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KAPOSI SARCOMA OF THE RECTUM IN THE INITIAL PRESENTATION OF HIV - A CASE REPORT

Joyce Regina da Cruz

Danilo Vidal Santos

Leonardo Meneguesso Bidoia

Letícia Godel

Maria Laura Rolim de Moura

Milena Lorenzetti

Ricardo Antônio Hoppen

Ruan Kelvin Wink Schell

Thabata Lechenski Membrive

Valentina Lima Zacharczuk

Wemerson Geraldo de Queiroz Filho

Abstract: This article aims to inform about Kaposi's Sarcoma of the rectum as an initial presentation in the context of HIV, in the form of a case report. This is based on information collected from the medical records of a patient at a Gastroenterology Clinic in the Southwest of Paraná. In this sense, descriptions are made about the tumor, as well as its etiology, development, clinical manifestations and treatment. These presentations are made through the study of scientific articles. In addition, the relationship of the tumor with the manifestation of the human immunodeficiency virus (HIV) and the actions taken individually in the case are addressed. The report will be a 68-year-old male patient, under investigation for anemia, with weight loss of 12 kg in 4 months, who discovered the diagnosis of HIV during a medical consultation with a hematologist. He was then referred to the colonoscopy service, where he was diagnosed with Kaposi's Sarcoma of the rectum. He started ART with good response and complete remission of the tumor without the need for chemotherapy or radiotherapy.
Keywords: Kaposi's sarcoma, HIV, AIDS, Rectum.

INTRODUCTION

Kaposi's sarcoma (KS) was first described in 1872 by dermatologist Mortiz Kaposi, who described the tumor as an "idiopathic multiple pigmented sarcoma of the skin" (1)(2)(3)(4)(5). This type of tumor originates in the vascular epithelium and presents varying degrees of malignancy (2). The etiology of Kaposi's sarcoma has not yet been fully elucidated; however, it is believed that the tumor has a strong relationship with the human immunodeficiency virus (HIV) and human herpesvirus-8 (HHV-8) (1)(2)(3)(4)(5)(6)(7)(8). Furthermore, some studies reveal that HHV-8 has homologues of cellular genes, which stimulate cell proliferation, inflammation and angiogenesis (2)(9). Furthermore, it is believed that other factors are involved in the etiology of KS, such as gender, which is more prevalent in men aged 18 to 65 with Mediterranean ancestry, immunosuppression and genetic predisposition. Furthermore, the activation of inflammatory cytokines appears to be related to the emergence of the tumor (2)(3) (10).

Kaposi's sarcoma is considered one of the multiple markers for the diagnosis of acquired immunodeficiency syndrome (AIDS), in addition to being considered the most common neoplastic complication of this syndrome (4)(11). According to the literature, HIV carriers have approximately 3,500 times more chances of developing the disease compared to the population without HIV (2);(10).

The clinical aspects of Kaposi's sarcoma vary according to the course of the disease and the morbid profile of the patient. The disease is most often benign, with a tendency to invade mainly the skin and subcutaneous tissue, and more rarely, in its severe form, it affects the viscera and mucous membranes (2)(3)(10). Diagnosis is made through anamnesis, physical examination and complementary tests such as simple chest x-ray, lymph node ultrasound and abdominal ultrasound, in addition to upper digestive endoscopy and colonoscopy (2)(5)(7).

Treatment is mainly aimed at relieving symptoms and reducing tumor progression and size. In cases of localized tumors, surgical removal may be performed, while in more disseminated cases, radiotherapy and chemotherapy combined with antiretroviral therapy (ART) are chosen (2)(6)(8).

METHODOLOGY

The case reported was obtained through the analysis of medical records filed in a private gastroenterology and digestive endoscopy clinic in the Southwest of Paraná. The Terms of Commitment for Use of Data and Medical Records and the Institutional Consent Term for the use of information from the medical records were duly completed and signed, as well as the Free and Informed Consent Term guaranteeing the patient's privacy, the preservation of their identification and informing that no type of benefit or loss will be linked to their participation in the study. After obtaining the patient's consent through the attending physician, a clinical interview was conducted with the patient.

