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Case Report

Congenital colorectal tubular duplication in an infant: A tale of radiological diagnostic challenges ☆

Selim Ahmed, MBBS, FCPS[a], *Mohd Syafiek Saifuddin, BSc, MB, BCh, BAO, MMED*[b], *Chiak Yot Ng, MBBS, MMED*[c,*], *Nik Fatimah Salwati, MBBS, MMED*[b], *Firdaus Hayati, MD, MMED*[d], *Suseela Ismail, MD, MMED*[e]

[a] *Paediatric Department, Medical Based Department, Faculty of Medicine & Health Science, Universiti Malaysia Sabah, Kota Kinabalu, Sabah, Malaysia*
[b] *Department of Radiology, Hospital Sultanah Bahiyah, Alor Setar, Kedah*
[c] *Department of Radiology, Medical Based Department, Faculty of Medicine & Health Science, Universiti Malaysia Sabah, Jalan UMS, 88400, Kota Kinabalu, Sabah, Malaysia*
[d] *Department of Surgery, Faculty of Medicine & Health Science, Universiti Malaysia Sabah, Kota Kinabalu, Sabah, Malaysia*
[e] *Department of Radiology, Queen Elizabeth Hospital, Ministry of Health Malaysia, Kota Kinabalu, Sabah, Malaysia*

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ABSTRACT

Enteric duplications (EDs) are rare congenital anomalies that result from defect during embryonic development of the gut. Although EDs can literally occur at any part of the gastrointestinal tract, ileocecal duplication is the commonest type followed by colorectal type. Morphologically, EDs are mostly cystic in nature; tubular duplications are uncommon. We report radiological diagnostic challenges encountered in dealing with a 10 month-old infant who presented with chronic constipation, progressive abdominal distension, and voiding difficulty for several weeks followed by colicky abdominal pain for three days. After a series of radiological procedures, a diagnosis of tubular colorectal duplication was made. The duplicated segment was loaded with impacted feces which exerted pressure effect on the rectum and urinary bladder. The case was treated surgically through laparoscopic procedure that included fenestration and stapling of the duplicated bowel followed by irrigation. Postoperatively, the child was followed up half-yearly for three years and was found to remain symptom-free. This case exemplifies the challenges a radiologist may experience while dealing with a case presenting with features of as acute-on-chronic intestinal obstruction and voiding difficulty.

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* Corresponding author.
E-mail addresses: ngchiakyot@ums.edu.my, ngchiakyot@gmail.com (C.Y. Ng).

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Introduction

Enteric duplications (ED) are uncommon congenital anomalies that originate as a result of caudal twinning of the primitive gut during development and differentiation in foetal life [1]. Although EDs can arise from anywhere along the gut, most commonly they stem from the ileal region. They are named according to the anatomical region they originate from [2-4]. Morphologically, EDs are commonly cystic (80%) and less commonly tubular (20%) in nature [5].

They are firmly attached to the mesenteric border of the intestine and share common blood supply with it. They are lined by cells representing the part of mature gut they derive from. Most EDs do not have a communication with the parent intestinal segment.

EDs may occur either as an isolated disorder or in association with uro-genital or spino-vertebral anomalies. They may remain asymptomatic, of which most are discovered incidentally as a soft abdominal mass during abdominal examination. Among the symptomatic cases, some may present with features of incomplete or complete intestinal obstruction, such as vomiting, constipation, abdominal pain and distention [6-10].

We share our experiences of radiological diagnostic challenges that we faced while evaluating an infant presented with chronic constipation, abdominal distension, and voiding difficulty for several weeks followed by colicky abdominal pain for three days.

Case Report

A 10 month-old boy with the history of chronic constipation, gradually increasing abdominal distension, and straining during micturition for 2 months followed by acute colicky abdominal pain for 3 days. The child did not have any vomiting and he could pass flatus and scanty stool. During each episode of abdominal pain, the infant screamed with curling the body and arching the back in attempt to reduce pain intensity. General examination showed a fretful infant with agony in facial expression. The abdomen was globally distended but nontender with palpable, indentable, mobile, soft tissue masses suggestive of colonic faecal impaction. Digital rectal examination identified no rectal grip and partially filled rectum. A soft, boggy mass was felt anterior to the rectum. No Genitourinary or spino-vertebral anomalies were found in the child. A clinical diagnosis of acute-on-chronic intestinal obstruction was made with a likely differential diagnosis of bladder neck mass. He was admitted in paediatric surgical ward for further investigations and management.

