

CASE REPORT

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Primary appendiceal lymphoma managed through videolaparoscopic appendectomy in an Amazonian center: Case report and review of literature

Thaís Caroline Sales Raposo, Gustavo Lopes de Castro, Maria Letícia da Silva Martins, Ana Elisa de Landa Moraes Teixeira Grossi, Jonathan William da Silva Rodrigues, Alice Brito Brandão

ABSTRACT

Introduction: Primary gastrointestinal lymphoma is rare and appendix involvement is rarer. Most cases of appendiceal lymphoma present as clinical acute appendicitis and are generally diagnosed intra-operatory. Some computed tomography findings in preoperative evaluation may suggest appendix neoplasm rather than appendicitis, such as appendix diameter larger than 15 mm, abdominal lymphadenopathy and aneurysmal dilatation of the lumen.

Case Report: We report a case of a 26-year-old Brazilian man first diagnosed with acute appendicitis and submitted to videolaparoscopic approach with appendectomies and posterior chemotherapy after histopathological reveal of primary appendiceal lymphoma.

Conclusion: Surgical resection followed by chemotherapy remains as standard management choice when possible. Adequate investigation and mandatory histopathological analysis in appendectomies are fundamental for diagnosis and proper treatment.

Keywords: Appendiceal lymphoma, Appendectomy, Appendix, Lymphoma

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INTRODUCTION

Primary gastrointestinal lymphoma is rare, accounting for 1–4% of all gastrointestinal neoplasms and about 0.5% of all colonic malignancies. Intestinal lymphoma presents two principal classifications in follicular B cell lymphomas and intestinal T cell lymphomas, with intestinal tract as the most common site of extranodal involvement for non-Hodgkin lymphomas [1–4].

Appendiceal lymphoma is extremely uncommon, representing about 0.015% of all gastrointestinal lymphomas. Secondary involvement of the gastrointestinal tract is more frequent than primary lymphoma, mainly affecting stomach (75%), small bowel (9%), and ileocecal topography (7%) [3, 5–7].

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 in

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intra-operative approach, supporting histopathological analysis as routine procedure for all appendectomies [3, 5].

We report a case of a 26-year-old Brazilian man diagnosed with acute appendicitis and submitted to videolaparoscopic approach with appendectomy.

CASE REPORT

A 26-year-old man was admitted in our emergency service with chief complaint of a three-day pain in right iliac region, with no history of irradiation, associated with anorexia and nausea. No vomiting, diarrhea, or fever were reported. He also commented previous chronic constipation, although physiologic functions were preserved. He had no significant past surgical history, comorbidities, or drug allergy.

At first examination, the patient was stable, afebrile, acyanotic, anicteric, hydrated, and vital signs in normal values. There were no respiratory or cardiac abnormal findings in physical examination. Abdomen was plan, flaccid, painful in right iliac region, localized peritoneal irritation with decompression pain, without any palpable mass.

Admissive laboratory tests showed absence of leukocytosis and inflammatory tests were normal. COVID-19 test was negative. He was submitted to an abdominal and pelvic computed tomography (CT) that revealed appendix with increased diameter of approximately 2.3 cm, hyperdense, no lumen visualized, discreet adjacent fat densification and no signs of obstruction. Additional findings included great quantity of stomach content (Figure 1).

Due to the strong suggestion of acute appendicitis and hemodynamic stability, the patient was submitted to videolaparoscopic approach under general anesthesia. Intra-operative findings included mass in middle third of appendix and peri-appendiceal fibrin (Figure 2). Appendectomy was performed without complications by two general surgeons.



Figure 1: Axial CT showing thickened appendix (red arrow) with approximately a 3 cm diameter and adjacent fat densification.