CASE REPORT

Male patient, 68 years old, divorced, heterosexual, Catholic. Presenting as comorbidities type 2 diabetes (DM2) using alogliptin benzoate in combination with pioglitazone hydrochloride and benign prostatic hyperplasia (BPH) using tamsulosin hydrochloride. He sought a hematologist due to complaints of weakness, fatigue, persistent diarrhea and weight loss of approximately 12 kg in 4 months. The hematologist requested laboratory tests and serology that showed a positive diagnosis of HIV, the patient was referred to the Serological Orientation and Reception Center (COAS) to start antiretroviral therapy. The chosen protocol for treatment was antiretroviral therapy using the following medications: dolutegravir sodium, tenofovir disoproxil fumarate + lamivudine and sulfametaxazole trimethoprim. He continued to have diarrhea while taking antiretroviral medication, and was then referred for gastroenterological investigation with colonoscopy.

A colonoscopy was performed on 09/12/2019, showing an ulcerated infiltrative vegetative lesion approximately 12 cm from the anal margin, suggestive of neoplasia (Figure 1). The lesion was biopsied and the results were: negative smooth muscle actin, positive CD31, inconclusive CD34, negative desmin, negative DOG 1, positive ERG 1, strong and diffuse in the lesion. Positive HHV 8, negative protein S 100. Thus, it is concluded that the immunohistochemical profile, together with the histopathological findings, are consistent with Kaposi's sarcoma.

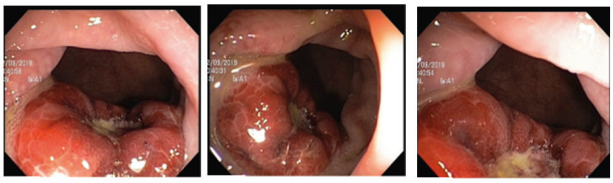

Figure 1: Ulcerated infiltrative vegetative lesion

Source: the authors

On 01/09/2020, a new colonoscopy was performed at the request of the attending physician for treatment control, describing the presence of an infiltrative ulcerated lesion with extensive central erosion and raised edges approximately 12/15 cm from the anal margin (Figure 2).

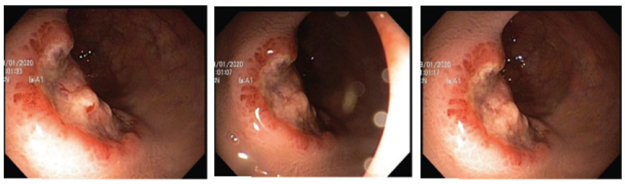

Figure 2: Infiltrative ulcerated lesion with extensive central erosion and raised edges

Source: the authors

After the second result, it was decided not to start chemotherapy and instead to continue therapy with antiretrovirals, as there were signs of partial remission of the lesion and response to HIV treatment.

RESULTS AND DISCUSSION

Kaposi's sarcoma can be classified into four types, which vary according to the patient's clinical condition. It can then be classified as classic, endemic, iatrogenic and epidemic (1)(2)(3)(4)(5) (11); (12). The sarcoma originally described by Kaposi in 1872 was called classic, and generally affects middle-aged or elderly men of Eastern European Jewish descent. The other form of Kaposi's sarcoma is the endemic form, which is very present in the sub-Saharan African population, with a higher morbidity and mortality rate in young patients. The iatrogenic form is associated with immunosuppressive drug therapy and is very prevalent in young people. Finally, the epidemic form is related to HIV infection, mainly affecting men aged between 18 and 65 years (2)(3) (10). Kaposi's sarcoma in the rectum is rare, with skin involvement being more common, manifesting as spots, papules, plaques or deep nodular lesions with a purplish-brown or reddish coloration that can spread to adjacent organs. These skin lesions can bleed, ulcerate and cause significant lymphedema, which causes a lot of pain to the patient (3)(5)(2) (10)(7) (13).

