

Acute Appendicitis Or Appendiceal Lymphoma: Diffuse Large B-Cell Lymphoma Presenting as Appendicitis: A Case Report and Review of the Literature

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ABSTRACT

Introduction: Primary appendiceal lymphoma is an extremely rare neoplasm, representing approximately 0.015% of gastrointestinal lymphomas. We present a case of a young woman with acute appendicitis whose surgical specimen revealed an atypical lymphoid proliferation.

Case Presentation: A 21-year-old female presented with colicky epigastric pain evolving over two weeks. Physical examination revealed right iliac fossa tenderness with a positive McBurney's sign. Laboratory studies showed leukocytosis (12,200/ μ L) with neutrophilia. Abdominal ultrasound demonstrated a hypoechoic lesion in the right iliac fossa (56 $\times$ 30 mm) with peripheral hypervascularity on Doppler. Laparoscopic appendectomy was performed, revealing a markedly edematous appendix with concentric wall thickening of neoplastic appearance. Histopathological examination confirmed diffuse large B-cell lymphoma, activated B-cell subtype, with a Ki 67 proliferation index of 75%, consistent with primary appendiceal lymphoma.

Discussion: Appendiceal lymphomas constitute fewer than 0.015% of gastrointestinal lymphomas and present a diagnostic challenge given their nonspecific clinical picture. B cell non Hodgkin lymphoma is the predominant subtype. Simple appendectomy may be curative in localized disease; however, multidisciplinary oncologic evaluation is essential.

Conclusion: Primary appendiceal lymphoma should be considered in the differential diagnosis of appendiceal tumors. Pathological examination of all appendectomy specimens is mandatory to ensure accurate diagnosis and appropriate oncologic management.

KEYWORDS: Appendiceal lymphoma; Non Hodgkin lymphoma; Diffuse large B cell lymphoma; Appendiceal neoplasm; Laparoscopic appendectomy; Gastrointestinal lymphoma

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1. INTRODUCTION

Acute appendicitis remains one of the most frequent surgical emergencies encountered in both adult and pediatric populations, with a lifetime risk of approximately 8.6% in males and 6.7% in females, most commonly affecting individuals between 10 and 20 years of age. Although the vast majority of cases are attributable to simple inflammatory disease, a growing body of evidence suggests that up to 2.9% of patients diagnosed with acute appendicitis may harbor an

underlying colorectal or appendiceal malignancy, compared with only 0.1% of the general population ^[1,2].

Appendiceal neoplasms are identified in approximately 0.2–3.09% of all appendectomy specimens. The World Health Organization (WHO) classifies appendiceal tumors into two broad categories: epithelial tumors, which include adenomas, adenocarcinomas, carcinoid tumors (well differentiated neuroendocrine tumors), tubular carcinoids, mucinous carcinoids, and mixed carcinoid adenocarcinomas; and non

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epithelial tumors, encompassing neuromas, lipomas, leiomyomas, gastrointestinal stromal tumors (GISTs), leiomyosarcomas, and Kaposi's sarcoma, among others [3]. Latin American series estimate the prevalence of appendiceal tumors at 0.8% of all appendectomy specimens [4].

Primary lymphoma of the appendix represents an exceptionally rare entity even within this already uncommon group. The gastrointestinal tract is the most frequent site of extranodal lymphomas, accounting for 30–45% of all extranodal cases, with the stomach being the most commonly involved organ, followed by the small intestine, colon, and esophagus [5]. However, primary appendiceal lymphoma accounts for only approximately 0.015% of all gastrointestinal lymphomas, with very few cases reported in the world literature [6]. Virtually all reported cases have corresponded to non Hodgkin lymphoma, with B cell lineage being the predominant subtype [7].

Given the extreme rarity of this entity and the diagnostic and therapeutic challenges it poses, we present the case of a young woman who underwent laparoscopic appendectomy for presumed acute appendicitis, with a subsequent histopathological diagnosis of primary appendiceal diffuse large B cell lymphoma. The objectives of this report are to describe the clinical presentation, intraoperative findings, histopathological characteristics, and surgical management of primary appendiceal lymphoma in a secondary level care setting, and to provide a concise review of the available literature.

2. CASE PRESENTATION

2.1. Patient History and Clinical Presentation}

A 21 year old female with no significant past medical history, no known drug allergies, no prior surgical interventions, and

no history of tobacco use, alcohol consumption, or blood transfusions presented to the emergency department. Her obstetric and gynecological history was unremarkable: menarche at age 13, regular menstrual cycles (6–7 days in length, 28–33 day intervals), and no prior gestations (G0P0A0C0). She reported initiating sexual activity at age 18 with one lifetime partner.