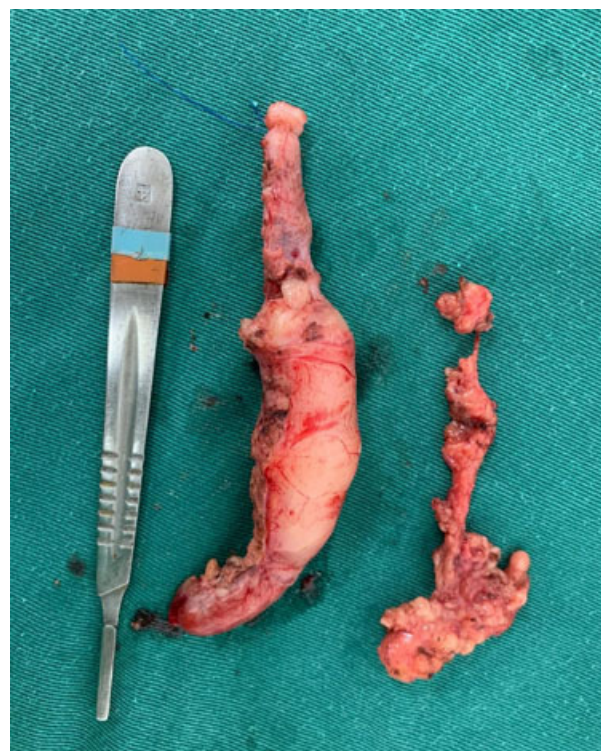


Figure 2: Appendix and mesoappendix as product of videolaparoscopic appendectomy.

Histopathological analysis was suggestive of primary appendiceal lymphoma. Immunohistochemistry was performed and revealed positivity to MUM1 antigen in 60%, CD20, BCL6, and elevated cell proliferation index (Ki-67) of 90%. BCL2 was negative. Diffuse large B-cell proliferation (CD20+) with nongerminal center phenotype (BCLE and MUM1 positives) and elevated cell proliferation index (Ki-67: 90%) confirmed diagnosis of diffuse large B cell lymphoma (DLBCL) classified as activated B-cell type (ABC) (Table 1).

Table 1: Immunohistochemistry analysis: diffuse large B-cell proliferation (CD20+) with nongerminal center phenotype (BCLE and MUM1 positives) and elevated cell proliferation index (Ki-67: 90%)

Antigen	Antibody (clone)	Result
AE1/AE3	AE1/AE3/PCK26	Negative
MUM1	MRQ-43	Positive (60%)
c-MYC	Y69	Negative
CYCLIN-D1	SP4-R	Negative
CD30	Ber-H2	Negative
CD20	L26	Positive
CD10	SP67	Negative
CD3	2GV6	Negative
BCL6	G1191E/AB	Positive
BCL2	124	Negative
Ki67	30-9	Positive (90%)

In postoperative recovery, the patient evolved without any complications. He was discharged three days after surgery. After immunohistochemistry result and further CT investigation, the patient was submitted to pelvic lymphadenectomy. He was also referred to oncologic evaluation and chemotherapy with CHOP scheme was initiated (cyclophosphamide, doxorubicin, vincristine, and prednisone). He had been submitted to four sessions until the present moment and further investigation showed no evidence of other tumor sites.

DISCUSSION

Primary appendiceal lymphoma presenting as acute appendicitis is extremely rare and it is more prevalent in white male around fifth decade of life. Non-Hodgkin lymphomas are the most common type of lymphoma, originating from lymphocytes proliferation, with DLBCL as main finding in anatomopathological result [3, 7].

Diffuse large B cell lymphoma is classified into two major distinct molecular subtypes of germinal center B-cell (GCB) and activated B-cell (ABC) according to cell-of-origin classification (COO) [8, 9]. Activated B-cell–Diffuse large B cell lymphomas are currently thought to be derived from B cells that have passage through germinal center and are committed to plasmablastic differentiation [9]. A recent study stands a new subclassification for DLBCL in five subtypes: ABC type, poor risk GCB with BCL2, poor risk GCB with alterations in PTEN gene and epigenetic enzymes, good-risk GCB with alterations in BCR/PI3K, JAK/STAT, and BRAF markers and cell-of-origin independent group of tumors [9]. Primary gastrointestinal DLBCLs are classified according to the germinal or non-germinal center B-cells [10].

Clinical presentation may present abdominal pain, constipation, diarrhea, or weight loss in symptomatic patients; however, asymptomatic cases are not unusual and diagnosis tends to be incidental. Some cases mimic acute appendicitis due to lumen obstruction and nonspecific gastrointestinal symptoms such as nausea, vomiting, and anorexia [6, 7, 11]. Our case did not differ from clinical presentation reported in the literature.

Diagnosis is generally intra-operative, nevertheless some CT findings may indicate appendiceal neoplasm, including appendix diameter larger than 15 mm, abdominal lymphadenopathy and aneurysmal dilatation of the lumen. Late diagnosis may be associated with complications as gastrointestinal bleeding, intussusception, intestinal and ureteral obstruction. Diffuse large B cell lymphoma is confirmed through postoperative histopathologic study, which makes this analysis mandatory in every appendectomy [3, 12].

