



## Multidisciplinary expertise in the diagnosis of cecal lymphoma

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### ARTICLE INFO

#### Keywords:

Intestinal lymphoma  
Pediatric oncology  
Pediatric colonoscopy

### ABSTRACT

This particular case highlights the importance of a multidisciplinary team expertise in the differential diagnostic process of acute abdominal pain. The case involves a 10-year old Chinese boy who presented with intermittent abdominal pain; the clinical and radiologic picture narrowed the differential diagnosis between an acute appendicitis and an intestinal lymphoma. Diagnosis of a high grade - B lymphoma was made by performing a colonoscopy; this procedure was deemed the best option to obtain a quick diagnosis with low invasiveness.

### 1. Introduction

Acute abdominal pain is one of the main reasons of access to the emergency department in pediatric as well as adult patients. Although acute appendicitis is one of the most frequent causes of pain localized in the right iliac fossa, differential diagnosis must comprehend, besides common diseases like gastroenteritis, other rarer causes such as intussusception, which however is commonly idiopathic in younger patients, or abdominal masses, which may occur at any age. The following case highlights the importance of the multidisciplinary pediatric team expertise in the differential diagnostic process of abdominal pain. Furthermore, it shows the usefulness and safety of colonoscopy in diagnosing pediatric intestinal lymphomas.

### 2. Case report

A 10-year old Chinese boy was referred to our Center for a suspected acute appendicitis. His clinical history consisted of intermittent abdominal pain localized in the right iliac fossa which had started two days before our evaluation, associated with vomit but no fever. The

routine blood tests performed upon admittance were normal, except for a mild hyperphosphatemia (1.94 mmol/L). Upon the physical exam there was mild tenderness localized in the right iliac fossa, but no signs of peritonitis nor a palpable mass indicating an abscess. Given the un-specific clinical picture, it was decided to perform an ultrasound that demonstrated the presence of an appendix with moderately thick walls (8 mm), associated with hyperechogenic pericecal adipose tissue, pericecal lymph nodes enlargement and free fluid. The exam also detected a short ileocolic intussusception localized inside the cecum, surrounded by dishomogeneous, hypoechoic and hypervascularized tissue, protruding into the lumen. After discussion with the pediatric radiologists, it was decided to perform a CT scan to better define the intussusception and the presence of dishomogeneous tissue inside the cecum, which could be a sign of another collateral disease. The exam confirmed the presence of multiple enlarged lymph nodes in the mesentery, associated with free fluid in the right iliac fossa, and indeed demonstrated a solid, contrast-enhanced lesion of 41 × 23 mm, localized at the level of the ileocecal valve, extending into the last ileal loop. The appendix was distended with no signs of inflammation (Fig. 1).

Given the high suspicion of an intestinal lymphoma, we needed to

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<https://doi.org/10.1016/j.epsc.2020.101489>

Received 29 April 2020; Received in revised form 9 May 2020; Accepted 11 May 2020

Available online 16 May 2020

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decide which option could be the easiest to obtain a quick diagnosis and solve the symptoms at the same time. A laparoscopic exploration could have allowed us to remove the appendix and a few lymph nodes of the mesentery, but it could have not been sufficient for the diagnosis. The alternative would have been the resection of the cecum, together with the ileum and ileocecal valve. After discussion with the Pediatric Oncologists and the GastroEnterologists, we opted for a colonoscopy, considering the presence of mild symptoms and the non-necessity to remove the appendix. The colonoscopy confirmed the presence of an occluding mass of 4 cm of diameter, localized in the cecum and involving the ileocecal valve (Fig. 2). The terminal ileum had a hyperemic, hypertrophic mucosa that determined a mild stenosis of two-three centimeters of length; above this lesion, the intestinal mucosa was characterized by a granular appearance in an area of about 15 mm in length.

The frozen biopsies were suggestive for a high-grade B lymphoma. After positioning of a central venous catheter, the patient was transferred to the Pediatric Oncology Unit, and chemotherapy was promptly started according to the AIEOP LNH 97 protocol. The definitive histologic exam confirmed Burkitt Lymphoma associated with nodular and diffuse lymphocytic infiltrate in the intestinal mucosa, consisting of large blasts in an active stage of mitosis, with dispersed chromatin, evident nucleoli and eosinophilic cytoplasm. The blasts were CD20<sup>+</sup>, Bcl6<sup>+</sup>, MUM1<sup>+</sup>, Bcl2<sup>+</sup>, c-myc<sup>+</sup>, CD 21<sup>+</sup>, CD23<sup>+</sup> and had a mitotic index of >95%. The FISH analysis detected a rearrangement of the IRF4 locus and no mutations of MYC. The patient underwent six cycles of chemotherapy for a total of three months. A CT scan was repeated four months after the diagnosis; the exam showed a slight decrease of the size of the cecal lesion (35 × 18 vs 41 × 23 mm), while the abdominal lymphadenopathies and the free abdominal fluid were no longer detectable. The patient was then monitored through monthly US: the last exam, performed eleven months after the initial diagnosis, showed no thickening of the intestinal mucosa nor abdominal lymphadenopathies. The patient is currently in the follow-up program that will last five years.

