ACG CASE REPORTS JOURNAL

CASE REPORT | COLON

Kaposi's Sarcoma of the Rectum in a Homosexual Male with HIV-AIDS

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ABSTRACT

Visceral involvement in AIDS-related Kaposi's sarcoma is common, but it is rarely seen in the absence of cutaneous disease. Most patients with gastrointestinal tract Kaposi's sarcoma remain asymptomatic and are often diagnosed on endoscopy or autopsy. We report a case of a 24-year-old homosexual man who presented with rectal pain and bleeding and was found to have skin-sparing, disseminated Kaposi's sarcoma with rectum, liver, lungs, and lymph node involvement. Despite treatment with highly active anti-retroviral therapy, he developed multiorgan failure resulting in death.

INTRODUCTION

Kaposi's sarcoma (KS) is an angioproliferative cancer derived from endothelial cell lineage.[1] It was an uncommon disease before the outbreak of human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS) in 1981. In the era of highly active anti-retroviral therapy (HAART), the incidence and severity of AIDS-related KS has substantially declined; nonetheless, it remains the second most common HIV-associated malignancy worldwide.[2,3] Although KS is primarily a cutaneous disease, it could involve virtually any visceral organ and could pose a serious diagnostic challenge to clinicians.

CASE REPORT

A 24-year-old black homosexual man with history of HIV-AIDS, cryptococcal meningitis, and hemorrhoids presented with severe rectal pain associated with bloody mucous discharge per-rectum for 2 weeks. He was not on anti-retroviral therapy. He reported intermittent fever and 7-kg weight loss over the past 3 months. On exam he appeared malnourished with oral thrush, and exhibited axillary and inguinal lymphadenopathy. Anal tenderness precluded a digital rectal exam. Skin did not show rash. Vital signs were normal, and initial tests revealed hemoglobin 10 g/dL, white blood cells 11 000/μL (85% neutrophils), platelets 30 000/μL, and normal liver tests. Computed tomography (CT) of abdomen demonstrated inflammation and fluid surrounding the rectum (proctitis) and diffuse adenopathy involving the retroperitoneal and mesenteric lymph nodes. Empirical antibiotics were initiated. Further tests showed CD4 cells 13/μL, HIV viral load 37 000 copies/mL, and negative blood, urine, and stool cultures. Colonoscopy revealed erythematous rectal mucosa with hemorrhagic nodules and a 3-cm friable necrotic ulcer in the rectum (Figure 1). Biopsy displayed active inflammation with negative immunostaining for herpes simplex virus, cytomegalovirus, and acid-fast bacilli. Stool tests for *Clostridium difficile*, chlamydia, gonorrhea, and parasites were negative.

Further, he developed hypoxia, jaundice, and abnormal liver tests (total bilirubin 10.2 mg/dL, alkaline phosphatase 344 U/L, aspartate transaminase 421 U/L, and alanine transaminase 116 U/L), and a repeat non-contrast CT scan demonstrated multiple new, bilateral, nodular opacities in the lungs and multiple new, hypoattenuated lesions in

ACG Case Rep J 2016;3(4):e192. doi:10.14309/crj.2016.165. Published online: December 21, 2016.

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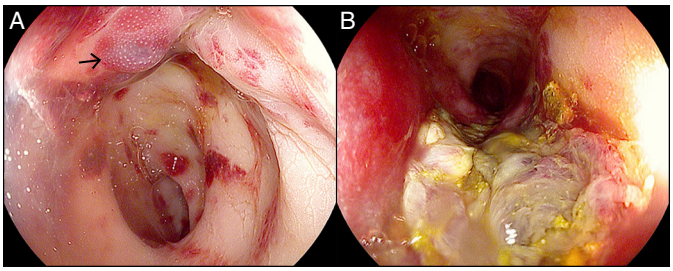

Figure 1. Colonoscopic image showing (A) erythematous rectal mucosa with hemorrhagic nodules (arrow) and (B) a 3-cm friable necrotic ulcer in the rectum.

the liver, along with worsening lymphadenopathy (Figure 2). An axillary lymph node biopsy revealed KS. No evidence of non-Hodgkin lymphoma was present on lymph node or bone marrow biopsies. On further review of rectal biopsies, few areas from the base of the ulcer displayed spindle cells in the lamina propria with extravasated red blood cells (Figure 3). Spindle cells stained positive for human herpes virus-8 (HHV-8), diagnostic of KS (Figure 3). Lung and liver lesions were suggestive of metastatic KS. Based on the AIDS Clinical Trials Group (ACTG) classification by the extent of tumor growth (T), immune status (I) and severity of systemic illness (S), KS was staged as T1I1S1, representing poor risk. Patient was started on HAART with emtricitabine, tenofovir, darunavir/ritonavir, and etravirine.

Despite aggressive measures, his condition continued to deteriorate, leading to pulmonary edema and pleural effusion requiring mechanical ventilation, multi-factorial kidney failure requiring dialysis, anemia, and coagulopathy from worsening liver failure. He died within 2 weeks of initiating HAART. No cutaneous KS lesions were identified until the end of life.

