

Case Report

Primary Ileal Diffuse Large B-Cell Lymphoma with Acute Intestinal Obstruction: Case Report and Narrative Literature Review

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Abstract: Diffuse large B-cell lymphoma (DLBCL) is the most common subtype of non-Hodgkin lymphoma, but primary ileal presentation is rare and may mimic abdominal emergencies. We report the case of a 40-year-old man with acute intestinal obstruction and no B symptoms. Abdominal CT revealed stenotic wall thickening at the jejunoileal transition and lymphadenopathy. Emergency laparotomy with segmental enterectomy was performed. Immunohistochemical analysis showed CD20+, Bcl-2+, PAX-5+, Ki-67 70% DLBCL, with CD3 expression in reactive T lymphocytes, and CD10-, Bcl-6-, and MUM-1-, classifying the tumor as the non-GCB subtype according to the Hans algorithm. Complete staging was limited by the emergency setting and the limited resources of the service. After six cycles of R-CHOP, the patient achieved complete metabolic remission (Deauville 2) on follow-up PET/CT, with no recurrence. This case highlights the diagnostic challenge of ileal DLBCL as a cause of acute obstructive abdomen and the effectiveness of emergency surgical resection followed by immunochemotherapy, even in aggressive non-GCB subtypes, while demonstrating the diagnostic limitations imposed by the emergency setting.

Keywords: Non-Hodgkin Lymphoma; B Lymphocytes; Intestinal Obstruction; Ileal Neoplasms; Rituximab.

Citation: Moura e Silva VA, Lucena LS, Gondim Neto ASB, Oliveira Filho FAG, Lira MAO, Dantas MEC, Matias MAP. Primary Ileal Diffuse Large B-Cell Lymphoma with Acute Intestinal Obstruction: Case Report and Narrative Literature Review. Brazilian Journal of Case Reports. 2026 Jan-Dec; 06(1):bjcr224.

<https://doi.org/10.52600/2163-583X.bjcr.2026.6.1.bjcr224>

Received: 14 August 2026

Accepted: 9 September 2026

Published: 16 September 2026



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1. Introduction

Non-Hodgkin lymphomas (NHL) comprise a heterogeneous group of malignant neoplasms originating from cells of the lymphoid system, with considerable variation in clinical presentation, biological behavior, and therapeutic response. Diffuse large B-cell lymphoma (DLBCL) is the most prevalent subtype, accounting for approximately 30 to 40% of NHL cases in adults [1]. Although most lymphomas initially manifest in peripheral lymph nodes, a significant proportion occurs at extranodal sites, with the gastrointestinal tract being the most commonly involved site [2]. Gastrointestinal NHL is predominantly classified as DLBCL, followed by mucosa-associated lymphoid tissue (MALT) lymphoma.

The pathogenesis of DLBCL is uncertain and may involve transformation from MALT lymphoma or represent a recurrent lymphoma. Primary DLBCL of the small intestine, particularly in the ileal region, is a rare presentation that may manifest in two ways: as an acute obstructive abdomen or as an incidental finding on imaging studies or during surgery. This rarity and the nonspecific nature of symptoms frequently hinder early diagnosis. Given this clinical complexity, the present case report, combined with a literature review, contributes to improving knowledge regarding the diagnostic and therapeutic management of primary ileal DLBCL with intestinal obstruction. Thus, this study aims to

evaluate enteric DLBCL (jejunum/ileum), including its presentation as an acute obstructive abdomen, the main symptoms, signs, and imaging findings indicative of tumor presence, staging, and possible therapeutic approaches. The importance of this analysis lies in disseminating the information available in the literature on this histological subtype, particularly as it concerns a primary extranodal lymphoma.

2. Materials and Methods

This study is a case report based on data collected from the electronic medical records of a referral emergency hospital, combined with a narrative literature review. The objective was to analyze an atypical case of primary intestinal diffuse large B-cell lymphoma (DLBCL) and contextualize it with the main scientific findings regarding this pathological condition. This methodological approach was chosen to provide an integrated understanding of the diagnostic and therapeutic particularities of the case in relation to established scientific knowledge.

