Kaposi Sarcoma in a Patient With Postpolio Syndrome

Melodi Javid Whitley, MD, PhD; Willis Barrow, MD; Oana I. Craciunescu, PhD; Michelle Pavlis, MD; David G. Kirsch, MD, PhD

PRACTICE **POINTS**

- Cutaneous Kaposi sarcoma (KS) is a human herpesvirus 8–positive tumor of endothelial origin typically seen in older men of Mediterranean or African descent and among immunosuppressed patients.
- In addition, patients with postpolio syndrome characterized by venous insufficiency may represent a population at increased risk for KS.
- Kaposi sarcoma is a radiosensitive vascular neoplasm, and radiation therapy can achieve local control.

Kaposi sarcoma (KS) is a low-grade vascular tumor that is rare among the general US population, with an incidence rate of less than 1 per 100,000.[1] The tumor is more common among certain groups of individuals due to geographic differences in the prevalence of KS-associated herpesvirus (also referred to as human herpesvirus 8) as well as host immune factors.[2] Kaposi sarcoma often is defined by the patient's predisposing characteristics yielding the following distinct epidemiologic subtypes: (1) classic KS is a rare disease affecting older men of Mediterranean descent; (2) African KS is an endemic cancer with male predominance in sub-Saharan Africa; (3) AIDS-associated KS is an often aggressive AIDS-defining illness; and (4) iatrogenic KS occurs in patients on immunosuppressive therapy.[3] When evaluating a patient without any of these risk factors, the clinical suspicion for KS may be low. We report a patient with postpolio syndrome (PPS) who presented with KS of the right leg, ankle, and foot.

A 77-year-old man with a distant history of paralytic poliomyelitis presented for an annual skin examination with concern for a new lesion on the right ankle. The patient had a history of PPS primarily affecting the right leg. Physical examination revealed residual weakness in an atrophic right lower extremity with a mottled appearance and mild pitting edema to the knee. Two red, dome-shaped, vascular papules were appreciated on the medial aspect of the right ankle (Figure 1), and a shave biopsy of the larger papule was performed. Microscopic examination of the biopsy specimen was consistent with KS (Figure 2). This patient had no history of human immunodeficiency virus or immunosuppressive therapy and was not of Mediterranean descent.

Because KS is a radiosensitive vascular neoplasm and radiation therapy (RT) alone can achieve local control,[4] the patient was treated with 6 megaelectron-volt electron-beam

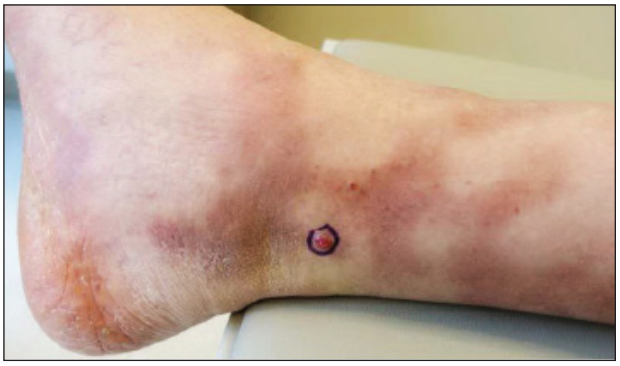

FIGURE 1. Vascular papules on the medial aspect of the right ankle with postpolio sequelae.

From Duke University Medical Center, Durham, North Carolina. Drs. Whitley and Pavlis are from the School of Medicine, Dr. Barrow was from the Department of Pathology, Dr. Craciunescu is from the Department of Radiation Oncology, and Dr. Kirsch is from the Departments of Radiation Oncology and Pharmacology and Cancer Biology.

The author reports no conflict of interest.

