An unexpected gangrenous duplication of ileum

Chiar Churn Inn, MD[1], Sivaneswaran Lechmiannadan, MS[1], Elango Thambusamy, MBBS[1], Umasangar Ramasamy, M.Med (Surg) USM[1], Mohan Nallusamy, FRCS[2]

[1]Department of Surgery, Hospital Taiping, Perak, [2]Paediatric Surgery, Department of Surgery, Hospital Sultanah Bahiyah, Alor Setar, Kedah

SUMMARY

Alimentary tract duplication is a rare congenital anomaly which may involve any part of the alimentary tract extending from stomach to rectum. Clinical presentation may mimic an inflamed appendix as described in this case. A 9-year-old boy with a clinical diagnosis of perforated appendix was noted to have a normal appendix intra-operatively. On further search for an underlying pathology, a gangrenous ileal duplication was discovered. En-bloc resection with primary bowel anastomosis was done. Histopathology report revealed a gangrenous small bowel duplication. We discuss the preoperative diagnostic dilemma and management options in approaching this rare entity.

KEY WORDS:
Duplication; Ileum; Gangrene; Perforated Appendicitis

INTRODUCTION

Alimentary tract duplication is a rare congenital anomaly which may involve gastrointestinal tract from oesophagus to rectum.[1]Aetiology is always related to embryogenic development.1 Clinical presentation varies from asymptomatic to obstruction, bleeding, perforation, gangrene or malignant changes.[1] Classification based on location, morphology (cystic or tubular)1 and blood supply pattern (type I and II)[2] has been proposed. Advance in imaging techniques have enabled early detection of asymptomatic duplications.[3] Recent reports suggest prevention of complication by early elective excision upon incidental radiological diagnosis.[4] Laparoscopic approach is equally successful as open surgery in uncomplicated cases.[4] Resection of the duplication alone is preferred whenever possible, rather than en-bloc resection with bowel anastomosis.[1] Prognosis of surgical resection is excellent especially in the uncomplicated elective setting. Here, we report a paediatric case of unexpected gangrenous ileal duplication with an initial diagnosis of perforated appendix with sepsis.

CASE REPORT

A 9-year-old Malay boy presented with three days history of right iliac fossa pain associated with fever and anorexia. There were no symptoms of urinary tract infection and trauma. There was no family history of inherited congenital abnormality. Clinically, he was febrile, mildly dehydrated and tachycardic. Systemic review was unremarkable. Physical examination revealed an unwell child with a tender, guarded right iliac fossa and localised peritonism signs. Blood counts revealed leukocytosis of 18.9 x 10^9 with predominant relative neutrophilia of 70%. He was adequately fluid resuscitated with crystalloids. Intravenous analgesia and a third-generation cephaolosporin were commenced.

A positive Mcburney's sign in a febrile and anorexic child with leukocytosis was suggestive of a perforated appendicitis. Thus, he was planned for open appendicectomy on the day of admission. Intra-operatively, a normal appendix was found. Further search for the underlying cause by medial extension of Lanz inscision was performed. Approximately 20 cm from ileocaecal junction, a 5 cm unhealthy gangrenous cystic duplication of ileum with its own mesentry was found twisted (Figure 1). There was no bleeding, perforation or peritoneal contamination. A 5 cm segment of ileum contiguous with the gangrenous duplication was resected and subsequent primary end to end anastomosis and appendicectomy was performed.

Histopathological examination demonstrated unilocular small intestine duplication cyst intimately associated with adjacent intestinal wall exhibiting extensive mucosal and transmural necrosis. There was no evidence of ectopic gastric, pancreatic mucosa or malignant changes within. The child was discharged well six days after surgery without complication.

DISCUSSION

In 1930, Ladd described duplication of the alimentary tract as a congenital lesion with the features of epithelial lining of gastrointestinal mucosa, presence of well-developed smooth muscle in the wall, gastrointestinal tract related. There is a male predominance.[3,5]

Embryologically, four theories have been postulated.[1] Partial twinning theory states that the alimentary organ may be double as a result of abnormal twining while split notochord theory states that endoderm may herniate through gaps formed when the notochord begins splitting. As described in canalisation theory, all hollow organs begin as solid organs which canalise later. However, some diverticula form in the foetal stage, which regress with development. External stress factors promote development of duplication as mentioned in the environmental factors theory. However, none of these has been scientifically proven as definitive contributing factors.

