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Appendicitis is like a box of chocolates

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ABSTRACT

Appendicitis is the most commonly encountered abdominal emergency in pediatric surgery. However, the presentation of acute appendicitis is as diverse as the patient population. We present here a case of appendicitis presenting as an intra-abdominal mass consistent with lymphoma. Our patient is a 3 year old male with an atypical presentation of acute appendicitis. Additionally, he was found to have malrotation at the time of surgery. A delayed presentation, coupled with anomalous laboratory findings and aberrant anatomy made for a difficult diagnosis and overall interesting case of appendicitis.

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Appendicitis is the most commonly encountered emergency in pediatric surgery [1]. However, the presentation of appendicitis varies greatly depending on the age and duration of disease. In a recent survey of 727 pediatrics practitioners, appendicitis was listed as one of the most commonly missed diagnoses [2]. Imaging modalities including ultrasonography and computed axial tomography (CT) have improved diagnostic accuracy [3–5]. However, despite these advances there is a defined negative appendectomy rate and error in diagnosis [6]. Here we present a case of perforated appendicitis in a patient with malrotation whose preoperative diagnosis was intra-abdominal lymphoma. We review the case and discuss other variable presentations of appendicitis.

1. Case report

A 3 year old otherwise healthy boy presented to an outside hospital with 3 days of abdominal pain and constipation. The pain was peri-umbilical, non-radiating and did not have alleviating or aggravating factors. An abdominal radiograph was negative for pneumoperitoneum, revealing only increased stool burden. An

abdominal ultrasound at the outside facility visualized a tubular soft tissue structure in the peri-umbilical region and was concerning for bowel wall thickening. The patient was discharged home with a bowel regimen. He returned to his pediatrician who repeated an abdominal X-ray which revealed an abnormal bowel pattern and he was transferred to our institution for further evaluation.

There was no report of recent illness, fever, night sweats, anorexia or weight loss. On initial exam he was fussy, but consolable and had a benign abdominal exam with palpable inguinal lymph nodes. His white blood cell count was normal at 9.18, but lactate dehydrogenase was elevated to 1318. A repeat ultrasound of the abdomen revealed a 5.1 × 2.3 cm thickened tubular structure in the left abdomen, indicating thickened bowel, with a differential diagnosis including intussusception with a pathologic lead point or an infiltrative, malignant process.

Based on these findings, a CT of the abdomen and pelvis with contrast (Fig. 1) was performed. It revealed a mass measuring approximately 5.6 × 2.4 × 3.0 cm adjacent to the transverse colon in the left upper abdomen and crossing the midline as well as multiple enlarged mesenteric lymph nodes. The majority of the small bowel was located in the right abdomen and the ascending colon and cecum were non-rotated. The appendix was not visualized and the transverse colon was thickened in the region of an adjacent mass. The primary differential diagnosis was intra-abdominal lymphoma. Discussion with our radiologist suggested an intra-abdominal process consistent with lymphoma (Fig. 2).

As the inguinal lymph nodes were quite small, laparoscopy was performed to obtain a diagnostic biopsy. This revealed a mass in proximity to the transverse colon displacing the surrounding tissue

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Fig. 1. Representative images: abdominal CT. The upper image shows the large left sided mass measuring $5.6 \times 2.4 \times 3.0$ cm with associated inflammatory changes. The lower image highlights a large mesenteric lymph node.

and a biopsy could not be performed safely. Consequently, we converted to an open procedure. On laparotomy, the mass seemed to involve the transverse colon, cecum and terminal ileum. Malrotation was present. A biopsy sent for frozen section pathology was negative for malignancy. The cecum and ileum were then successfully dissected away from the mass and upon doing so, the appendix was noted to be thickened and enlarged. An appendectomy was performed and the dissection of the mass continued. Upon separation from the transverse colon, it appeared that the mass was densely adherent and inflamed omentum. It was excised and a Ladd's procedure performed.

Pathology revealed acute and sub-acute appendicitis with peri-appendicitis and peri-appendiceal fibrosis. The mass was indeed omentum with inflammatory changes. The patient was discharged on post-operative day 1 in good condition.

2. Discussion

Abdominal pain in children is a common indication for a visit to the emergency room [7], with appendicitis being the culprit only 32% of the time [8]. The diagnosis of appendicitis however, can be quite challenging, the list of differentials extensive and changing with age and gender [9,10]. Maladies of the gastrointestinal tract, reproductive system, urinary tract and even the pulmonary system can all produce abdominal pain similar to appendicitis.

