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Gastrointestinal Kaposi Sarcoma without Dermatological Lesions: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

AEF 1 Michael Imeh
AEF 1 Jorge O. Gutierrez
EF 1 Rinku Bhatija
EF 1 Saher Shiza
EF 2 Brian A. Murillo
EF 2 Ryan Lubarsky
AEF 1 Rajan Khanna

1 Department of Internal Medicine, Lincoln Medical Center, Bronx, NY, USA
2 Department of Internal Medicine, New York Medical College, Valhalla, NY, USA

Corresponding Author: Michael Imeh, e-mail: imehm@nychhc.org
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Patient: Male, 35-year-old
Final Diagnosis: Gastrointestinal Kaposi sarcoma
Symptoms: Abdominal pain • fatigue • melena
Clinical Procedure: —
Specialty: Infectious Diseases • General and Internal Medicine • Oncology

Objective: Unusual clinical course

Background: Kaposi sarcoma is a malignancy of the vascular endothelium. It is associated with human herpesvirus 8 (HHV-8) infection, typically found with HIV/AIDS. It is rarely seen presenting as visceral involvement without any cutaneous lesions. Few case reports have described this.

Case Report: We report a case of visceral Kaposi sarcoma (specifically, gastrointestinal lesions) without any cutaneous lesions in a 35-year-old man with HIV/AIDS who presented with abdominal pain, fatigue, and melena of a 15-day duration. Physical examination revealed tachycardia and hypertension, with a negative orthostatic sign. There were no visible signs of bleeding or cutaneous lesions, no abdominal pain, and a digital rectal examination was negative. Laboratory test results were significant for severe microcytic anemia, with hemoglobin level of 3.3 g/dL, decreased ferritin and iron levels, high red cell distribution width, and reticulocyte index lower than appropriate for anemia level. The absolute CD4 count was 33/uL, and the viral load was 56 895 copies/mL. Hemoglobin was optimized with packed red cells prior to endoscopy, and Pneumocystis jirovecii pneumonia prophylaxis was started. Esophagogastroduodenoscopy and colonoscopy revealed small and large bowel hemorrhagic stellate and annular lesions of varying sizes. Pathology reports from biopsy of the lesions seen in the procedure reported Kaposi sarcoma positive for HHV-8. He underwent chemotherapy with doxorubicin and showed clinical and laboratory improvement after treatment.

Conclusions: Kaposi sarcoma should be considered and investigated in patients with HIV/AIDS who are not on highly active antiretroviral therapy and present with gastrointestinal bleeding as an initial symptom, without any cutaneous lesions.

Keywords: HIV • AIDS-Related Kaposi Sarcoma

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/941815

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Background

Kaposi sarcoma is a low-grade neoplasm of vascular endothelial cells [1]. It is associated with human herpesvirus 8 (HHV-8) infection typically found in immunocompromised patients, most commonly those with HIV/AIDS [2].

Up to 20% of patients with HIV/AIDS develop Kaposi sarcoma, which manifests as cutaneous, mucosal, or visceral lesions. Cutaneous lesions are the most common, presenting as smooth, round, pink-purplish painless papules involving the face, upper extremities, and trunk. Visceral lesions mostly involve the gastrointestinal tract, and endoscopy with biopsy is the criterion standard treatment [3]. There are 3 types of endoscopic lesions: type 1, or maculopapular lesions, are characterized by raised scarlet-like lesions; type 2, or polypoid/nodular papular lesions, are darker than type 1; and type 3, or volcano-like nodular lesions, have a central depression [4]. Although these lesions are easily recognized by gastroenterologists, sometimes gastrointestinal Kaposi sarcoma lesions can present as ulcer-like lesions, and histological and immunohistochemical testing allow for the confirmation of the diagnosis [5].

AIDS-associated Kaposi sarcoma has no preferred location, it is widely scattered, and involvement of the lymph nodes and intestine usually occurs early [6]. Visceral involvement of Kaposi sarcoma without cutaneous lesions is rare in patients with AIDS/HIV.

We describe the case of a 35-year-old man with AIDS and visceral Kaposi sarcoma without cutaneous involvement presenting as abdominal pain and melena.

Case Report

A 35-year-old man with a past medical history of HIV and alcohol use disorder presented to the Emergency Department (ED) for abdominal pain, fatigue, and melena. His symptoms started 15 days earlier and progressively got worse. The character of pain was described poorly, with no radiation and localization to the epigastrium. There were no other associated symptoms. There was no ingestion of nonsteroidal anti-inflammatory drugs or any over-the-counter medications prior to symptom onset. He stopped drinking alcohol more than 4 months prior and stopped highly active antiretroviral therapy (HAART) for more than a year because he had not seen a primary care provider. He was meant to be on bictegravir-emtricitabine-tenofovir-alafenamide (BIKTARVY).