The patient's symptoms began approximately two weeks prior to admission with intermittent colicky epigastric pain of moderate intensity, for which she self medicated without significant relief. On February 9, 2026, she experienced a recurrence of colicky pain that originated in the epigastrium and migrated to the periumbilical region. The absence of additional symptoms such as fever, nausea, vomiting, or changes in bowel habits prompted her to seek evaluation at our emergency department.

2.2. Physical Examination

On physical examination, the abdomen was soft and non tender to palpation, except for tenderness in the right iliac fossa and hypogastrium. McBurney's point was positive. The Rovsing, obturator, and psoas signs were negative. Bowel sounds were present, and there were no signs of peritoneal irritation or rebound tenderness. Vital signs were within normal limits.

2.3. Diagnostic Studies

Laboratory results obtained on February 13, 2026 are summarized in Table 1. Notably, the patient presented with mild leukocytosis (12,200/ μ L) with neutrophilic predominance (76%) and an elevated C reactive protein (CRP) of 4.15 mg/dL, consistent with an acute inflammatory process. The remaining biochemical parameters were within normal ranges.

Table 1. Laboratory Results on Admission (February 13, 2026)

Parameter	Result	Reference Range	Interpretation
White Blood Cell Count	12,200 / μ L	4,500–11,000 / μ L	Elevated ↑
Neutrophils	76%	50–70%	Elevated ↑
Hemoglobin	12.0 g/dL	12.0–16.0 g/dL (F)	Low-normal
Hematocrit	36%	36–46% (F)	Normal
Platelets	392,000 / μ L	150,000–400,000 / μ L	Normal
C-Reactive Protein	4.15 mg/dL	<0.5 mg/dL	Elevated ↑
Creatinine	0.72 mg/dL	0.5–1.1 mg/dL	Normal
Glucose	103 mg/dL	70–100 mg/dL	Borderline
Total Bilirubin	0.46 mg/dL	0.2–1.2 mg/dL	Normal
Prothrombin Time	13.9 s	11.0–14.5 s	Normal
INR	1.27	<1.5	Normal

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Parameter	Result	Reference Range	Interpretation
Sodium	138 mEq/L	136–145 mEq/L	Normal
Potassium	3.81 mEq/L	3.5–5.1 mEq/L	Normal
Chloride	102 mEq/L	98–107 mEq/L	Normal

F = female reference range; ↑ = elevated above reference range.

Abdominal ultrasound (February 13, 2026) demonstrated, at the level of the right iliac fossa, a hypoechoic, poorly defined lesion measuring 56 × 18 × 30 mm (volume approximately 16 mL), with avid peripheral vascularity on Doppler imaging and surrounding echogenic fat, consistent with inflammatory change. These findings raised concern for a peri appendiceal inflammatory process, though a neoplastic etiology could not be excluded.

A simple computed tomography of the abdomen and pelvis was performed (February 13, 2026), in which a tubular structure was identified in the right iliac fossa, enlarged in caliber and with parietal thickening. Significant striation of the periappendicular fat was observed, associated with an adjacent collection with heterogeneous density. Reactive thickening of the cecal wall and a low amount of free fluid in the pelvis were also documented.



