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Primary Appendiceal Diffuse Large B-Cell Lymphoma Mimicking Acute Appendicitis: A Case Report

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Abstract

Primary appendiceal lymphoma is an exceptionally rare malignancy that can closely resemble acute appendicitis, making preoperative diagnosis difficult. We report the case of a 24-year-old man who presented with right lower quadrant abdominal pain and imaging findings suggestive of an appendiceal mucocele. Open appendectomy revealed a markedly enlarged, firm appendix. Histopathological examination demonstrated diffuse large B-cell lymphoma of the germinal center B-cell subtype. Postoperative positron emission tomography/computed tomography showed no evidence of nodal or extranodal disease. The patient received four cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) and achieved complete remission. This case emphasizes the importance of routine histopathological evaluation of appendectomy specimens and early multidisciplinary management in atypical appendiceal presentations.

Keywords: appendix, appendicitis, lymphoma, diffuse large B-cell lymphoma, non-Hodgkin lymphoma

Ethical Approval
In accordance with the policies of Al-Ahli Hospital, single-patient case reports are exempt from formal ethics committee approval

Informed Consent
Written informed consent was obtained directly from the patient for the publication of this case report and all accompanying anonymized radiological and histopathological images.

Conflict of Interest
The authors declare no conflicts of interest relevant to the content of this manuscript.

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OA: Conceptualization, Data curation, Investigation, Writing – review & editing; ABAJQ: Investigation, Resources, Validation, Writing – review & editing; ATH: Methodology, Visualization, Writing – original draft, Writing – review & editing; MN: Supervision, Project administration, Validation, Writing – review & editing. All authors have read and approved the final version of the manuscript.

Use of Artificial Intelligence (AI) Statement
During the preparation of this work, the authors used Google Gemini solely to improve English language readability, grammar, and figure layout formatting. After using this tool, the authors reviewed and edited all content and take full responsibility for the scientific accuracy and integrity of the publication.

Data Availability
The clinical data, imaging, and pathology records supporting the findings of this case report are restricted due to patient privacy and institutional confidentiality regulations, but anonymized datasets are available from the corresponding author upon reasonable request.

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Introduction

Lymphoid malignancies are broadly divided into Hodgkin lymphoma and non-Hodgkin lymphoma (NHL), with NHL accounting for most cases [1]. Extranodal involvement is a well-recognized feature of NHL, and the gastrointestinal tract is the most frequently affected extranodal site [1, 2]. Nevertheless, primary lymphomas arising within the gastrointestinal tract remain uncommon and account for only a small proportion of all gastrointestinal neoplasms [1, 2].

Within this context, appendiceal involvement is particularly unusual. Primary appendiceal lymphoma is an exceedingly rare clinical entity, representing only a minute fraction of gastrointestinal lymphomas and an even smaller proportion of appendectomy specimens [3, 4]. Among its histological subtypes, diffuse large B-cell lymphoma (DLBCL) is the most commonly reported subtype at this location [3].

A major diagnostic challenge is that appendiceal lymphoma often presents with symptoms that are indistinguishable from acute appendicitis. Consequently, the diagnosis is rarely suspected before surgery and is typically established only after histopathological evaluation [1–3]. We report a case of primary appendiceal DLBCL in a young adult and emphasize the importance of maintaining a broad differential diagnosis in atypical appendiceal presentations.

Case Presentation

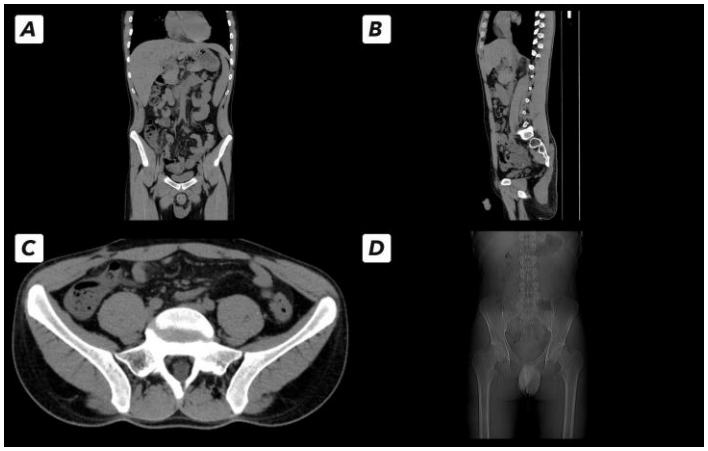
A 24-year-old male presented with a 2-day history of right lower quadrant abdominal pain associated with vomiting and diarrhea. He was afebrile and hemodynamically stable. Physical examination revealed localized tenderness without signs of peritonitis. Laboratory investigations showed a hemoglobin level of 13.4 g/dL and a white blood cell count of 8,400/μL.

