Not So Classic, Classic Kaposi Sarcoma

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PRACTICE **POINTS**

- Classic Kaposi sarcoma is an indolent neoplasm that can be diagnosed in immunocompetent females of Mexican descent.
- Common diagnoses appearing in skin of color may appear morphologically disparate, and a high index of suspicion is needed for correct diagnosis.

To the Editor:

Kaposi sarcoma (KS) is a lymphatic endothelial cell neoplasm that frequently presents as multiple vascular cutaneous and mucosal nodules.[1] The classic KS variant typically is described as the presentation of KS in otherwise healthy elderly men of Jewish or Mediterranean descent.[2] We present 2 cases of classic KS presenting in Mexican women living in Los Angeles County, California, with atypical clinical features.

A 65-year-old woman of Mexican descent presented to the dermatology clinic for evaluation of an asymptomatic growth on the right ventral forearm of 4 months' duration. Biopsy of the lesion demonstrated a spindle cell proliferation suspicious for KS. Physical examination several months following the initial biopsy revealed a mildly indurated scar on the right ventral forearm with an adjacent faintly erythematous papule (Figure 1A). Repeat biopsy revealed a dermal spindle cell proliferation suggestive of KS (Figures 1B and 1C). S-100 and cytokeratin stains were negative, but a latent nuclear antigen 1 stain for human herpesvirus 8 was positive. Human immunodeficiency virus screening also was negative. Given the isolated findings, the oncology service determined that observation alone was the most appropriate management. Over time, the patient developed several similar scattered erythematous papules, and treatment with imiquimod cream 5% was initiated for 1 month without improvement. She was subsequently given a trial of alitretinoin ointment 0.1% twice daily. The lesions improved, and she continues to be well controlled on this topical therapy alone.

A 62-year-old woman of Mexican descent with end-stage cryptogenic cirrhosis was admitted to the hospital for evaluation of transplant candidacy. Dermatology was consulted to assess a 5×3-cm, asymptomatic, solitary, violaceous plaque on the right plantar foot (Figure 2A) with no palpable lymph nodes. Biopsy revealed a dermal proliferation of slit-like vascular channels infiltrating through the collagen and surrounding preexisting vascular spaces and adnexal structures (Figure 2B). Extravasation of erythrocytes and plasma cells also was appreciated. Latent nuclear antigen 1 staining showed strong nuclear positivity consistent with KS (Figure 2C), and a human immunodeficiency virus test and workup for underlying immunosuppression were negative. The patient had no history of treatment with immunosuppressive medications. Further workup revealed involvement of the lymphatic system. The patient was removed from the transplant list and was not a candidate for chemotherapy due to liver failure.

Kaposi sarcoma is a vascular neoplasm associated with human herpesvirus 8. It typically presents as erythematous to violaceous papules and plaques on the extremities.[1] At least 10 morphologic variants of KS have been identified.[3] The indolent classic variant of KS most commonly is found in immunocompetent individuals and has been reported to primarily affect elderly men of Jewish or Mediterranean descent.

Epidemiologic analyses of this disease in the South American population are rare. More than 250 cases have

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The author reports no conflict of interest.

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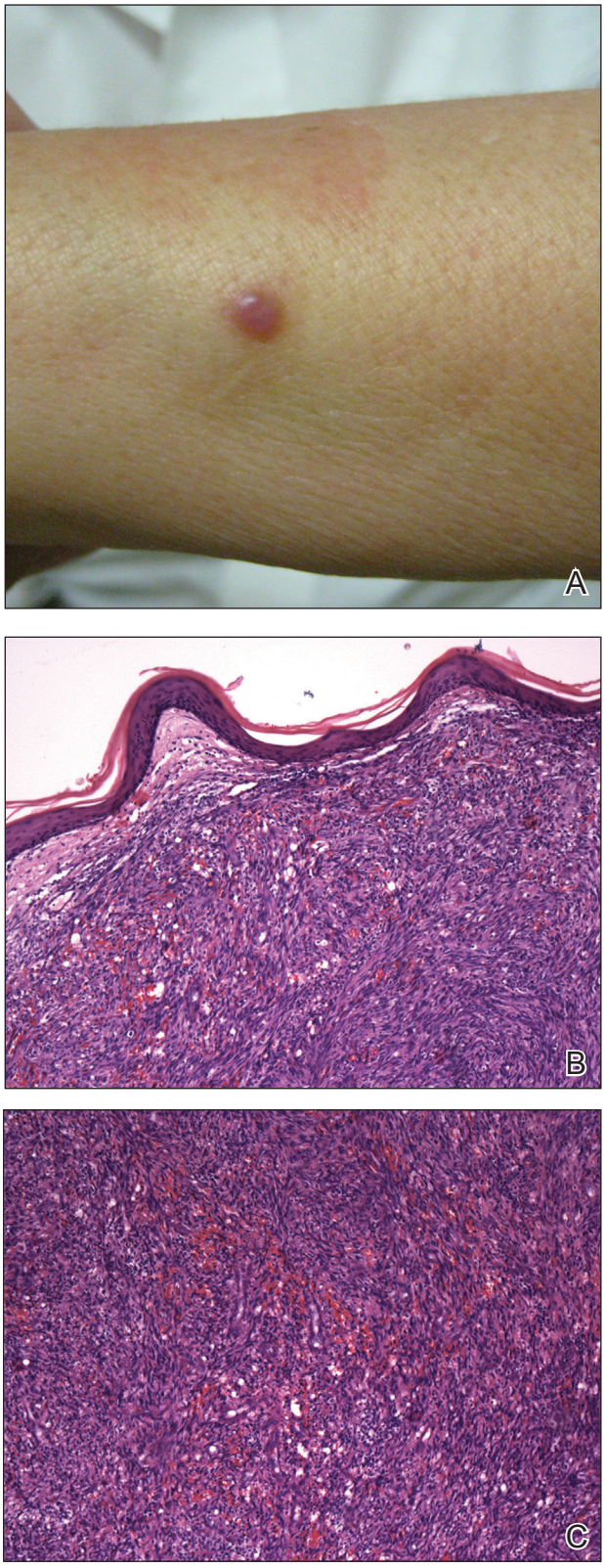

