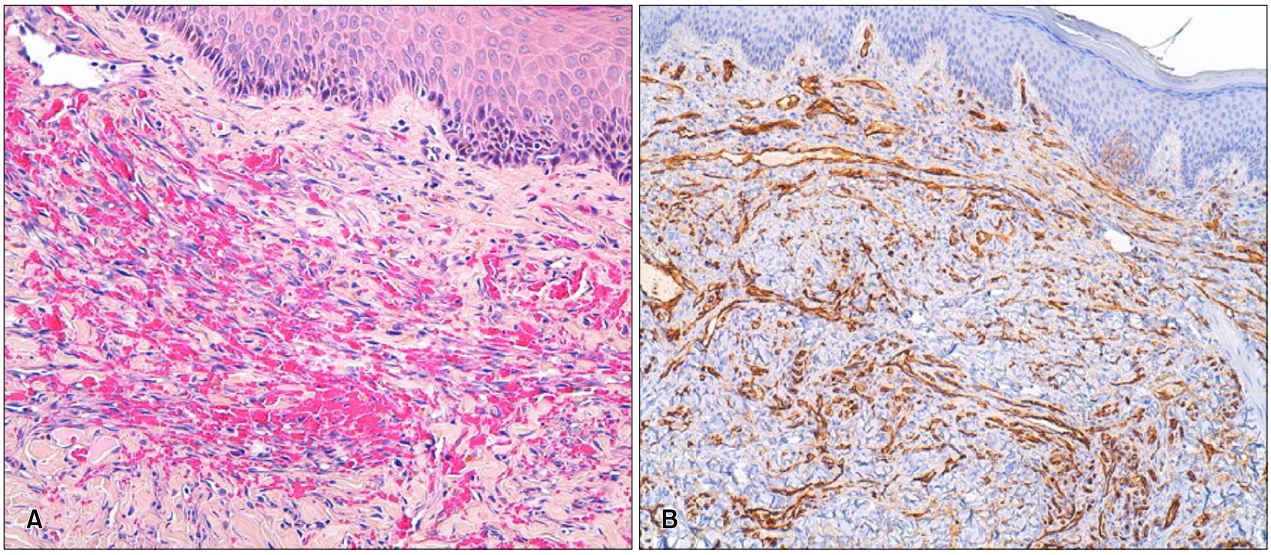

Fig. 2. (A) Dermal proliferation of spindle cells, slit-like vascular spaces, and extravasated red blood cells (H&E, ×200), (B) immunohistochemical positivity for vascular marker CD31 (×100).

lesions were observed, and there was no evidence of inguinal lymphadenopathy. Furthermore, no other signs or symptoms suggestive of HIV infection or immunosuppression were present. The patient denied other health problems and drug use. Skin biopsy from a lesion on the right labia majora was performed for histological examination. The clinical diagnosis of KS was confirmed on the basis of revealing dermal proliferation of spindle cells, slit-like vascular spaces, and extravasated red blood cells (Fig. 2A). Immunohistochemical investigation also demonstrated positivity for D2-40, CD34, and CD31 (Fig. 2B). Moreover, polymerase chain reaction assay revealed HHV-8 DNA sequences in the lesional skin tissue; positive and negative controls were also tested and yielded the expected results. HHV-8 in the lesional tissue was also detected by in situ hybridization. Serological test for HIV was negative; chest x-ray as well as abdominal and pelvic computed tomography showed no visceral lesions. The patient refused all proposed treatments for the vulvar lesions, including intralesional therapy with bleomycin, cryotherapy, and laser photocoagulation. The 6-month follow-up did not show any disease progression.

Overall, KS restricted to the external genitalia is a very rare finding that is more frequent in men, particularly on the penis; approximately 50 cases have been reported in the English-language literature[3]. Meanwhile, it is very uncommon in women, with only 9 reported cases, all of which involved the vulva[2,4]. Most cases of KS of the external genitalia are related to HIV infection. In fact, to our knowledge, only 15 well-documented cases of primary penile KS in HIV-negative patients have been reported[5], while there are no instances of KS confined to female external genitalia not associated with HIV infection. All previous cases of vulvar KS were monolesional forms, clinically presenting as a non-specific mass, nodule, papilloma, or abscess; none were initially suspected to be KS[2].

The present case of classic KS restricted to the vulva is of particular interest because of its more typical clinical appearance than previous cases, with multiple lesions showing an angiomatous aspect as well as onset in an HIV-negative patient; the latter finding emphasizes the need to include KS in the differential diagnosis of vulvar disorders in immunocompetent women, especially elderly women.

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