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Plasmablastic Lymphoma of the Anal Canal in an HIV-Infected Patient

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Conflict of interest: None declared

Patient: Female, 46
Final Diagnosis: Plasmablastic lymphoma
Symptoms: Fecal incontinence
Medication: Chemotherapy
Clinical Procedure: Anal canal biopsy
Specialty: Internal medicine • Oncology • Radiology

Objective: Unusual clinical course





Background: The advent of antiretroviral therapy increased the life expectancy of human immunodeficiency virus (HIV)-positive patients and, consequently, the morbidity and mortality due to neoplasms. Plasmablastic lymphoma is one such neoplasm that generally presents with involvement of the oral cavity; cases of extra-oral involvement are rare.

Case Report: We report a case of plasmablastic lymphoma in a 46-year-old woman for whom the initial clinical manifestation was a painless perineal tumor accompanied by fecal incontinence.

Conclusions: The possibility of this neoplasm should be considered in patients with HIV/acquired immune deficiency syndrome (HIV/AIDS) because its early diagnosis is essential so that the start of the treatment is not delayed.

MeSH Keywords: AIDS Serodiagnosis • Anti-Retroviral Agents • Anus Neoplasms • Lymphoma, AIDS-Related

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/892313>

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Background

Human immunodeficiency virus (HIV) infection and the occurrence of acquired immune deficiency syndrome (AIDS) cause cellular immune deficiency, which leads to the development of opportunistic diseases. Neoplasms are among these diseases, especially lymphoproliferative disorders, which are more aggressive in such patients [1]. Plasmablastic lymphoma is a subtype of diffuse large B-cell lymphoma (5% of cases) and is considered an AIDS-defining tumor [2]. It most commonly occurs in the oral cavity (90%), with cases of extra-oral involvement being rare [1–3]. Herein, we report a case of a patient with HIV/AIDS presenting with plasmablastic lymphoma of the anal canal.

Case Report

A 46-year-old woman being treated for panic disorder without known comorbidities developed a painless perineal tumor with

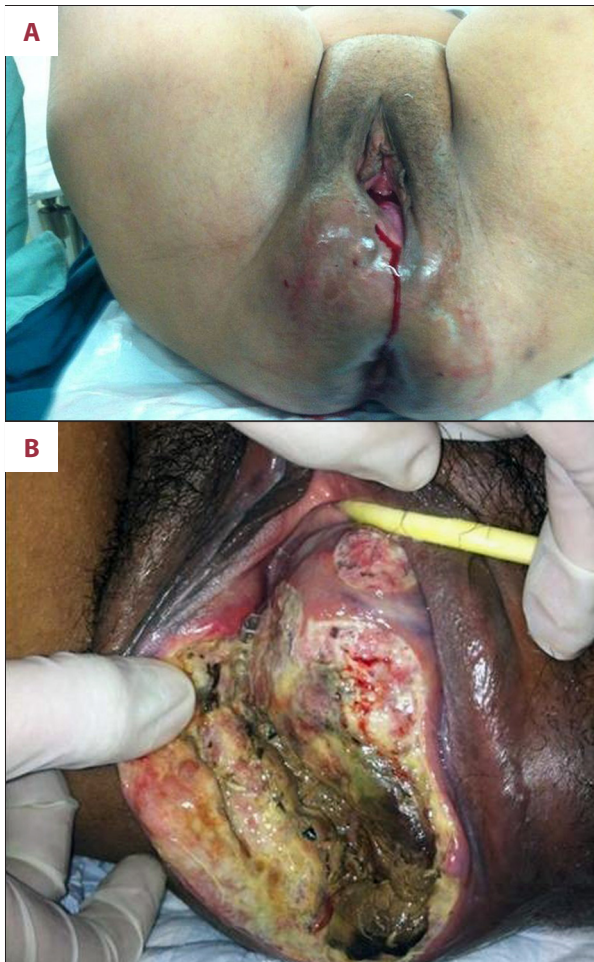


Figure 1. Aspect of the lesion at admission (A) and after 1 month of chemotherapy (B).



Figure 2. T2-weighted sagittal (A) and coronal (B) sections, without contrast agent, showing a lesion with a slightly hyperintense signal.

