

Cecal duplication cyst associated with a malfixed right colon presenting as an abdominal mouse in an infant causing a diagnostic dilemma: a case report

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Duplication cysts (DCs) of the cecum are extremely rare anomalies. A DC causing luminal obstruction and presenting as a recurrent intestinal obstruction in infants is quite a rare occurrence. They usually present with a palpable lump, but when associated with malrotation, can result in a diagnostic dilemma. We report a rare case of a cecal DC associated with a malfixed right colon presenting with a wandering intra-abdominal lump and recurrent intestinal obstruction, resulting in a diagnostic dilemma in an infant. *Ann Pediatr Surg* 14:101–104 © 2018 Annals of Pediatric Surgery.

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Introduction

Enteric duplication cysts (DCs) are rare anomalies; among these, cecal DCs are the rarest [1]. They have highly variable clinical presentations; they can present with abdominal mass, palpable intussusception [2], intestinal obstruction, gastrointestinal hemorrhage, can mimic acute appendicitis [3], disappearing intra-abdominal mass [4], and recurrent intestinal obstruction [5]. The majority (50%) of cecal DC present with a palpable mass [6]. However, in cases of cecal DCs, when associated with a malfixed right colon, the palpable lump can present as a wandering intra-abdominal lump, mimicking an 'abdominal mouse'. We report a rare case of a cecal DC associated with a malfixed cecum and right colon presenting as an 'abdominal mouse' in an infant, resulting in a diagnostic dilemma.

Case report

Fifty two day old, male term baby, weighing 4 kg, presented to us with bilious vomiting, mild abdominal distension, and constipation for 3 days. The baby had passed meconium on day 1 of life. He had been hospitalized elsewhere once at the age of 1 month for similar complaints. There was no melena or hematochezia. A freely mobile firm intra-abdominal lump measuring $9 \times 5 \text{ cm}^2$ was palpable in the right paraumbilical region (Fig. 1a). The lump was highly mobile, slippery, and was easily movable to all the quadrants similar to an 'abdominal mouse'. Per-rectal examination was normal. Abdominal radiography showed multiple dilated small bowel loops and a round 'crescent'-shaped radio-opaque shadow in the distended distal ileal loop (Renu's sign) (Fig. 1b). There was absolutely no colonic gas suggesting a complete ileocecal obstruction. Abdominal ultrasound showed a thin-walled cyst in the left hypochondrium and suggested a mesenteric cyst or an omental cyst. Abdominal contrast-enhanced computed tomography showed a partially enhancing cystic lesion in the left iliac fossa, which suggested a DC of the sigmoid colon or

an inflammatory loculation (Fig. 2a). Various intraperitoneal cysts causing bowel obstruction were considered in the differential diagnoses. Laparotomy indicated a freely mobile cecal mass measuring $9 \times 7 \times 5 \text{ cm}^3$ with a grossly dilated ileum and a malfixed, narrow, and collapsed ascending colon (Fig. 2b). The cecum was grossly enlarged with a luminal cystic mass, causing complete ileocecal junction obstruction (Fig. 2b). However, the duodenojejunal flexure was located in the normal position to the left of the spine. Ileocecal resection with ileo-ascending anastomosis was performed. Gross specimen showed a $7.5 \times 5.5 \times 4.7 \text{ cm}^3$, intact tense luminal submucosal cystic mass, arising from the mesenteric side of the cecum and extending into the ileum (Fig. 2b). The cyst was filled with whitish mucoid material. Microscopy of the cyst showed denuded colonic epithelium and two distinct muscle layers with focal attenuation suggestive of an enteric DC (Fig. 3b and c). The baby had an uneventful recovery.