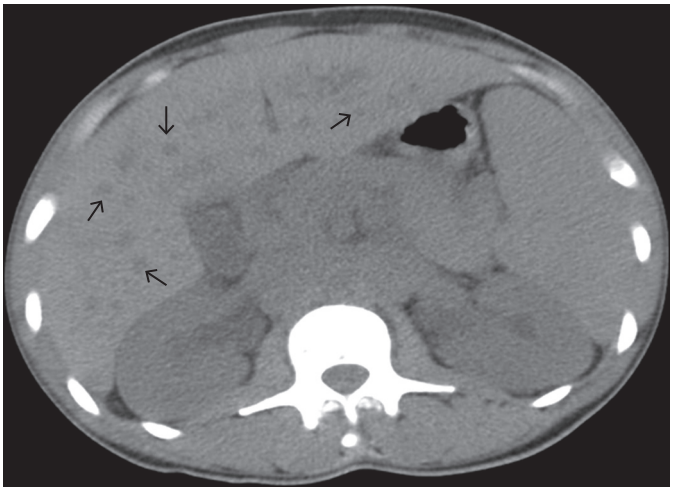

Figure 2. Non-contrast abdominal CT showing multiple hypoattenuated lesions in the liver (arrows).

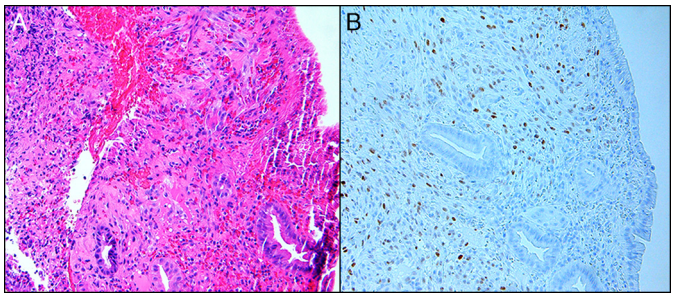

Figure 3. Histopathology image from the base of rectal ulcer. (A) Hematoxylin and eosin stain showing spindle cells in the lamina propria with extravasated red blood cells. (B) Immunohistochemical stain revealing HHV-8 positive spindle cells indicative of KS.

DISCUSSION

KS is a vascular tumor of endothelial origin, caused by HHV-8, typically occurring in the setting of immunodeficiency states.[1,2] There are 4 epidemiological variants of KS: classic, African or endemic, iatrogenic or related to immunosuppressive treatment, and epidemic or AIDS-related.[1-3] Classic KS, first described in 1872 by Moriz Kaposi, is primarily a skin disease affecting elderly men of Mediterranean and east-European heritage and runs a relatively benign course.[1] African KS is endemic in native populations of sub-Saharan Africa, and iatrogenic KS is described in patients receiving chronic immunosuppressive therapy, typically after organ transplants.[2,3] AIDS-related KS is the most common and most aggressive form of KS in the United States, predominantly seen among homosexual/bisexual men with low CD4 counts.[2] Through unclear mechanisms, co-infection with HIV and HHV-8 dramatically increases the incidence and progression of KS.[4] During the early 1980s HIV epidemic, KS was a common manifestation of HIV-AIDS, affecting >50% patients.[5] Initial presentation of KS is usually with mucocutaneous lesions, characterized by reddish-purple or dark black macules, plaques, and nodules.[1,6] Lymph node and visceral involvement is often seen in advanced cases of iatrogenic and AIDS-associated KS.[6]

Of patients with AIDS-related KS involving the skin and lymph nodes, 40–50% of them also have KS lesions in the gastrointestinal (GI) tract (GI-KS).[6,7] Seldom will GI-KS precede the appearance of cutaneous lesions, but rare cases of KS isolated to the GI tract have been reported.[8-11] To our knowledge, we have reported the first case of disseminated, multicentric KS without any identifiable skin involvement until the end of the patient's life. While most patients with GI-KS remain asymptomatic, depending on the location and size of the lesions, they may present with pain, bleeding, or obstruction.[5,7] Diagnosis is through endoscopic visualization of characteristic red, nodular lesions, confirmed with histology demonstrating spindle cells along with inflammatory infiltrate, ill-defined vascular channels, and hemorrhage.[7] Spindle cells

are proliferating HHV-8–infected endothelial cells and stain positive for endothelial markers CD31 and CD34.[1,7] Most GI-KS lesions, including anorectal KS, are submucosal and often difficult to sample by endoscopic biopsy. In such cases, transrectal sonography or echoendoscopy-guided needle biopsies could be used for pathological diagnosis.[7,8,12]

To date, there is no curative treatment for KS, and the goal of therapy is symptom palliation.[13] Management of AIDS-related KS is based on the ACTG staging. HAART remains the mainstay of treatment and represents the first-line therapy for limited cutaneous and slowly progressive disease. HAART with concomitant systemic chemotherapy with drugs like liposomal doxorubicin is indicated in selected patients with visceral, disseminated, or rapidly progressive disease.[1,3,13] The dismal clinical status with multiorgan failure precluded safe administration of chemotherapy in our patient. Ten to 30% of patients may develop paradoxical worsening of existing infections and malignancies following successful immune restoration with HAART, a phenomenon called immune reconstitution inflammatory syndrome (IRIS).[3,13] It is unlikely that our patient had developed IRIS as his KS was already advanced prior to initiating HAART and repeat viral load test failed to suggest significant immune reconstitution within the timeframe of his death.

Rare cases of primary GI-KS continue to surprise physicians, warranting a high index of suspicion for its diagnosis in patients with HIV-AIDS, even without skin lesions. Early endoscopic evaluation with adequate biopsies could help in timely diagnosis and management of GI-KS. Once disseminated, KS is often fatal.

DISCLOSURES

Author contributions: A. Kumar collected the data, wrote the manuscript, and is the article guarantor. D. Nautsch obtained the histopathological images.

Acknowledgements: The authors thank Dipti Sagar, MD, for providing colonoscopic images.

Financial disclosure: None to report.

Informed consent was obtained from the deceased patient's next of kin for this case report.

Received June 3, 2016; Accepted September 16, 2016

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