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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.

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 antalgic 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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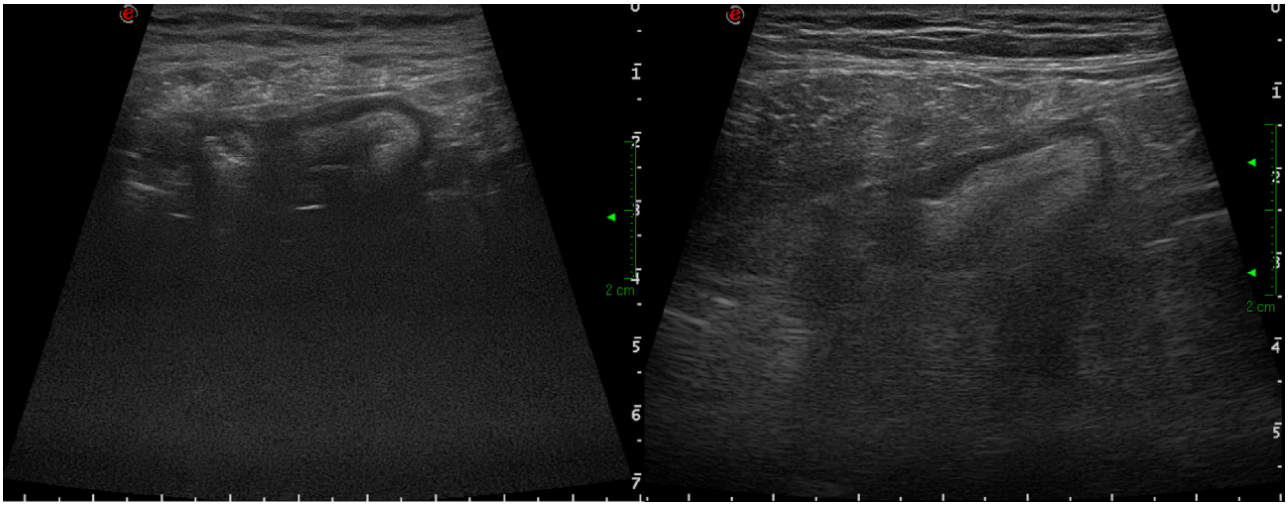

Figs. 1 and 2. Ultrasound: small bowel loop of the iliac or cecal region with thickened walls, not peristaltic, with surrounding free liquid and an important hyperemia of the adjacent mesenteric adipose tissue.

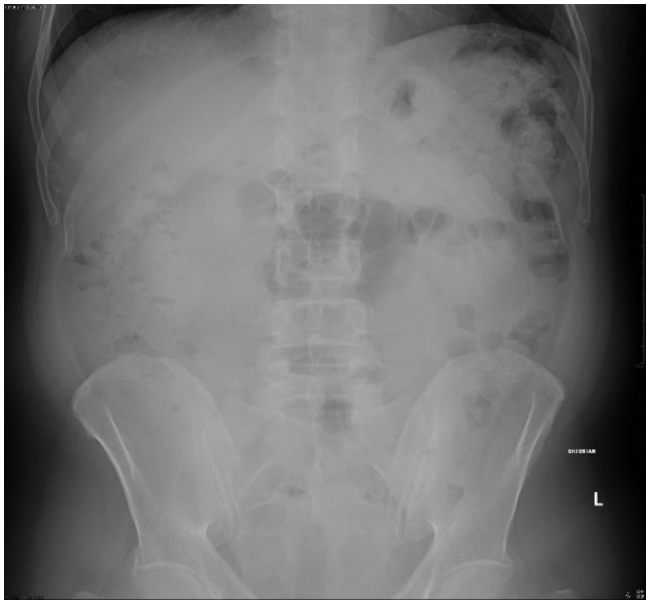

Fig. 3. Radiograph of the abdomen revealed isolated air-fluid levels without pathological significance.

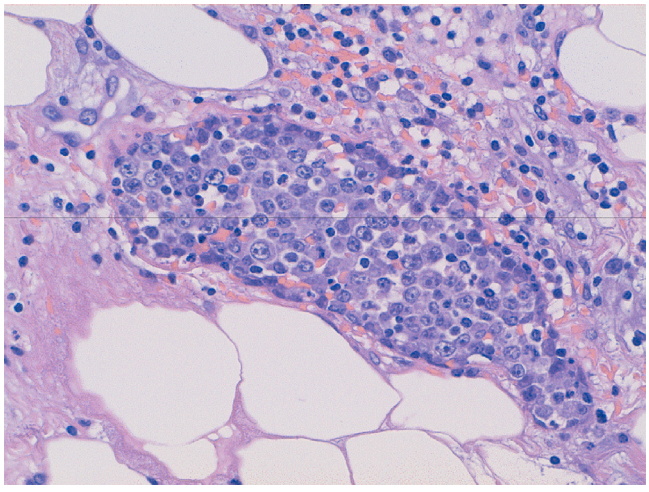

Fig. 4. EE Large lymphoid elements with a blastic habitus.

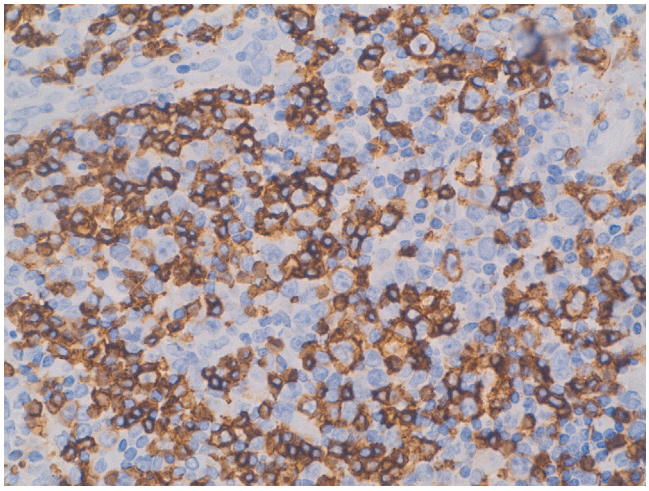

Fig. 5. CD20+ in Large lymphoid elements.

Immunophenotipic 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