In surgical training we are taught to look for certain classic signs and symptoms, commonly represented in the Alvarado Score or



Fig. 2. Representative images: abdominal CT. Other significant findings included non-rotated intestines with ileocecal valve in the upper left quadrant, upper image and the superior mesenteric artery oriented to the right of the superior mesenteric vein (arrow) in the lower image.

Pediatric Appendicitis Score [11,12]. Unfortunately the classic story of peri-umbilical abdominal pain migrating to the right lower quadrant with associated fever, nausea, vomiting and anorexia is seen in less than 50% of pediatric patients [13]. Diagnosis is further complicated in younger patients (<3 years of age) who tend to present more commonly with perforation (80–100% perforation rate) versus older patient (10–17 years of age) who are not usually perforated at initial presentation (20% perforation rate) [14].

Imaging modalities have reduced the rate of missed diagnosis; however a negative appendectomy rate persists. A recent study of forty children's hospitals in the United States found that the negative appendectomy rate varies significantly between imaging modalities, age and gender. Notably, children under 5 years of age and girls older than ten had the highest rates, and while preoperative CT lowered the negative appendectomy rate in boys and girls less than 5 years old it did not affect the rate in girls older than ten [6].

Anomalies of the gastrointestinal tract, specifically malrotation can make diagnosing appendicitis more difficult. Intestinal malrotation is a congenital anomaly whereby the intestines fail to rotate around the superior mesenteric artery during the 10th–12th weeks of development. As a result, the majority of small bowel is located to the right with the cecum and ascending colon on the left.

Consequently these children develop left sided appendicitis, presenting a diagnostic challenge for emergency physicians and surgeons alike. It is worth noting that this is not limited to children, a recent review of 95 cases of left sided appendicitis identified patients ranging from 8 to 82 years of age, 66 had left sided pain and 23 had malrotation [15]. Acknowledging that malrotation leads to an atypical presentation of acute appendicitis, an appendectomy has become a standard element of the Ladd's procedure. Although interestingly it was not an element of the initially described procedure [16].

Beyond the dilemma of making the initial diagnosis in our patient, we were faced with another decision; should a Ladd's procedure be performed in an asymptomatic patient with an incidental discovery of malrotation? While symptomatic malrotation is more prevalent in young children, particularly those younger than 3 years old, there is evidence to suggest that the benefit of operating for incidentally found malrotation outweighs the potential risks of waiting for symptoms to develop. Specifically, duodenal obstruction and intestinal ischemia secondary to midgut volvulus [17,18].

Abdominal masses in the pediatric population require thorough evaluation with history and physical exam, laboratory studies and appropriate imaging modalities. Our patient's presentation and findings were concerning for malignancy, specifically lymphoma. While non-Hodgkins lymphoma is a common malignancy of childhood, it is relatively rare in children younger than 5 years of age [19]. Consequently, we had to consider the possibility of missed appendicitis with associated mass. At the time of operation we were unable to proceed safely with a laparoscopic approach and converted to open laparotomy. A review of the recent literature suggests that while early appendectomy in children presenting with an appendiceal mass is safe, they have significantly longer operative times, delayed time to ambulation and return to normal diet, as well as longer hospital stays. However, when compared to laparoscopic appendectomy for appendicitis without mass, the conversion to open rate is not significantly different [20].

Appendicitis presenting as an abdominal mass suggestive of tumor has not been described in the literature and our case is unique. However, there are cases of lymphoma and other malignancies presenting as acute appendicitis in both adults and children. Specifically, a recent review of published cases found that 1% of all surgical specimens contained an unexpected pathology; including, but not limited to amoebic infection, tuberculosis, carcinoid, lymphoma, adenoma and mucocele [21]. There is also a published report of omental abscess presenting as an intra-abdominal tumor, however this was in a 12 year old patient who had previously had an appendectomy [22]. Our patient had no prior surgeries, although he did have a delayed presentation.

3. Conclusion

We report here a case of appendicitis masquerading as intra-abdominal lymphoma in a patient with malrotation. Appendicitis

should be part of the differential when malrotation is known to exist.

Conflict of interest

The authors have no financial or personal relationships to disclose.

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