Journal of Medical Surgical and Allied Sciences
Case Report

Acute appendicitis as the initial presentation of primary appendiceal diffuse large b-cell lymphoma: a diagnostic challenge

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Volume: 3, Issue: 1, Pages: 6-9
DOI: https://doi.org/10.37446/jmedsurg/cr/3.1.2025.6-9
Received: 21 November 2024 / Accepted: 22 March 2025 / Published: 30 June 2025

Background: Primary appendiceal lymphoma is exceedingly rare, representing less than 1% of appendiceal neoplasms. Its clinical presentation is usually indistinguishable from acute appendicitis, often leading to a diagnosis only after histopathological evaluation.

Case presentation: We report the case of a 14-year-old male who presented with acute right iliac fossa pain and underwent appendectomy for suspected appendicitis. The patient was operated on, and the tissue was sent for histopathology, where a diagnosis of primary appendiceal lymphoma was made. Histopathological analysis and immunohistochemistry revealed a diffuse large B-cell lymphoma that was immunopositive for CD20. The patient succumbed to death on the seventh post-operative day.

Conclusion: This case highlights the importance of routine histopathological examination of appendectomy specimens and the need for multidisciplinary management. A high suspicion of non-Hodgkin lymphoma is important for early diagnosis, so that patients will receive treatment in the early stage, which can lead to a better prognosis.

Keywords: *Appendicitis, Gastrointestinal, Non-Hodgkin Lymphoma, Rare, Diffuse large B-cell lymphoma*

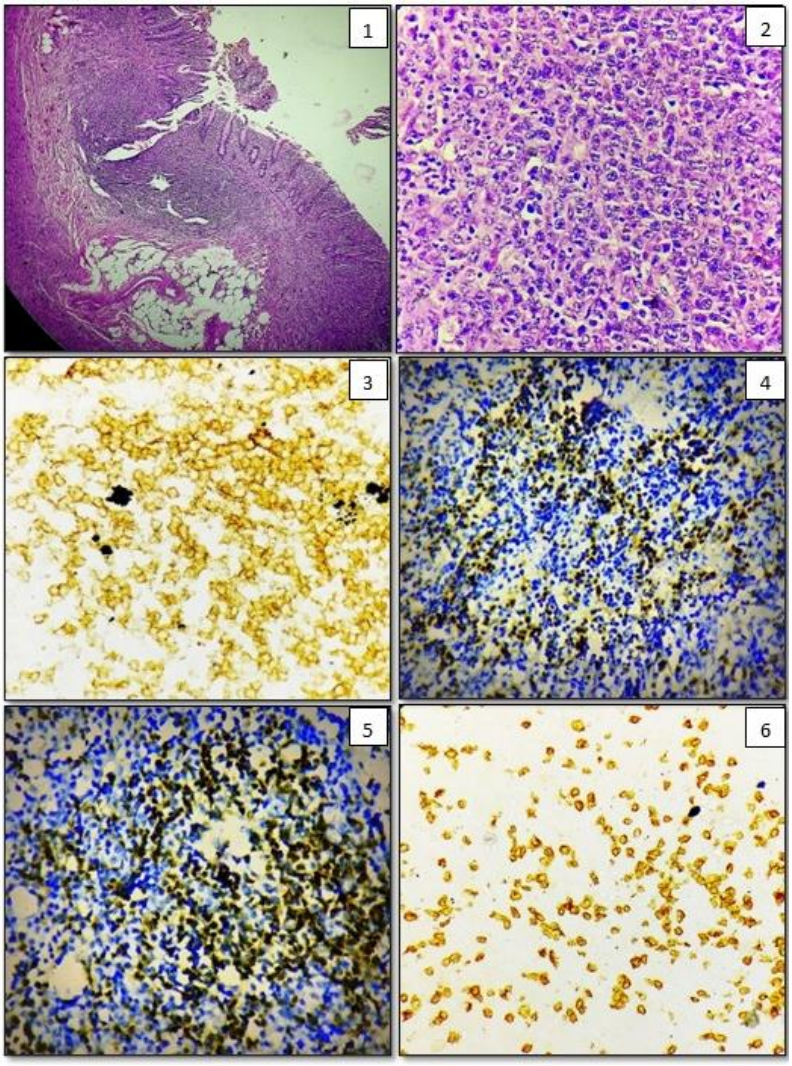
Introduction

Primary gastrointestinal non-Hodgkin lymphoma (GI-NHL) constitutes some of the most common types of extra-nodal non-Hodgkin lymphomas (NHL), accounting for 30%-40% of cases (Hawkes et al., 2012). However, these lymphomas are relatively rare, accounting for only 1-4% of all gastrointestinal (GI) malignancies (McSwain & Beal, 1944). Primary appendiceal lymphoma (PAL) is extremely uncommon, representing about 0.015% of all GI lymphomas (García-Norzagaray et al., 2019; Bugeja & Micallef, 2019; Ayub et al., 2017). Acute appendicitis is the most common clinical presentation of appendiceal lymphoma, including anorexia or hyporexia, weight loss, pain, and intestinal bleeding. Appendiceal neoplasms are generally diagnosed intraoperatively, supporting histopathological analysis as a routine procedure for all appendectomies. We report a rare case of primary appendiceal lymphoma in a 14-year-old boy presenting as acute appendicitis and review relevant literature to emphasize diagnostic challenges and management strategies.

