

Kaposi Sarcoma in Rectum as a Cause of Lower Digestive Tract Bleeding

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Abstract: *Kaposi sarcoma KS is a rare, angioproliferative disorder primarily associated with human herpesvirus 8 HHV8. This article discusses a case involving a 29 year old man diagnosed with HIV and KS, with unusual colorectal and anal canal involvement but no skin lesions. Despite antiretroviral treatment, the patient experienced clinical deterioration, ultimately leading to a fatal outcome. The case emphasizes the rarity and complexity of KS in the gastrointestinal tract, particularly in the absence of external manifestations, and discusses the clinical and histological challenges in diagnosing and managing such cases.*

Keywords: Kaposi sarcoma, HIV associated Kaposi sarcoma, gastrointestinal Kaposi sarcoma, rectal bleeding, angioproliferative disorder.

1. Introduction

Kaposi sarcoma (KS) is a low - grade vascular neoplasm associated with human herpesvirus 8 (HHV - 8) infection. There are four forms of presentation, with the most prevalent clinical variant in our setting being that associated with human immunodeficiency virus (HIV) infection. The morphology, number, color, size, and distribution of endoscopic lesions affecting the gastrointestinal tract are highly variable. It typically presents in men (and women, although less frequently) in their 60s and 70s, but has been described in adolescent patients and those under 20 years of age [4, 5] Only 4 to 8 percent of cases develop in individuals under 50 years of age [6]. Data suggest that HIV - negative men who have sex with men are at higher risk of developing Kaposi sarcoma than men in the general population. It is unclear whether these patients represent a distinct clinical variant of Kaposi sarcoma or whether they should simply be considered part of the clinical spectrum of Kaposi sarcoma. Diagnosis requires histological confirmation, with immunohistochemical testing with antibodies confirming the vascular origin of the neoplastic cells as well as the presence of HHV - 8 being particularly relevant ¹. Treatment is based on a combination of antiretroviral therapy and systemic chemotherapy, using radiotherapy and other local treatments in selected cases. ²

2. Case Presentation

Male, diagnosed with Epilepsy at the age of 15 under treatment with phenytoin, secondary syphilis diagnosed during his current hospitalization under treatment with benzathine penicillin G, diagnosed with human immunodeficiency virus in August 2022 without treatment, who comes to the emergency service of our unit in August 2023 due to dyspnea, odynophagia, fever predominantly at night, rectal bleeding, diarrheal stools in multiple episodes,

weight loss of 10 kilograms in three months prior to admission, physical examination shows fever, hyperemic pharynx, hypertrophic tonsils with violaceous characteristics, multiple bilateral cervical adenopathy, at the level of the penis genitals with an ulcerative lesion on the back of 1x1 cm, not painful, during his diagnostic approach a viral panel with VDRL is requested which is reported positive, a biopsy of the right tonsil is taken with a report of sarcoma of Kaposi's disease associated with HIV. The laboratory findings were as follows: creatinine 1.1 mg/dl; sodium 130 mmol/l; potassium 5.3 mmol/l; glutamate - oxaloacetic transaminase (AST) 20 U/l; glutamate - pyruvate transaminase (ALT) 10 U/l; total bilirubin 0.41 mg/dl; lactate dehydrogenase 135 U/l; the complete blood count showed a hemoglobin of 7.9 g/dl; a white blood cell count of $4.7 \times 10^3 / \mu\text{l}$ with normal formula (67% neutrophils), and a platelet count of $156 \times 10^3 / \mu\text{l}$. The serological study confirmed HIV infection.

Continuing with the approach to anemia associated with rectal bleeding, a colonoscopy was requested, reporting: Perianal region without alterations, upon digital rectal examination: external anal sphincter with preserved tone, rectal ampulla with liquid fecal matter in explorer glove, indurated circular lesion, it is thoroughly examined in anterograde and retrograde manner, with content of little liquid fecal matter, mucosa of ascending, transverse, descending and sigmoid colon, multiple maculopapular, erythematous vascular lesions with fibrin and other violaceous pseudopolypoid lesions between 3 - 9 mm in diameter are observed, in the rectum, thickening of the wall is observed that narrows the lumen by 30%, with a cobblestone and indurated appearance without evidence of hemorrhoidal packages. 15 biopsies of colon and rectum are taken. Pathology report of Kaposi sarcoma in colonic mucosa. In immunohistochemical study, these tumor cells were positive for CD31, CD34 (endothelial cell markers) and HHV - 8 (viral marker).