The initial abdominal plain radiograph showed prominent bowel loops displaced superiorly by a soft tissue opacity seemingly arising from the pelvic cavity (Fig. 1). No pneumo-peritoneum noted. Our clinical impression started favoring the probability of the urinary bladder neck mass with Hirschsprung disease as an alternative possible diagnosis. Pelvic ultrasound detected normal pelvic organs including normal configuration of the urinary bladder with no bladder wall outpouching reaching the posteriorly located rectum. However, sonological evaluation of the large gut was limited due to presence of colonic gas. Therefore, a lower gastrointestinal (LGI) contrast study and a micturating cystourethrogram (MCU) were performed. The LGI contrast study (Fig. 2A and B) demonstrated anterior indentation and compression of the contrast-opacified rectum complicated with mild colonic dilatation proximal to the compression. The rectum was displaced posteriorly to the right and the sigmoid colon was displaced superiorly. However, despite external compression on the rectum, contrast opacification was seen all the way from the rectum to the level of descending colon. The MCU study (Fig. 3) demonstrated displacement of the contrast-filled urinary bladder anteriorly with posterior indentations of the urinary bladder wall. No fistulous communication between bowel and urinary bladder was identified. The urethra was normal. From this series of fluoroscopic studies, a conclusion of a non-communicating rectovesical mass causing compression onto the rectum and urinary bladder was made. Subsequently, for further evaluation of the rectovesical mass, a contrast computed tomography (CT) scan of abdomen was performed. The CT images (Fig. 4A, B, C) revealed a long, tubular, faecal-laden, bowel-like structure sharing a common wall with the rectum and colon. It spanned from anorectal junction to the splenic flexure. The imaging features were highly suggestive of a colonic duplication which exerted mass effect onto the rectum, sigmoid colon, and urinary bladder supporting the earlier imaging studies. The widest diameter of the bowel-like structure measured 5 cm, however, there was absence of demonstrable contrast within it that led us to

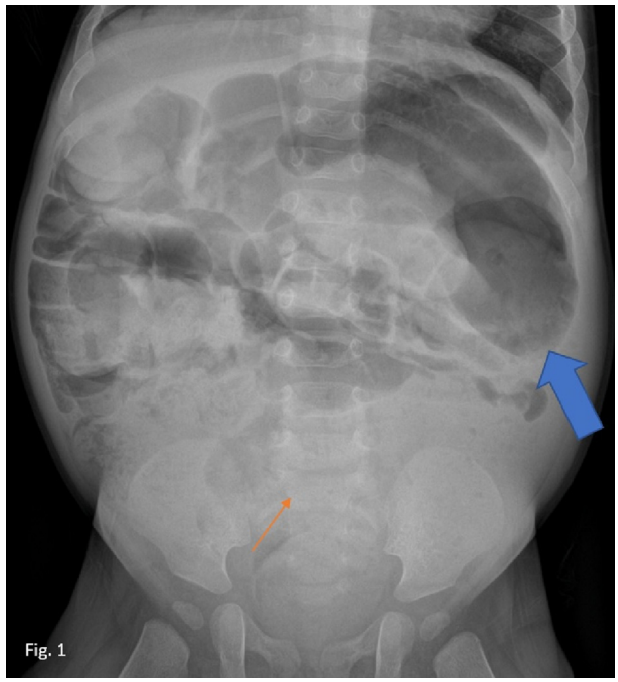

Fig. 1 – Abdominal radiograph revealed superiorly displaced distended bowel loops (blue arrow) by a soft tissue opacity seen likely arising from the pelvic region (Orange arrow). Color version of figure is available online.