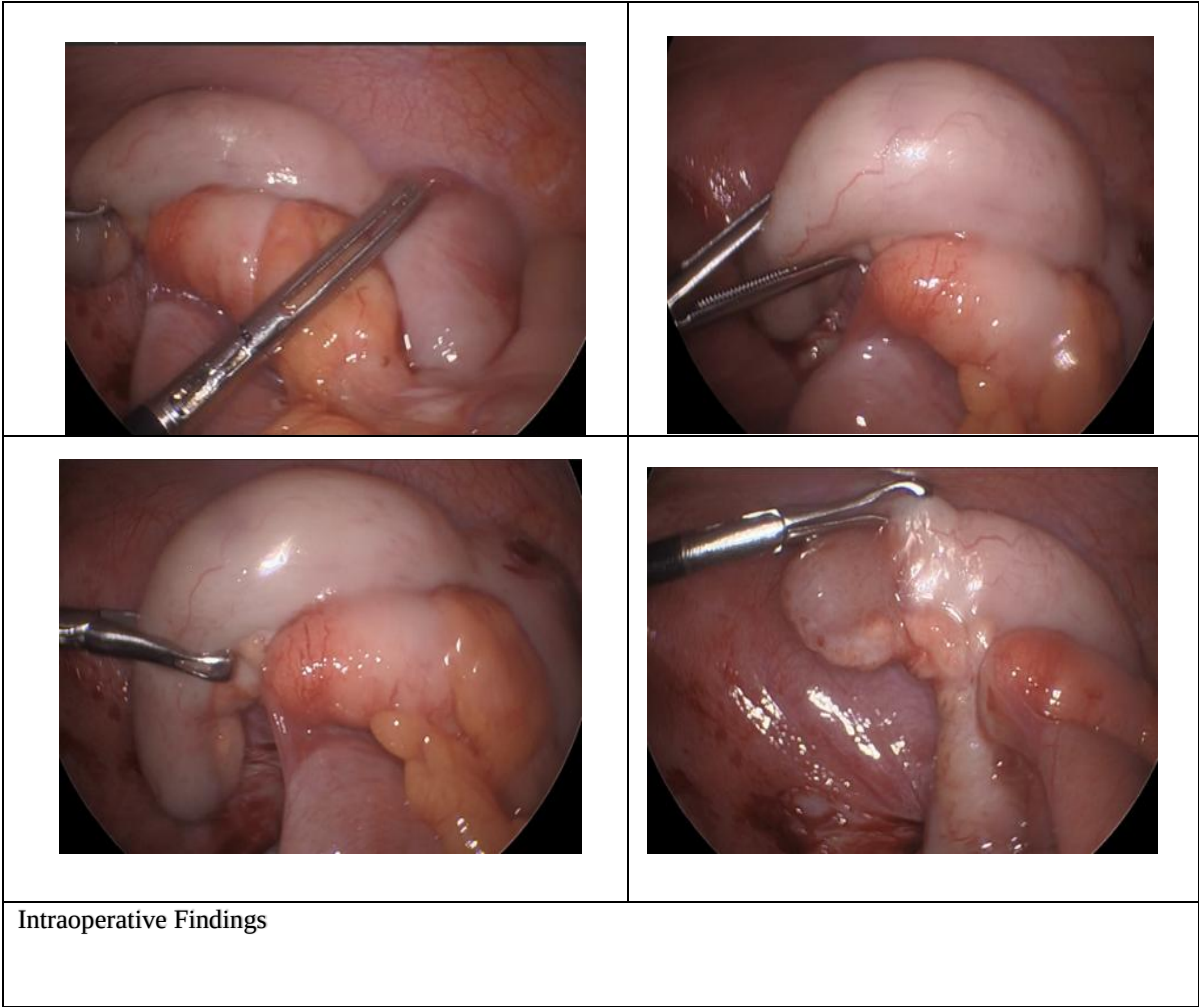
2.4. Intraoperative Findings and Surgical Technique

After obtaining written informed consent, the patient was taken to the operating room. Under general balanced anesthesia, the patient was positioned supine, and the surgical field was prepared with standard aseptic technique. A laparoscopic approach was performed using a three trocar technique: a 10 mm supraumbilical trocar was placed via the Hasson open technique, followed by two additional trocars (5 mm suprapubic and 5 mm left iliac fossa) under direct vision. Pneumoperitoneum was established and maintained at 12 mmHg.

Systematic diagnostic laparoscopy revealed a peri appendiceal plastron with a markedly edematous appendix exhibiting significant concentric wall thickening and a friable base, with no active bleeding. The macroscopic appearance

was highly suspicious for a neoplastic process, raising the intraoperative differential diagnosis of carcinoid tumor or other appendiceal neoplasm. The mesoappendix was divided using LigaSure™ (Medtronic). The appendiceal base was managed with a Z stitch using 2 0 silk suture. The appendix was excised and externalized via the umbilical port in an endobag, without specimen contamination.

A thorough review of the peritoneal cavity was performed, including examination of the terminal ileum and pelvis. No additional lesions were identified, and no macroscopic leaks or free fluid were noted. The abdominal cavity was irrigated, and residual fluid was aspirated. Hemostasis was confirmed. Trocars were removed under direct vision, and port sites were closed in layers. Total operative time was 150 minutes. Estimated blood loss was minimal. The patient was transferred to the ward in stable condition.



