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Primary lymphoma of appendix presenting as acute appendicitis: A case report

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ABSTRACT

INTRODUCTION: Primary lymphomas of appendix are extremely rare tumors. The incidence is 0.015% of all gastrointestinal lymphomas.

PRESENTATION OF CASE: We present a case of a 75 year-old male patient who presented with acute abdominal pain in the lower right quadrant and fever.

DISCUSSION: The patient received laparotomic appendectomy. The definitive histopathological examination revealed the presence of diffuse large cell B-lymphoma of the appendix. The neoplasms of appendix usually manifest clinically with sign and symptoms of acute appendicitis from luminal obstruction (30–50%). Preoperative diagnosis is difficult and often occurs through histopathological examination.

CONCLUSION: Primary appendiceal lymphoma is rare and there are no clear guidelines for therapy. Primary surgical resection followed by post-operative chemotherapy showed high efficacy. The histopathological examination of all appendectomy is essential.

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1. Introduction

Primitive gastrointestinal lymphomas are rare and represent only 1–4% of all tumors of the gastrointestinal tract [1]. Gastrointestinal tract is the most common location for extranodal lymphomas [2]. The most affected organs are the stomach and the small intestine [3]. The primary lymphoma of the appendix is extremely rare. We present a case of primary diffuse large B-cell lymphoma of the appendix manifest clinically with sign and symptoms of acute appendicitis. This case is reported in line with the SCARE criteria [4].

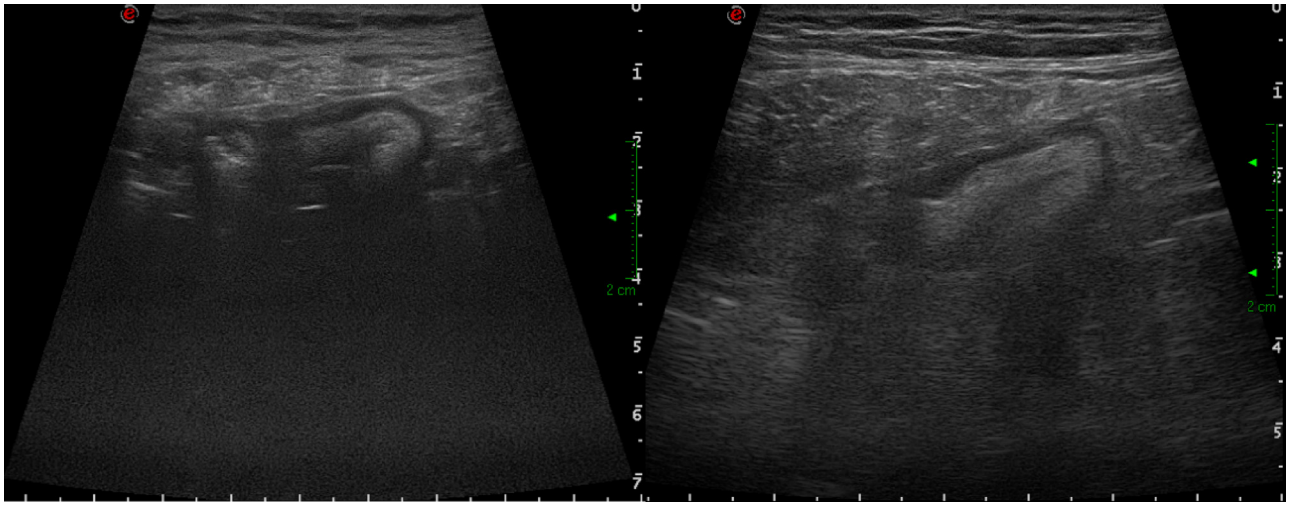
2. Case report

A 75-year-old man, without major medical history, went to our emergency room for abdominal pain in the lower right quadrant and fever (axillary temperature 38 °C) for about 1 day. Physical

