

Anal canal lymphoma: Case report and literature review

Julián Andrés Romo-Sánchez,^{1*} Laura X. Covelli,² Laura A. López,³ Carlos Figueroa,⁴ David Baquero,⁵ Horacio Garzón,⁵ Andrea Recaman.³

OPEN ACCESS

Citation:

Romo-Sánchez JA, Covelli L, López L, Figueroa C, Baquero D, Garzón H, Recaman A. Anal canal lymphoma: Case report and literature review. *Rev Colomb Gastroenterol.* 2022;37(2):233-236. <https://doi.org/10.22516/25007440.777>

¹ MD, General Surgeon, Universidad del Rosario. Bogotá, D. C., Colombia.

² MD, Third-year General Surgery Resident, Universidad del Rosario. Bogotá D. C., Colombia.

³ MD, Universidad del Rosario. Bogotá D. C., Colombia.

⁴ MD, General Surgeon and Coloproctologist, Head of the Coloproctology Service, Hospital Universitario Mayor Méderi. Bogotá D. C., Colombia.

⁵ MD, General Surgeon and Coloproctologist, Hospital Universitario Mayor Méderi. Bogotá D. C., Colombia.

*Correspondence: Julián Romo S.
doctorromo22@gmail.com

Received: 12/05/2021

Accepted: 11/06/2021



Abstract

Introduction: Primary tumors of the anal canal other than carcinomas are rare entities; among them, anal canal lymphomas are extremely unusual and pose both a diagnostic and therapeutic challenge for the coloproctologist. **Case presentation:** A male patient with positive human immunodeficiency virus (HIV) with proctalgia and mass sensation at the perianal level. A concentric thickening of the walls of the lower rectum was documented by magnetic resonance imaging, with colonoscopy and biopsies with histopathology compatible with plasmablastic lymphoma. Therefore, a diverting colostomy was performed and, subsequently, the hematology service indicated chemotherapy with the EPOCH scheme. **Discussion:** Lymphoma of the anus represents 0.2% of anorectal tumors, most of these are non-Hodgkin's lymphomas; Hodgkin's disease at the anorectal level is even rarer. The population with the highest risk of this entity is HIV-positive patients, such as the patient in this case, although other associated factors are described in the literature.

Keywords

Anal lymphoma, plasmablastic lymphoma, colorectal cancer, anorectal cancer.

INTRODUCTION

Malignant neoplasms of the anal canal are rare diseases, and anal lymphoma is one of the least frequent, comprising approximately 0.2% of anorectal tumors. It affects primarily young adult men in a 6:1 ratio; a critical factor associated with these tumors is human immunodeficiency virus (HIV) infection⁽¹⁾. The low incidence of this pathology represents a diagnostic and therapeutic challenge for both clinical oncology and colorectal surgery groups. Cancers of the anus and anal canal represent approximately 2.4% of all gastrointestinal neoplasms⁽¹⁾. They are the majority

of these squamous cell cancers; however, there are other types of less frequent tumors, including lymphoma of the anus, which is a diagnostic and therapeutic challenge.

CLINICAL CASE

A 33-year-old male patient with a history of stage C3 HIV infection was admitted for a clinical picture of approximately three months of evolution consisting of sensation of perianal mass and pain, anal discharge, and intermittent rectal bleeding. On physical examination, a mass of approximately 4 x 5 cm was observed at the level of the anal

canal, protruding through it. Digital rectal examination revealed an ulcerated, exophytic, friable mass that extended from the anal ridge up to 5 cm, infiltrated the external anal sphincter, and was in contact with the puborectalis muscle.

Within the extension studies, he had a contrast-enhanced magnetic resonance imaging (MRI) of the pelvis that showed a concentric thickening of the walls of the lower rectum, with an exophytic lesion that compromised the anal canal and extended towards the intergluteal soft tissues. The anal canal lesion had a longitudinal extension of 63 mm with an anteroposterior (AP) diameter of 73 mm and a transverse diameter of 40 mm with an adequate cleavage plane between the walls of the rectum and the seminal vesicles. The image described in the lower rectum made contact with the prostate posterior contour with nodes in the mesorectal fat with a diameter of up to 7 mm.

