Ileocecal burkitt's lymphoma presenting as acute appendicitis: A case report

Jean-Pierre Gonçalves, Arnaldo Cerqueira, Henedina Antunes, Íris Maia, Susana Carvalho

ABSTRACT

Introduction: Burkitt's lymphoma is a highly aggressive non-Hodgkin lymphoma and occurs predominantly in the first decades of life in males. Often the clinical presentation is an abdominal mass associated with abdominal pain, nausea, intestinal obstruction or intussusceptions. The association between Burkitt's lymphoma and histological proven acute appendicitis is rare, and usually is secondary to intussusceptions. Case Report: We present a case of a 14-year-old boy with an ileocecal Burkitt's lymphoma presented as acute appendicitis. Conclusion: This report emphasis the importance of the histopathological exam of the appendix in all patients with acute appendicitis suspicious, even those with typical clinical presentation.

Keywords: Burkitt's lymphoma, Acute appendicitis


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INTRODUCTION

Lymphomas are a group of malignant tumors involving cells of the lymphoreticular or immune system [1]. Burkitt's lymphoma (BL) accounts for 30% of non-endemic pediatric lymphoma and is a highly aggressive non-Hodgkin lymphoma. It is also the faster growing human tumor with a doubling time of the less than 24 hours [2] and has two major forms, the endemic (African) and non-endemic (sporadic) form.

Burkitt's lymphoma occurs predominantly in the first decades of life, in males and might present primarily as an abdominal mass associated with abdominal pain, nausea, intestinal obstruction or intussusceptions [3]. The association between BL and histological proven acute appendicitis (AA) is rare [4] and usually is secondary to intussusceptions [3]. We present a rare case of ileocecal BL presenting as AA.

CASE REPORT

A 14-year-old Caucasian boy presented to our Emergency Unit with a six-day history of periumbilical pain associated with fever. He had anorexia but nausea or vomiting were not reported. There was no diarrhea, melena or hematemesis. Past medical, surgical and drug histories were unremarkable.

Physical examination revealed tenderness over the right iliac fossa, but there were no palpable masses or abdominal distension. Blood analysis showed an increased white blood cell counts (11500 cells/μL),
neutrophilia (9000 cells/μL) and raised C-reactive protein (172 mg/L). Abdominal ultrasound scan (USS) results have supported the clinical AA diagnosis and showed an enlarged appendix and a periappendiceal abscess formation (3x4 cm).

A diagnosis of AA was made and the patient was prepared for an open appendectomy. The findings at surgery were macroscopically suggestive of AA with periappendicular abscess and no adjacent lymph nodes. His postoperative course was uneventful and he was discharged home with antibiotics and no gastrointestinal symptoms.

Fourteen days later, the histopathological examinations showed appendicitis and high grade B cell non-Hodgkin lymphoma consistent with BL in the thick-walled appendix (Figure 1).

The patient was referred to a specialist pediatric oncology unit where abdominal computerized tomography (CT) scan (Figure 2), positron emission tomography (PET) scan (Figure 3) and immunohistochemical studies were performed. These exams confirmed the histological diagnosis of abdominal BL. Immunohistochemical study showed the malignant cell population was positive for CD20, CD10 and BCL6 and negative for CD3 and MUM1. Chromosomal translocation 8q24 (myc gene) was reported.

Polychemotherapy according to the B-non-Hodgkin’s lymphoma chemotherapy protocols (group B LMB protocol 2001) were administered [5]. Follow-up abdominal CT at four months, six months and one year was performed, and the patient is disease free and shows no signs of recurrence or metastasis.

DISCUSSION

Appendiceal lymphomas are exceedingly rare and often diagnosed postoperatively. In a review of 29 patients with appendiceal lymphoma, Burkitt’s lymphoma was diagnosed in 9 cases [6]. The clinical findings are nonspecific, leading to a delay in diagnosis. Primary appendiceal lymphoma may present clinically as acute appendicitis [7,8]. Besides this association, the precise mechanism for the AA occurrence in patients with appendiceal lymphoma is not known, but obstruction could play a role in the pathogenesis of the AA [6].

BL is a rare and rapidly progressive tumor and commonly presents as an abdominal mass and long duration of symptoms and weight loss were two important clinical clues to the presence of gastrointestinal lymphoma. These symptoms were not present in our patient.

AA is the initial preoperative diagnosis of similar clinical presentation of periumbilical pain, low grade fever and anorexia. The clinical presentation and operative findings were highly suggestive of AA with periappendicular abscess. The preliminary macroscopic pathological examination of terminal ileum revealed only a thickened mucosa. No periappendicular mass was apparent. The diagnosis of AA and LB were made in the
A histopathological exam of biopsy fragment. There are already other cases of BL described presenting as suspicious AA, but usually it is secondary to ileoceleal intussusceptions [9].

In this case, examination of the specimen never revealed discernible tumor. The entire appendix was submitted for microscopic examination. The fact that this lymphoma was discovered only in the histopathological exam raises questions about the importance of complete examination and acquisition of adequate appendix biopsies in children more than five years of age [10].

This tumor responds well to chemotherapy and the role of surgery remains controversial [11, 12]. In this case the tumor regressed completely after the chemotherapy and there are no signs of recurrence at 1 year of follow-up.

CONCLUSION

AA is mostly diagnosed preoperatively but the histology is the gold standard exam for the diagnosis. The histopathological exam of the appendix is vital and should be mandatory for all patients with AA suspicious, even those with typical clinical presentation.

In this patient with histologically proved acute appendicitis the unexpected diagnosis of LB was made by the histology. This diagnosis was early and allowed a prompt treatment of the tumor which might have been life saving for this boy.

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Author Contributions

Jean-Pierre Gonçalves – Conception and design, Acquisition of data, interpretation of data, Drafting the article, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES