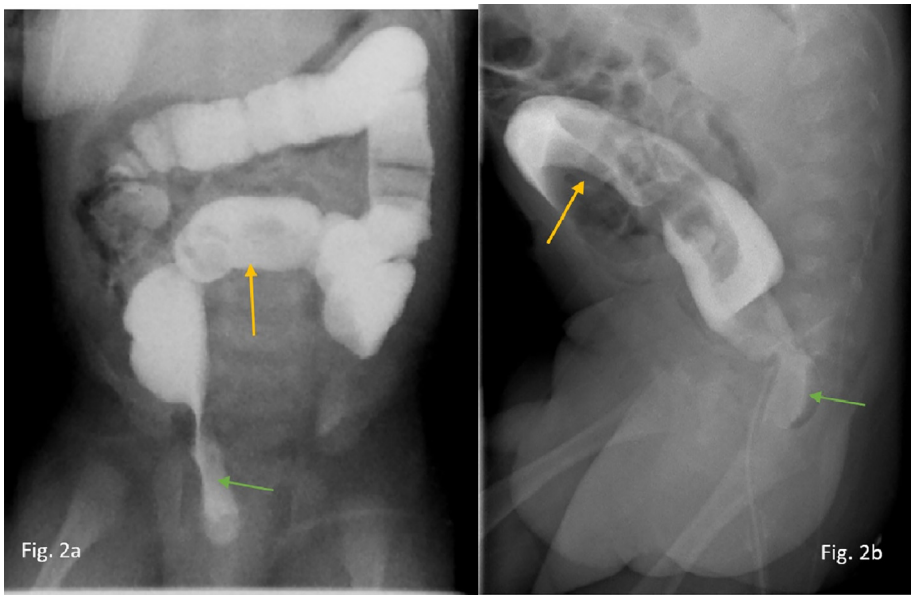

Fig. 2 – LGI contrast study in anterior posterior view (Fig. 2A) and lateral view (Fig. 2B) shows abnormal displacement of the rectum posteriorly (green arrow) and sigmoid colon superiorly (yellow arrow). Color version of figure is available online.

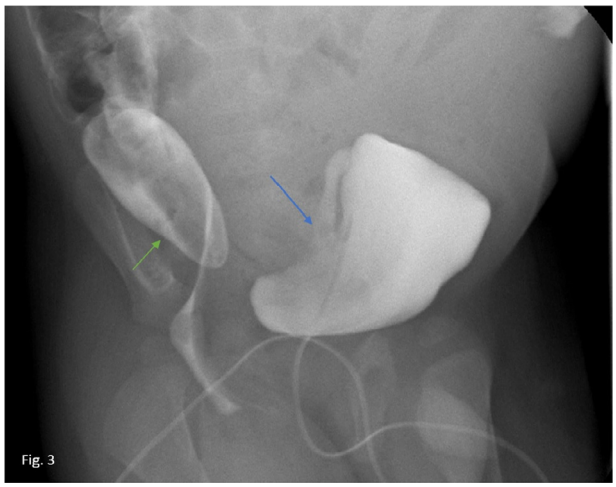

Fig. 3 – Right anterior oblique view of micturating cystourethrogram (MCU) demonstrated abnormally gapped urinary bladder and rectum (green arrow) with posterior indentation of a well distended urinary bladder (blue arrow) suggesting a rectovesical mass causing compression onto the urinary bladder and rectum. Color version of figure is available online.

uncertainty about the presence of a communication between the native colon and the duplicated part.

An agreement was achieved for laparoscopic exploration after a multidisciplinary discussion was conducted involving the managing surgeon and radiologist. Intraoperatively, the team found a tubular, faecal-laden colonic duplication arising from the caecum at the mesenteric site, extending up to the rectum pushing the rectum posteriorly. The tubular colonic duplication shared the same wall with the native bowel with no demonstrable luminal communication between the tubular colonic duplication with the native colon. Single appendix and terminal ileum visualized attaching to the cecum. Laparoscopic assisted fenestration and stapling of the duplicated part followed by irrigation of impacted faecal materials was performed. Biopsy of the duplicated bowel was taken, and the biopsy sample showed submucosa, mucosal and smooth muscle layers mimicking large bowel in keeping with colonic duplication. Post-operatively, the boy recovered well without any complication. He was followed up every 6 monthly for 3 years and was found symptom-free, well and healthy.