2 months of evolution that was accompanied by fecal incontinence with ribbon-like feces. The patient denied fever, weight loss, or any other associated clinical manifestations. At another institution, the patient was subjected to attempted lesion drainage, without success. Physical examination revealed voluminous bulging in the perineal region with vulvar ulceration and no phlogistic signs. Digital vaginal examination revealed bulging of the back wall of the vagina extending to the end of the vaginal cul-de-sac (Figure 1).

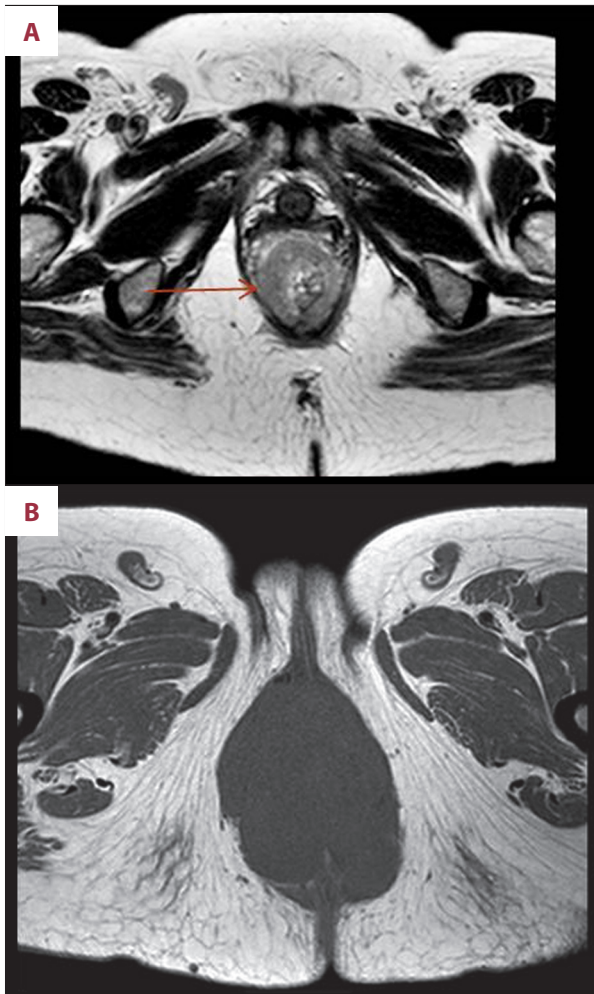


Figure 3. T2-weighted (A) and T1-weighted (B) axial sections, both without fat suppression, showing a slightly hyperintense signal on T2 and an isointense signal on T1, which are common in tumors with high cellularity. Note the infiltration in the lower rectum (arrow).

Following admission to our institution, laboratory exams were performed. Laboratory evaluation revealed a leukocyte count of 4.4×10^9 cells/L (74% neutrophils, 16% lymphocytes, 7.5% monocytes, and 2% eosinophils). She had a lactate dehydrogenase of 305 IU/L. The HIV Western blot test was positive. The CD4⁺ and CD8⁺ counts were 67/mm³ and 602/mm³, respectively, with a low CD4⁺/CD8⁺ T cell ratio of 0.11. In blood, the serology test for Epstein-Barr virus with IgG was positive, while IgM was negative. Once the HIV/AIDS diagnosis was made, an appropriate treatment was initiated. The patient was unable to identify a specific risk exposure to HIV, and thus it was not possible to estimate the time of infection.

The patient underwent nuclear magnetic resonance imaging, which revealed a voluminous expansive formation in the anal canal, with descending exophytic appearance, measuring



Figure 4. T1-weighted sagittal section with fat saturation (A) showing a hyperintense focus, indicating bleeding (arrow), and a T1-weighted axial section with post-contrast fat saturation (B) showing a gadolinium-enhanced lesion.

13.3×8.8×7.6 cm and associated with multiple enlarged lymph nodes in the retroperitoneum, hepatic hilum, and mesentery and iliac chains bilaterally (Figures 2–5). A biopsy of the lesion and immunohistopathological analysis of the fragment were performed, and plasmablastic lymphoma was diagnosed (Figures 6 and 7). In histopathological analysis, the Epstein-Barr virus polymerase chain reaction was negative. Hyper-CVAD chemotherapy (cyclophosphamide, vincristine, doxorubicin, dexamethasone/methotrexate, cytarabine) was then initiated.

