Rare Pediatric Cecal Anomalies: A Lesson in Pediatric Surgery

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ABSTRACT **Background:** Rare incidence cases are part of the routine work of pediatric surgeons. Cecal anomalies in children are an example of such cases.

Objectives: To describe the presentation, workup, management and outcome of rare cecal anomalies in children and to analyze the skills needed for their successful treatment.

Methods: A retrospective chart review was conducted of all cases of cecal anomalies managed by the pediatric surgical service at a tertiary hospital from June 2017 to January 2020. Data regarding demographics, clinical presentation, radiological studies, surgical treatment, pathology, complications, and outcome were collected.

Results: Five cases of cecal anomalies were encountered over a period of 32 months, including a cecal volvulus, cecal duplication, cecal intussusception, and two cecal masses (one ulcerated lipoma and one polyp). All patients, except the patient with cecal duplication, presented acutely and were managed surgically. Long-term follow-up of 17–24 months was unremarkable in all cases.

Conclusions: A wide knowledge base, careful judgment, and creativity enable pediatric surgeons to successfully treat rare conditions such as rare cecal anomalies. These skills should be part of the education of pediatric surgery trainees.

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KEY WORDS: cecum, children, pediatric cecal anomalies, pediatric surgery

Rare incidence cases are part of the routine work of pediatric surgeons. These cases may lead to challenging diagnoses and lack of procedure specific operative learning curves [1]. A wide knowledge base, careful judgment, and creativity are therefore needed in such cases for appropriate management.

Cecal anomalies in children are an example of such rare cases. Among these cases are cecal duplications [2], cecal volvulus [3-5], and cecal masses [6-8].

In this study we described our experience with a series of five cases of rare cecal anomalies to demonstrate implications of the fundamental principles of pediatric surgery.

PATIENTS AND METHODS

We conducted a retrospective chart review of all cecal anomalies encountered by the pediatric surgery service in a tertiary medical center over a 32-month period (June 2017–January 2019). The cases were identified through a database of the department of pediatric surgery starting at its initiation on June 2017. We used the search terms: *cecum and cecal in the diagnoses*, *cecum*, *cecal*, *cecectomy*, and *ileocecectomy* under the category *procedure names*. Data were collected from patient charts and included demographics, clinical presentation, radiological studies, surgical treatment, pathology, complications, and outcome.

RESULTS

Five patients who were diagnosed with cecal anomalies were identified during the study period. Three cases were clustered within a 3-week period (December 2018). As 2073 patients were hospitalized in the pediatric surgery ward during this period, the occurrence of such anomalies within the population of children admitted to the hospital's pediatric surgery department over the study period was 1:414. Table 1 summarizes the pertinent data for the series.

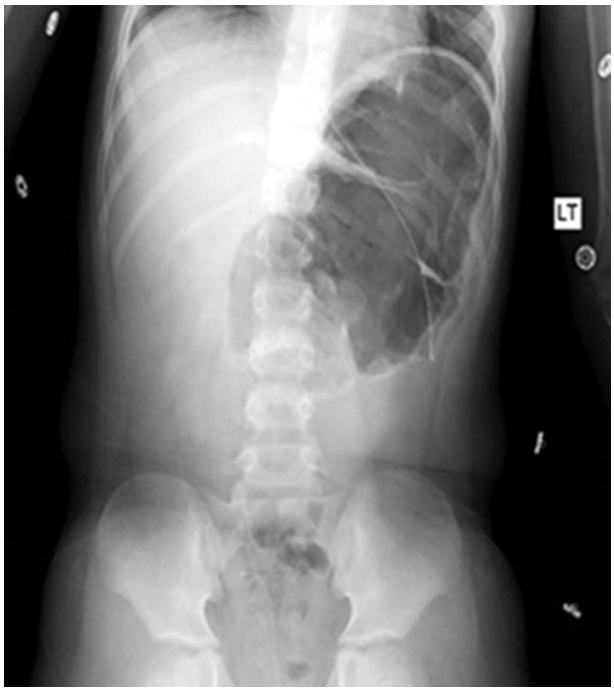
CASE 1

An 8-year-old boy presented to the emergency department with acute onset abdominal pain. He was pale, sweating, and hypertensive in agonizing pain. A physical examination revealed a mildly distended non-tender epigastrium and hyperperistalsis. The initial laboratory tests were normal. Plain abdominal plain film revealed a gas bubble on left upper quadrant suggesting cecal volvulus [Figure 1]. Abdominal computed tomography (CT) was conducted and confirmed the diagnosis. The patient was given fluid resuscitation and taken to the operating room for laparoscopy. Laparoscopy confirmed the diagnosis and reduction of volvulus and cecopexy were completed laparoscopically. Appendectomy was not performed. The patient recovered well. Nevertheless, due to continued upper abdominal fullness on