Ultrasonography demonstrated a 5.7 × 2.0 cm tubular, fluid-filled structure with echogenic periappendiceal fat. Computed tomography revealed an enlarged appendix measuring 2.5 cm in diameter, raising suspicion for an appendiceal mucocele; this imaging pattern has been described in appendiceal lymphoma (Figure 1) [1]. Given the bulky appearance of the appendix, open appendectomy was performed.

Intraoperatively, the appendix was markedly enlarged, firm, and adherent to adjacent tissues (Figure 2). Histopathological examination revealed diffuse infiltration by large atypical lymphoid cells (Figures 3, 4). Immunohistochemical analysis showed positivity for CD20, CD10, BCL6, and BCL2, with a Ki-67 proliferation index of approximately 40%, consistent with germinal center B-cell subtype DLBCL (Figures 5-7). Tumor cells were negative for CD3, TdT, and Cyclin D1. These findings are consistent with previously reported immunophenotypic profiles of appendiceal DLBCL [2, 3].

Postoperative positron emission tomography/computed tomography demonstrated no evidence of nodal or extranodal involvement, consistent with stage IE primary appendiceal lymphoma [4]. The patient received four cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP), the standard first-line regimen for localized DLBCL [5, 6]. Follow-up imaging at three and six months confirmed complete remission, and the patient remains asymptomatic and fully active.

Figure 1. Representative CT images of the abdomen and pelvis. (A) Coronal CT reconstruction. (B) Sagittal CT reconstruction. (C) Axial CT image through the lower abdomen/pelvis. (D) Frontal CT scout (topogram) projection radiograph. The diagnostic CT showed a markedly dilated appendix (approximately 2.5 cm in diameter) with wall thickening and periappendiceal fat stranding, without perforation or abscess.



Abbreviations: CT: computed tomography.

Figure 2. Gross intraoperative appearance of the appendix.

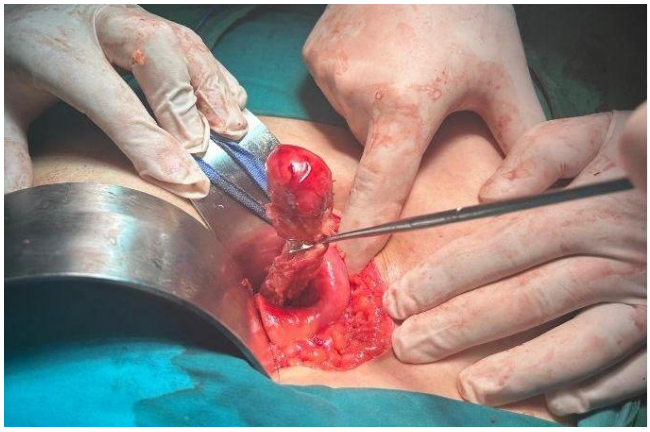


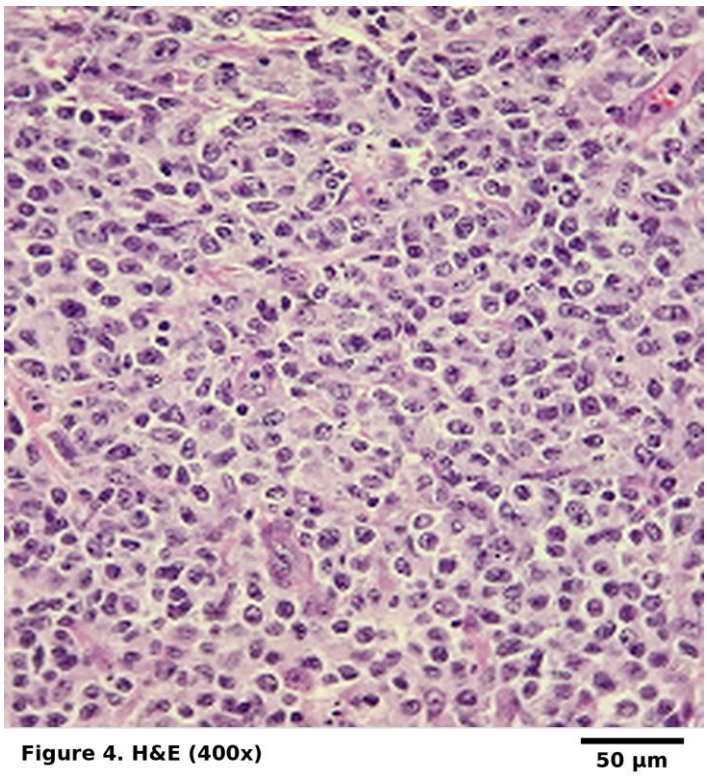
Figure 3. Whole-mount H&E section showing effacement of the appendiceal lumen by proliferation of atypical lymphoid cells.



