



Pediatric mucinous neoplasm of the appendix presenting as a mucocele: A case report and review of the literature



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ABSTRACT

Mucocele of the appendix is a rare entity in pediatrics, with few case reports in the literature. We present a case of a 17-year-old male with right lower quadrant pain who was found to have an appendiceal mucocele while being evaluated for appendicitis. Laparoscopic-assisted resection was performed, with pathology confirming a low-grade mucinous neoplasm of the appendix, a tumor scarcely described in the pediatric population. He underwent complete resection with negative margins, rendering him cured without the need for any further resection or treatment. We review the literature on this topic.

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1. Introduction

Laparoscopic appendectomy remains one of the most frequently performed procedures by pediatric surgeons, with acute appendicitis being the most common pathology identified. Unexpected pathology findings after appendectomy in children are present in approximately 2% of specimens, and include pinworm, granulomas, eosinophilic infiltrates and neoplasms [1]. Tumors of the appendix are rare in the pediatric population, found in less than 0.5% of pathology specimens in this age group, with neuroendocrine tumors representing the most common tumor-type [1]. Mucoceles due to appendiceal mucinous neoplasms are exceedingly rare in this population. We describe a case of a 17-year-old boy with right lower quadrant pain found to have a mucocele secondary to a mucinous neoplasm of the appendix, successfully treated by laparoscopic assisted resection.

2. Case report

A 17-year-old boy, with no significant medical history, presented to the emergency department with one day of abdominal pain. The

pain was initially diffuse, and subsequently localized to the right lower quadrant. The pain was described as sharp and constant. It was associated with emesis, but no fevers, diarrhea, or urinary symptoms. The remainder of review of systems was within normal limits. He had no family history of malignancy.

On physical exam, he was well-appearing and afebrile with normal vital signs. His abdomen was soft and tender in the right lower quadrant with localized rebound tenderness. His laboratory values were normal, including a white blood cell count of 7.6 K/ μ L with an Auto Neutrophil % of 53.3%. An ultrasound of his abdomen demonstrated a blind-ending tubular structure measuring upwards of 2.5 cm. This structure was seen to extend from the cecum, but did not demonstrate any wall thickening or increased vascularity. The luminal contents appeared hypoechoic. The impression by the pediatric radiologist was appendicitis versus a mucocele of the appendix (Fig. 1). Given these findings, the patient was taken to the operating room for laparoscopic appendectomy using a standard three-port technique. The appendix was easily identified in the right lower quadrant. It was significantly dilated and tense (Fig. 2a and b). There was no evidence of any free fluid or any mucinous deposits throughout the abdomen and pelvis, including the peritoneal surfaces. In order to better examine the appendix and assure adequate resection, the appendix and cecum were carefully mobilized bluntly from their attachments to the right lower quadrant and eviscerated out of the umbilical incision using a wound protector/retractor (Fig. 2c).

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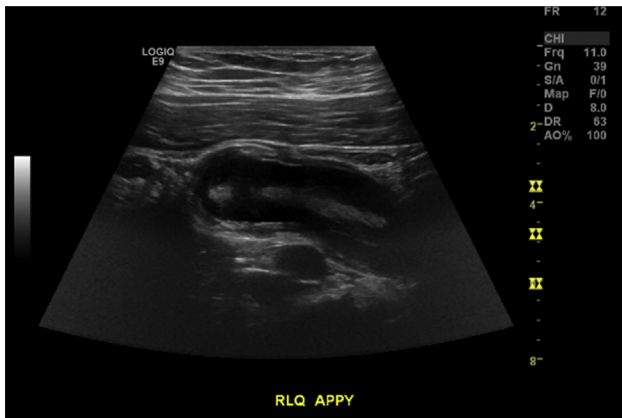


Fig. 1. Ultrasound of right lower quadrant showing 2.5 cm appendix with hypoechoic contents.

The appendix was diffusely firm to the base of the cecum. An appendectomy with partial cecectomy was performed using a 3.5 mm Endo GIA stapler, taking care to resect the specimen at an area of normal cecum (Fig. 2d, e, f). There was no violation or spillage of the appendiceal contents and the intact specimen was submitted for intraoperative pathologic consultation. The frozen section diagnosis was a low-grade appendiceal mucinous neoplasm (LAMN), and after a multidisciplinary intraoperative discussion, the decision was made to conclude the procedure with appendectomy only.

