Journal of Pediatric Surgery Case Reports 54 (2020) 101388

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: http://www.elsevier.com/locate/epsc

Cecal duplication cyst complicated by prolapsed ileocolic intussusception

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ARTICLE INFO

Keywords:
Cecal duplication cyst
Ileocolic intussusception
Prolapsed intussusception

ABSTRACT

Cecal duplication cysts are very rare including 0.4% of all GI tract duplications. The ultrasound (US) is the imaging of choice for duplication cysts detection in pediatrics. Cyst's wall is made up of an inner mucosal layer, which is echogenic and an outer muscular layer, which is hypoechoic at US (called "pseudo kidney" appearance on longitudinal view or "doughnut" appearance on transverse view). Intussusception is one of the duplication cyst's complications. Intussusception presented with *trans*-anal protrusion (prolapsed intussusception) is a rare and confusing condition which can cause delayed diagnosis and further complications.We present an 18-month old boy with Cecal duplication cyst causing intussusception, which protruded from anus.

1. Introduction

One of uncommon inherited diseases is enteric duplication cyst, its incidence is 1 per 4500 live births [1]. It is more common in boys under 2 years old [2]. It can be present in all parts of alimentary tract from the mouth to anus. The most prevalent site is the small bowel. In order of frequency, it occurs in the ileum, jejunum and duodenum [3]. Cecum is one of the rarest locations.[2]Intussusception is a complication of enteric duplication cysts, which leads to bowel obstruction [1]. We present an 18-month old boy with Cecal duplication cyst causing intussusception, which was protruded from anus. Each of Cecal duplication cyst and intussusception prolapse is extremely a rare entity. This is the first reported case of prolapsed intussusception of Cecal duplication cyst.

2. Case presentation

The 18-month old boy was brought to our emergency department with complaints of colic abdominal pain from 2 weeks ago, fever and multiple episodes of non-bloody and non-bilious vomiting from one day ago.

This patient was admitted to another small general hospital the day before he had been referred to us. His referral form mentioned that during the admission protrusion of a mass from the anus occurred after defecation and that they had performed abdominopelvic ultrasound (US) for finding the cause. There was an ileocolic intussusception extended to the rectum and protruded from the anus with no pathologic lead point; as expected owing to the fact that the patient was in 6_36 months age group in which most of the intussusceptions occur idiopathically. The prolapse was managed and reduced successfully before the patient's referral, but intussusception had not responded to reduction, and so the patient was referred to our pediatric specialized hospital for further investigations.

On physical examination low grade fever was detected. His abdomen was distended, soft, and nontender. A palpable left lower quadrant mass was found. His complete blood count showed leukocytosis (WBC = 14.7 * 10^9/L, Polymorphonuclear = 65%). The remainder of the physical examination and lab data were unremarkable.

US performed in our hospital showed ileocolic intussusception. One large (37*28 mm), thick-walled cyst with a "double layered" appearance was observed in front of intussusception which was acting as a lead point (Fig. 1). (video 1).

Abbreviations: US, ultrasound, GI, gastrointestinal, NEC, necrotizing enterocolitis, DRE, Digital Rectal exam

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https://doi.org/10.1016/j.epsc.2020.101388
Received 11 January 2020; Accepted 18 January 2020
Available online 22 January 2020

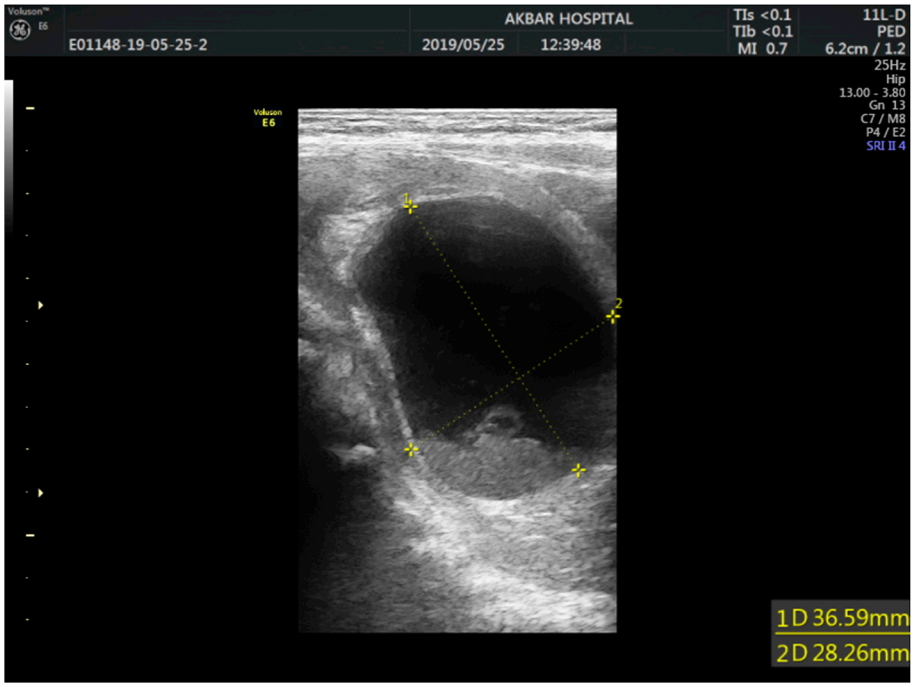

Fig. 1. Large, thick-walled cyst with double layered appearance and mucinous content in an 18-month old boy's ultrasound.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.epsc.2020.101388.