With this result and the findings upon physical examination, we decided to perform a rectosigmoidoscopy, finding a circumferential mass that extended from the anal rim to 5 cm, compromising 100% of the lumen and 95% of the circumference. It was obstructive and did not allow the team to advance, from which biopsies were taken (**Figures 1 and 2**). The histopathological report was a plasmablastic lymphoma with a positive immunophenotype for CD38, MUM-1, Ki67 95%, and CMYC in 70% of the tumor cells; and negative for cytokeratins, AE1/AE3, CD20, CD3, CD5, CD30, cyclin D1, Bcl2, Bcl6, PAX5, and CD10.

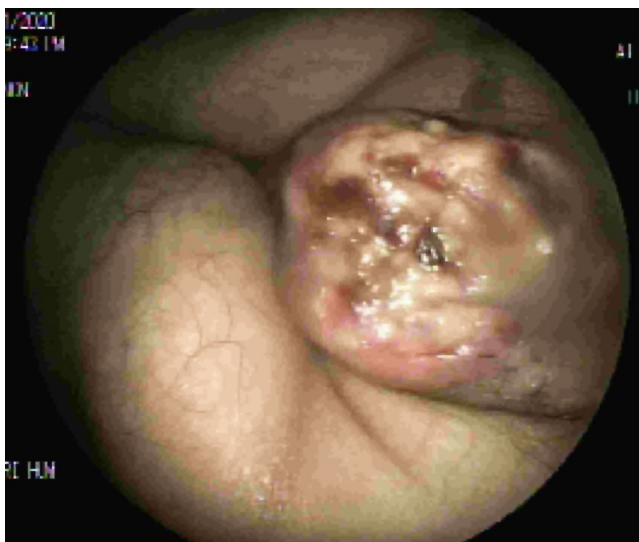


Figure 1. Mass at the level of the anal rim.

The diagnosis of plasmablastic lymphoma was made. We decided to perform a laparoscopic diverting colostomy, without complications during the postoperative period. Due to the patient's history of HIV, in-hospital studies were performed that showed a CD4+ lymphocyte count:

23 cells/mm³ and a viral load of 16,803 replicates, with an indication to start chemoprophylaxis with trimethoprim/sulfamethoxazole and antiretroviral therapy with tenofovir, emtricitabine, and efavirenz.

Staging studies showed an abdominal computerized axial tomography (CT) scan with hepatosplenomegaly and a contrast-enhanced chest CT without evidence of lymphadenopathy or pulmonary nodules suggestive of secondary neoplastic disease. He was assessed by the haemato-oncology department, which considered that the patient had a stage IV BX plasmablastic lymphoma according to the Ann Arbor classification. We opted for management with systemic chemotherapy DA-EPOCH scheme for six cycles, with a partial response after the third cycle.

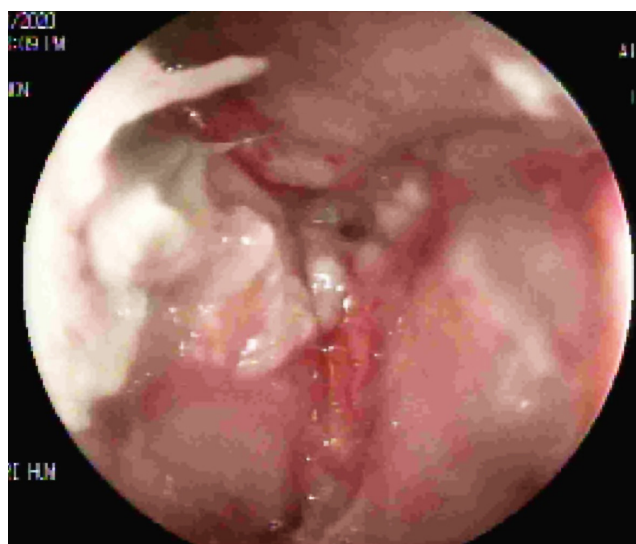


Figure 2. Mass with complete obstructive effect in the anal canal.