2.5. Histopathological Findings

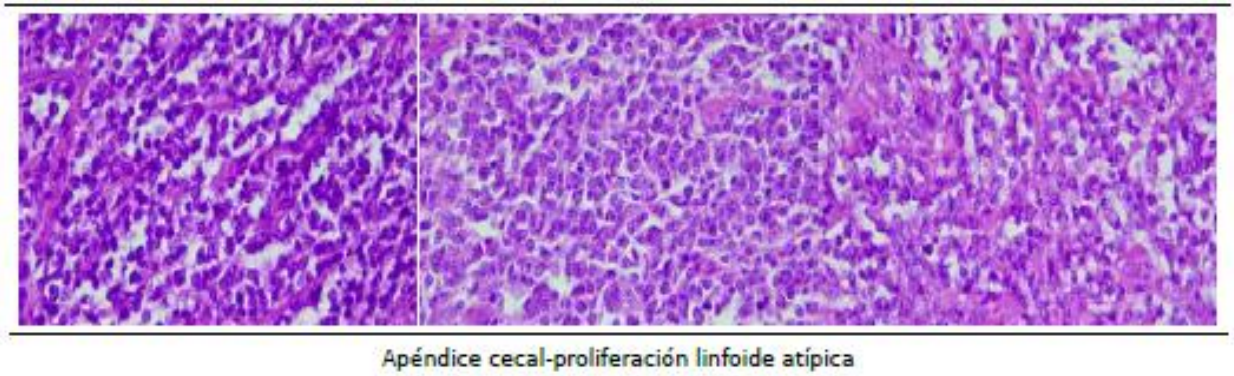
- Gross Description:** The specimen consisted of an appendiceal fragment measuring 5.0 × 2.0 × 1.0 cm, which was firm and whitish on cut section. Two additional irregular, firm, fibrous fragments measuring 1.8 × 1.0 cm in aggregate were

also identified. The proximal margin was submitted separately.

- Microscopic Description:** Histologic sections showed a diffuse atypical lymphoid proliferation infiltrating the appendiceal wall from the serosa to the mucosa. The neoplastic cells were large with vesicular

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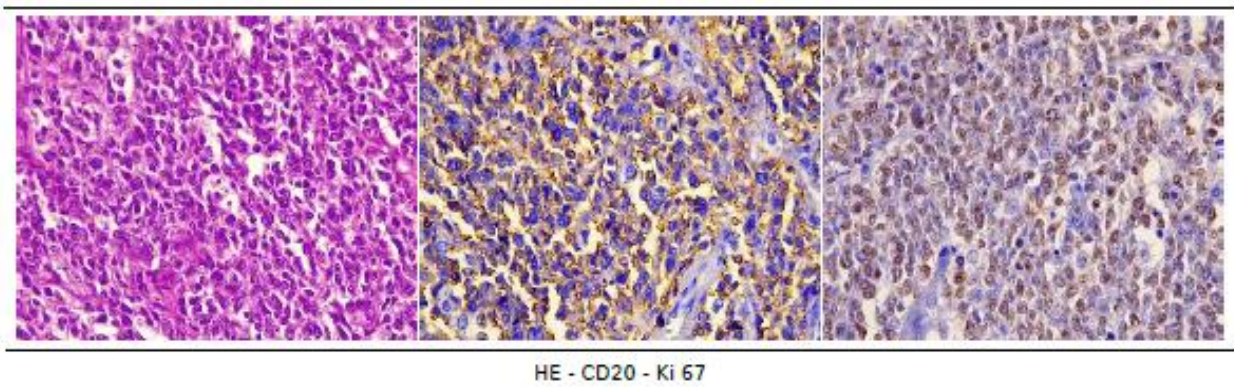
chromatin and evident nucleoli. Areas of zonal necrosis were observed.



Immunohistochemistry: Immunohistochemical studies demonstrated the following results:

Antibody	Result	Interpretation
CD20	Positive	B-cell marker
PAX5	Positive	B-cell marker
CD3	Negative	T-cell marker
CD30	Negative	Excludes anaplastic lymphoma
CD10	Negative	Non-germinal center phenotype
Bcl-6	Negative	Non-germinal center phenotype
Bcl-2	Positive	Anti-apoptotic protein
MUM1	Positive (>30%)	Activated B-cell phenotype
c-MYC	Positive (>40%)	High proliferative activity
Ki-67	75%	High proliferation index

Final Diagnosis: Non-Hodgkin lymphoma, diffuse large B-cell lymphoma (DLBCL), activated B-cell (ABC) subtype (CD20+, MUM1+, Bcl-2+, CD10–, Bcl-6–). High proliferation index of 75%.



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Pathological Stage: Ann Arbor stage IE (disease confined to the appendix).

2.6. Postoperative Course and Follow-up

The patient's postoperative course was uneventful. Oral intake was initiated 24 hours after surgery, and she was discharged on the second postoperative day. She was evaluated by the Hematology and Medical Oncology services. Staging computed tomography of the chest, abdomen, and pelvis showed no evidence of residual or distant disease.

Adjuvant immunochemotherapy was initiated with R CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) for a total of four cycles. At six month follow up, the patient remained asymptomatic with no clinical or radiological evidence of recurrence.

