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A case of disseminated angiosarcoma-like Kaposi sarcoma presenting with unusual vesiculobullous lesions

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Abstract

Angiosarcoma-like Kaposi sarcoma represents a recently delineated rare histomorphologic variant of Kaposi sarcoma that can be difficult to distinguish from other vasoproliferative lesions. Conventional lesions of Kaposi sarcoma encompass patches, plaques, and nodules; however, rarely vesiculobullous lesions have been described. Angiosarcoma-like Kaposi sarcoma has never been previously reported to present with vesiculobullous lesions. Herein, we describe a unique case of disseminated angiosarcoma-like Kaposi sarcoma with vesiculobullous lesions as the initial manifestation of human immunodeficiency virus infection.

Keywords

Angiosarcoma-like Kaposi sarcoma, vesiculobullous, disseminated, cutaneous vascular tumors, human immunodeficiency virus

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Introduction

Kaposi sarcoma is a human herpesvirus-8 (HHV-8)-associated vascular proliferation often occurring in the context of acquired immunodeficiency syndrome (AIDS).[1,2] Owing to its vascular origin, lesions may present as pink, purple, red, or brown plaques, patches, and nodules, often involving the lower extremities and face.[3] The conventional histopathological subtypes include the patch-stage, plaque-stage, and well-developed nodular-stage KS.[4,5] Recently, a new distinctive variant known as angiosarcoma-like Kaposi sarcoma (ASL-KS) has been described in the literature. Among the handful of reported cases of ASL-KS, none of these cases were described with vesiculobullous lesions, nor did they present with an aggressive, disseminated form of disease.[6–8]

Case summary

A previously healthy 33-year-old male presented with an 8-month history of a large vegetative right inner thigh wound. The wound began as four small vesicular lesions that progressively developed into numerous pink papules, nodules, and vesiculobullous lesions extending from his right thigh to the dorsal foot, with significant associated serosanguinous and purulent drainage (Figure 1). His social history was remarkable for a history of unprotected sex with two male partners in the preceding 2 years.

He was initially seen at a primary care clinic and treated with oral vancomycin and topical Fucidin with no improvement. A month later, he re-presented to a community hospital and was transferred to a tertiary care center for possible necrotizing fasciitis, where he was assessed initially by plastic surgery and empirically treated with broad-spectrum antibiotics. A CT pelvis showed soft tissue thickening and swelling of the right upper leg and flank without abscess formation. Examination by dermatology revealed a large,

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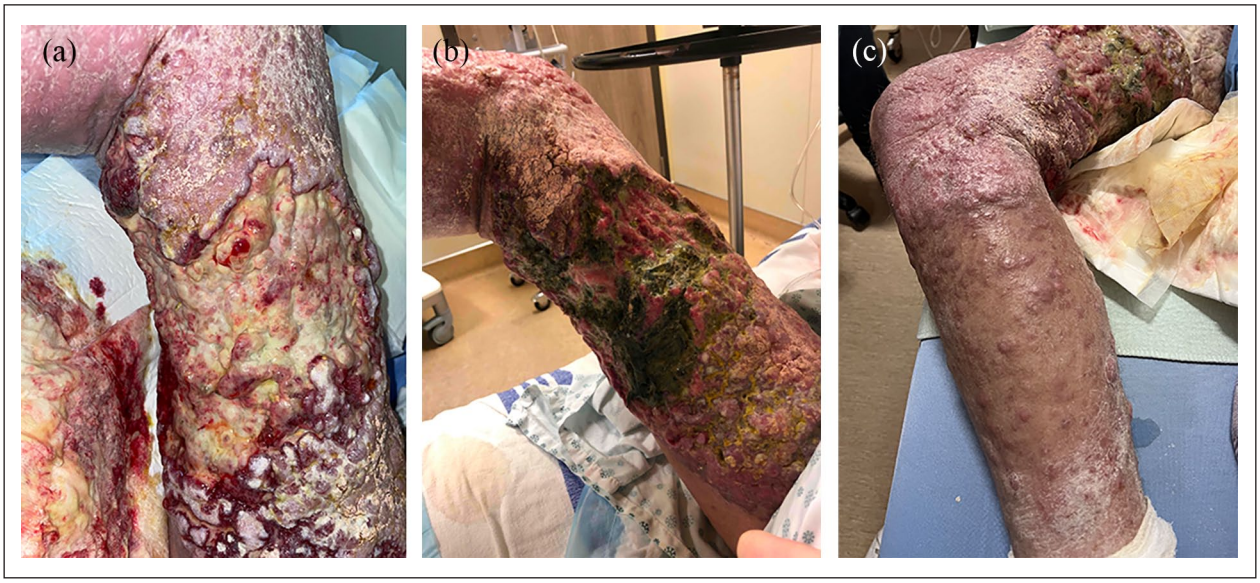