Discussion

The incidence is estimated to be 1 in 4500 live births. They are primarily encountered in children and rarely in adults [11]. The exact aetiology of EDs is yet unknown, however, several theories are postulated of which in-utero vascular accident theory is the most popular 1 [12,13]. In terms of site of origin, jejuno-ileal duplication tops the list accounting for as high as 61% of all cases [14]. Hindgut duplications are reported to comprise 6.8%-13% of all cases of EDs. The colorectal tubular duplications, as was our case, are further rare; they are double-barrelled duplication that can have associated rectogenital or recto-urinary fistula, duplication of internal or external genitalia, bladder anomalies or vertebral anomalies [15]. Our reported case, fortunately, did not have any.

Clinical presentations of EDs depend on several factors including age, size and location of the duplication, type of mucosal lining, communication with the bowel lumen, and associated anomalies [16,17].

Our case presented with chronic constipation, progressively increasing abdominal distension and voiding difficulty

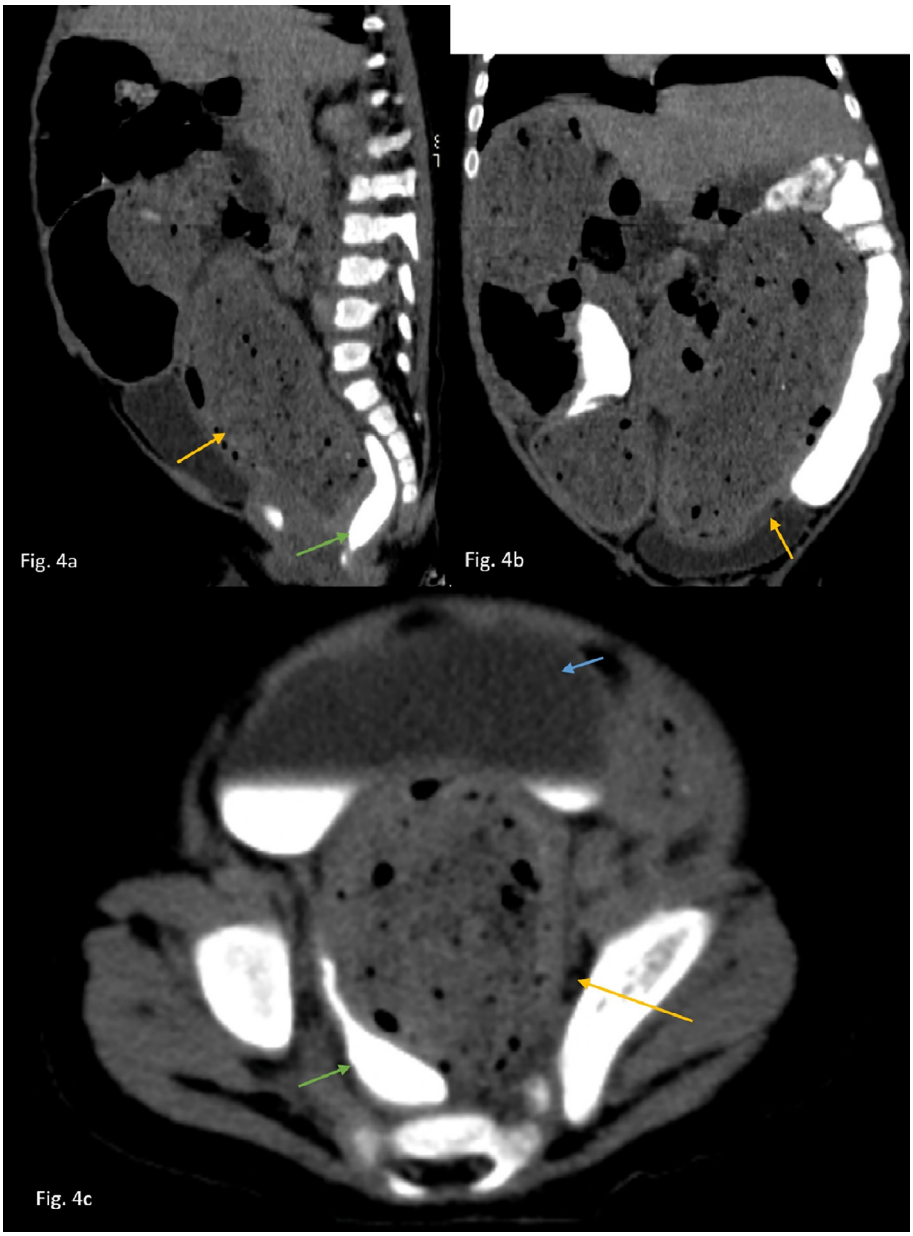

Fig. 4 – (A-C) – At sagittal (Fig. 4A), coronal (Fig. 4B), and axial view (Fig. 4C) respectively, the CT showed faecal-laden duplication cyst (yellow arrow) causing mass effect onto the contrast filled rectum (green arrow) and contrast-layered urinary bladder (blue arrow). Color version of figure is available online.

(straining to complete the voiding) indicating chronic, partial, large gut obstruction with pressure symptoms on bladder. The parents sought medical treatment for these complaints in the local community clinic without any mentionable improvement. However, sudden development of severe colicky abdominal pain brought this child to paediatric surgeon's care in the hospital. The attending paediatric surgeon made a provisional diagnosis of acute-on-chronic intestinal obstruction keeping high probability of Hirschsprung disease in mind and requested for radiological evaluation of the case to find out the probable cause. The radiology team had to proceed through several diagnostic steps before obtaining a concluding diagnosis of tubular, colorectal duplication through contrast CT scan of abdomen.

Ultrasound remains the most preferable, initial non-invasive imaging modality of choice for evaluating paediatric abdominal pathologies and anatomical anomalies [18] Ultrasound may display a cystic structure with 'gut signature' of hyperechoic mucosal layer and hypoechoic smooth muscle layer [19]; however, we failed to detect these features in our case during initial ultrasound presumably because of huge bowel gas which markedly limited sonographic evaluation. In recent