Correspondence: David G. Kirsch, MD, PhD, Duke University Medical Center, Box 91006, Durham, NC 27708 (david.kirsch@duke.edu)

RT. He received 30 Gy in 10 fractions to the affected area of the medial ankle. The patient tolerated RT well. Three weeks after completing treatment, he was found to have mild lichenification on the right medial ankle with no clinical evidence of disease. Four months later, he presented with multiple additional vascular papules on the right third toe and in the interdigital web space (Figure 3). Shave biopsy of one of these lesions was consistent with KS. Contrast computed tomography of the chest, abdomen, and pelvis was performed, revealing no evidence of metastatic disease. The patient was treated with 30 Gy in 15 fractions using opposed lateral 6 megaelectron-volt photon fields to the entire right lower extremity below the knee to treat all of the skin affected by the PPS. His posttreatment course was complicated by edema in the affected leg that resolved after daily pneumatic compression. He had no evidence of residual or recurrent disease 6 months after completing RT (Figure 4).

Cutaneous KS is a human herpesvirus 8–positive tumor of endothelial origin typically seen in older men of Mediterranean or African descent and among immunosuppressed patients.[4] Our patient did not have any classic risk factors for KS, but his disease did arise in the setting of a right lower extremity that was notably affected by PPS. Postpolio syndrome is characterized by

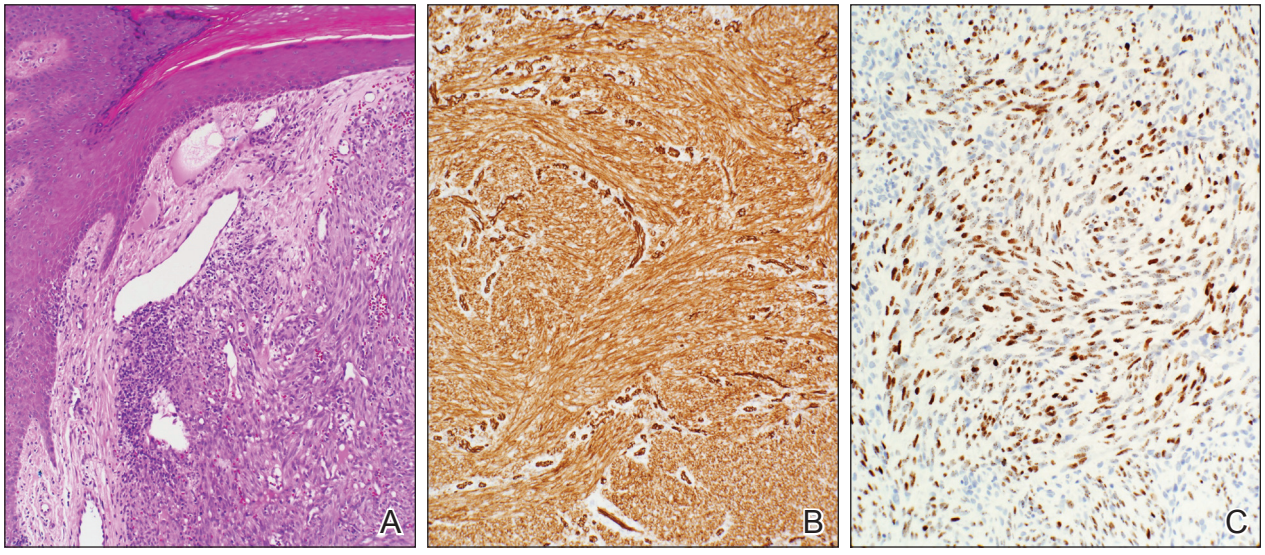

FIGURE 2. A, Microscopic examination showed a well-circumscribed hypercellular spindle cell dermal lesion with an epidermal collarette and peripheral vascular dilation (H&E, medium power). B, Immunohistochemical studies showed lesional cells positive for CD31 (medium power). C, Immunohistochemical studies showed lesional cells positive for human herpesvirus 8 (medium power).