Surgical treatment is controversial in the available literature due to its rarity and the difficulty in establishing guidelines for this pathology. Surgical resection followed by postoperative chemotherapy with CHOP scheme has demonstrated high efficacy in intestinal diffuse large

B cell lymphoma (DLBCL) and remains as standard management when possible. Isolated chemotherapy and radiotherapy have also been considered as a second treatment option [5, 12, 13]. Our patient was managed through videolaparoscopic appendectomy and posterior pelvic lymphadenectomy, referred to our oncologic service and treated with four sessions of CHOP chemotherapy so far.

Prognosis in DLBCL and all cases of lymphomas is based on the stage of the disease. Patients in early-stage condition have a better prognosis than those with advanced disease. Close follow-up is essential and highly recommended [7, 12].

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. Further studies and establishing guidelines are fundamental for management of appendiceal lymphoma to avoid poor prognosis and complications. Preoperative diagnosis is difficult and attention to CT findings may suggest neoplasm rather than appendicitis.

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Author Contributions

Thaís Caroline Sales Raposo – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Gustavo Lopes de Castro – Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Maria Letícia da Silva Martins – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Ana Elisa de Landa Moraes Teixeira Grossi – Conception of the work, Acquisition of data, Analysis of data,

Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Jonathan William da Silva Rodrigues – Conception of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Alice Brito Brandão – Conception of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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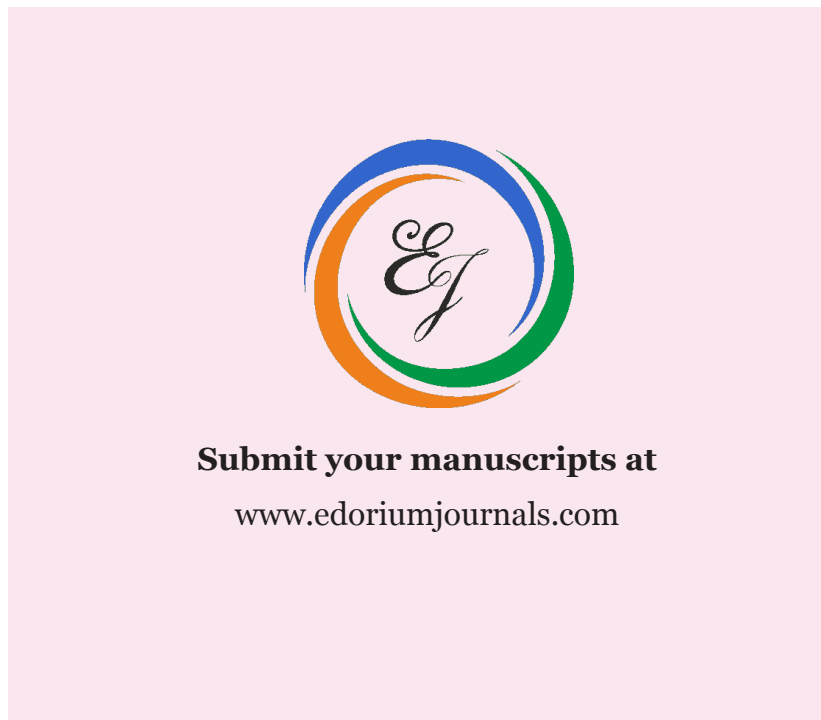
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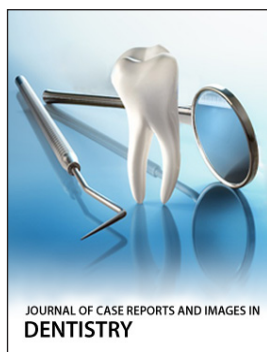
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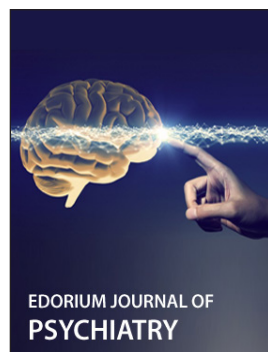
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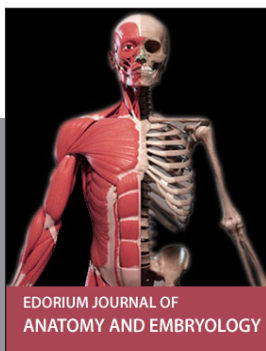
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