FIGURE 1. A, Mildly erythematous papule on the right ventral forearm. B and C, Histopathology showed a spindle cell proliferation in the dermis suggestive of classic Kaposi sarcoma (H&E, original magnifications ×40 and ×100).

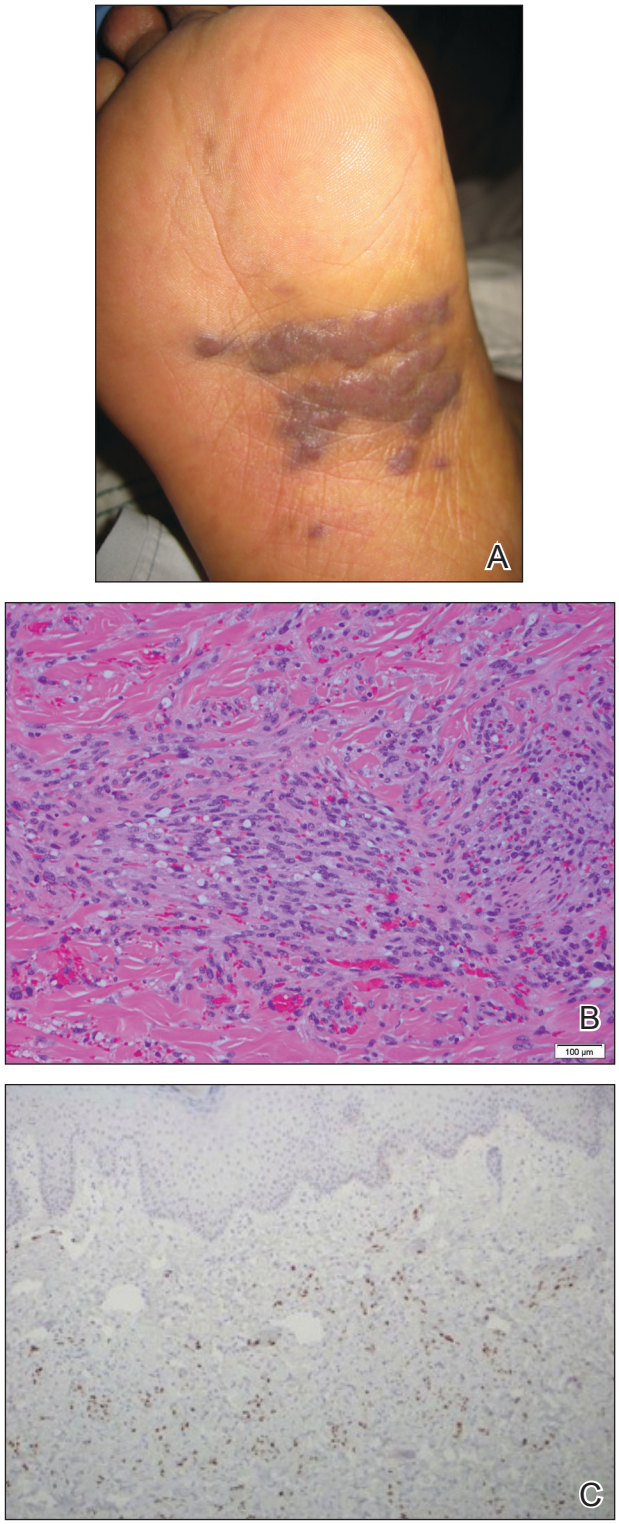

FIGURE 2. A, Violaceous plaque on the right plantar foot. B, Histopathology showed slit-like vascular channels infiltrating through the collagen and surrounding vascular spaces and adnexal structures with extravasation of erythrocytes and plasma cells (H&E, original magnification ×400). C, Latent nuclear antigen 1 staining revealed strong nuclear positivity consistent with Kaposi sarcoma (original magnification ×10).

been published from South American countries with the largest series published from patients in Argentina, Peru, and Colombia.[4] The incidence of classic KS in Peru is 2.54 per 10,000 individuals,[5] and the disease has been diagnosed in 1 of 1000 malignant neoplasms at the National Cancer Institute of Colombia.[6]

A search of the Armed Forces Institute of Pathology Soft Tissue Pathology registry for classic KS patients (1980-2000) showed that 18% of 438 cases of classic KS were diagnosed in South American Hispanics,[7] a percentage that nearly approximates the proportion of patients with KS of Mediterranean descent. Interestingly, in an analysis conducted within Los Angeles County, classic KS was most frequently diagnosed in Jewish, Eastern European–born men who were 55 years or older, followed by European-born women. In this study, white, Spanish, and black populations were diagnosed with classic KS with equal frequency.[8]

Our 2 cases of classic KS from Los Angeles County had several notable features. Both patients were women from Mexico, a demographic not previously associated with classic KS,[8] and they did not have risk factors commonly associated with classic KS. We emphasize that classic KS likely is underreported and understudied in the Mexican and South American populations with need for further epidemiological and clinical analyses.

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