DISCUSSION

Cancers of the anus and anal canal represent approximately 2.4% of all gastrointestinal neoplasms⁽¹⁾. They are the majority of these squamous cell cancers; however, there are other types of less frequent tumors, including lymphoma of the anus, which poses a diagnostic and therapeutic challenge.

Lymphoma of the anus represents 0.2% of anorectal tumors, most of which correspond to non-Hodgkin's lymphomas; Hodgkin's disease at the anorectal level is even rarer⁽¹⁾. This pathology affects more young adult men in a ratio of 6:1; an important associated factor is HIV infection, mainly when the CD4 count is < 100 mm³. Nevertheless, in recent decades, active antiretroviral therapy has decreased the incidence of HIV-associated lymphoma⁽²⁾. Anorectal lymphoma can be classified as primary if it refers to a primary neoplastic process of anorectal origin and secondary

if it is a generalized neoplastic process in which anorectal involvement is caused by lymph node metastases⁽³⁾.

As already mentioned, primary colorectal lymphomas usually tend to be of the non-Hodgkin type; however, in some patients immunocompromised by HIV, the anorectal involvement could be of the Hodgkin type⁽⁴⁾. In this particular population, the involvement of the anal canal can reach up to 26%^(3,5).

The clinical manifestations of anorectal lymphoma are similar to those of adenocarcinoma of the rectum. The most frequent is the sensation of a perianal mass, chronic ulceration, and bleeding; in turn, B symptoms such as weight loss, abdominal pain, or fever may occur⁽⁴⁾. It is a problematic pathology; sometimes, the symptoms or diagnostic studies are misinterpreted. The clinical manifestation described in the world literature is diverse, from cases diagnosed postoperatively after hemorrhoidectomy⁽⁶⁾ to perianal abscesses⁽⁷⁾. Therefore, obtaining adequate samples that allow histopathological diagnosis is vital for establishing appropriate treatment.

Anal canal lymphoma is usually a tumor associated with HIV infection; however, other risk factors are described in the literature for its development, such as long-standing ulcerative colitis, transplant patients, pelvic radiotherapy, and immunodeficiencies of other causes⁽⁵⁾. Regarding diagnosis, imaging studies impede adequate differentiation from other types of lesions and most of the time, they are performed to evaluate the regional extension of the disease and rule out extranodal involvement⁽⁴⁾. The histopathological study provides the definitive diagnosis. Nonetheless, it continues to be a challenge because there are more than 60 varieties of non-Hodgkin's lymphomas with different histological provide. It is often necessary to obtain multiple biopsies and various immunohistochemical markers that allow a definitive diagnosis^(4,8); the most common are diffuse large B-cell non-Hodgkin's lymphoma and plasmablastic lymphoma, as in the previously described case.

Currently, the treatment is controversial, and there is no consensus about it; the most important thing is to define the extent of the disease, classify the type of lymphoma, and whether or not it is associated with HIV. Most studies suggest that the treatment of primary anorectal lymphoma

could be surgical through oncological resection when it is a localized disease (Ann Arbor stages I–II), in which imaging findings classify it as potentially resectable. For example, high lesions could be treated with anterior resections of the rectum, while lower lesions that compromise the sphincters can be treated with abdominoperineal resections^(9,10). For advanced lymphomas (Ann Arbor stages III–IV), the treatment is systemic chemotherapy, of which there are multiple schemes⁽¹¹⁾. Traditionally, multiple chemotherapy regimens have been administered for anal lymphomas, and the most used is CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone); nonetheless, adequate response and survival rates have not been achieved with this scheme^(10,12).

Due to the low frequency of this pathology, most studies conducted to date are retrospective and with a relatively low number of cases, without showing significant differences in overall survival and response to treatment; however, more recent studies suggest an improvement in survival with more intensive chemotherapy schemes such as EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin)⁽¹³⁾.

Radiotherapy treatment in lymphomas of the anus can be complementary, especially in local recurrences or in patients who underwent local resections with positive margins⁽⁴⁾. The overall survival rate is poor for anal lymphoma. Before highly active antiretroviral therapies (HAART), survival was eight months; now, the 5-year overall survival is 50% in localized disease, while for advanced disease with regional or distant metastatic lymph node involvement is 24% at five years⁽⁴⁾.