For the narrative review, an exploratory search was conducted in the PubMed/MEDLINE, Embase, SciELO, LILACS, and Cochrane Library databases, using the following search strategy, adapted for each database: ("Lymphoma, Non-Hodgkin" OR "Diffuse Large B-Cell Lymphoma" OR "DLBCL") AND ("Intestinal Obstruction" OR "Ileal Neoplasms" OR "Small Intestine") AND ("Case Reports" OR "Case Series"). Filters were applied for language (Portuguese, English, and Spanish) and publication period (2015-2024). Inclusion criteria were: case reports and case series of primary intestinal DLBCL with complete clinicopathological descriptions; studies presenting data on demographic characteristics, clinical presentation, histopathological and immunohistochemical findings; and cases with a minimum follow-up of 12 months (for outcome analysis). Exclusion criteria were: gastric or colorectal lymphomas without small intestinal involvement; studies based exclusively on epidemiological data without case descriptions; reports with incomplete clinical or pathological data; and case series with fewer than 5 published cases.

After screening titles and abstracts and subsequently reviewing the full texts, 14 studies were selected to form the discussion base for this report. The article selection process is summarized in Figure 1. Data were synthesized descriptively (narratively), grouping the main findings regarding clinical presentation, diagnosis, treatment, and outcomes, as presented in Table 1. The search results were documented in a PRISMA flowchart (Figure 1). Data synthesis was narrative, with descriptive aggregation of the variables.

Table 1. Data extraction from the narrative literature review.

No.	Type of study	Main contributions to the case
[1]	Narrative review	Pathophysiological and therapeutic basis of DLBCL; R-CHOP as the gold-standard treatment; GC/non-GCB classification.
[2]	Narrative review	Epidemiology of gastrointestinal lymphomas: the small intestine is a rare site, and DLBCL is the most common subtype.
[3]	Retrospective study	Diagnostic delay in small intestinal tumors; obstruction as a late presentation.
[4]	Retrospective cohort	Intestinal DLBCL: obstruction is an indication for surgery; surgery + chemotherapy associated with better survival in localized stages.
[5]	Prospective cohort	Intestinal obstruction in 28% of intestinal lymphoma cases; DLBCL is the most common subtype (43%).
[6]	Cohort study	Validation of prognostic classification by IHC (CD10, Bcl-6, MUM-1); non-GCB associated with poorer prognosis.
[7]	Cohort study	Hans algorithm for GC/non-GCB classification; basis for classification of the present case.
[8]	Retrospective cohort	Prognostic score based on inflammatory markers for gastrointestinal DLBCL; complications and survival.

[9] Case report	DLBCL with jejunal obstruction and perforation; emergency surgery + chemotherapy with complete remission.
[10] Clinical guideline	Recommends R-CHOP for CD20+ DLBCL; stratification using IPI and FISH for MYC/BCL2.
[11] Systematic review	Obstruction/perforation are major complications; resection + chemotherapy associated with better outcomes.
[12] Systematic review	Staging (Lugano) and treatment of gastric DLBCL; principles applicable to intestinal disease.
[13] Narrative review	Duodenal lymphoma: DLBCL is the second most common subtype; surgery reserved for complications.
[14] Narrative review	Brazilian context of gastrointestinal lymphoma; importance of early diagnosis.

3. Case Report

A 40-year-old man, previously healthy, with an allergy to dipyrone, presented to the emergency department on May 13, 2025, with diffuse abdominal pain, nausea, vomiting (4 episodes), marked abdominal distension, and absence of bowel movements for 1 day. On admission, he was dehydrated and eupneic, with a painful facial expression and a distended abdomen diffusely tender to palpation, without signs of peritonitis. Acute intestinal obstruction was suspected. Abdominal computed tomography (CT) scans with and without contrast revealed diffuse distension of the small bowel loops with air-fluid levels and a transition point in the right iliac fossa. The contrast-enhanced examination demonstrated irregular, stenotic mural thickening in a small bowel loop at the jejunoileal transition, associated with intestinal obstruction. Mesenteric lymphadenopathy was also observed, with no free intraperitoneal fluid.

Given these findings, the patient underwent exploratory laparotomy with segmental enterectomy and side-to-side enteroenteric anastomosis. Intraoperatively, a tumor lesion was identified in a small bowel loop approximately 100 cm from the ileocecal valve, associated with lymphadenopathy extending to the root of the mesentery. Partial resection of the affected lymph nodes was performed. The surgical specimen was sent for anatomicopathological analysis.