Kaposi's sarcoma involving the gastrointestinal tract was very common before the introduction of antiretroviral therapy, accounting for up to 40% of initial diagnoses of the disease. The appearance of lesions in the gastrointestinal tract occurs in the absence of skin lesions. These lesions in the gastrointestinal tract may be asymptomatic or may cause symptoms such as significant weight loss, abdominal pain, nausea and vomiting, diarrhea, upper or lower gastrointestinal bleeding, malabsorption and intestinal obstruction, which in most cases leads to the development of anemia (14)(15)(16). Lesions in the gastrointestinal tract can be visualized by endoscopy or colonoscopy, in which it is possible to visualize hemorrhagic nodules, which may be isolated or confluent and occur in any part of the gastrointestinal system (17)(18). The diagnosis is always confirmed with biopsy and immunohistochemical study. However, in some lesions, it is not possible to confirm Kaposi's sarcoma because the lesions are present in the submucosal layer. Furthermore, high-grade lesions have a high chance of dissemination and tissue invasion. Treatment is performed according to the presence or absence of symptoms (14).

According to the Clinical Trial Group (ACTG) of the National Institute of Health, Kaposi's sarcoma can be staged according to a good or poor prognosis (8) (19). Thus, it is possible to stage Kaposi's sarcoma taking into consideration, the following parameters: tumor extension (T), in which a favorable prognosis is based on the disease being limited to the skin with minimal involvement of the cavity; however, those who present lymphedema associated with extensive involvement of the cavity or invasion of viscera are considered to have a poor prognosis (T1). The immunological status (I) is also considered; patients with a CD4 greater than 200 cells/microL have a favorable prognosis; however, those with a reduced CD4 count are classified as having a poor prognosis (I1). The severity of the systemic disease (S) should not be forgotten. Patients who do not present any systemic manifestations have a favorable prognosis (S0), while those with a history of opportunistic infection, mouth ulcers, symptoms of fever, night sweats, significant weight loss and diarrhea for more than two weeks have a poor prognosis (S1). (8); (20)

Treatment options aim to control both the symptoms and the sarcoma. Therefore, the main therapeutic objectives are to relieve lymphedema when present, improve function, reduce the size of skin or visceral lesions and prevent or delay the progression of the disease (8); (21). The use of antiretroviral

therapy (ART) is indicated for all patients with HIV-related KS and this may be the only treatment when there is no recommendation to start chemotherapy. However, patients who are eligible for chemotherapy can do so by combining treatment with ART in a safe manner (8).

Surgery is prescribed only in some cases of KS, such as in the classic form. Therefore, if there is a single symptomatic lesion that presents bleeding, excision alone may provide sustained local tumor control, but new lesions commonly develop at other sites (12); (22). Finally, radiotherapy may be indicated in all forms of KS (12).

CONCLUSIONS

The main interest of this case report is to explain the possible manifestations of HIV, and mainly, the rare manifestation of KS in the rectum, which, according to the literature, is more common in the cutaneous form, but can rarely affect viscera and mucous membranes. Despite all the knowledge about HIV, its incidence is still high and the recognition of possible manifestations is of utmost importance, since early diagnosis and institution of treatment with ART prevents rare complications of HIV such as Kaposi's sarcoma in the rectum.

REFERENCES

1. Iscovich J, Boffetta P, Franceschi S, Azizi E, Sarid R. Classic Kaposi sarcoma: Epidemiology and risk factors. Cancer. 2000;88(3):500–17.

2. Wagner AK, Tozo JP, Abreu MAMM de, Wedy GF. Sarcoma de Kaposi em paciente portador de HIV: relato de caso. Med (Ribeirao Preto Online). 2018;51(2):157–61.

3. Ruiz MÁG, Rivero JQ, García JLM, Rodríguez JJC, Kavadoy YR, Carmona MFR, et al. High-dose-rate brachytherapy in the treatment of skin Kaposi sarcoma. J Contemp Brachytherapy. 2017;9(6):561–5.

4. Borges SAL, Real LHG, Schreiner RB. Sarcoma de Kaposi em pacientes HIV: novamente uma realidade. Rev Eletrônica Acervo Saúde. 2019;11(6):e352.

5. Sarcoma S, Report C, Patrocínio J, Rita Espírito Santo A, Patrocínio J, Pereira R, et al. SARCOMA DE KAPOSI CLÁSSICO, A PROPÓSITO DE UM CASO CLÍNICO. Vol. 4, Rev Clin Hosp Prof Dr Fernando Fonseca. 2016.

6. Kak I, Salama S, Gohla G, Naqvi A, Alowami S. A case of patch stage of Kaposi's sarcoma and discussion of the differential diagnosis. Rare Tumors. 2016;8(1):30–2.