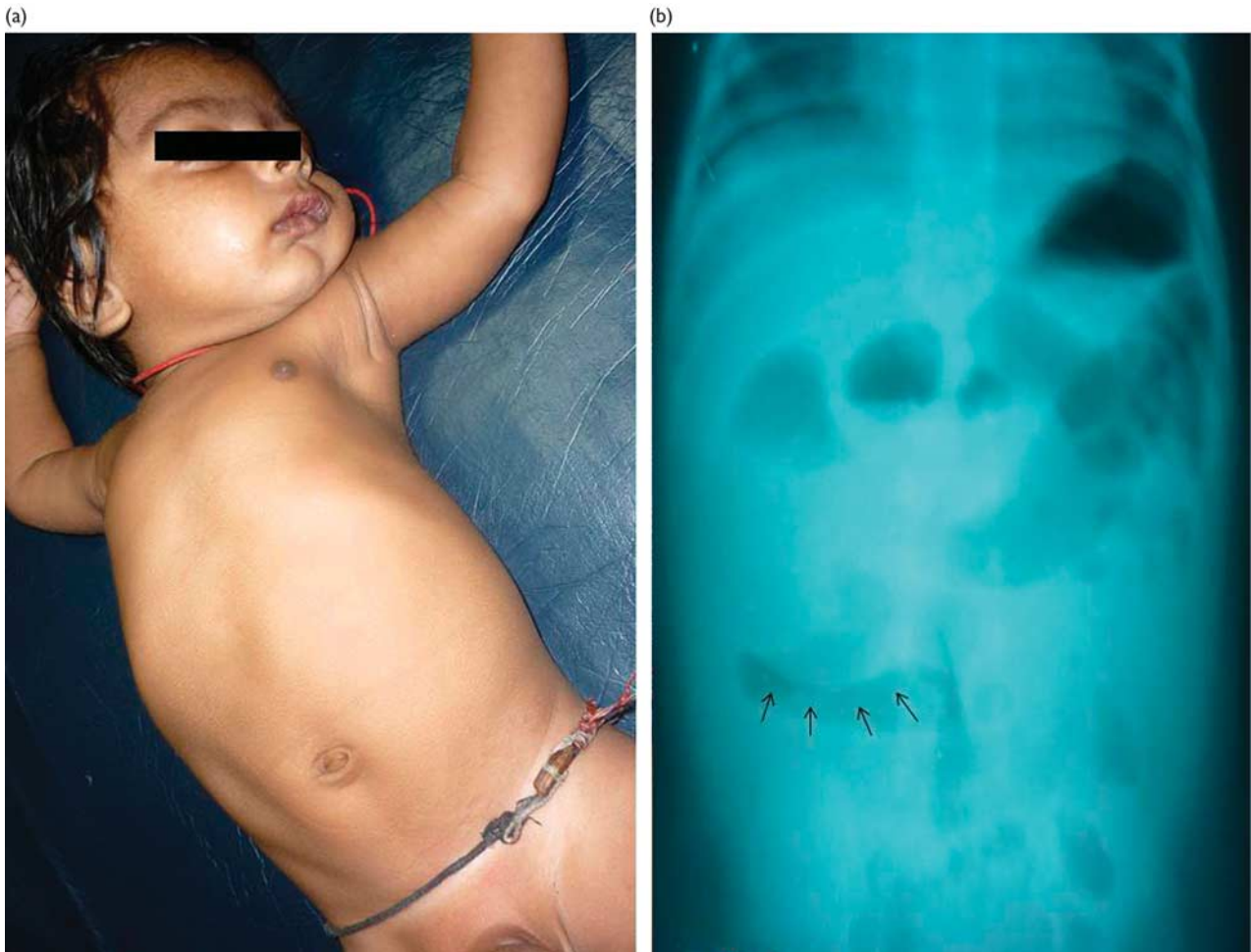
Discussion

Enteric DCs are frequently located around the jejunoileal mesentery. The esophagus and colon are other rare sites, whereas rectal, duodenal, gastric, thoracoabdominal, and cecal DCs are extremely rare [7]. Colonic duplications contribute to only 13% of all DCs [8]. Among these, cecal DCs are the rarest congenital anomalies [1].

The 'aberrant luminal recanalization theory' adequately explains the development of cecal DCs where persistence of a vacuole during the recanalization of the 'solid stage' of the gut can result in a DC in the submucosal plane. This theory holds true for DCs of the esophagus, small bowel, and colon as they also pass through the 'solid stage' during their development. However, it fails to explain DCs at other sites [9].