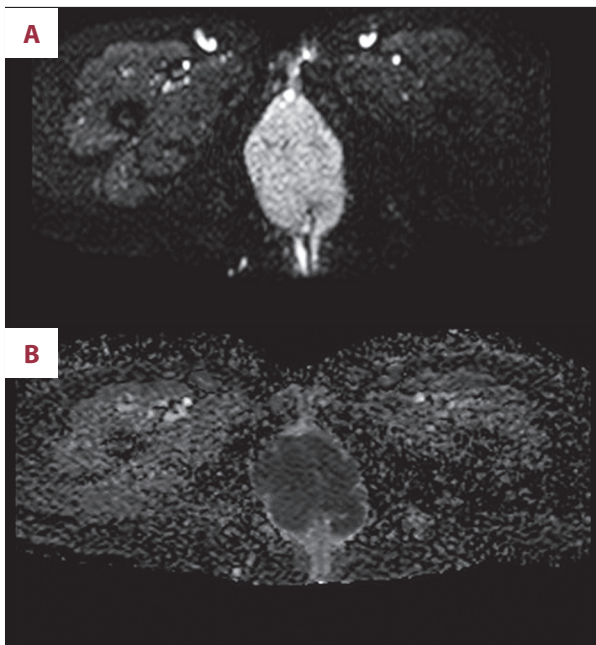


Figure 5. Lesion presenting a high B-value diffusion-weighted signal (A) and a strongly hypointense signal on the apparent diffusion coefficient (ADC) map (B), indicating restriction to water diffusion due to the increased cellularity characteristic of lymphomas.

