

Obstructed jejunal duplication cyst in an infant

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SUMMARY

Intestinal obstruction in infancy due to duplication cyst is rare. This is a case of a three-month old boy presented to the hospital with symptoms and signs of intermittent intestinal obstruction for three-week duration. Investigation with ultrasound revealed a small bowel duplication cyst. Patient underwent successful segmental jejunal resection and made an uneventful recovery. He made significant weight gain at one-year follow-up. The diagnostic approach to infant with intestinal obstruction is described with special emphasis on ultrasonographic features of jejunal duplication cyst.

INTRODUCTION

Enteric duplication cysts (EDCs) are rare congenital malformation of the gastrointestinal tract (GIT). The incidence of EDCs is reported to be 1 in every 4,500 live births.^{1,2} The pathophysiology of EDCs remained unclear. It is a challenge to the medical practitioner when dealing with an infant with duplication cyst because of the intermittent nature of the disease. With the wide-spread use of ultrasound in the diagnostic work-up of infants with symptoms of intestinal obstruction, the diagnosis of duplication cyst can be made with high degree of accuracy. The purpose of this case report was to describe the ultrasonographic features of duplication cyst in an infant and the importance of US as a diagnostic tool in this clinical scenario.

CASE REPORT

A three-month old boy from Indonesia presented to Hospital Lam Wah Ee, Penang with three weeks history of bilious intermittent vomiting. The patient was able to tolerate small and frequent feeds with total of 100mls per day. There was no significant past medical history. Pertinent physical examination revealed the infant to be small for age with weight of 4.8kg (5th centile). Abdominal examination revealed a sausage-like mass in the right iliac fossa (RIF). Examinations of other systems were unremarkable. Blood tests were essentially normal. A working diagnosis of intestinal obstruction was made. An initial ultrasound examination revealed a cystic mass in the RIF. The cystic mass showed 'gut signature sign'^{2,3} with multiple alternating hyperechoic and hypoechoic layers in its wall. The cystic mass was adjacent to a loop of dilated fluid-filled jejunum with its contents 'yo-yoing' back and forth on real time ultrasound (Figure 1a & 1b). Subsequent upper GIT barium study showed near complete obstruction at proximal jejunum. An oval shape soft tissue density noted at the level of obstruction with 'coil-spring' appearance of the dilated proximal segment (Figure 2a). With the clinical history,

physical examination, ultrasound and barium findings, a diagnosis of jejunal duplication cyst was made. Patient underwent an emergency laparotomy. Operative findings showed a grossly distended segment of proximal jejunum due to obstruction by a cyst with collapsed distal segment (Figure 2b). A cyst measuring 3x2.5cm was noted at the mesenteric border of the jejunum. Segmental resection of the jejunum containing the cyst with end-to-end anastomosis performed. Post operation recovery was uneventful and the patient was discharged on the 7th post-operative day. Patient remained asymptomatic throughout the follow-up period. His last visit was 1 year 2 months later with his weight at 9.7kg (10-25th centile). The gross pathological specimen revealed a cyst at the wall of the jejunum protruding into the lumen. Microscopic examination revealed the cyst with small intestine mucosal lining. It is separated from the small bowel mucosa by a band of muscle fibres with no direct communication between them. The final diagnosis was jejunal duplication cyst.

DISCUSSION

Enteric duplication cysts (EDCs) is a group of heterogeneous clinical entity with varied clinical presentations due to the size, location, type and mucosal pattern of the cysts. They are thought to occur between the 4th and 8th week of embryonic development.² The aetiology of EDCs remained unclear. Duplication cyst in the ileum (33%) is the most common, followed by oesophagus (20%), colon (13%), jejunum (10%), stomach (7%) and duodenum (5%).^{1,2} Most cases of EDCs were diagnosed when complications like obstruction, bleeding or perforation occurred,^{3,4} as in this case.

When confronted with an infant with intestinal obstruction, ultrasound should always be done first. Ultrasonography is a simple, easy, fast, portable and non-invasive examination. More importantly ultrasound does not involve ionising radiation. The importance of no ionizing radiation involved in ultrasound should be emphasized as children especially infants are extremely sensitive to the long-term effects of radiation, hence the use of diagnostic radiation on infants should be kept to the minimum.