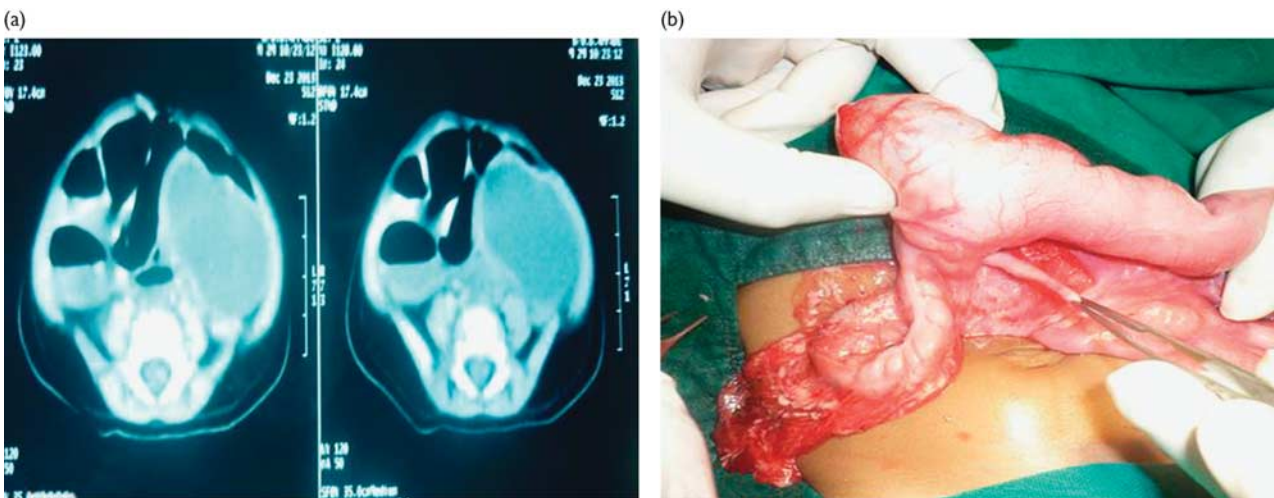
Classically, DCs lie and grow in the mesentery, and share a common muscle wall and blood supply with the

Fig. 1



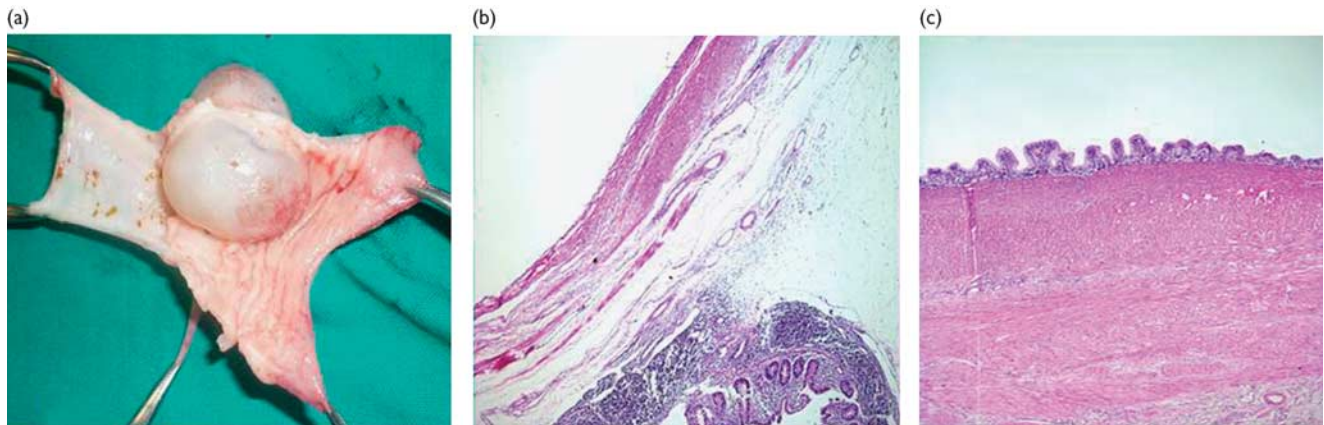
(a) Clinical photograph of an infant showing mild left-sided distension. (b) Erect abdominal radiography showing a round 'crescent-shaped radio-opaque shadow' in the distended terminal ileal loop (Renu's sign marked by black arrows).

Fig. 2



(a) Contrast-enhanced computed tomography image showing a partially enhancing cystic lesion with intermittent wall attenuation in continuation with the bowel wall in the left iliac fossa, which suggested a duplication cyst of the sigmoid colon. (b) Intraoperative photograph showing a freely mobile mass with a grossly dilated ileum, and a malfixed and collapsed ascending colon. The cecum was grossly enlarged with a luminal cystic mass, causing complete ileocecal junction obstruction.