CONCLUSION

Primary lymphoma of the anus is an infrequent entity, representing approximately 0.02% of all anorectal tumors; primary lymphomas of this location are usually non-Hodgkin's type, the population with the most significant relationship to this entity are HIV positive patients, and its management is surgical in early stages (Ann Arbor I and II). For more advanced lesions (Ann Arbor III and IV), treatment must involve systemic chemotherapy with EPOCH.

REFERENCES

1. Hoff PM, Coudry R, Moniz CM. Pathology of Anal Cancer. *Surg Oncol Clin N Am*. 2017;26(1):57-71. <https://doi.org/10.1016/j.soc.2016.07.013>
2. Machalek DA, Poynten M, Jin F, Fairley CK, Farnsworth A, Garland SM, et al. Anal human papillomavirus infection and associated neoplastic lesions in men who have sex with men: a systematic review and meta-analysis. *Lancet Oncol*. 2012;13(5):487-500. [https://doi.org/10.1016/S1470-2045\(12\)70080-3](https://doi.org/10.1016/S1470-2045(12)70080-3)

3. Cuffy M, Abir F, Longo WE. Management of less common tumors of the colon, rectum, and anus. *Clin Colorectal Cancer*. 2006;5(5):327-37.
<https://doi.org/10.3816/CCC.2006.n.002>
4. Simpson L, Taylor S, Piotrowski A. Uncommon presentations of Hodgkin's disease. Case 3. Hodgkin's disease: rectal presentation. *J Clin Oncol*. 2004;22(1):196-8.
<https://doi.org/10.1200/JCO.2004.02.144>
5. Peralta EA. Rare anorectal neoplasms: gastrointestinal stromal tumor, carcinoid, and lymphoma. *Clin Colon Rectal Surg*. 2009;22(2):107-14.
<https://doi.org/10.1055/s-0029-1223842>
6. Díaz Barrientos CZ, Osorio Hernández RM, Navarrete Cruces T. Clinical image in gastroenterology. Primary anal lymphoma in a patient that underwent hemorrhoidectomy. *Rev Gastroenterol Mex*. 2014;79(4):292-3.
<https://doi.org/10.1016/j.rgm.2014.09.002>
7. Jayasekera H, Gorissen K, Francis L, Chow C. Diffuse large B cell lymphoma presenting as a peri-anal abscess. *J Surg Case Rep*. 2014;2014(6):rju035.
<https://doi.org/10.1093/jscr/rju035>
8. Ambrosio MR, Rocca BJ, Barone A, Mastrogiulio MG, Costa A, Bellan C, et al. Primary anorectal Hodgkin lymphoma: report of a case and review of the literature. *Hum Pathol*. 2014;45(3):648-52.
<https://doi.org/10.1016/j.humpath.2013.09.015>
9. Zigelboim J, Larson MV. Primary colonic lymphoma. Clinical presentation, histopathologic features, and outcome with combination chemotherapy. *J Clin Gastroenterol*. 1994;18(4):291-7.
<https://doi.org/10.1097/00004836-199406000-00007>
10. Errasti Alustiza J, Espín Basany E, Reina Duarte A. Rare tumors of the rectum. Narrative review. *Cir Esp*. 2014;92(9):579-88. English, Spanish.
<https://doi.org/10.1016/j.ciresp.2013.06.019>
11. Jeong JH, Kim S, Kim JE, Yoon DH, Lee SW, Huh J, et al. Clinical characteristics, treatment, and outcome of primary rectal lymphoma: a single center experience of 16 patients. *Blood Res*. 2017;52(2):125-129.
<https://doi.org/10.5045/br.2017.52.2.125>
12. Fernández-Álvarez R, Sancho JM, Ribera JM. Linfoma plasmablastico. *Med Clin (Barc)*. 2016;147(9):399-404.
<https://doi.org/10.1016/j.medcli.2016.06.036>
13. Arnett J, Henry B, Fankell A. Managing late effects of breast cancer treatment. *Clin J Oncol Nurs*. 2014;18 Suppl:32-5.
<https://doi.org/10.1188/14.CJON.S2.32-35>