In the ED, vital signs were temperature of 37.8°C, heart rate of 109 beats per min, blood pressure of 145/76 mmHg, respiratory rate of 18 breaths per min, and SaO2 of 98% on room air.

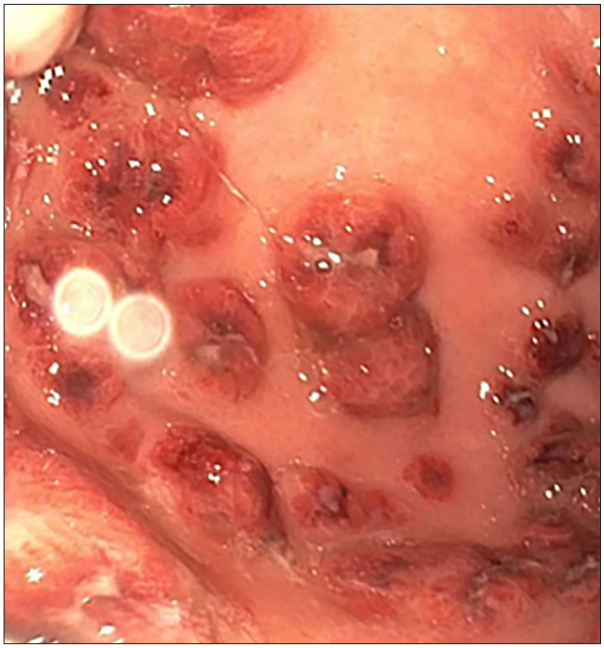

Figure 1. Endoscopic picture of the gastric body showing several hemorrhagic, annular lesions.

The orthostatic sign was negative. The patient presented with a normal appearance, without signs of distress. There was no visible bleeding. Physical examination revealed no dermatological lesions, a normal heart rate and rhythm without murmur, and no abnormal breath sounds. The abdomen was soft, depressible, not distended, and without tenderness. The digital rectal examination was negative for blood. Laboratory test results were significant for severe microcytic anemia, with a hemoglobin level of 3.3g/dL, decreased ferritin and iron levels, high red cell distribution width, and reticulocyte index lower than appropriate for anemia level. A repeated hemoglobin level remained the same. The absolute CD4 count was 33/uL, and viral load was 56 895 copies/mL. He received 2 units of packed red blood cells (RBC) and intravenous pantoprazole, while the Gastroenterology Department was consulted for endoscopy.

During the patient's first day of hospitalization, a repeated complete blood count after 2 packed RBC units showed a hemoglobin level of 6.3 g/dL. One additional unit of RBC was given prior to the endoscopic procedure. The patient had esophagogastroduodenoscopy and colonoscopy, which showed diffuse small-to-large hemorrhagic stellate, annular mucosal lesions seen in the entire stomach examined (**Figure 1**), terminal ileum, colon, and rectum. The lesions were biopsied. As per the gastroenterologist, the findings were suggestive of Kaposi sarcoma. Two additional RBC units were given for the bleeding during the procedure and biopsy. The Infectious Disease Department was consulted for further recommendations and suggested to start HAART therapy, give Bactrim for Pneumocystis jirovecii pneumonia prophylaxis, and wait for pathology results for further

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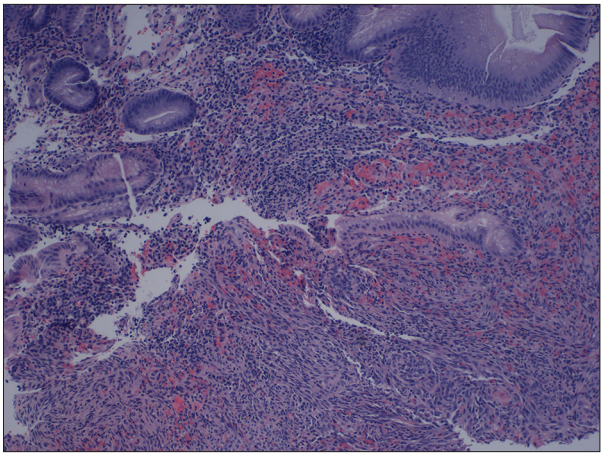

Figure 2. Gastric body biopsy shows atypical spindle cells with mild to moderate nuclear atypia, extravasated red blood cells and rare mitoses (hematoxylin and eosin, original magnification ×100).

