Classification of Kaposi Sarcoma Subtype in a Patient from Cape Verde

ALYSSA M. IURILLO; VICTORIA HOFFMAN; MEGAN HOANG; LAURA BURNS, MD; JACLYN ANDERSON, MD; LESLIE ROBINSON-BOSTOM, MD; OLIVER J. WISCO, DO

An immunocompetent 72-year-old male from Cape Verde presented with a four-year history of progressively spreading violaceous plaques and papules localized to the hands, feet, and lower extremities. His past medical history is significant for coronary artery disease (CAD), essential hypertension, type 2 diabetes mellitus, moderate non-proliferative diabetic retinopathy, dyslipidemia, and status post-coronary artery bypass grafting (CABG) ×4. His current medications include aspirin, atorvastatin, glipizide, sitagliptin, losartan, metformin, metoprolol, valacyclovir, and triamcinolone 0.1% ointment. The lesions were initially asymptomatic, with no systemic symptoms such as fever, weight loss, or lymphadenopathy. Over time, the affected areas became painful and swollen.

On examination, the left medial palm exhibited annular violaceous to purple, non-blanching plaques, while the left fourth digit displayed annular purple to dark brown plaques and papules. The bilateral dorsal feet had well-circumscribed violaceous plaques, with the left side more severely affected. The right plantar surface showed some violaceous plaques and patches. Additional findings included two isolated plaques on the left medial leg and right inner arm. No oral lesions were present. A biopsy of a left plantar foot lesion was diagnostic for Kaposi sarcoma (KS). HIV testing was negative. Initial CT showed no metastatic disease, and follow-up CT remained negative. Histologically, KS lesions demonstrate spindle-shaped endothelial cells forming irregular, slit-like vascular spaces filled with extravasated red blood cells.[1] Hemosiderin deposits, lymphocytic infiltrates, and hyaline globules are frequently observed. Early macular lesions exhibit mild vascular proliferation, whereas nodular lesions display dense spindle cell proliferation and extensive vascular growth. Immunohistochemical staining for HHV-8 latency-associated nuclear antigen-1 (LANA-1) and vascular markers (CD34, CD31 or ERG) are diagnostic for KS.

Given the absence of human immunodeficiency virus (HIV/AIDS) or medication-induced immunosuppression, the diagnosis was classified as classic Kaposi's sarcoma. Treatment options, including observation, surgical excision, intralesional chemotherapy, and radiation therapy, were discussed. The patient elected to proceed with radiation as the primary treatment. He was prescribed and completed five radiation treatments to the left foot and eight to the left fingers. He remains without systemic progression and follows medical oncology.

Figure 1. Kaposi sarcoma affecting the left lower extremity, primarily the dorsal aspect of the left foot, (October 2024).

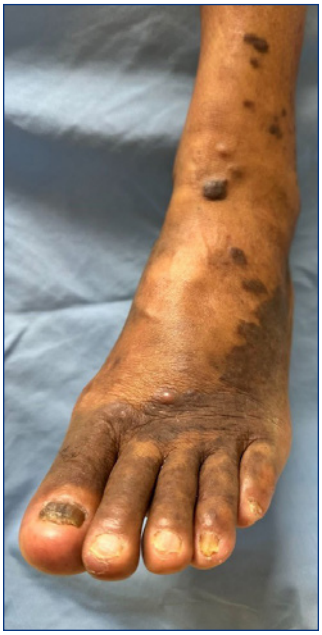
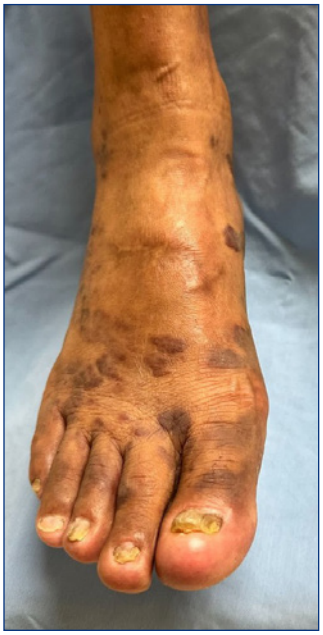


Figure 2. Purple to dark brown macules, patches, and plaques, showing Kaposi sarcoma, observed on the dorsal aspect of the right foot (October 2024).