3. DISCUSSION

Primary appendiceal lymphoma is among the rarest appendiceal neoplasms, representing less than 0.015% of all gastrointestinal lymphomas^[6]. The paucity of reported cases makes it a significant diagnostic challenge for both clinicians and pathologists. To our knowledge, the present case adds to fewer than 100 well documented cases in the world literature and represents one of the youngest patients described with this diagnosis in the Latin American context.

The clinical presentation of appendiceal lymphoma closely mimics that of benign acute appendicitis, as observed in our patient: progressive right iliac fossa pain, localized tenderness with a positive McBurney's sign, mild leukocytosis, and elevated inflammatory markers^[8]. This overlap in presentation makes preoperative diagnosis virtually impossible without advanced imaging. While abdominal computed tomography (CT) may suggest appendiceal wall thickening or a mass like lesion, the findings are nonspecific and often interpreted as complicated appendicitis^[9]. In our case, Doppler ultrasound showed peripheral hypervascularity and perienteric fat stranding. In retrospect, these findings could have suggested a neoplastic process, although carcinoid tumor was the primary intraoperative suspicion. The diagnosis was ultimately established by histopathology.

Non Hodgkin lymphoma of B cell lineage accounts for the overwhelming majority of reported appendiceal lymphomas^[7]. The most frequently identified subtypes in published series include diffuse large B cell lymphoma (DLBCL), Burkitt lymphoma—particularly in pediatric and young adult patients—and mucosa associated lymphoid tissue (MALT) lymphoma. Our case was classified as DLBCL of the activated B cell (ABC) subtype, characterized by MUM1 positivity, CD10 negativity, and a high proliferation index (Ki 67 75%). This subtype is associated with a more aggressive course and typically requires systemic immunochemotherapy^[8].

Regarding surgical management, simple appendectomy is generally considered curative for early stage, localized appendiceal lymphoma confined to the appendix without peritoneal dissemination^[8,9]. Right hemicolectomy has been advocated in cases with involvement of the mesoappendix, regional lymph nodes, or cecal base, and for high grade histological subtypes, although evidence supporting this approach over appendectomy alone remains limited given the rarity of the disease^[10]. Cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (HIPEC) is reserved for cases with peritoneal dissemination, particularly in the context of mucinous tumors^[8]. In our case, laparoscopic appendectomy was technically feasible despite the inflammatory plastron, and no evidence of regional or distant disease was identified intraoperatively or on staging imaging.

Postoperative oncologic management depends critically on the histological subtype, stage, and molecular profile of the tumor. Most reported cases of appendiceal lymphoma have been managed with adjuvant chemotherapy—most commonly rituximab based regimens (R CHOP) for B cell lymphomas—in close coordination with hematology and oncology teams^[9]. Surveillance endoscopy and CT imaging are typically recommended, though no standardized follow up protocols exist due to the rarity of this entity.

This case reinforces the critical importance of routine histopathological examination of all appendectomy specimens, regardless of the presumed etiology. In clinical settings where intraoperative frozen sections are available, their use may be considered when macroscopic findings are atypical, as was the case here. The finding of a thickened, friable appendiceal wall without the classic features of phlegmonous appendicitis should alert the surgeon to the possibility of an underlying neoplasm and prompt appropriate specimen handling and expedited pathological evaluation.

4. CONCLUSION

Primary appendiceal lymphoma is a rare but clinically significant entity that can present indistinguishably from acute appendicitis. The diagnosis is invariably made postoperatively upon histopathological examination of the appendectomy specimen. Surgeons must maintain a heightened index of suspicion when intraoperative findings deviate from the typical features of benign appendicitis. This is particularly important in young patients presenting with appendiceal wall thickening and a neoplastic appearance, where the differential diagnosis includes aggressive lymphomas such as DLBCL. Routine pathological analysis of all resected appendices is mandatory. Once the diagnosis is established, a multidisciplinary oncological approach—encompassing hematology, medical oncology, and radiology—is essential to determine staging and define the optimal adjuvant treatment strategy. Further case reports and

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multicenter series are needed to better characterize the natural history, optimal surgical approach, and long term outcomes of this rare condition.

Ethical Considerations

The patient was fully informed about the diagnosis, procedures, and potential risks. Written informed consent was obtained for the surgical and anesthetic procedures as well as for publication of this case report, in accordance with the ethical principles of the Declaration of Helsinki and the institutional regulations of the hospital

Conflict of Interest

The authors declare no conflict of interest.

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Reporting Compliance.

This case report was prepared in accordance with the CARE (CAse REport) 2013 guidelines. The CARE checklist has been completed and is available as supplementary material upon request.