Histopathological examination revealed diffuse lymphoid neoplasia composed of loosely cohesive cells infiltrating the mucosa, intestinal wall, and adjacent adipose tissue, with free surgical margins. Three of the eight resected lymph nodes showed neoplastic involvement. Immunohistochemical analysis demonstrated positivity for CD20, PAX-5, and Bcl-2, with a Ki-67 proliferation index of approximately 70%. The CD3 result was reported as "POLYCLONAL POSITIVE"; however, the report did not specify whether the staining occurred in neoplastic cells or reactive lymphocytes, as reports from emergency pathology services are generally objective and concise, prioritizing the primary diagnosis. CD30, CD10, Bcl-6, MUM-1, cyclin D1, CD15, EBV, C-MYC, and cytokeratins were negative. The morphological and immunophenotypic profile was compatible with diffuse large B-cell lymphoma (DLBCL). Applying the Hans algorithm to the available markers (CD10-, Bcl-6-, MUM-1-), the case was classified as a non-germinal center B-cell (non-GCB/ABC) subtype, which, according to the literature, is associated with a less favorable prognosis compared with the germinal center B-cell subtype. Photomicrographs of the histological slides were not obtained because the pathology service of the emergency hospital does not routinely capture and archive images.

FISH testing for gene rearrangements involving MYC, BCL2, and BCL6 was not performed because this high-complexity molecular biology method is not available in the routine workflow of the emergency service and requires prior planning and referral to a reference laboratory, which was incompatible with the immediate surgical indication. Similarly, oncological staging was limited by the absence of baseline PET/CT, bone marrow biopsy, lactate dehydrogenase (LDH) measurement, beta-2 microglobulin, and viral serologies (HIV, hepatitis B/C, and HTLV). Given the emergency nature of the condition,

with acute obstructive abdomen requiring transfer to the operating room within a few hours, laboratory investigation was focused on priority preoperative tests (complete blood count, coagulation profile, renal function, and electrolytes), with the oncological workup deferred to outpatient follow-up. Consequently, it was not possible to calculate the International Prognostic Index (IPI) or apply the Dawson criteria, as both require data including imaging-based staging, bone marrow evaluation, and serum LDH levels, which could not be obtained before emergency laparotomy.

After surgical recovery and hospital discharge on May 18, 2025, the patient underwent six cycles of chemotherapy, as documented in the follow-up PET/CT examination. Although the specific chemotherapy regimen was not documented in the records of the emergency hospital, as treatment was provided by an oncology service at another healthcare facility without integration of the medical records, it is important to note that the combination of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) constitutes the standard first-line regimen for CD20-positive diffuse large B-cell lymphoma (DLBCL) and is therefore fully compatible with the treatment administered. In January 2026, follow-up PET/CT demonstrated no metabolic activity suggestive of lymphoproliferative disease, with a Deauville score of 2 according to the Lugano criteria, indicating complete metabolic remission, with no evidence of recurrence at the time of this report.