Figure 1. ((a) and (b)) Large vegetative ulcer with malodorous, purulent, yellow/green drainage, extending from the proximal inner right thigh to the popliteal fossa. (c) Numerous pink to erythematous, indurated plaques and nodules extend from the right thigh to the dorsal aspect of the foot.

deep, malodorous ulcerative wound with a pink to violaceous border on the right medial thigh. Granulation tissue was at the ulcer base with overlying yellow and green drainage. In addition, numerous pink to erythematous, indurated plaques and nodules extended from the right thigh to the dorsal foot, with grouped bullae on the medial right ankle (Figure 1). The right leg was considerably edematous in contrast to his largely unaffected left leg. A preliminary clinical diagnosis of pustular pyoderma gangrenosum was made with other considerations on the differential, including atypical infectious etiology and vesiculobullous Sweet syndrome.

Human immunodeficiency virus (HIV)-1, Chlamydia, and Syphilis serology subsequently came back positive, and he was treated with doxycycline, penicillin G, and Biktarvy. HIV viral load at the diagnosis was 157,285 copies/mL with an absolute CD4 count of 19×10^6/L and a CD percentage of 14%. Two skin punch biopsies from the anterior thigh revealed poorly circumscribed dermal based vascular proliferation composed of a meshwork of numerous anastomosing vascular channels dissecting the collagen bundles and lined by a single layer of endothelium without cellular stratification (Figure 2(a) and (b)). Only minimal cytologic atypia was noted in the lining endothelium. The vascular channels had varying caliber, and the proliferation infiltrated the underlying subcutis. A spindle cell component, solid growth pattern, or promontory sign were not seen. In the background, extravasated red blood cells and mild lymphoplasmacytic infiltrate were noted. One of the biopsies showed corresponding papillary dermal edema in the clinical vesiculobullous area (Figure 2(c)). HHV-8 stain showed strong and diffuse nuclear positivity in lesional cells (Figure 2(d)). CD31 highlighted the lining cells (Figure 2(e)); however, smooth muscle actin staining was lost around the proliferating channels. Direct immunofluorescence studies and immunohistochemical stains for cytomegalovirus, Ziehl-Neelsen, and Periodic Acid-Schiff were all negative. Histopathologic findings were compatible with the angiosarcoma-like histomorphological variant of KS.

Subsequent imaging showed widespread lymphadenopathy and lung involvement consistent with disseminated KS. Following discharge from the hospital, the patient underwent palliative radiation at a dose of 20 Gray in five fractions to the wounds on the right thigh and calf. Treatment was well-tolerated, with noted improvement of the right thigh wound (Figure 3).

There was worsening involvement of the right foot and ankle, which were subsequently treated with palliative radiation (20 Gray in five fractions at each site). Upon review by the sarcoma multidisciplinary team, he was initiated on chemotherapy with liposomal doxorubicin. His most recent absolute CD4 count was 153×10^6/L, with an undetectable HIV viral load.

Discussion

Four primary epidemiologic-clinical variants of KS have been established, including classic, African (endemic), and AIDS-associated (epidemic) which are seen predominately in men who have sex with men (MSM), and iatrogenic (including post-transplant) KS.[1–4] AIDS-associated disseminated KS, though rare, is an aggressive entity that often manifests with progressive widespread tumors predominately involving the lymph nodes, and gastrointestinal and