Much information can be gleaned from this examination, especially when a mass was palpable in the abdomen. The presence of a cyst with 'gut signature sign' in this case, instead of the 'doughnut' or 'pseudo-kidney' signs practically exclude intussusception. To establish the diagnosis of duplication cyst, there must be presence of mucosa with smooth muscle coat and it must be attached to the GIT.^{3,5} On high-frequency ultrasound imaging, the gastrointestinal wall

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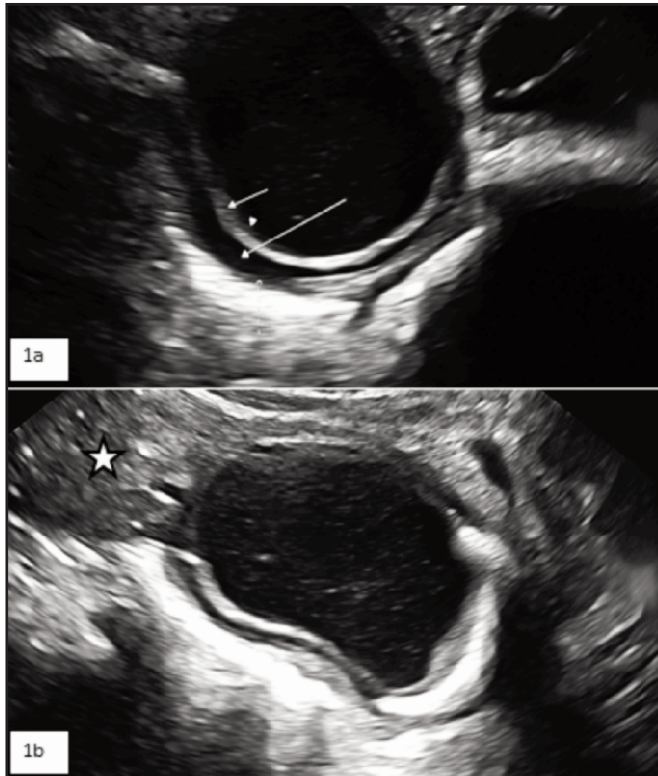


Fig. 1: a) This ultrasound picture of the cyst showed the ultrasonographic ‘gut signature sign’ depicting from inner to outer, mucosa layer (arrow head), the echogenic submucosa layer (short arrow), hypoechoic muscularis propria layer (long arrow) and the serosa layer (broken arrow).
 b) A dilated loop of bowel (star) is seen in close relationship with the ‘gut signature sign’ on this US picture.

can be divided into 5-layer of alternating echogenic and hypoechoic layers. The inner most layer is a thin interface between the content of the lumen and the mucosa. This is followed by the hypoechoic muscularis layer. The 3rd layer is the echogenic submucosal layer which consists of fat and collagen. The 4th layer is the hypoechoic muscularis propria and the 5th or the outer most layer is the echogenic serosa.^{1,2,4} In this case, the inner most interface between the content of the lumen and mucosa was not depicted (Figure 1a & 1b), likely because a convex probe with lower frequency was used. With ‘gut signature sign’ and the presence of adjacent dilated fluid-filled loops of bowel (Figure 1a & 1b) made the diagnosis of duplication cyst almost certain. Other cystic lesions in the abdomen like omental cyst or mesenteric cyst do not demonstrate this characteristic multi-layered wall. At this juncture, by using ultrasound we were able to diagnose duplication cyst but not absolutely certain about the level of obstruction, hence the upper GIT barium study. It should be reiterated here that upper GIT study is not necessary if ultrasound alone or ultrasound with a simple

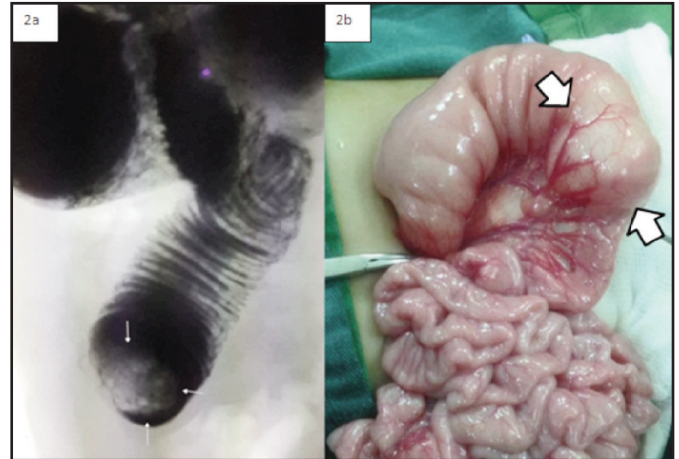


Fig. 2: a) Barium study demonstrated a filling defect (arrows) within the lumen of the jejunum with ‘coil-spring’ appearance of the dilated proximal segment.
 b) Operative findings of a cyst at the mesenteric border of the jejunum (arrows) with dilated proximal segment and collapsed distal segment.

abdominal radiograph are able to diagnose duplication cyst together with the level of obstruction, so as to minimise the radiation burden to the infant. The use of barium in this case was unconventional as most paediatric centres are moving towards water-soluble contrast medium.

In conclusion, the use of ultrasound as the investigation of choice and the recognition of the ‘gut signature sign’ may provide a fast diagnostic approach in an infant with obstructed duplication cyst so as to prevent the potential catastrophic complications.

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