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CASE REPORT | ENDOSCOPY

Gastrointestinal Kaposi Sarcoma: A Rare Case of an Isolated Rectal Lesion in an Immunocompetent HIV-Negative Patient

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ABSTRACT

Kaposi sarcoma (KS) is a pathological endothelial growth associated with human herpes virus-8 which primarily affects the skin. In HIV-negative men who have sex with men, the clinical presentation of KS resembles the classic form limited to cutaneous or multifocal disease. In this report, we present a unique case of a healthy 61-year-old man who has sex with men with an isolated gastrointestinal KS who does not meet criteria for any of the typical KS clinical variants. Proper follow-up and regular HIV screenings are needed to evaluate the potential progression course of the disease in these patients.

KEYWORDS: rectal kaposi sarcoma; non-AIDS; HHV-8

INTRODUCTION

Kaposi sarcoma (KS) is a pathological condition characterized by a multifocal growth of endothelial cells associated with human herpes virus-8 (HHV-8). KS primarily affects the skin; however, it can be present in lymph nodes, gastrointestinal (GI) tract, and other mucous membranes.[1,2] KS presents in 4 clinical forms. The endemic form is native to equatorial African countries. The iatrogenic form is common in organ transplant patients. The classic form is characterized mainly by skin involvement of lower limbs in Mediterranean-descent patients (Greece, Italy), and the HIV-associated form usually presents with visceral and GI disease.[3] Sporadic cases of KS have been described in HIV-negative men who have sex with men (MSM). However, the clinical presentation of these reports resembled the classic form, limited to cutaneous or multifocal disease.[4] In this article, we report an interesting case of an isolated GI KS in a healthy patient who does not meet criteria for any of the typical KS clinical variants.

CASE REPORT

A 61-year-old HIV-negative White Irish-descent man on pre-exposure prophylaxis (PrEP) for HIV with a history of gastro-esophageal reflux disease and sexual history of MSM presented to the clinic for a screening colonoscopy. At presentation, the patient denied abdominal symptoms such as discomfort, diarrhea, constipation, melena, or hematochezia. He also denied any recent significant weight loss, drug consumption, or travel outside the United States.

Systemic evaluation ruled out any skin lesion, hepatomegaly, splenomegaly, or lymph node enlargement. Routine blood tests, such as complete blood count, liver enzymes, and renal function, were within normal limits. Additional tests for infectious diseases, such as

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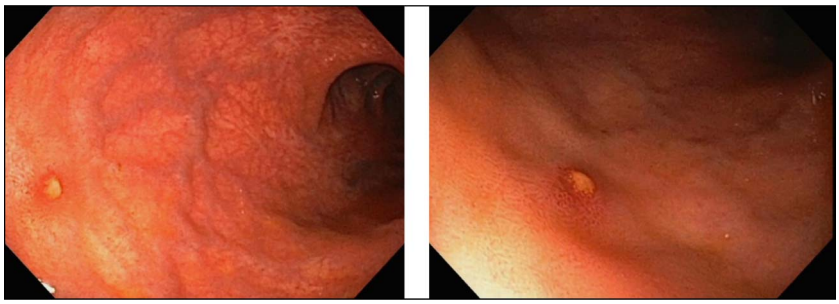

Figure 1. Endoscopic visualizations of KS lesion in the rectum. KS, Kaposi sarcoma.

HIV serology, hepatitis C virus antibody, gonorrhea and chlamydia polymerase chain reaction, and syphilis serology, were negative. An HIV RNA quantitative real-time polymerase chain reaction was also negative.

The colonoscopy showed a single sessile 6 mm subepithelial nodule of benign appearance in the distal rectum (Figure 1). Band endoscopic mucosal resection of the polyp was undertaken. Pathology results demonstrated a mildly atypical spindle cell proliferation forming slit-like vascular channels admixed with granulation tissue most consistent with KS (Figure 2). Immunohistochemical staining for CD31 and CD34 highlighted abundant vascular channels (Figure 3a). HHV-8 was also positive in a large population of lesional cells, and c-KIT was weakly positive in a subset of spindled cells (Figure 3b).

To assess for any other lesions, a PET/CT was ordered showing an increased circumferential F-fluorodeoxyglucose avidity in the rectum related to the patient's biopsy-proven KS. However, no other suspicious uptake outside the rectum was found. The patient is currently followed longitudinally by oncology and gastroenterology every 6 months.

DISCUSSION

Typically, GI KS presents along with skin, mucosal, or visceral lesions. Although isolated GI KS has been reported in the past, the great majority of these cases have been a manifestation of HIV/AIDS-related KS[2,5,6] or iatrogenic KS.[7,8] Sporadic cases of isolated GI KS in immunocompetent, HIV-negative patients have also been reported; however, they have been considered unusual or initial forms of classic KS.[9,10] Based on clinical classification, we present a unique case of an isolated KS lesion in the rectum that does not match the traditional forms of this disease.