Table 1. Case characteristics

Case	Age	Sex	Diagnosis	Acute/Elective	Surgery	Complications	Follow-up time	Outcome
1	8 years	Male	Cecal volvulus	acute	Laparoscopic reduction and cecopexy	ileus	24 months	normal
2	4 years	Male	Cecal polyp, florid follicular hyperplasia	acute	Open partial cecectomy	none	24 months	normal
3	4 months	Female	Cecal duplication	elective	Laparoscopic ileocecectomy	none	24 months	normal
4	4 years	Female	Cecal mass follicular hyperplasia	acute	Laparoscopic partial cecectomy	none	20 months	normal
5	17 years	Female	Cecal mass lipoma	acute	Open partial cecectomy	none	17 months	normal

Figure 1. Plain abdominal film demonstrating a distended cecum in the left upper quadrant (Case 1)



physical examination, repeat plain films were obtained. Distended cecal bulb at the right abdomen was demonstrated. Contrast enema CT demonstrated passage of contrast into the distended cecum with an area of narrowing and thickened walls just distal to it, which was suspected as an area of bowel wall edema. Due to passage of normal gas and stool, diet was advanced and the patient was discharged home. Outcome at 24 months follow-up was normal, including a normal abdominal X-ray.

CASE 2

A 4-year-old boy presented to the emergency department with acute onset abdominal pain. Physical examination, laboratory findings and ultrasound were consistent with acute appendicitis. The patient was taken for an open appendectomy. Findings at surgery included a very thickened and distended non-inflamed appendix. A firm mass was palpated in the cecum next to the appendiceal origin [Figure 2]. Partial cecectomy enblock with the appendix using an Echelon® device (Ethicon US, LLC., USA) was performed. On opening the resected cecum, a large polyp was found. Pathology revealed florid follicular hyperplasia in both the polyp and the appendix [Figure 3]. Due to suspicion for Burkitt lymphoma, polymerase chain reaction (PCR) was performed and was negative for B-cell monoclonality. Flourescence in situ hybridization (FISH) was also performed and no cells with rearrangement of *C-MYC* gene were seen. The patient recovered uneventfully. Outcome at 24 months follow-up was normal.

CASE 3

Prenatal ultrasound anatomical scan and fetal magnetic resonance imaging revealed a right abdominal cystic mass on a female fetus, suspected for intestinal duplication cyst. After delivery, the newborn was asymptomatic. Initial ultrasound revealed a 1.2 cm cystic mass suspected for a mesenteric cyst. Follow-up sonography was initially without changes. However, ultrasound at the age of 4 months revealed a larger 2.7 cm right abdominal mass suspected to be a duplication cyst. An elective diagnostic laparoscopy was performed. A cecal duplication cyst was demonstrated and an ileocecectomy was performed. Pathology confirmed a cecal duplication cyst. The patient recovered uneventfully. Outcome at 24 months follow-up was normal.

CASE 4

A 4-year-old girl presented with colicky right lower quadrant abdominal pain. Physical examination was normal. Blood tests revealed elevated C-reactive protein (CRP). Ultrasound was equivocal for either appendicitis or a short ileocecal intussusception. Abdominal CT demonstrated a short ileocecal intussusception. Air enema demonstrated normal passage of air into the small bowel, with a cecal filling defect still suspected. Repeat

Figure 2. Cecal polyp between fingers (Case 2)

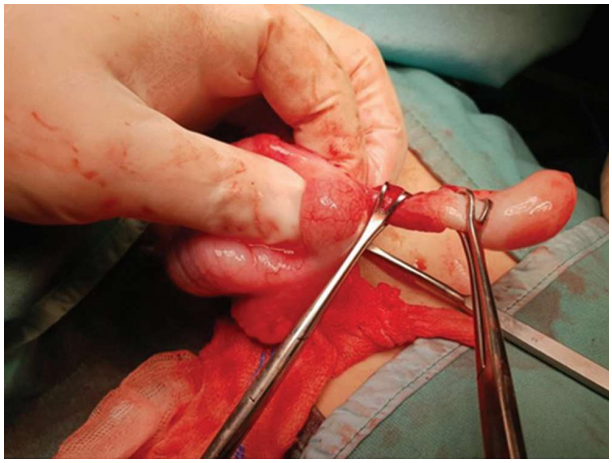
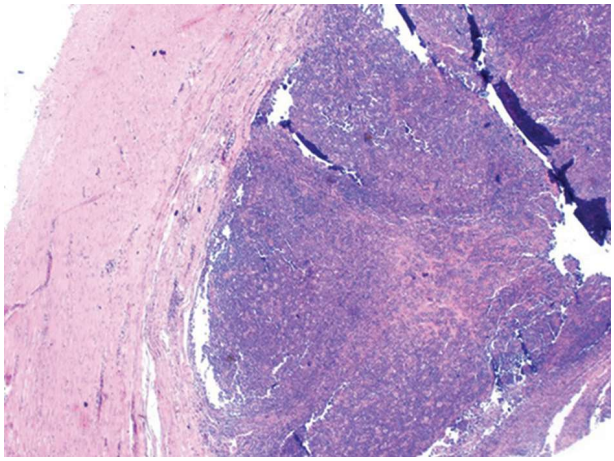


Figure 3. Appendix lumen occupied with lymphatic tissue (Case 2)



ultrasound demonstrated unchanged findings. Therefore, the patient was taken for diagnostic laparoscopy with the suspected diagnosis of intussusception of the appendix. On laparoscopy, a thickened appendix was demonstrated, without other signs of inflammation. The base of the appendix was sunken into the cecum. A circumferential cecal mass was palpated around the orifice of the appendix. A partial cecectomy using an Echelon® device was performed en block with the appendix. On opening the resected cecum an ulcerated circular mass was seen around the base of the appendix. Pathology revealed an ulcerated florid follicular hyperplasia around the base of an appendix with a sign of subsiding acute appendicitis. The patient recovered uneventfully. Outcome at 20 months follow-up was normal.