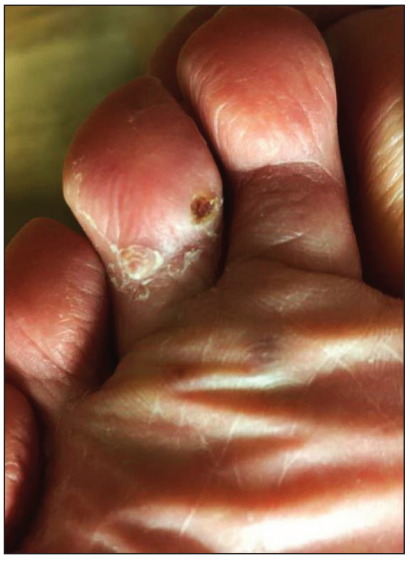

FIGURE 3. Recurrent vascular papules on the third toe before second course of radiotherapy.

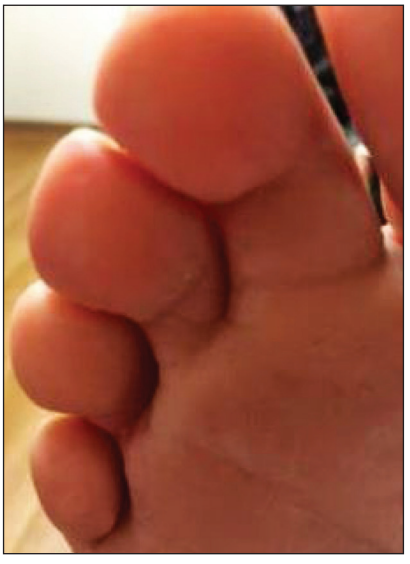

FIGURE 4. No evidence of disease 6 months after the second course of radiotherapy.

muscle atrophy due to denervation of the motor unit.[5] Bruno et al[6] found that such deficits in motor innervation could lead to impairments in venous outflow causing cutaneous venous congestion. Acroangiodermatitis clinically resembles KS but is a benign reactive vasoproliferative disorder and is well known to occur in the lower extremities as a sequela of chronic venous insufficiency.[7] A case of bilateral lower extremity pseudo-KS was reported in a patient with notable PPS.[8] A report of 2 patients describes KS arising in the setting of chronic venous insufficiency without any classic risk factors.[9] Therefore, patients with PPS characterized by venous insufficiency may represent a population at increased risk for KS.

REFERENCES

1. Surveillance, Epidemiology, and End Results (SEER) Program. *US Population Data—1969-2017*. https://seer.cancer.gov/popdata/. Published January 2019. Accessed November 25, 2019.
2. Uldrick TS, Whitby D. Update on KSHV epidemiology, kaposi sarcoma pathogenesis, and treatment of saposi sarcoma. *Cancer Lett.* 2011;305:150-162.
3. Schwartz RA, Micali G, Nasca MR, et al. Kaposi sarcoma: a continuing conundrum. *J Am Acad Dermatol.* 2008;59:179-206.
4. Arnold HL, Odom RB, James WD, et al. *Andrews' Diseases of the Skin: Clinical Dermatology.* Philadelphia, PA: Saunders; 1990.
5. Boyer FV, Tiffreau V, Rapin A, et al. Post-polio syndrome: pathophysiological hypotheses, diagnosis criteria, drug therapy. *Ann Phys Rehabil Med.* 2010;53:34-41.
6. Bruno RL, Johnson JC, Berman WS. Vasomotor abnormalities as post-polio sequelae: functional and clinical implications. *Orthopedics.* 1985;8:865-869.
7. Palmer B, Xia Y, Cho S, Lewis FS. Acroangiodermatitis secondary to chronic venous insufficiency. *Cutis.* 2010;86:239-240.
8. Rotbart G. Kaposi's disease and venous insufficiency. *Phlebologie.* 1978;31:439-443.
9. Que SK, DeFelice T, Abdulla FR, et al. Non-HIV-related kaposi sarcoma in 2 Hispanic patients arising in the setting of chronic venous insufficiency. *Cutis.* 2015;95:E30-E33.