examination was significant only for lower abdominal tenderness with more on the right iliac fossa without clear signs of peritoneal irritation. The peristalsis was poor. Laboratory examination revealed white cell count of 15610/mm³ with 12,720 neutrophils/mm³ and elevated inflammation index (C-reactive Protein 12.65 mg/dl). The ultrasound revealed the presence of a small bowel loop of the iliac or cecal region with thickened walls, not peristaltic, with surrounding free liquid and an important hyperemia of the adjacent mesenteric adipose tissue. No free liquid in the Douglas (Figs. 1 and 2). The radiograph of the abdomen revealed isolated air-fluid levels without pathological significance (Fig. 3). Pain was resistant to analgic drugs. The patient then received laparotomic appendectomy under clinical diagnosis of acute appendicitis. A laparoscopic approach was interrupted due to excessive intestinal distension. The appendix appeared enlarged, folded, phlogosated with gangrene at the tip. The postoperative period was regular. Antibiotic therapy (piperacillin-tazobactam), intravenous fluid, analgesics (paracetamol), anti-emetics and antithrombotics were administered. The discharge was on the 5th postoperative day. The definitive histopathological examination revealed, in the context of perforated appendicitis with acute inflammation, presence in the wall of habitus blastic lymphoid elements (Fig. 4), also in small aggregates in vascular-like spaces (Fig. 5), with an unusual immunophenotype due to incomplete expression of CD20.

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Figs. 1 and 2. Ultrasound: small bowel loop of the iliac or cecal region with thickened walls, not peristaltic, with surrounding free liquid and an important hyperemia of the adjacent mesenteric adipose tissue.



Fig. 3. Radiograph of the abdomen revealed isolated air-fluid levels without pathological significance.

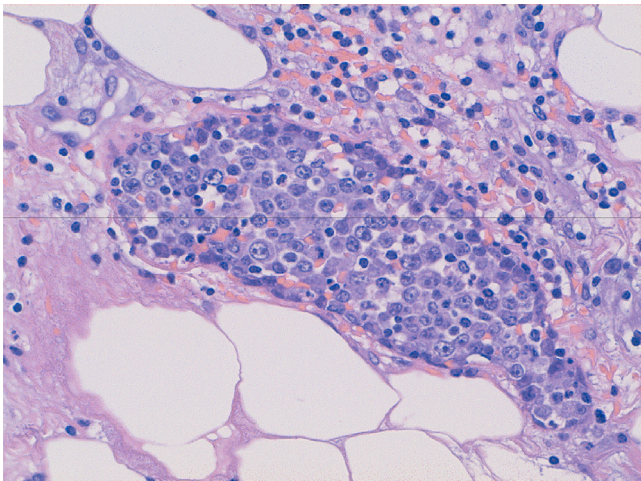


Fig. 4. EE Large lymphoid elements with a blastic habitus.

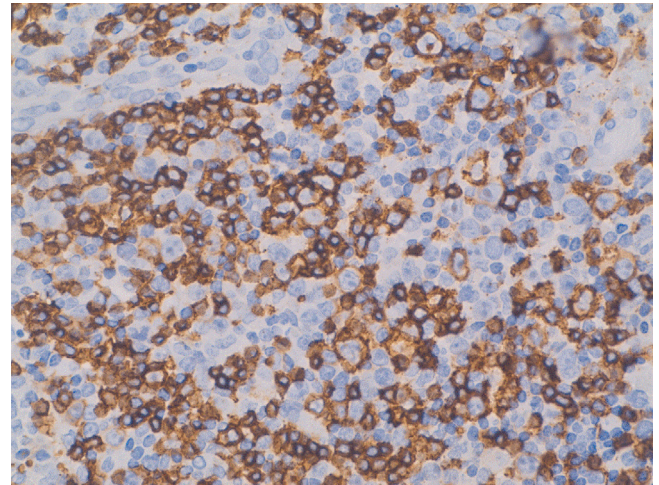


Fig. 5. CD20+ in Large lymphoid elements.

Immunophenotypic findings: CD45+, CD 20+ (in some elements), Pax 5+ (in some elements), MUM1+, OCT2-, BOB1+ (in some elements), CD3+ (in rare elements), CD5+ (in rare elements), CD 10-, Bcl2-, Bcl6-, CD 138-, CD68-, CD30+, (in some elements), S100-, MPO-, ALK1-, light chains K and lambda not strains, CKAE1AE3-, EMA-, Chromogranin-, Ki67 (Mib1)+ in most of the elements (80%). Monoclonal rearrangement was not found (Study method is performed on the DNA extracted from the sample): lymphocyte B polyclonal pattern and lymphocyte T oligoclonal pattern. PET-CT did not demonstrate metabolically active sites in the investigated body segments. The patient was assigned to hematology for the continuation of the diagnostic and therapeutic process.