KS is an angioproliferative neoplasm caused by infection human herpesvirus 8 (HHV-8).[1] KS is classified into four clinical subtypes: classic KS (CKS), African endemic KS, immunosuppression-related KS, and AIDS-related KS. Classic Kaposi Sarcoma (CKS) affects older men of Mediterranean, Eastern European, or Middle Eastern descent, typically during their sixth to seventh decades of life.[2] Although our patient does not have the expected demographic for the classic subtype, his physical exam findings and lack of immunosuppression support CKS. CKS presents as violaceous macules, patches, or nodules localized to the lower extremities, particularly the feet and ankles [**Figure 1**].[3] KS manifests as cutaneous lesions with a wide range of presentations, beginning as scattered pink to purple or dark brown macules and papules [**Figure 2**] and, over time, progressing into larger plaques and nodules, which may ulcerate, become multicentric, and cause significant discomfort [**Figure 3**].[3] While CKS progresses slowly, advanced cases may involve visceral organs. African Endemic KS is prevalent in sub-

Figure 3. Multicentric ulcerated nodules and violaceous patches on the medial longitudinal arch of the left foot (December 2023).

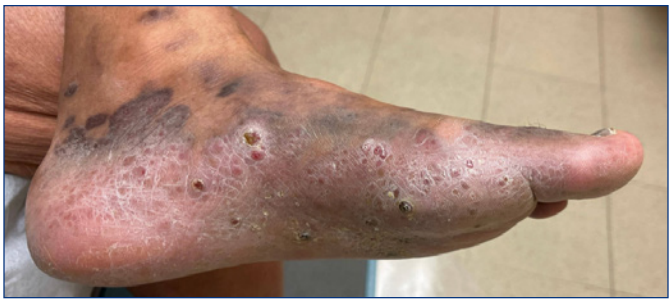
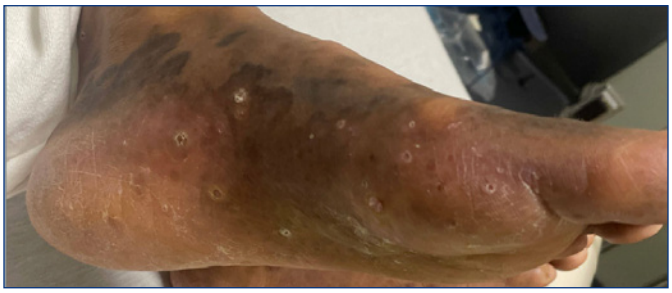


Figure 4. Kaposi sarcoma lesions at various stages of development on the left foot (February 2024).



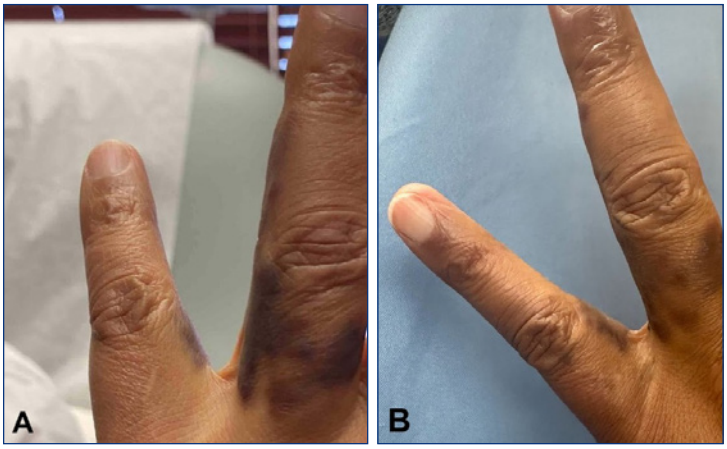
Saharan Africa, where KSHV infection rates are high.[4] It disproportionately affects younger individuals and includes a particularly aggressive variant called lymphadenopathic KS, primarily seen in children.[4] Since the patient was born in Cape Verde and goes back once every few years, the African Endemic subtype cannot be entirely ruled out.