Abbreviations: H&E: hematoxylin and eosin.

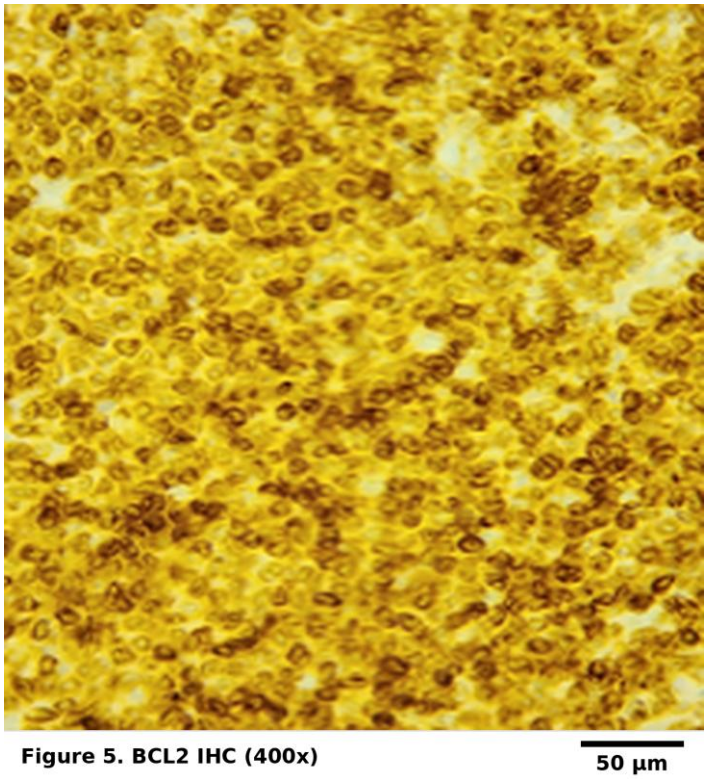
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Figure 4. H&E section showing neoplastic lymphoid cells with large, irregular nuclei, open chromatin, prominent nucleoli, and a moderate amount of pale cytoplasm.



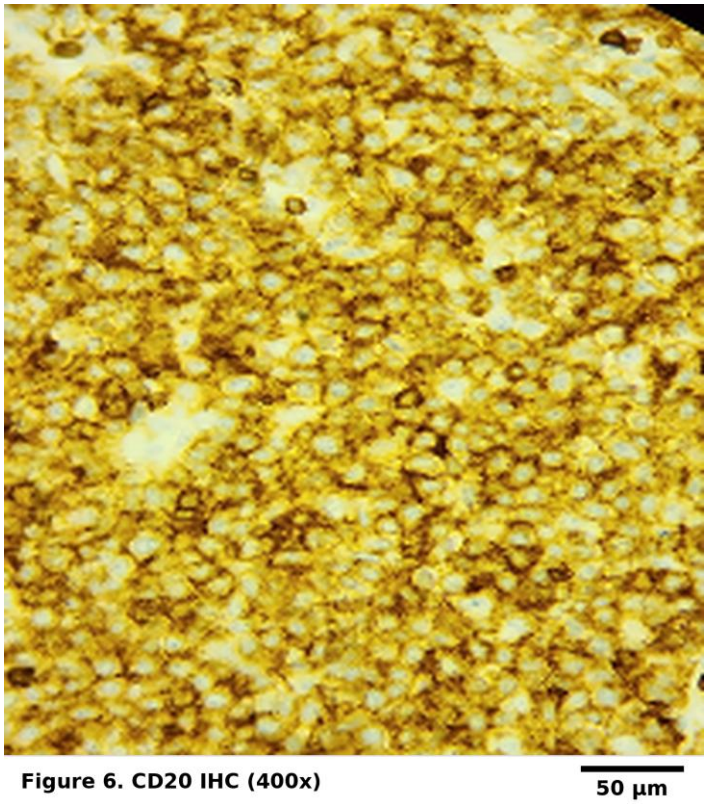
Abbreviations: H&E: hematoxylin and eosin.