The patient got ready for operation and then abdomen opened with a transverse incision above the umbilicus. There was reactive fluid within abdomen and ileocolic intussusception extending to the rectum. After intussusception reduction, a mass was palpated in the cecum (lead point) and then ileocecal resection and end to end anastomosis performed. Post operative period was uneventful diagnosis after pathologic examination was a 40*30 mm mucin filled duplication cyst originating from cecum and extending to the ileocecal valve (Fig. 2).

3. Discussion

Cecal duplication cysts are very rare, including 0.4% of all GI tract duplications.[2]They can be found incidentally. Half of the Cecal duplication cysts are correctly diagnosed pre operatively [2]. Our preoperative ultrasound showed its origin correctly. Appendicular mass, tumor and necrotizing enterocolitis (NEC) are its differential diagnoses [2].

The clinical presentation is mainly contingent upon the size and site. Small cysts usually present in the role of leading point for intussusception or volvulus. Bigger cysts can cause a compressive effect, which leads to GI tract obstruction or ischemia [3].

Histologically have cystic structures and share their muscular wall and blood supply with the adjacent intestine [3]. Amount of mucin in Cecal duplication cyst is related to duration of obstruction. In this case cyst's mucosa was lined by mucinous epithelium and cyst's lumen was partially loaded by mucin.

The US is the imaging of choice for duplication cysts detection in pediatrics. Its specificity is reported as high as 95% and its positive predictive value is 85–100% [4]. The colonic duplication cysts have spherical (>80% of cases) or tubular appearance [5]. Cyst's wall is made up of an inner mucosal layer, which is echogenic and an outer muscular layer, which is hypoechoic at US (called “pseudo kidney” appearance on longitudinal view or “doughnut” appearance on transverse view) [6]. The twofold layered wall appearance is demonstrated in more than half of the cases [2].

Intussusception is one of the duplication cysts' complications, which is an invagination of a part of GI tract into the more distal part. They are usually idiopathic in pediatric patients but can sometimes

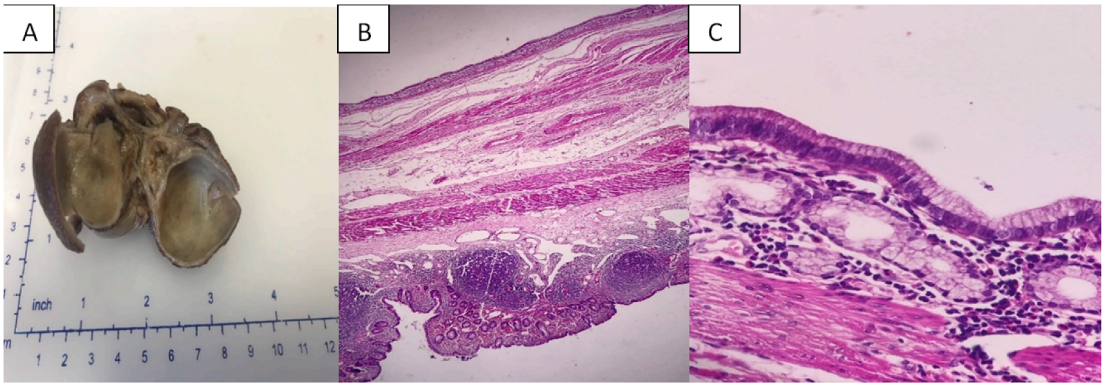

Fig. 2. A. gross pathology (unilocular intramural Cecal cyst) B. True duplication cyst of Cecum with separate muscularis propria (H&E staining, HPF) C. Duplication cyst mucosa lined by mucinous epithelium (H&E staining, HPF).

have lead points (2.2–15%) [7]. The most common lead points in children are Meckel's diverticulum, Duplication cyst and intestinal polyps [7]. Timely diagnosis of these secondary intussusceptions is very important due to their potential to cause complications and need for surgery [5].

Intussusception presented with *trans*-anal protrusion (prolapsed intussusception) is a rare and confusing condition, which usually leads to delayed diagnosis, high morbidity and mortality [8]. Most of previous case reports are from low and middle income African and Asian countries and neglected cases [8]. It is usually confused with rectal prolapse in the first clinical encounter as our case. After the lack of improvement with conservative management of rectal prolapse, further examination leads to the diagnosis of intussusception with *trans*-anal protrusion. [8]Prolapsing rectal mass with intestinal obstruction is also another clinical clue; Although in one third of cases it can happen without typical presentations of intussusception. Digital Rectal exam (DRE) helps to differentiate rectal prolapse from intussusception prolapse. In DRE of intussusception prolapse, the finger can insert parallel to intussuscepted bowel and the anorectal is fixed in its position [8]. Also, using the US can help differentiating these two entities.

4. Conclusion

Despite Cecal duplication cyst rarity, it is easily diagnosed by clinical suspicion and ultrasound. Thick-walled cyst or double-layered appearance is the most important ultrasound finding. Quick detection and management is essential in order to improve the post-op outcomes; so it is important to keep the diagnosis in mind for the patients with similar presentations.

Consent to participate

Formal written consent was obtained from the patient's legal guardian (his father in this case as the patient was under 18). Patient's father agreed on publication of this case report and accompanying images.

Consent for publication

The patient's father has given oral and written consent for use of the ultrasound and pathology images and he is aware that the report will be published in a medical journal which is distributed worldwide for educational purposes.

Funding

The authors do not receive any funding.

Declaration of competing interest

The authors declare no competing interest.

This study was done with no specific funding or non-funding support. The authors have no conflicts of interest relevant to this article to disclose.

Acknowledgements

not applicable to this article.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.epsc.2020.101388.

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