Common locations of duplication include ileum 30%, ileocecal valve 30%, duodenum 10%, stomach 8%, jejunum 8%, colon 7%, rectum 5%. Other than anatomy location,

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Corresponding Author: Chiar Churn Inn, 1-5-6, Kuarters Kakitangan Hospital Taiping, Jalan Stesen, 34000 Taiping, Perak.
Email: chiarchurninn@gmail.com

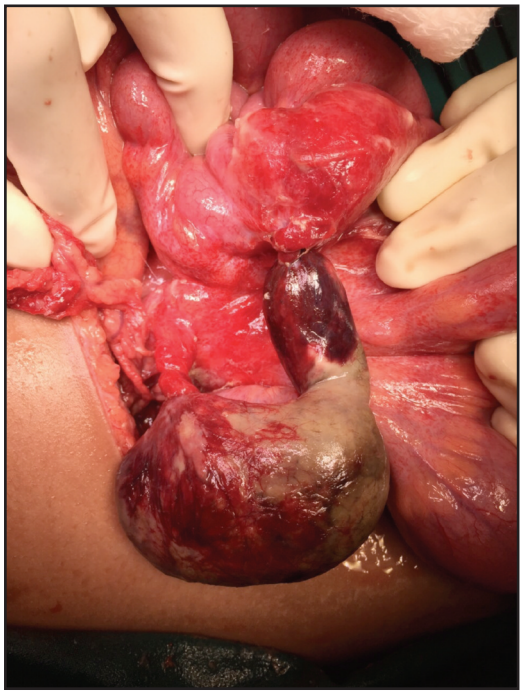

Fig. 1: Shows a gangrenous duplication ileal cyst with its own mesentery and blood supply twisted in our patient.

nomenclature based on vascular supply has been proposed as described later in management part. Morphologically, duplication can either be cystic (82%) or tubular which is more frequent in colon than small intestine. Tubular duplications may communicate with the intestinal lumen at multiple points.[1]

The presentation of duplicated small bowel varies, ranging from asymptomatic to simple inflammation, gangrene or complicated conditions such as obstruction and perforation. Rare presentation such as anorectal malformations and hydrocele has been reported as well. Duplication arising from the midgut may cause pain in right lower quadrant mimicking appendicitis[1] as seen in this patient. Another rare possibility is the presence of ectopic gastric mucosa within the duplication where the patient may present with abnormal gastrointestinal haemorrhage. In a study, among 11 patients with midgut and hindgut duplication, eight were discovered as incidental finding intra-operatively after an initial clinical diagnosis of acute appendicitis.[5] In contrast, the majority (67%) of foregut duplication was successfully diagnosed pre-operatively via imaging.[5]

A duplicated small bowel cyst mucosa which has undergone malignant transformation to adenocarcinoma has also been reported. Unfortunately, due to its rarity and non-specific symptoms, malignant duplicated cysts are found at a late stage with poor prognosis.

A preoperative diagnosis of duplication of the small intestine is not easy. With advancement in ultrasonography, pre-natal diagnosis was reported.[3] Ultrasonography may detect duplication cyst as an echogenic inner mucosal layer and hypoechoeic outer muscular layer. CT and MRI remained as non-first line imaging and only considered when confusion arises. In our patient, initial diagnosis was perforated appendix in sepsis as evidenced clinically by localised peritonism, tachycardia, dehydration and leukocytosis, Hence, he was wheeled in early for surgery without imaging to prevent delay in definitive operative intervention. However, the intra-operative finding of a normal appendix in a child warranted further search for other causes of localised septic peritonitis such as Merkel's diverticulitis, especially when pre-operative imaging is not done.

In terms of treatment, whenever possible, preservation of normal small bowel with resection of duplication is preferred. However, it might be extremely difficult as frequently, the duplication is intimately adherent to a common bowel wall, sharing a common blood supply. Thus, en-bloc resection can be performed followed by primary anastomosis as performed in our patient. Nevertheless, Long Li et al. [2] proposed the possibility of resection of duplicated structure alone, based on classification of vascular supply. The majority (76.6%) are the parallel type (Type 1), in which duplication is located on one of the mesenteric leaf. Thus, the straight arteries to duplication are separated from straight arteries supplying bowel. Whereas in type 2 (Intra-mesenteric type, 24.4%), duplication is found in between two mesenteric leaves and straight arteries pass over both the surfaces of the duplication to supply the normal bowel. Resection usually requires removal of the contiguous normal bowel segment. There is a role for elective resection of asymptomatic duplication to avoid complication, provided that the child is in optimal condition for surgery.[4] In fact, one of the six reported case in 2006 described a refusal of elective resection at diagnosis complicated with bowel perforation four months later.[4] Laparoscopic assisted resection has been reported as well in non-complicated elective cases.[4] However, there is lack of controlled studies suggesting quality superiority of laparoscopic approach compared to open. Thus, the methods of approach are always determined by the experience of the attending team.[3] Uncomplicated early elective intervention yields favourable outcome and prognosis in such cases.

CONCLUSION

Alimentary tract duplication is almost always found in paediatric age group, mimicking acute appendicitis. CT Imaging may not be readily available in some centres, and high resolution ultrasound may help in identifying a duplication in the hands of those aware of this condition Hence, a wide range of differential diagnosis such as Merkel's diverticulitis, torsion of greater omentum, torsion of ovarian cyst in females, ceacal diverticulitis, or duplicated alimentary tract as in this patient, should be considered. Intra-operatively, a normal appendix necessitates further search for an underlying cause.

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