3. Discussion and Conclusion

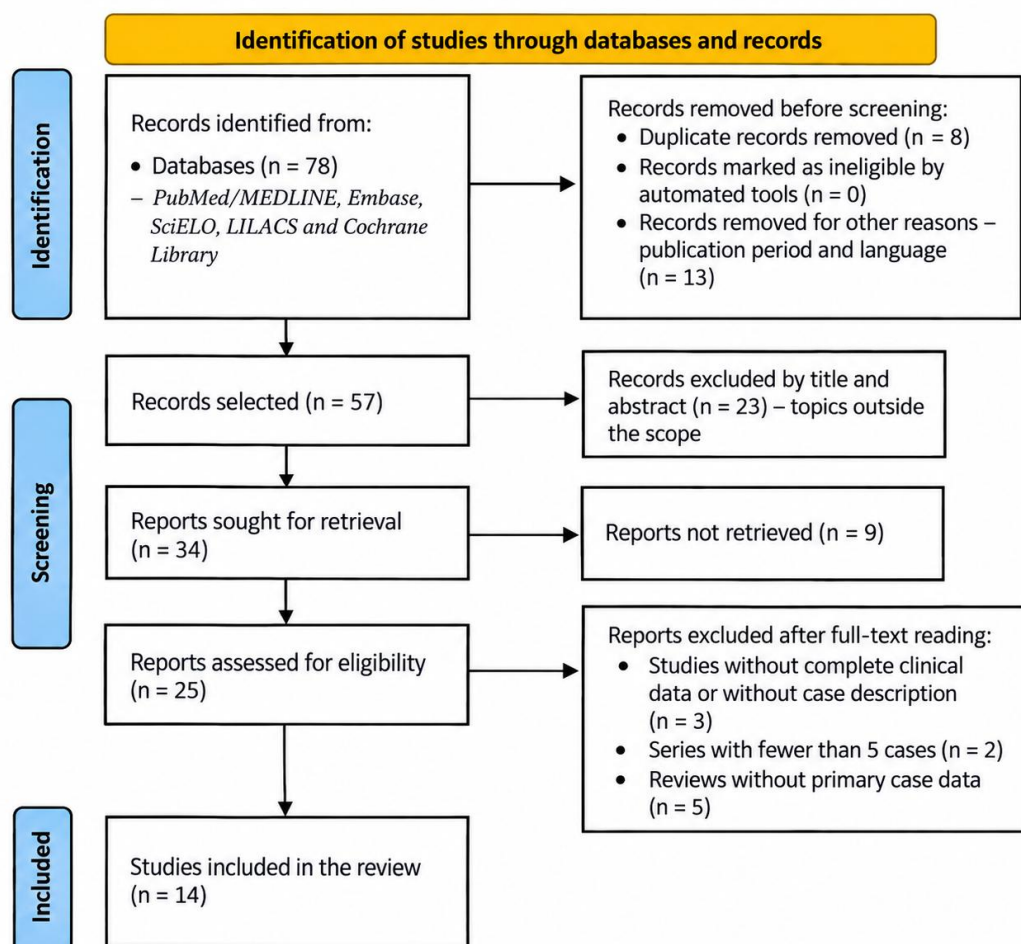
The database search (PubMed/MEDLINE, Embase, SciELO, LILACS, and Cochrane Library) identified 78 relevant studies. After applying the period (2015–2024) and language (Portuguese, English, and Spanish) criteria, 21 studies were excluded, leaving 57 for screening. After screening the titles and abstracts, 23 studies were excluded because they did not meet the inclusion criteria (out-of-scope topics, incomplete data, or duplicates), leaving 25 studies for full-text assessment. After full-text analysis, 9 studies were excluded because they did not meet all eligibility criteria (absence of a minimum follow-up of 12 months, incomplete clinical or pathological data, or case series with fewer than 5 cases), resulting in 14 studies included in the narrative review. The selection process is presented in Figure 1. After application of the eligibility criteria, five studies comprised the final sample of this review. Table 1 presents the characteristics of the included studies, including information on authorship, year of publication, country of origin, methodological design, sample characteristics, clinical presentation, tumor location, immunophenotype (Hans), treatment administered, and main outcomes.

The case reported here clearly illustrates the diagnostic challenges associated with primary small intestinal lymphomas. The patient presented with a classic picture of acute intestinal obstruction, diffuse abdominal pain, distension, vomiting, and cessation of bowel movements, a manifestation that, as reported by Daum et al. [5] and Park et al. [4], is not uncommon in ileal DLBCL and may constitute the first clinical manifestation of the disease. The absence of classic B symptoms, such as fever, night sweats, and weight loss, which are more frequently absent in extranodal gastrointestinal lymphomas than in nodal lymphomas, contributed to the initial diagnostic hypothesis being directed exclusively toward benign causes of intestinal obstruction, thereby delaying suspicion of an underlying malignancy.

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lymphomas, contributed to the initial diagnostic hypothesis being directed exclusively toward benign causes of intestinal obstruction, thereby delaying suspicion of an underlying malignancy.

Figure 1. Flowchart of the identification, screening, and selection process of studies included in the narrative literature review.



This pattern of presentation supports the secondary hypothesis formulated in this study, that primary ileal DLBCL predominantly manifests as an obstructive syndrome, differing significantly from gastric lymphomas, in which dyspeptic symptoms and weight loss tend to be more prominent. The rarity of primary small intestinal tumors, which account for less than 10% of all gastrointestinal neoplasms, with malignant tumors representing only 1% to 3% of these cases [2,3], further reinforces the low level of initial clinical suspicion.