3. Discussion

Malignant neoplasm of the gastrointestinal tract are common and the most diffuse type is adenocarcinoma. Malignant lymphoma is rare and comprises 1–4% of the malignant gastrointestinal neoplasms [1]. The lymphomas can be divided in Hodgkin and non-Hodgkin, the last type being classified in B cell or T cell lymphomas [2]. The lymphomas of the gastrointestinal tract mainly affect the stomach followed by the small intestine, pharynx, colon and esophagus. Men are more affected and the median age at diagnosis for

lymphomas non-Hodgkin of gastrointestinal tract was 55 years [3]. Primary lymphomas of appendix are extremely rare tumors. The incidence of primary lymphoma of appendix is 0.015% of all gastrointestinal lymphomas [5].

The clinical onset of the disease is often constituted by a surgical complication that requires urgent intervention [6]. The neoplasms of appendix usually manifest clinically with sign and symptoms of acute appendicitis from luminal obstruction (30–50%). Other important clinical manifestations may be an asymptomatic palpable mass, incidental imaging findings, intussusception, gastrointestinal bleeding and ureteral obstruction or hematuria and increasing abdominal girth from rupture of a malignant mucocele, resulting in pseudomyxoma peritonei [7].

The surgical approach and obviate additional surgery may change with detection of these neoplasms at preoperative imaging [7]. In the past, preoperative diagnosis of appendiceal neoplasms was rare but is becoming more common with the increased use of computed tomography (CT). A peculiar diagnostic element for lymphoma is diffuse enlargement of the appendix from lymphomatous infiltration on both ultrasound and CT. Diffuse appendiceal enlargement (diameter of 6–7 mm) is also well established as the diagnostic threshold for acute appendicitis, especially if it is associated with stranding of the periappendiceal fat. The inflamed appendix without neoplasm will usually not exceed 15 mm in diameter on CT, enlargement beyond this size should be viewed with suspicion [3]. Although the CT finding of enlargement of the appendix seems to be fairly characteristic of non-Hodgkin's lymphoma, such a finding is not pathognomonic. Neuroendocrine tumors can show an infiltrative pattern of growth. Specificity for lymphoma will be increased in the setting of abdominal lymphadenopathy or aneurysmal dilatation of the appendiceal lumen [3]. One apparent error in the ultrasound or tomographic diagnosis of appendiceal lymphoma is the erroneous interpretation of the grossly abnormal appendix as a thickened small-bowel loop. In our case, the ultrasound interpreted the enlarged appendix as a small bowel loop thickened with free fluid around and hyperemia of periappendiceal fat. PET/CT is important for staging lymphoma. PET/CT has greater sensitivity, compared to CT alone, in identifying lymph node, extranodal sites and bone marrow involvement by lymphoma. PET/CT can indicate the metabolic response to therapy, earlier detection of disease recurrence and also the overall level of metabolic activity of lymphoma, which correlates with level of aggressiveness and represent a prognostic predictor [8]. In our case, PET/CT, after appendectomy, did not identify areas of increased metabolic activity which did not require further surgical measures.

Primary appendiceal lymphoma is rare and there are no clear guidelines for therapy. In the literature primary surgical resection followed by post-operative CHOP (cyclophosphamide, hydroxydoxorubicin, vincristine, prednisolone) chemotherapy showed high efficacy in patients with localized intestinal diffuse large B cell lymphoma [9]. There was no evidence of recurrence 6 months after operation [10]. However, close follow-up is certainly recommended.

4. Summary

Lymphoma of the appendix is rare. Usually manifest clinically with sign and symptoms of acute appendicitis. The detection of this neoplasm at preoperative imaging is very difficult although a characteristic CT scan appearance may lead to preoperative diagnosis. PET/CT is important for staging lymphoma. The histopathological examination of all appendectomy is essential and should be mandatory. In the literature, the combination of surgery and chemotherapy is the best treatment for appendiceal lymphomas.

Conflicts of interest

There is no conflict of interest to declare.

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Ethical approval

The paper is not a research study. For case report our Institute exempted to take ethical approval.

Consent

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

Author contribution

Giuseppe Caristo idealizes study, design, data analysis, critical review and writes the article.

Guido Griseri idealizes study, data analysis and critical review of important intellectual content.

Rosario Fornaro reviews it critically for important intellectual contents.

Antonio Langone reviews it critically for important intellectual contents and participates in data acquisition.

Angelo Franceschi reviews it critically for important intellectual contents.

Veronica Errigo reviews critically the anatomical pathological evaluation.

Cecilia Ferrari participates in data acquisition.

Marco Casaccia reviews it critically for important intellectual contents.

Marco Frascio reviews it critically for important intellectual contents.

Angelo Schirru idealizes study and reviews it critically for important intellectual contents.

Registration of research studies

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Guarantor

Dr Giuseppe Caristo.

Dr Guido Griseri.

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