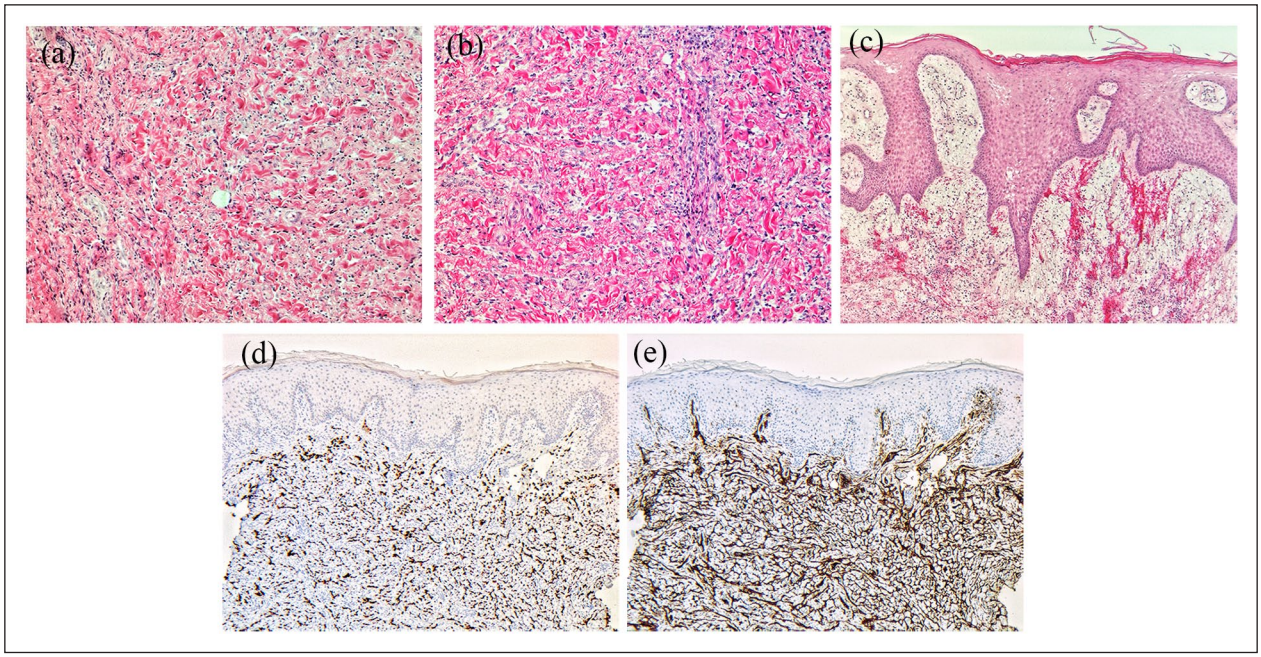

Figure 2. ((a) and (b)) The punch biopsy of skin showing dermal-based infiltrating vascular proliferation formed by meshwork of anastomosing vascular channels dissecting the collagen and lined by a single endothelial layer with minimal atypia (Hematoxylin & Eosin, right thigh, ×100 and ×200). (c) Clinically vesiculobullous area disclosed papillary dermal edema, dilated vascularity, and hemorrhage (Hematoxylin & Eosin, right thigh, ×50). (d) Immunohistochemical stain for human herpesvirus-8 (HHV-8) displayed strong nuclear positivity in the endothelial lining cells (HHV-8, right thigh, ×50). (e) Immunohistochemical stain for CD31 displayed strong positivity in the endothelial lining cells highlighting the meshwork of anastomosing vessels (CD31, right thigh, ×50).