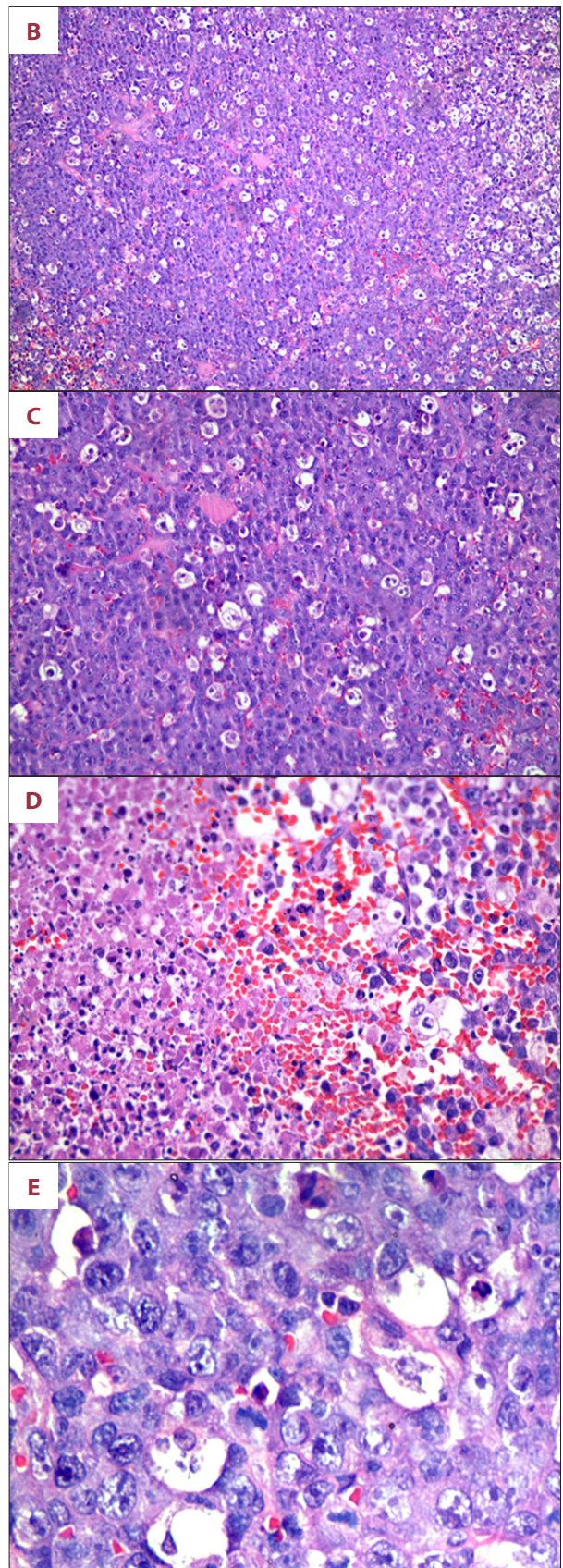


Figure 6. Biopsy of the anal canal (hematoxylin and eosin [H&E]). At lower magnification (A), diffuse proliferation of blue cells is observed. At medium magnifications (B and C), a "starry-sky" pattern is observed, typical of high-grade lymphoid neoplasms, with the figures consisting of histiocytes phagocytizing apoptotic cell remains. Morphologically, cells of different sizes are observed: smaller cells with plasmacytoid characteristics and larger, more atypical cells. At medium magnification (D), there is evidence of necrotic areas and apoptotic figures, which are characteristic of malignant tumors. At higher magnification (E), diffuse and monomorphically proliferating atypical cells can be observed. These cells are of medium and large sizes, with moderate cytoplasm and large irregular nuclei with vesicular chromatin and evident, sometimes multiple, nucleoli. Apoptotic and mitotic figures are emphasized.

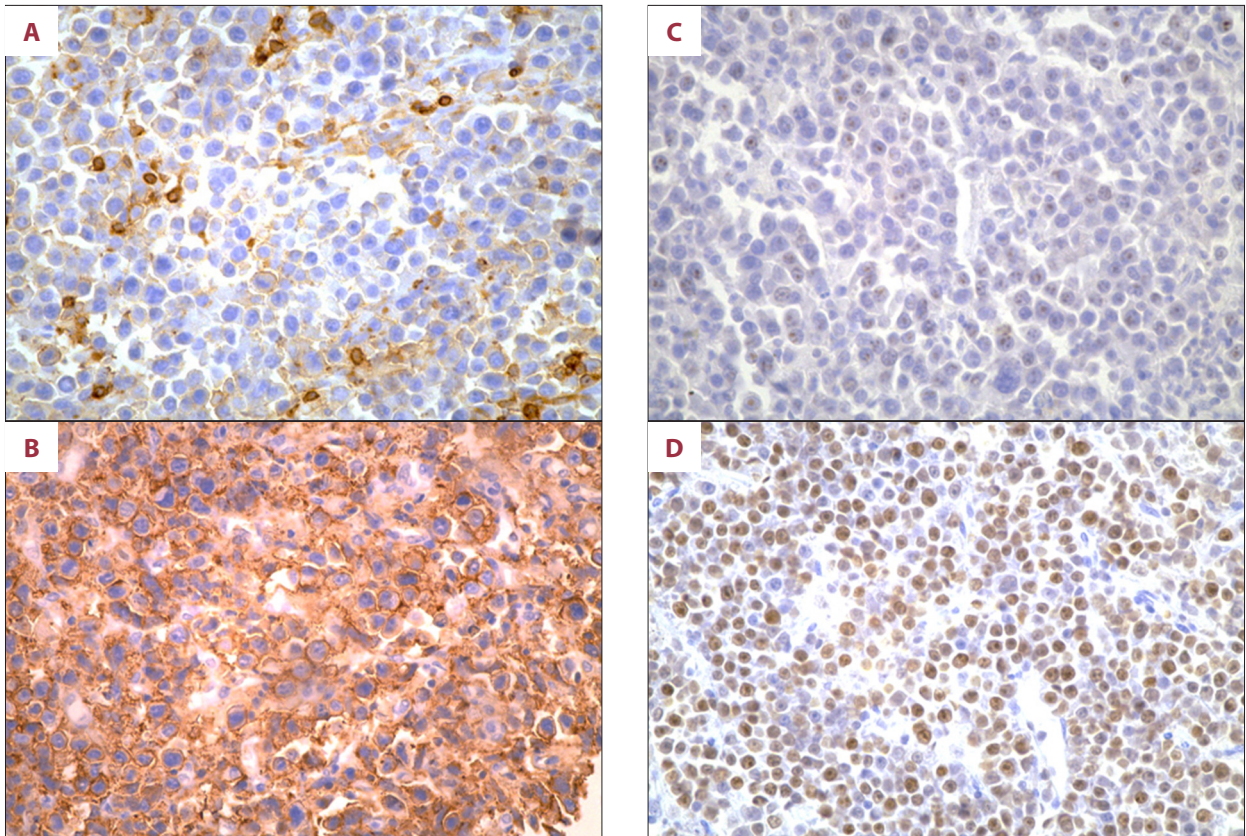


Figure 7. Biopsy of the anal canal (immunohistochemistry). (A) LCA (CD45): positive marker of cells of lymphoid origin in sparse cells. (B) CD138: positive marker of B-cells at the final stages of differentiation (plasma cells). (C) CD20: negative marker of B lymphocytes. (D) Ki67: marker of the cell proliferation index, which is approximately 90%, reflecting a high tumor grade.