Figure 5. BCL2 immunostaining.



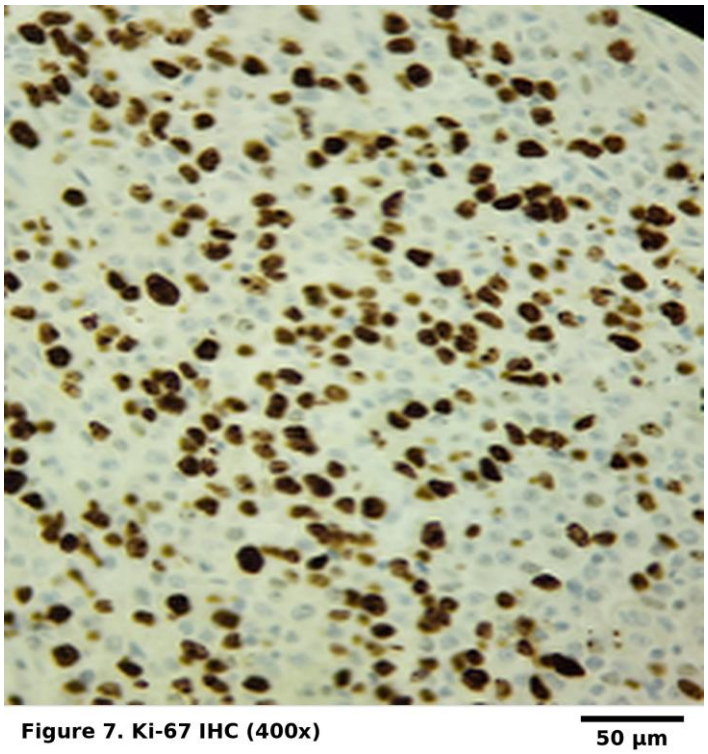
Abbreviations: BCL2: B-cell lymphoma 2.

Figure 6. CD20 immunostaining.



Abbreviations: CD20: cluster of differentiation 20.

Figure 7. Ki-67 proliferation index.



Abbreviations: Ki-67: proliferation marker Ki-67.

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Discussion

Primary appendiceal lymphoma is an exceptionally uncommon malignancy, and its clinical presentation often mirrors that of acute appendicitis, which can obscure the underlying diagnosis [2, 7, 8]. From a radiological perspective, certain features may raise suspicion for a lymphoproliferative process, including marked appendiceal enlargement, luminal dilatation exceeding 15 mm, and periappendiceal lymphadenopathy [1]. However, these findings are not specific and may overlap with more common conditions, such as appendiceal mucocele or uncomplicated appendicitis. This overlap reinforces the pivotal role of histopathological examination in establishing the diagnosis [7, 9, 10].

Diffuse large B-cell lymphoma constitutes a substantial proportion of reported cases of primary appendiceal lymphoma, accounting for approximately one-third of cases in published series [8, 11]. The condition usually affects older individuals, with a median age in the sixth decade of life [8]. In this context, the diagnosis in a 24-year-old patient is noteworthy and contributes to the limited number of similar cases reported in younger populations [12].

Management strategies for localized disease have evolved toward a combined approach. Surgical resection followed by systemic chemotherapy has been associated with favorable outcomes in early-stage disease. In particular, immunochemotherapy with rituximab combined with cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) remains the standard first-line regimen for DLBCL [5, 6]. Reported survival outcomes in stage IE disease are encouraging, with high rates of long-term remission observed in multiple studies [8, 13, 14].

Recent advances in lymphoma treatment have broadened the therapeutic landscape for relapsed or refractory DLBCL, including the use of novel agents such as bispecific antibodies [6]. Although the role of these therapies in primary appendiceal lymphoma remains to be fully defined, they may further improve outcomes in selected patient populations.

Conclusion

This case underscores that primary appendiceal lymphoma, although rare, should be considered when appendiceal imaging or intraoperative findings are atypical. Routine histopathological assessment of appendectomy specimens remains essential, and early multidisciplinary coordination is critical to ensure accurate staging, timely chemotherapy, and appropriate surveillance.

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