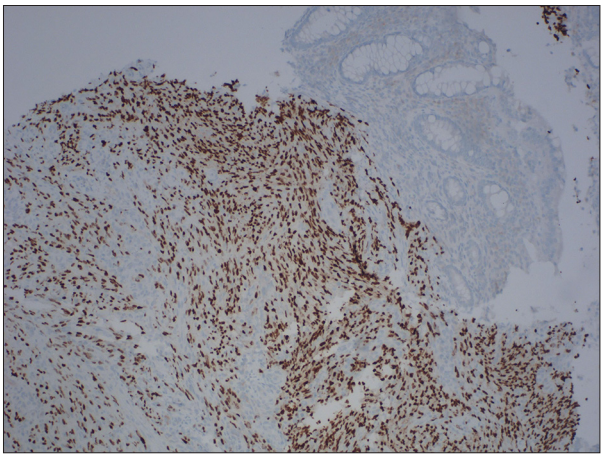

Figure 3. HHV-8 immunohistochemical stain shows nuclear positivity in atypical spindle cells, supporting the diagnosis of Kaposi sarcoma.

workup and treatment. He was restarted on BIKTARVY and is currently still on it. Prior to discharge, the Hematology-Oncology Department was consulted and recommended to give intravenous iron and follow up at the clinic once the results were available. Approximately 1 week after discharge from the hospital, pathology results reported Kaposi sarcoma in the biopsy of the stomach (**Figure 2**) and the colon positive for HHV-8 (**Figure 3**). The patient was contacted to discuss findings and set up appointments with Hematology-Oncology. He is currently under treatment with doxorubicin. Since then, he has completed 6 cycles of doxorubicin, with clinical and laboratory improvements. Computed tomography of the abdomen and pelvis after treatment showed no evidence of metastases or progression of disease. Subsequent arrangements for repeat endoscopy/colonoscopy has been limited by the patient's poor adherence to visit schedules.

Discussion

A huge number of patients with gastrointestinal Kaposi sarcoma are asymptomatic, and therefore, most of these cases are undiagnosed [4]. With continued tumor growth, considerable variation in clinical presentation can be seen, including abdominal pain, nausea, vomiting, iron-deficiency anemia (either chronic or frank gastrointestinal bleeding), and rarely mechanical obstruction alone or combined with bowel perforation [1].

AIDS-Kaposi sarcoma presenting exclusively with gastrointestinal lesions is seldom described in the literature. It is reported that 90% of Kaposi sarcoma presentations involve cutaneous lesions [6], with dark, violaceous plaques and nodules on the epidermis and mucosa. Few case reports have shown Kaposi sarcoma lesions in the setting of gastrointestinal bleeding; however, in most of these case reports, gastrointestinal lesions were also accompanied by dark violaceous cutaneous lesions on the face, trunk, or extremities [7,8].

In the last 10 years, only 3 case reports have described patients with gastrointestinal lesions in the absence of cutaneous manifestations, as in our patient [6,9,10]. One case report by Daar et al in 2017 showed a similar presentation in a 65-year-old man with upper gastrointestinal lesions in the absence of cutaneous lesions, following a sharp drop in the hemoglobin level and findings of stool occult blood on admission. Biopsy showed proliferation of neoplastic spindle-shaped cells arranged in bundles with slit-like capillary spaces containing erythrocytes, and a positive HHV-8 biopsy was able to confirm Kaposi sarcoma [6]. This case was comparable to that of our patient, whose biopsy report showed diffuse annular mucosal lesions, atypical spindle cells with mild-to-moderate nuclear atypia, extravasated RBC, and rare mitoses. Similar findings were also seen in the case reported by Kumar et al in 2016, but unfortunately outcomes in that case differed, as the patient died from complications of multi-organ failure despite HAART initiation [10].

The treatment of choice is HAART and systemic chemotherapy [8]. Gastrointestinal Kaposi sarcoma is an indicator of negative outcomes, and its treatment improves morbidity and mortality [8]. Investigators have suggested screening endoscopies in a selected group of patients for early detection and treatment to improve patient outcomes [4].

Conclusions

This case shows an atypical presentation of Kaposi sarcoma in a patient with AIDS; however, because endoscopy was done timely enough, a proper diagnosis and treatment was done. For an early intervention, physicians must consider Kaposi sarcoma

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in their differential diagnosis of patients with HIV/AIDS with gastrointestinal bleeding, even in the absence of the classic purple patches and nodules in the skin or mucus membranes. Further research still needs to be done to assess the prevalence of such lesions in the absence of symptoms. It would be a steppingstone to further stratify screening procedures to determine which procedures would be impactful in preventing morbidity and mortality from gastrointestinal bleeding in the AIDS-affected population.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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