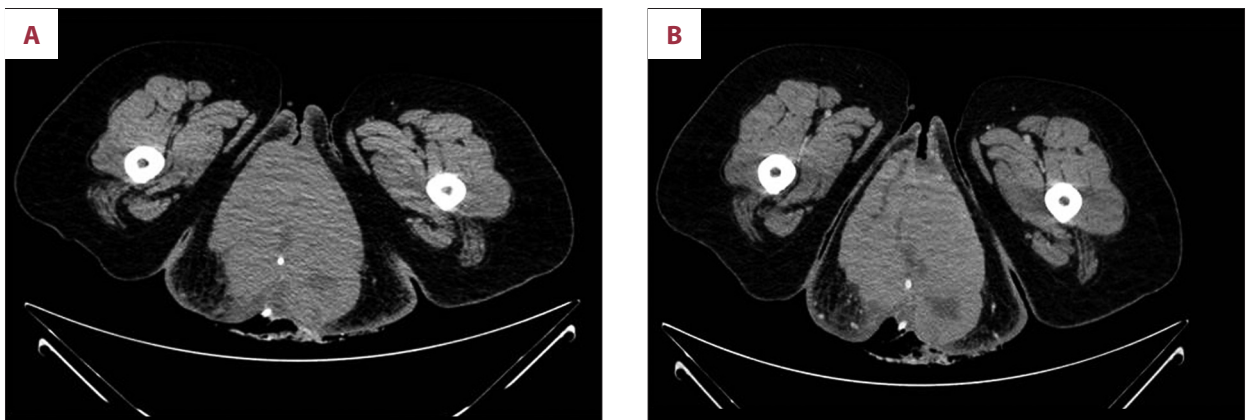


Figure 8. (A) Axial section of computed tomography without administration of intravenous contrast, showing a voluminous expansive and infiltrating formation with an epicenter in the anal canal and extending to the vagina and the anal cutaneous surface. (B) Following administration of intravenous contrast, a slight heterogeneous contrast uptake by the mass with an epicenter in the anal canal is observed.

The patient underwent computed tomography of the abdomen and pelvis for evaluation of the clinical condition approximately 1 month following the performance of the nuclear magnetic resonance imaging. The lesion had grown significantly and measured approximately 19.5×15.2×11.5 cm at this

time, with invasion of the lower rectum, vagina, and uterus (Figures 8 and 9). Formation of an abscess in the anterior peritoneal region was observed, along with increased numbers of enlarged lymph nodes in the retroperitoneum, hepatic hilum, and mesentery and iliac chains bilaterally.