CASE 5

A 17-year-old girl presented with right lower abdominal pain. History, physical examination, laboratory results, and abdominal sonography were all supporting acute appendicitis. The patient was taken to the operating room for an open appendectomy. At surgery, the appendix appeared normal, and a 2 cm cecal mass was palpated adjacent to the base of the appendix. A partial cecectomy using an Echelon® device was performed. Pathology revealed a 2 cm ulcerated lipoma with degenerative changes with free surgical margins and an unremarkable appendix. The patient recovered uneventfully. Outcome at 17 months follow-up was normal.

DISCUSSION

We presented a cluster of five cases of individually rare pediatric cecal anomalies over a period of 32 months, which were treated successfully by pediatric surgeons. Unfortunately, most cases presented acutely with no prior complaints and therefore an elective management plan was not possible.

Cecal volvulus is a rare condition, reported both in adults and children. Its occurrence in children is so rare that its true incidence is unknown [3-5]. Suspicion for this rare entity should arise when the patient is complaining of epigastric pain, and when epigastric fullness is demonstrated on physical examination. A plain abdominal film usually suffices for the diagnosis. However, abdominal CT may assist with diagnosis in equivocal cases [9]. Prompt surgery should follow to relieve the obstruction and prevent bowel necrosis. In surgery, reduction of volvulus is performed. Some advocate additional cecal resection [9,10] as the most efficient way to prevent recurrence. Others recommend cecopexy [11], which entails a much lower risk for morbidity than resection. Reports of laparoscopy for the management of this pathology are scarce, and we found no reports of laparoscopic management of cecal volvulus in the pediatric population [12,13]. In contrast to the suggestion of some authors against laparoscopy as an impractical approach due to bowel obstruction with significant bowel distention [14], we demonstrated that laparoscopy can be a safe and efficient mode of treatment, even in a child. The extended postoperative distention of the cecum, which resolved without intervention, should be considered as a possible postoperative course. Colonoscopic reduction of cecal volvulus in children has also been documented [4], although long-term follow-up for possible recurrence is not reported.

Enteral duplications are a group of rare malformation, estimated to occur in 1:100,000 births [15]. Cecal duplications are very rare, and we found only two dozen cases reported in the literature [2]. Complications associated with enteric duplications include perforation, bleeding, and bowel obstruction [15,16]. In recent years, prenatal diagnosis has become more common. The recommendations for the management of asymptomatic duplications diagnosed prenatally are resection in infancy to avoid complications [17].

Large cecal polyps are usually encountered during routine screening colonoscopy procedures in adults [18]. These polyps are usually adenomas, but can also sometimes be adenocar-

cinomas [18]. Intestinal nodal lymphoid hyperplasia, despite being rare in adults [19], is common in the pediatric population [20-22]. Large intestinal lymphoid masses, if present, are usually reported in the rectum (rectal tonsils) [8]. Nevertheless, it is not surprising to encounter lymphoid hyperplasia as cecal masses in children. Other etiologies for intra-cecal masses in otherwise healthy children, such as lipomas [6] and carcinomas [7], are rare. Cecal lipomas are rare even among adults. We found only 26 cases described in the English-language literature between 2000 and 2017, all in adults. The predominant clinical symptom was abdominal pain (80.8%), and the most common preoperative diagnosis was acute or chronic recurrent intussusception (46.2%) [23]. Intussusception of the appendix, a rare entity, can be secondary the cecal masses [24]. In our case, it was secondary to follicular hyperplasia. We think that partial cecectomy, as long as the ileocecal valve is preserved and performed during either an open or a laparoscopic appendectomy using an endoscopic linear stapler as described for some adult conditions [18,25], is a good treatment option. The results of three of our case demonstrated such partial cecectomies, revealed no complications, and concluded with a benign pathology in all cases.

CONCLUSIONS

Our series of rare pediatric cecal anomalies demonstrated that a wide knowledge base, careful judgment, and creativity are all essential components when facing children presenting with rare cecal pathologies. These skills should be part of the education of pediatric surgery trainees, a discipline that may commonly present a challenge of managing rare conditions.

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There may be times when we are powerless to prevent injustice, but there must never be a time when we fail to protest.

Elie Wiesel (1928–2016), Romanian-born American Jewish writer, professor, political activist, Nobel Laureate and Holocaust survivor