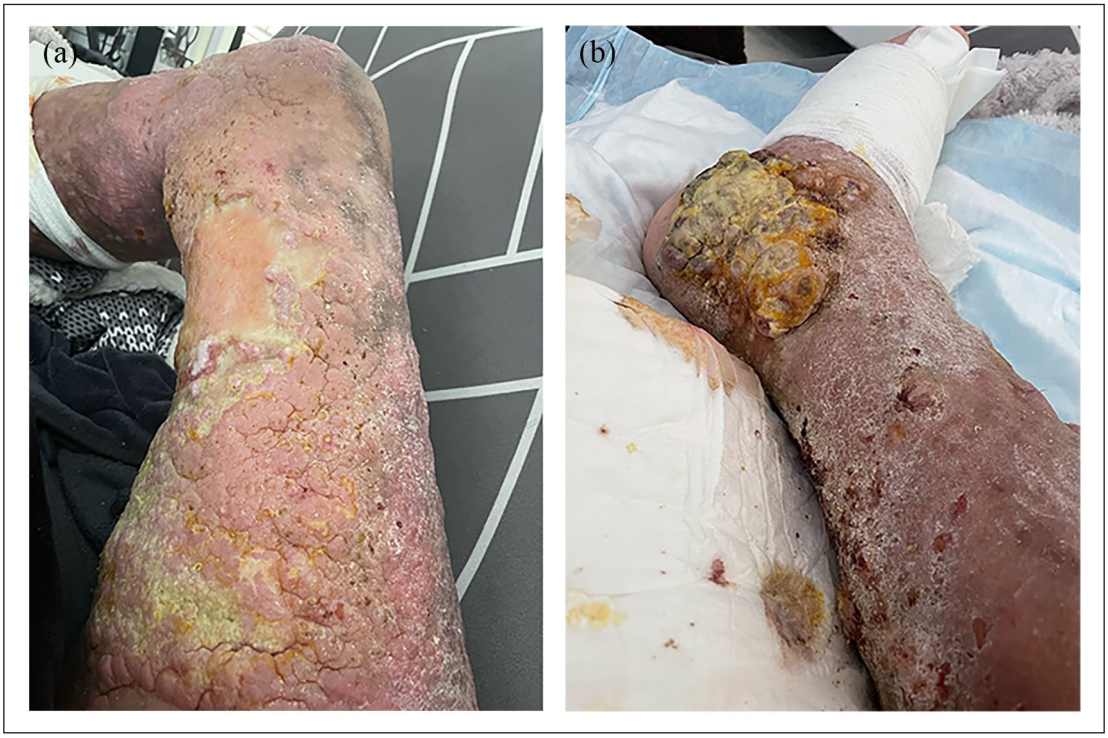

Figure 3. (a) Appearance of right inner thigh subsequent to palliative radiation and initiation of liposomal doxorubicin. (b) Progression of grouped vesiculobullous lesions with surrounding indurated papulonodules involving the medial right ankle which was not initially treated with radiation.

respiratory tracts with an unfavorable prognosis in the absence of therapy.[6]

Microscopic features of KS include spindle-shaped cells and ill-defined vascular channels associated with extravasated erythrocytes and hemosiderin.[2,3] While all conventional subtypes of KS share the microscopic features, several less common histopathological variants have been described in the literature. These variants include anaplastic (pleomorphic), hyperkeratotic (verrucous), lymphangioma-like (lymphangiomatous), bullous, telangiectatic, lymphangiectatic, regressing/regressed, ecchymotic, keloidal, pyogenic granuloma-like, micronodular, intravascular, glomeruloid, cavernous hemangioma-like, pigmented, and KS with myoid nodules.[4,6,7–10] AIDS-related bullous KS tends to present at an earlier age, and dissemination with an aggressive clinical course can be observed.[7] The bullous morphology is attributed to marked papillary dermal edema. Kandemir et al. reviewed a series of 178 lesions from 75 cases and observed vesiculobullous changes in 14%.[7] Lesions showing bullous features were in the nodular stage and showed marked edema and enlarged lymphatic vessels within the papillary dermis.

Angiosarcoma-like KS is a newly characterized histopathologic variant which expands the morphological spectrum of KS. 14 cases of ASL-KS were reviewed by Plaza et al., with 12-HIV positive and 2 HIV-negative patients (transplant-associated and endemic KS). All cases were located on the extremities, mainly on the lower limbs. Histopathologic findings included poorly circumscribed dermal-based tumors infiltrating into the subcutaneous fat, composed of a meshwork of anastomosing vessels dissecting the collagen bundles. Vascular channels showed no cellular stratification and were lined by a single layer of endothelium with minimal pleomorphism. A solid growth pattern and promontory signs were not seen. Findings were reminiscent of a well-differentiated angiosarcoma, and HHV-8 immunostaining was suggested as the key ancillary diagnostic tool to distinguish between ASL-KS, angiosarcoma, and benign lymphangioendothelioma. Notably, none of the reported cases of ASL-KS showed bullous lesions or disseminated disease.[8]

In summary, we describe a unique case of disseminated ASL-KS with vesiculobullous lesions as the initial presentation of HIV infection. KS lesions with a bullous appearance may be confused with other vesiculobullous disorders, potentially delaying diagnosis and management. Therefore, in patients with risk factors, it is important to recognize vesiculobullous lesions as a potential primary clinical presentation of KS. To the authors' best knowledge, this is the first case of ASL-KS displaying the presence of vesiculobullous lesions and disseminated aggressive disease. As such, we encourage dermatologists to maintain a broad differential with the goal of obtaining tissue diagnosis for accurate identification. Additionally, pathologists should include the well-differentiated angiosarcoma-like appearance in the histomorphologic spectrum of KS.

Declaration of conflicting interests

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Ethical approval

No institutional approval is required to publish the case details.

Informed consent

A formal written consent was obtained from the patient for publication of the case details and associated images.

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