Fig. 3



(a) Gross specimen showing an intact tense luminal submucosal cystic mass, arising from the mesenteric side of the cecum and extending into the dilated ileum. The cyst was filled with whitish mucoid material. (b) Microscopy of the cyst indicated denuded colonic epithelium and two distinct muscle layers with focal attenuation suggestive of an enteric duplication cyst. (c) Photomicrograph of the cyst wall showing well-formed colonic mucosa and distinct musculature on the representative section.

adjacent bowel. In the case of cecal DC, the cyst lies in the cecal wall usually on the mesenteric side and grows submucosally toward the lumen (area of least resistance) and is filled by the pent up mucoid secretions and causes ileocecal obstruction [8]. Such recurrent episodes can occur because of the initial slow enlargement of the cyst sufficiently to just obstruct the ileocecal valve, and then the ileum overcoming the obstruction by vigorous peristalsis to settle the obstruction, but only to recur again [5]. Several such episodes of subacute obstruction with vigorous peristalsis can leave the ileum significantly dilated. Once the cyst completely fills up the cecum, with no further room for distension, it culminates in an acute intestinal obstruction [5].

In contrast to the other reported cases, the absence of gross abdominal distension in our case made the mobile lump palpable. Unlike this case, most cystic DCs are preoperatively diagnosed as Intussusceptions. However, in this case, the cystic nature observed on preoperative ultrasound and partially enhancing cyst walls on the CT scan led us to consider DC as a remote differential diagnosis.

The history of recurrent episodes of subacute intestinal obstruction in our case did suggest Hirschsprung's disease, but a palpable intra-abdominal mass helped us rule this out. However, in the case of recurrent intestinal obstructions in the presence of a palpable lump, the most probable diagnosis should be recurrent intussusceptions, but this was ruled out by sonogram. Cecal DCs are known to cause recurrent intestinal obstruction with or without a palpable mass [5]. Abdominal radiography showed a round radio-opaque shadow within a dilated ileal loop (Renu's sign) in the right ileac fossa, which led us to suspect a luminal cyst [5], but extreme mobility of the mass kept us in dilemma. Because of extreme mobility and a thin cyst wall, ultrasound scan misinterpreted the cecal DC as a mesenteric cyst, further confusing the diagnosis. Computed tomography scan showed a partially enhancing thin-walled cyst in the left lumbar region which suggested a sigmoid DC leading to further dilemma.

A correct preoperative diagnosis of enteric DCs is made only in less than 25% of cases [4]. Preoperative abdominal radiography in cecal DC may show a 'round' or a 'crescent'-shaped radio-opaque shadow in a dilated terminal small bowel loop in the right iliac fossa (Renu's sign) [5], but in the presence of malrotation, the site may vary. The Renu's sign rounded or crescent-shaped radio-opaque shadow in a dilated terminal small bowel loop suggests a luminal cystic lesion; especially in the presence of ileocecal obstruction in infants, this is suggestive of a 'cecal DC', as it was in this case [5]. Ultrasound scan has shown a cystic lesion in the left pelvis instead of the right side as the right colon was not fixed in our case. Contrast-enhanced computed tomography in our case suggested a partially enhancing cystic lesion in continuity with the bowel in the left iliac region contrary to the abdominal radiography findings. This suggested a sigmoid DC; hence, cecal DC associated with malrotation can be misdiagnosed as sigmoid DC. Cecal DC may not show a 'classically well enhancing cyst wall' all around like other classical enteric DCs because of the thin cyst wall, especially on the luminal (submucosal) aspect, as the cyst wall and mucosa become thinned out because of pressure atrophy and the muscle layer may also become atrophied. The mucosa may be eroded and musculature may be attenuated enough to cause perforation.

The differentials for a highly mobile intra-abdominal lump in males are a ptotic kidney, a mesenteric cyst, and an omental cyst. However, enteric DC have limited mobility. Rarely DC in the sigmoid mesentery can be highly mobile because of the redundant mesosigmoid. However, cecal DC, when associated with a malfixed right colon as in this case, can present with a highly mobile intra-abdominal lump resembling an 'abdominal mouse'.

Unlike tubular variants of DC, cystic DC lack luminal communication with the adjacent bowel; hence present early with their mass effect and cecal DC located in the lumen can present very early in the infancy and majority like this case present during first 3 months of life [5]. Therefore, DCs represent an important differential

diagnosis for a palpable lump in young infants. Cecal DC can have a bizarre presentation such as a disappearing intra-abdominal mass [4], recurrent intestinal obstruction [5], and, when associated with a malfixed right colon, can present with a 'wandering intra-abdominal lump' resembling an 'abdominal mouse'.

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Conflicts of interest

There are no conflicts of interest.

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