The seroprevalence of HHV-8 is estimated to be 1% to 5% in the general US population. Among MSM without HIV infection, the seroprevalence increases to 13% to 20%.[11] Therefore, MSM behavior is a key factor in KS in HIV-negative patients. Friedman et al[12] and Lanternier et al[4] reported 6 and 28 case series of KS in HIV-negative MSM, respectively, mainly describing skin lesions with sporadic concomitant visceral manifestations. Although KS in MSM resembles the classic form with cutaneous affection and, more rarely, the upper GI tract, to this date, there are no descriptions of isolated sigmoid or rectal involvement in non-AIDS MSM related to this variant. In the current era of PrEP, there is concern that an increase in unprotected sexual practices may contribute to increasing exposure to different HHV-8 serotypes with varying oncogenic potential. Interestingly, time of exposure is important as well. As shown by Janis et al, duration of PrEP longer than 3.5 months was associated with HHV-8 infection. By itself, HIV PrEp does not have a direct effect on the life cycle of HHV-8.[14]

In this scenario, since the rectal nodule was an incidental finding in an asymptomatic patient, we can infer an early diagnosis of KS. F-fluorodeoxyglucose PET/CT proves valuable as a noninvasive tool to assess the extent of the disease (lymph nodes, bone, and muscle lesions) guiding further explorations and localizing occult lesions. Likewise, direct endoscopic visualization and sampling of the GI mucosa should be performed if other lesions are suspected due to

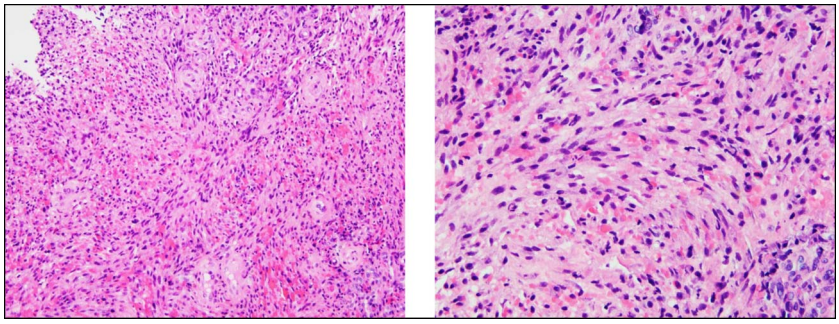

Figure 2. Intermediate to high-power microscopy shows spindle cells arranged in vague fascicles, separated by slit-like vessels with extravasated red blood cells.

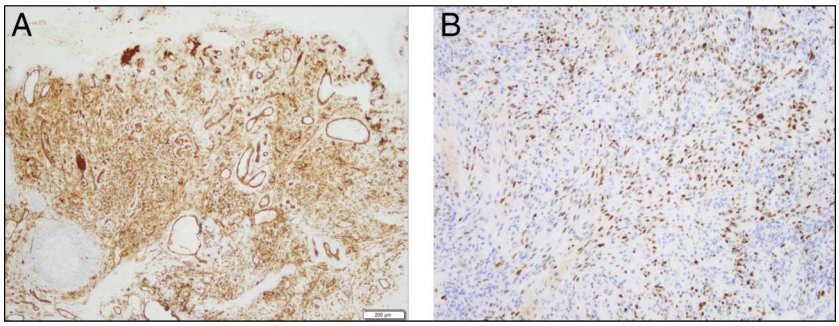

Figure 3. (A) CD31 immunostaining demonstrating the highly vascularized lesion. (B) HHV-8 immunohistochemical stain highlighting the lesional cells.

the potential progression of KS.[15,16] One year after diagnosis, our patient remains disease-free after resection of the rectal nodule.

Compared with prior reports, our case describes mucosal KS in the rectum which differs from most cases of HIV-negative patients in which oral cavity and glans of the penis were the most frequently involved anatomical sites.[17] KS tends not to present as an isolated GI disease in which asymptomatic lesions are usually left untreated. For symptomatic patients, treatment involves local therapy (excision, radiation, cryotherapy) and systemic chemotherapy for systemic advanced disease.[18] In previous cases of HIV-negative MSM with KS, a second neoplasia was found in 14% of patients in the form of lymphoproliferative disorder which highlights the importance of follow-up.[13] Therefore, proper follow-up and regular HIV screenings will determine the course of the disease in this patient.[15,19]

DISCLOSURES

Author contributions: Study concept and design: Sultan, Silva-Santisteban, Berzin. Acquisition of data: Silva-Santisteban. Drafting of the manuscript: Silva-Santisteban, Sultan, Diaz, Berzin. Critical revision of the manuscript for important intellectual content: Silva-Santisteban, Sultan, Diaz, Igbinedion, Holzwanger, Sawhney, Pleskow, Gabr, Berzin. Study supervision: Berzin.

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Informed consent was obtained for this case report.

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