7. El-Mallawany NK, Kamiyango W, Slone JS, Villiera J, Kovarik CL, Cox CM, et al. Clinical factors associated with long-term complete remission versus poor response to chemotherapy in HIV-Infected children and adolescents with kaposi sarcoma receiving bleomycin and vincristine: a retrospective observational study. PLoS One. 2016;11(4):1–17.

8. Clinical A, Group T. Sarcoma de Kaposi relacionado à AIDS : estadiamento e tratamento. 2021;1–23.

9. Yenice MG, Varnalı E, Şeker KG, Kavak A, Tuğcu V. Scrotal Kaposi's Sarcoma in HIV-negative patient: A case report and review of the literature. Turkish J Urol. 2018;44(2):182–4.

10. Akinde O, Obadofin O, Adeyemo T, Omoseebi O, Ikeri N, Okonkwo I, et al. Kaposi Sarcoma among HIV Infected Patients in Lagos University Teaching Hospital, Nigeria: A 14-Year Retrospective Clinicopathological Study. J Skin Cancer. 2016;2016.

11. Eduardo C, Barros P De, Duarte SC, Shimoda E. Sarcoma de Kaposi como diagnóstico diferencial de lesões vulvares : relato de caso e revisão bibliográfica. 2013;8:26–30.

12. Krown SE, Chadha J. Classic Kaposi sarcoma: Clinical features, staging, diagnosis, and treatment. UptoDate [Internet]. 2019;1–16. Available from: http://www.uptodate.com/contents/classic-kaposi-sarcoma-clinical-features-staging-diagnosis-and-treatment

13. Bhutani M, Polizzotto MN, Uldrick TS, Yarchoan R. KSHV-associated Malignancies: Epidemiology, Pathogenesis, and Advances in Treatment. Semin Oncol. 2018;42(2):223–46.

14. Sarcoma de Kaposi relacionado à AIDS : manifestações clínicas e diagnóstico. 2021;(Cdc):1–28.

15. Ho-Yen C, Chang F, van der Walt J, Lucas S. Gastrointestinal Malignancies in HIV-infected or Immunosuppressed Patients: Pathologic Features and Review of the Literature. Adv Anat Pathol [Internet]. 2007;14(6). Available from: https://journals.lww.com/anatomicpathology/Fulltext/2007/11000/Gastrointestinal_Malignancies_in_HIV_infected_or.5.aspx

16. Laine L, Amerian J, Rarick M, Harb M, Gill PS. The response of symptomatic gastrointestinal Kaposi's sarcoma to chemotherapy: a prospective evaluation using an endoscopic method of disease quantification. Am J Gastroenterol. 1990 Aug;85(8):959–61.

17. Friedman SL, Wright TL, Altman DF. Gastrointestinal Kaposi's sarcoma in patients with acquired immunodeficiency syndrome. Endoscopic and autopsy findings. Gastroenterology. 1985;89(1):102–8.

18. Dorsett B. Kaposi ' s Sarcoma of Internal Organs A Multiparameter Study of 86 Cases. :1376–85.

19. Krown SE, Metroka C, Wernz JC. Kaposi's sarcoma in the acquired immune deficiency syndrome: a proposal for uniform evaluation, response, and staging criteria. AIDS Clinical Trials Group Oncology Committee. J Clin Oncol Off J Am Soc Clin Oncol. 1989 Sep;7(9):1201–7.

20. Boffi El Amari E, Toutous-Trellu L, Gayet-Ageron A, Baumann M, Cathomas G, Steffen I, et al. Predicting the evolution of Kaposi sarcoma, in the highly active antiretroviral therapy era. Aids. 2008;22(9):1019–28.

21. Dezube BJ, Pantanowitz L, Aboulafia DM. Management of AIDS-related Kaposi sarcoma: advances in target discovery and treatment. AIDS Read. 2004 May;14(5):236-238,243-244,251-253.

22. Weintraub CM, Skudowitz RB. Excision of 1,674 classic Kaposi's sarcomas. Vol. 40, South African journal of surgery. Suid-Afrikaanse tydskrif vir chirurgie. South Africa; 2002. p. 80.