Case presentation

A 14-year-old male child presented with acute right iliac fossa pain and clinical manifestations of acute abdomen for which emergency abdominal surgery was planned. During surgery swollen appendix was noted. Based on clinical

examination, a diagnosis of acute appendicitis was made, and appendectomy was performed. We received a bulky appendix measuring 7 cm in length. On cutting open, the lumen was obliterated by a grey white friable growth. Histopathology sections from the appendix showed dense and diffuse infiltration of atypical large lymphoid cells in the submucosa. These cells showed vesicular nuclei, prominent nucleoli, and scant to moderate amount of cytoplasm, suggestive of a non-Hodgkin lymphoma [Figures 1 and 2]. Sections from the mesenteric lymph nodes showed tumor metastasis. Immunohistochemistry was performed to determine the type of lymphoma. CD20 [Figure 3], which determines B lymphoid cell lineage, and CD10 and bcl6 [Figure 4 and 5], which determine a germinal centre phenotype, were positive. Ki67%, which determines the proliferative index, was 80%. CD3 was positive in scattered cells [Figure 6]; bcl2, CD5, CD23, MUM1, and cMYC were negative. A diagnosis of diffuse large B-cell lymphoma, germinal centre B phenotype, was made.



Discussion

Malignant neoplasm of the GI tract is common, and the most diffuse type is adenocarcinoma. Malignant lymphoma is rare and comprises 1–4% of the malignant GI neoplasms (Hawkes et al., 2012). The most common sites of extra nodal NHL are the GI tract, lung, liver, spleen, bone, and skin. In the GI tract, the most commonly involved organs are the stomach, small bowel, large bowel, and oesophagus, in decreasing order of frequency (García-Norzagaray et al., 2019; Bugeja & Micallef, 2019). PALs are extremely rare tumors. Secondary involvement of the gastrointestinal tract is more frequent than primary lymphoma, mainly affecting the stomach (75%), small bowel (9%), and ileocecal topography (7%) Bugeja & Micallef, 2019; Ayub et al., 2017). PALs present with symptoms of acute appendicitis, perforated appendix, palpable mass, or incidental imaging findings (Hawkes et al., 2012; McSwain & Beal, 1944; García-Norzagaray et al., 2019; Bugeja & Micallef, 2019; Ayub et al., 2017). Imaging findings are not specific. An enlarged appendix raises suspicion of malignancy, especially if the diameter is more than 1.5 cm and associated with appendiceal fat stranding (Hawkes et al., 2012; McSwain & Beal, 1944; García-Norzagaray et al., 2019; Ayub et al., 2017). Late

diagnosis may be associated with complications as gastrointestinal bleeding, intussusception, intestinal and ureteral obstruction (McSwain & Beal, 1944; García-Norzagaray et al., 2019). Our case had no different clinical presentation from classic appendicitis. While appendicular lymphoma may present as appendicitis, early-stage appendiceal lymphomas are commonly diagnosed incidentally during resection for suspected appendicitis (McSwain & Beal, 1944). Features of acute appendicitis-like pain in the right lower quadrant is the most common clinical presentation. Advanced-stage appendiceal lymphomas may penetrate the visceral peritoneum and invade adjacent organs, leading to serious complications (McSwain & Beal, 1944; Ibrahim, M. et al., 2022; Malaguarnera, M., 2011). PAL cases mimic acute appendicitis due to lumen obstruction and nonspecific gastrointestinal symptoms such as nausea, vomiting, and anorexia (Solis-Pazmino, P. et al., 2023; Ibrahim, M. et al., 2022).

Appendiceal neoplasms are more frequently observed in females during the sixth decade of life. A retrospective analysis of 116 patients with PAL by Ayub et al. revealed an average age at diagnosis of 48 years, average overall survival of 185 months, and a 5-year survival rate of 67% (Ayub et al., 2017). In the present case, the patient was a 14-year-old male child and succumbed to death on the seventh post-operative day. The most common diagnosis of primary GI lymphoma is non-Hodgkin's lymphoma, with Hodgkin's lymphoma being rare (Solis-Pazmino, P. et al., 2023). DLBCL is the most common subtype, with scattered incidences of marginal zone lymphoma, follicular lymphoma, mantle cell lymphoma, and Burkitt's lymphoma. Most cases occur between the age group of 65 to 69 years (Bugeja & Micallef, 2019; Ayub et al., 2017; Solis-Pazmino, P. et al., 2023; Ibrahim, M. et al., 2022; Malaguarnera, M., 2011). In unexplained GI problems that are not resolving with conventional treatment modalities, NHL should be considered as a differential diagnosis and should be ruled out (Solis-Pazmino, P. et al., 2023). The majority of studies have inferred that early surgery followed by adjuvant chemotherapy results in the best overall survival, while a few others have proposed that surgical excision or chemotherapy alone extends optimum results (Ibrahim, M. et al., 2022; Malaguarnera, M., 2011; Rossi, A., & Maloney Patel, N., 2023). Major prognosticators for survival in NHL are histological grade, depth of infiltration, stage, and radical tumour resectability. Low grade, minimal depth of invasion, early stage, and complete resectability of tumour are highly significant for better survival in cases of GI lymphoma (Sinha, B. et al., 2024). Close follow-up is essential and highly recommended.

Conclusion

Primary appendiceal lymphoma is an extremely rare entity. Clinical presentation is similar to acute appendicitis; therefore, high suspicion and proper investigation are required for diagnosis and management. Further studies and establishing guidelines are fundamental for the management of appendiceal lymphoma to avoid poor prognosis and complications.

Author contribution

Nayantrishna Nath: Contributed to Case and Manuscript writing and proofreading. Prashant Rajput: Contributed to Cases and Manuscript writing.

Acknowledgment

NIL.

Funding

NIL.

Conflict of interest

The author declares no conflict of interest. The manuscript has not been submitted for publication in other journal.

Informed consent

Approval taken from the institution and Informed consent taken.

AI tool declaration

The authors declare that no AI and associated tools are used for writing scientific content in the article.

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