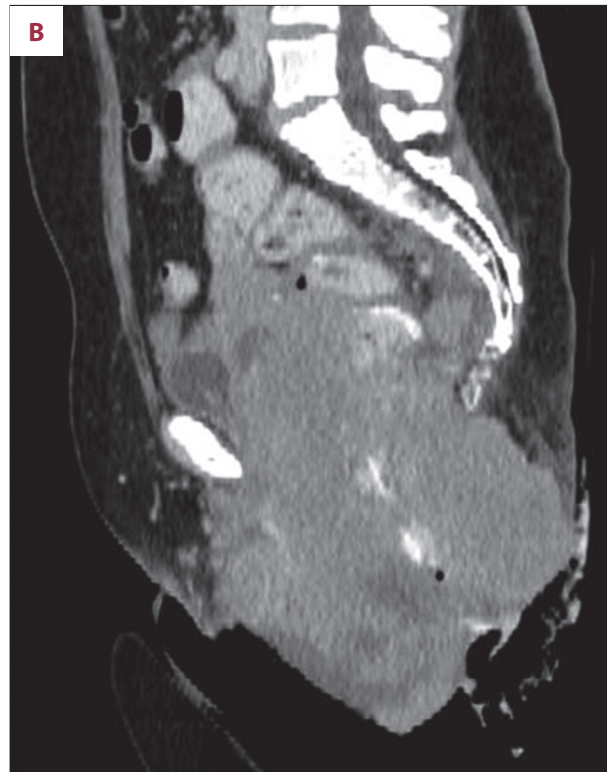


Figure 9. Computed tomography coronal (A) and sagittal (B) sections following administration of intravenous contrast showing low heterogeneous contrast uptake by the mass with an epicenter in the anal canal. Note the cranial extension of the lesion to the lower rectum, vagina, uterus, and bladder that is indissociable from these structures and the lower extension to the anal cutaneous surface.

A surgical drainage of the anterior perineal collection was performed, and a rectovaginal fistula was identified. Due to the occurrence of recurrent local infection, a transversostomy was performed in an attempt to control the infection to continue the chemotherapy. The patient presented several infectious complications during chemotherapy cycles, including pneumonia, oropharyngeal and esophageal candidiasis, cytomegalovirus infections, and aspergillosis, progressing to death.

Discussion

Non-Hodgkin lymphomas are the second most common malignancy in HIV-positive patients. One such lymphoma is the plasmablastic lymphoma, first described in 1997 as a variant of diffuse large B-cell lymphoma, strongly associated with Epstein-Barr virus infection and characteristically located in the oral cavity [1,3–5]. Plasmablastic lymphoma is rare and aggressive, with plasmablastic differentiation, and is responsible for approximately 2.6% of all AIDS-related lymphomas [6]. It predominates in adults, without clear racial differences [1,7]. Most patients are male, with an average age of presentation of 39 years [5,8].

The plasmablastic lymphoma is mainly located in the oral cavity, with fast local invasion and dissemination. However,

appearance in other locations has become more common over the last few years [3]. Involvements of the gastrointestinal tract, lymph nodes, and skin have been reported [8]. The lungs, orbits, liver, testicles, sinuses, and anal canal can also be affected, though rarely [2,9–11].

The main morphological and histochemical characteristics of the plasmablastic lymphoma are the presence of immature cells with abundant cytoplasm; eccentric nucleus with prominent nucleolus; the persistence of plasma cell markers, such as CD38 and CD138; and the absence of B cell markers, such as CD4 and CD20. In addition, high proliferation rates (Ki67 >60%) and positive results for Epstein-Barr virus infection assist in the diagnosis [1,3].

Computed tomography is the imaging method most commonly used for lymphomas due to its wide availability and relative low cost [12]. However, nuclear magnetic resonance imaging is considered the criterion standard examination due to the better spatial resolution, which allows better characterization of the tissue infiltration caused by the lesion [13]. With both imaging methods, the presence of a mural infiltrating mass that is concentric and homogeneous, with or without luminal occlusion, is the most common aspect of intestinal involvement in lymphoma. The presence of polypoid lesions, thickening of the levator ani, and local adenopathy are other related findings. In nuclear magnetic resonance imaging, the lesion presents with a generally homogeneous intermediate T1 signal and an intermediate/high T2 signal, with low/moderate contrast

uptake and restriction to water diffusion due to hypercellularity [1,12,13]. Fistula formation is common due to transmural involvement of the gastrointestinal tract.

Differential diagnosis of tumoral lesions in the perianal region should mainly include neoplasms of epithelial lineage, with epidermoid carcinoma being the main representative of this group [14]. Lymphoma, adenocarcinoma, basaloid carcinoma, cloacogenic carcinoma, melanoma, and metastasis should be included in the differential diagnosis [13–15].

In general, the prognosis of patients with plasmablastic lymphoma is poor, with most dying in the first year following diagnosis. However, good therapy results may be obtained in

patients who begin chemotherapy in the early disease stages [1,3,13]. The use of antiretroviral regimens in HIV-positive patients contributes to a better prognosis [1,3].

Conclusions

The present report describes an uncommon lymphoma subtype of the anal canal. The patient presented clinical and radiological characteristics suggestive of plasmablastic lymphoma, but this condition was not initially included in the differential diagnosis. Therefore, we wish to emphasize the possibility of this neoplasm in HIV/AIDS patients, because early diagnosis is important to avoid delay in the start of treatment.

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