### 3. Discussion

Normally, lymphoproliferative lesions of the abdomen manifest in children as huge masses, often encasing the bowel. Sometimes, they are characterized by an ileal intussusception which almost always necessitates a bowel resection with all the risks related to possible seeding.

The incidence of intussusception associated to a specific intestinal

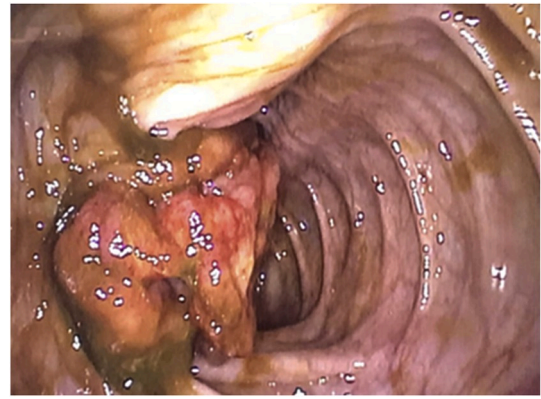


Fig. 2. The cecal mass found at colonoscopy (colored).

cause is reported between 1.5% and 20% [1], with a higher incidence in males [2]. Intestinal lymphomas represent approximately 6.5% of cases of secondary intussusception, the peak age of incidence being between 5 and 15 years, with a male to female ratio of 2:1. Burkitt and MALT lymphomas are the most common subtypes found, representing 42.5% of all intestinal lymphomas [3]. The presentation in our case was quite atypical, being localized in the cecum and associated with a very short intussusception, which did not seem the cause of the pain. In literature, similar features have been reported in only 17.5% of patients with abdominal Burkitt lymphoma [4], a huge abdominal mass being the first clinical presentation in the majority of cases.

When evaluating a patient with abdominal pain, ultrasound is the first diagnostic tool. Besides the classical signs of appendicitis or other non-surgical conditions, it may also identify a secondary intussusception by detecting a hypoechoic, hypervascularized mass with loss of the intestinal wall stratification [5]. These features are not always clear, and in these cases, a CT scan may be considered in the diagnostic process: the imaging normally shows segments of dilated intestine with a nodular contour and bowel wall thickening (the so-called aneurysmal dilatation sign) or a polypoid mass projecting into the lumen [6,7].

When dealing with a patient with an abdominal lymphoma, a prompt diagnosis is mandatory to start treatment as soon as possible, considering the high doubling time of these tumors. While in most cases with huge masses a biopsy is the only surgical procedure required [8], when an intussusception is present a more aggressive approach is needed, which normally includes the complete resection of the bowel involved. During these manoeuvres, surgeons do not see the tumor inside nor are allowed to explore the intestine, to avoid spilling of malignant cells, and they only need to perform the complete resection of the bowel segment followed by an end-to-end anastomosis. In our patient, even if the intussusception was involving a short portion of the ileocecal intestine, the proper procedure for diagnosis would have required the resection of the terminal ileum, the cecum and appendix and ileocecal valve. On the other hand, approaching just the appendix through a laparoscopy would have allowed us to remove it together with some lymph nodes, without any information about the intracecal lesion and uncertain histologic information on lymph nodes. Therefore, since the intussusception was not symptomatic and the CT scan had excluded an inflamed appendix, we considered to perform the colonoscopy. It seemed to be the best and less invasive option to ensure a correct diagnosis by sampling the cecal lesion directly. The procedure was performed 24 hours after the admission of the patient, due to the availability of an expert gastroenterologist-endoscopist in our Center. If we consider the adult population, colonoscopy is routinely used for colorectal malignancies as an efficacious diagnostic tool; a retrospective study on adult patients with intestinal follicular lymphoma demonstrated that the accuracy of the endoscopic evaluation of ileal lesions was 68.8% [9]. Few cases of diagnosis of pediatric Burkitt lymphoma through colonoscopy are



Fig. 1. CT scan showing a hyperdense cecal lesion (indicated by the black arrow).

reported in literature [10], but we need to consider that this kind of procedure is possible only if the tumor is localized in the cecum or terminal ileum. Of course, the availability of an expert endoscopist is also needed.

#### 4. Conclusion

This case highlights various aspects in term of diagnosis and further treatment of a child with intestinal lymphoma: a careful physical examination, the choice of the more appropriate diagnostic procedures and the importance of an experienced multidisciplinary team in the assessment of the disease are all fundamental for the success. Further studies would be useful to better evaluate the role of diagnostic colonoscopy in the pediatric oncologic population.

#### Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

#### Funding

No funding or grant support.

#### Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

#### Declaration of competing interest

The authors declare that they have no known competing financial

interests or personal relationships that could have appeared to influence the work reported in this paper.

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