Although abdominal computed tomography correctly identified the transition point in the right iliac fossa and segmental stenotic mural thickening, it was insufficient to establish the neoplastic nature of the lesion before surgery. This finding is consistent with the literature, which highlights the nonspecific nature of radiological examinations in the preoperative diagnosis of intestinal lymphomas [6]. The pattern of segmental mural thickening with mesenteric lymphadenopathy, although suggestive, may mimic other conditions, such as Crohn's disease, intestinal tuberculosis, or adenocarcinoma, with definitive diagnosis ultimately depending on histopathological and immunohistochemical analysis [6].

The immunohistochemical profile of the tumor, with positivity for CD20 and Bcl-2, a Ki-67 proliferation index of approximately 70%, and negativity for CD10, Bcl-6, and

MUM-1, is consistent with a non-germinal center B-cell (non-GCB) phenotype according to the Hans algorithm [7]. This classification, as demonstrated by Sehn and Salles [1], is associated with a poorer prognosis compared with the germinal center B-cell (GCB) subtype. The high cellular proliferation rate (Ki-67 of 70%) indicates a biologically aggressive tumor, a finding consistent with the acute and obstructive clinical presentation of the case. CD3 positivity observed in the immunohistochemical panel was attributed to reactive infiltrating T lymphocytes, given the unequivocal diagnosis of DLBCL with B-cell markers (CD20, PAX-5), thereby ruling out a mixed phenotype.

The involvement of three of eight resected lymph nodes demonstrates locoregional dissemination; however, the absence of metabolically active disease on post-treatment PET/CT demonstrates the characteristic chemosensitivity of DLBCL, even in aggressive extranodal presentations. This favorable outcome suggests that, despite the unfavorable immunohistochemical profile, multimodal treatment with surgery followed by R-CHOP chemotherapy may achieve complete remission [8].

Management of the case followed the approach recommended for complicated gastrointestinal lymphomas: surgery was indicated because of the emergency condition, namely intestinal obstruction, rather than because of the malignancy itself. Segmental resection with free surgical margins and primary enteroenteric anastomosis proved safe and effective, allowing both resolution of the obstructive condition and acquisition of tissue for definitive histopathological diagnosis [9]. This finding is consistent with the results of the narrative review, which showed that emergency surgery followed by chemotherapy is associated with high remission rates in patients with intestinal obstruction caused by DLBCL [11].

The question of the optimal timing of surgery in relation to chemotherapy remains unresolved in both the literature and this specific case. Surgery was mandated by the emergency condition, and postoperative adjuvant chemotherapy with R-CHOP, as recommended by current guidelines [10], was initiated at an appropriate postoperative time. The favorable outcome, with sustained complete remission, does not allow us to determine whether surgery before chemotherapy provided an advantage over treatment with R-CHOP alone. Nevertheless, this case demonstrates that, in situations of acute obstructive abdomen, surgical resection followed by chemotherapy is a feasible and potentially curative strategy [9].

The limitations inherent to a single-case report design, including the impossibility of statistical generalization, selection bias, and the absence of a control group, are acknowledged. In addition, although the patient's follow-up demonstrates complete remission to date, it remains relatively short for a definitive assessment of long-term outcomes, such as progression-free survival and overall survival [10]. The absence of baseline PET/CT, bone marrow biopsy, LDH measurement, and viral serologies constitutes an additional limitation, precluding the exact calculation of the IPI and full confirmation of the Dawson criteria. These limitations resulted from the emergency surgical setting, in which priority was given to management of the obstruction, and from the absence of institutional protocols for a complete oncological workup at admission.

Nevertheless, this report contributes to the literature by documenting a rare presentation of ileal DLBCL and highlighting the importance of including intestinal lymphomas in the differential diagnosis of acute obstructive abdomen. The findings of the narrative review reinforce that the combination of emergency surgery and R-CHOP is an effective strategy in patients with intestinal obstruction caused by DLBCL. The development of diagnostic and therapeutic algorithms adapted to the surgical and oncological setting remains a goal to be achieved through multicenter studies involving larger numbers of patients, capable of generating more robust evidence for the standardization of management.

Funding: None.

Research Ethics Committee Approval: The present study was approved by the Research Ethics Committee and conducted in accordance with Brazilian National Health Council Resolution No. 466/2012 (CAAE: 95669526.0.0000.5186).

Acknowledgments: None.

Conflicts of Interest: All other authors declare no conflicts of interest.

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