decades, endoscopic ultrasound has revolutionized the sonological evaluation of the EDs [5]. However, we could not do it primarily due to lack of technical skill and expertise. Hence, we proceeded to LGI contrast study. The decision for MCU, despite being an uncomfortable and disliked investigation by the paediatric patients and parents, was made to address the voiding difficulty encountered by our patient who was a boy and the probability of a posterior urethral valve or a bladder neck mass required to be excluded. Furthermore, MCU helped us to rule out any fistulous connection between the gut and urinary bladder which could be present in cases of colorectal duplication cyst [20]. In the final step, an abdominal contrast CT scan was performed to get a near-conclusive radiological diagnosis. It identified the presence of a tubular enteric duplication filled with faecal-like material. Being convinced by the CT scan report, the paediatric surgical team planned for laparoscopic surgical exploration for definite diagnosis and further management because surgical resection is the most widely-accepted method for majority, if not all, cases of gastrointestinal duplications even if the patient is aymptomatic [21].

During operation, the duplicated part was found loaded with faecal material; however, the surgical team could not depict a communication between gut and the duplicated part. This could be explained by the hypothesis that functional closure of the communication pathway might have happened, as postulated by Kimura S et al in 2018 [22].

Conclusion

Overall, EDs are uncommon congenital malformations of the gut and a colorectal tubular duplication is even a rarer anomaly. Although many cases of EDs remain asymptomatic, evaluation of a symptomatic case is quite challenging because the usual list of differential diagnosis does not include ED in the first place. When the initial radiological evaluations fail to explain the clinical vignette, a further step-by-step imaging procedure are required to get a near-definite diagnosis before surgical exploration. Our case is expected to remind our fellow radiologists about the patience and perseverance required to deal with an uncommon pathology, such as an enteric duplication.

Limitations

Intraoperative images were initially archived however were unfortunately lost due to technical issues on the archiving device.

Patient consent

Written informed consent was obtained from the patient's parents for the publication of this case report.

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