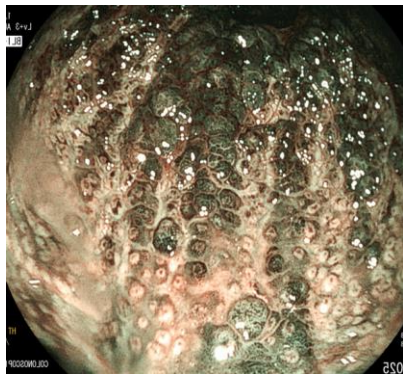


Figure 1

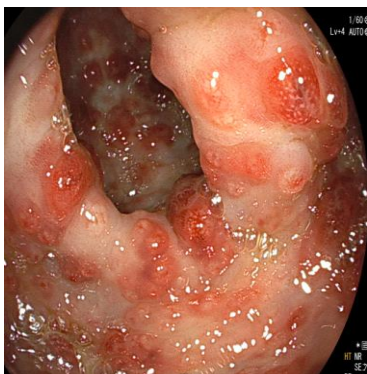


Figure 2

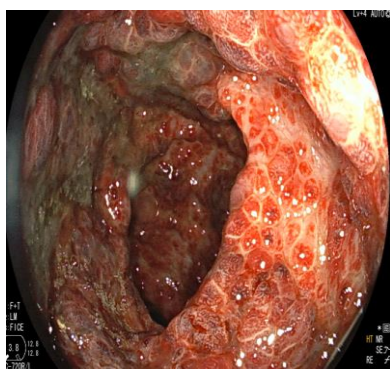


Figure 3

Figure 1, 2, 3: maculopapular, erythematous vascular lesions with fibrin and other violaceous pseudopolypoid lesions between 3 – 9 mm in diameter. Thickening of the wall is observed in the rectum, which narrows the lumen by 30%, with a cobbled and indurated appearance.

In summary, this case report highlights the rare and complex presentation of Kaposi's sarcoma with rectal involvement in an HIV positive patient, emphasizing the importance of early recognition and comprehensive management. Despite the use of antiretroviral therapy, the patient's condition deteriorated, underlining the challenges associated with treating KS in the gastrointestinal tract.

3. Discussion

Kaposi's sarcoma (KS) is an angioproliferative disorder that requires infection with human herpesvirus 8 (HHV - 8), also known as Kaposi's sarcoma - associated herpesvirus (KSHV), for its development [1 - 3]. The disease is named after Moritz Kaposi, a Hungarian dermatologist on the faculty of the

University of Vienna, who first described the entity in 1872 as "idiopathic multiple pigmented sarcoma of the skin" [4]. KS is classified into four types according to the clinical circumstances in which it develops: classic (the type originally described by Kaposi, which typically presents in middle or old age), endemic (various forms described in sub-Saharan African populations before the AIDS epidemic), iatrogenic (a type associated with immunosuppressive drug therapy, typically seen in renal allograft recipients), and AIDS-associated (epidemic KS).

The evolution of HIV-associated KS is extremely variable, oscillating between the minimal expression of the disease found incidentally and a rapidly progressive neoplasia. Its evolution depends fundamentally on factors related to the host's immunity and the extent of the disease. Skin and lymph node involvement is the most common, but the appearance of visceral lesions in the oral cavity, digestive tract and/or respiratory system is not uncommon. Visceral involvement of the gastrointestinal tract affects 40% of patients at the time of diagnosis and can occur in the absence of skin lesions. KS can appear in any location of the digestive tract, although it predominantly affects the upper gastrointestinal tract at the gastric level⁷. In a study with 50 patients with KS with gastrointestinal involvement, only 8% presented colorectal involvement⁸. Rectal involvement is exceptional⁹. Most lesions of the gastrointestinal tract are silent, although they may manifest clinically as weight loss, abdominal pain, vomiting and diarrhea, malabsorption, intestinal obstruction or digestive bleeding.

The morphology, number, color, size and endoscopic distribution of the lesions of the gastrointestinal tract are very variable, and nodular lesions with a hemorrhagic appearance, bluish-purple pseudopolyps or wine-red maculopapules may be found. Histological findings include proliferation of spindle cells, central mitosis, vascular clefts with erythrocytes inside, accompanied by macrophages loaded with hemosiderin and, occasionally, a lymphocytic inflammatory infiltrate. However, it is important to keep in mind that lesions can involve the submucosa, making histological study difficult; For this reason, the immunohistochemical study is essential and helps confirm the pathological suspicion through positive immunoreactivity to markers of vascular lineage (CD31, CD34) and immunohistochemical labeling of viral components of HHV - 8.

The treatment of KS in HIV-infected patients is HAART. To this, adjuvant therapies (systemic chemotherapy, local/topical treatment, radiotherapy, etc.) will be added or not depending on the visceral involvement and extent of the disease⁹. The initiation of HAART can be followed by immune reconstitution syndrome, a paradoxical effect consisting of a worsening of the clinical picture and an accelerated progression of the disease in the first 4–6 weeks as a consequence of the recovery of the patient's immune system⁹. Systemic chemotherapy is generally used in patients with advanced disease or evidence of rapid progression (extensive skin involvement with poor response to local/topical treatment, immune reconstitution syndrome, generalized edema, and symptomatic visceral involvement). The most commonly used chemotherapeutic agents are liposomal anthracyclines (doxorubicin and daunorubicin). Radiotherapy

administered prior to HAART is used, as in our patient, to reduce the volume of localized symptomatic lesions¹⁰. Currently there is no specific treatment directed against HHV-8.

The authors consider the clinical case previously presented relevant for several reasons: This case is significant as it highlights the atypical presentation of Kaposi sarcoma in the gastrointestinal tract, which is uncommon and presents unique diagnostic and therapeutic challenges, particularly in HIV-positive patients without skin manifestations.

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