The patient did well post-operatively and was discharged home on post-operative day 2. Gross pathologic examination revealed a dilated, intact appendix measuring $10 \times 3.5 \times 1.0$ cm and cecum measuring $3.5 \times 1.5 \times 0.5$ cm (Fig. 3a). The appendix mucosa was flat and the lumen was filled with tenacious mucin. Microscopic examination revealed a low-grade appendiceal mucinous neoplasm (LAMN), consisting of flat to villiform mucinous epithelium with basal nuclei and abundant apical cytoplasmic mucin, without nuclear atypia, evidence of invasion, or

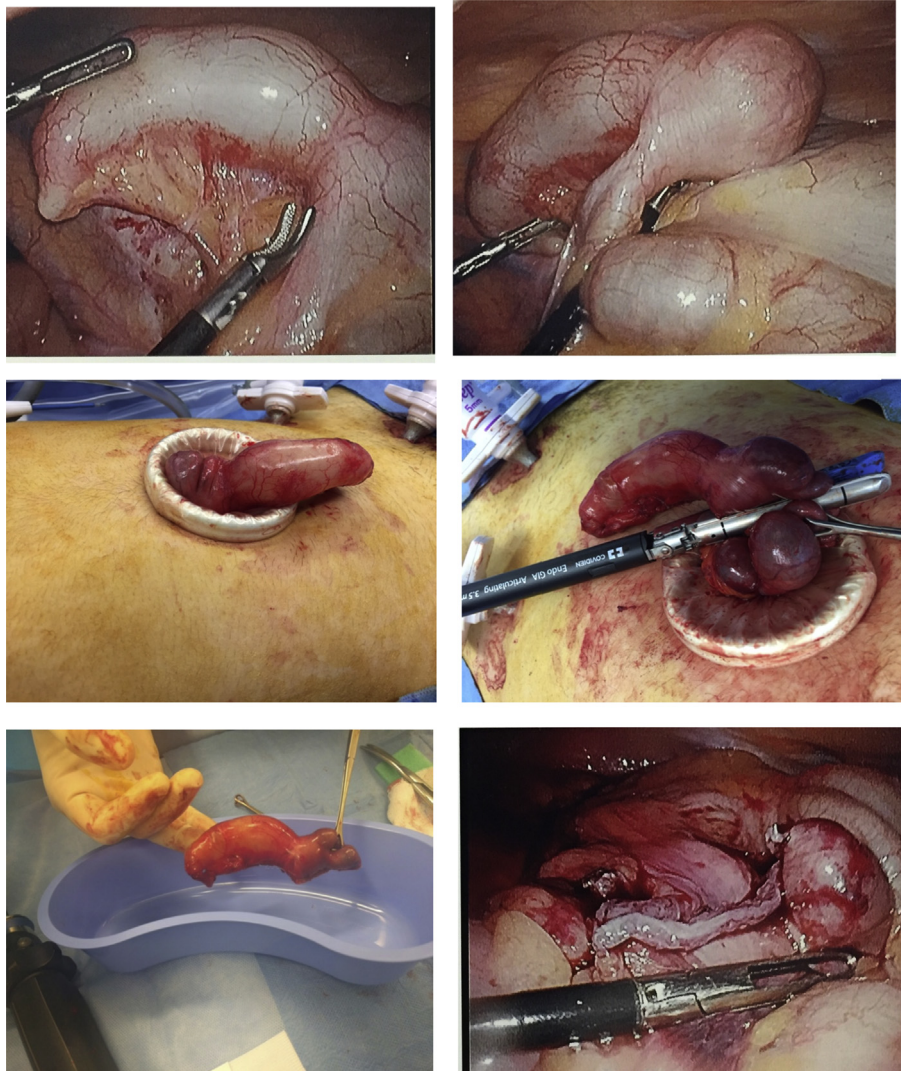


Fig. 2. a. Laparoscopic visualization of mucocele in right lower quadrant, b. laparoscopic mobilization of appendix and cecum c. extracorporealization of appendix and cecum through umbilical port site using wound protector d. stapled resection of appendix and partial cecectomy e. Resected specimen grossly intact f. Staple line.

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