Immunosuppression-related KS occurs in organ transplant recipients and individuals receiving long-term immunosuppressive therapy.[5] Lesions are often cutaneous but can spread to visceral organs. Adjusting or reducing immunosuppressive therapy can lead to disease regression. AIDS-related KS occurs in individuals with HIV infection.[6] Clinically, it presents as multifocal cutaneous lesions and frequently involves visceral sites. Before combined antiretroviral therapy (cART), AIDS-related KS was associated with significant morbidity and mortality.

KS lesions often progress in a chronic, multifocal pattern, with patches, macules, plaques, and nodules commonly appearing at different stages simultaneously [**Figure 4**]. Some local therapies include surgical excision, external beam radiation, and laser therapy.[4] Systemic therapies are reserved for widespread or symptomatic disease. Radiotherapy is particularly effective for localized lesions. A retrospective study analyzing 711 classic KS lesions and 771 HIV-related KS lesions demonstrated traditional X-ray radiotherapy as a safe and effective treatment modality for symptom relief and lesion control [**Figure 5A,B**].[5]

KS is a multifaceted disease with diverse clinical presentations and histopathological features. Subtype identification, early diagnosis, and tailored treatment strategies are essential for improving prognosis and enhancing quality of life.

Figure 5A,B. Comparison of the left hand before (**A**) and after (**B**) radiotherapy treatment for Kaposi sarcoma (April 2024).



References

1. Etemad SA, Dewan AK. Kaposi Sarcoma Updates. Dermatol Clin. 2019 Oct;37(4):505-517. doi: 10.1016/j.det.2019.05.008. Epub 2019 Jul 10. PMID: 31466590.
2. Marcoval J, Bonfill-Orti M, Martinez-Molina L, et. al. Evolution of Kaposi sarcoma in the past 30 years in a tertiary hospital of the European Mediterranean basin. Clin Exp Dermatol. 2019.
3. Anderson LA, Lauria C, Romano N, et al. Risk factors for classical Kaposi sarcoma in a population-based case-control study in Sicily. Cancer Epidemiol Biomarkers Prev. 2008; 17:3435-3443.
4. Radu O, Pantanowitz L. Kaposi sarcoma. Arch Pathol Lab Med. 2013 Feb;137(2):289-94. doi: 10.5858/arpa.2012-0101-RS. PMID: 23368874.
5. Caccialanza M, Marca S, Piccinno R, Eulisse G. Radiotherapy of classic and human immunodeficiency virus-related Kaposi's sarcoma: results in 1482 lesions. J Eur Acad Dermatol Venereol. 2008 Mar;22(3):297-302. doi: 10.1111/j.1468-3083.2007.02405.x. PMID: 18269597.
6. Lebbe C, Garbe C, Stratigos AJ, et al. Diagnosis and treatment of Kaposi's sarcoma: European consensus-based interdisciplinary guideline (EDF/EADO/EORTC). Eur J Cancer. 2019 Jun;114:117-127. doi: 10.1016/j.ejca.2018.12.036. Epub 2019 May 13. PMID: 31096150.

Authors

Alyssa M. Iurillo, Indiana University School of Medicine, Indianapolis, IN.

Victoria Hoffman, Jacobs School of Medicine, Buffalo, NY.

Megan Hoang, The Warren Alpert Medical School of Brown University, Providence, RI.

Laura Burns, MD, Department of Dermatology, The Warren Alpert Medical School of Brown University, Providence, RI.

Jaclyn Anderson, MD, Department of Dermatopathology, The Warren Alpert Medical School of Brown University, Providence, RI.

Leslie Robinson-Bostom, MD, Department of Dermatopathology and Dermatology, The Warren Alpert Medical School of Brown University, Providence, RI.

Oliver J. Wisco, DO, Department of Dermatology, The Warren Alpert Medical School of Brown University, Providence, RI.

Conflicts of interest

None disclosed.

Correspondence

Alyssa M. Iurillo
aiurillo@iu.edu