Perforated Ileal Duplication Cyst Presenting with Right Iliac Fossa Pain Mimicking Perforated Appendicitis

M A R Islah, MS*, T Hafizan, MS**

*Department of Surgery, Hospital Universiti Kebangsaan Malaysia, Jalan Yaakub Latif, 56000 Cheras, Kuala Lumpur, **Department of Surgery, Hospital Kuala Lumpur, Jalan Pahang, 50586 Kuala Lumpur

SUMMARY

Enteric duplication is an uncommon malformation of the gastrointestinal tract which is either asymptomatic or presents with vague symptoms mimicking other more common pathology. It is most commonly diagnosed when complications such as bleeding, intestinal obstruction or perforation occurs. This is a case report of a patient with this condition presenting with right iliac fossa pain and localised peritonitis mimicking acute appendicitis.

KEY WORDS:
Perforated Ileal Duplication Cyst, Perforated Appendicitis

INTRODUCTION

Enteric duplication is a rare form of congenital malformation which varies in its presentation and presents a tough challenge to those involved in its management. In 1733, Calder described the first case of enteric duplication. Synonyms include enteric or enterogenous cyst, giant diverticula, ileum, jejunum, or colon duplex, and unusual Meckel's diverticula[1]. By definition, enteric duplication comprises a group of lesions that contains smooth muscle wall and enteric mucosa and are found commonly on the mesenteric border of the intestine[2].

These features differentiate it from Meckel's diverticula which are present at the anti mesenteric border, and from other intra-abdominal cyst which are lined by non-enteric lining. It applies to congenital malformations that involve the mesenteric side of the associated alimentary tract and share a common blood supply with the native bowel[1].

CASE REPORT

A fourteen year old Malay boy presented to us with a week history of burning epigastric pain associated with vomiting. He was previously diagnosed with gastro-esophageal reflux disease and was on regular omeprazole. On admission, he was febrile and the abdomen was tender and guarded. The white blood cell count was raised. There was no pneumoperitoneum detected on erect chest x-ray. Based on the clinical findings, a diagnosis of acute peritonitis was made and an exploratory laparotomy was planned.

However, while waiting for surgery, the pain localised to the right iliac fossa. The diagnosis was changed to appendicitis and appendicectomy was planned. At operation, we found 500cc of pus within the peritoneal cavity. The appendix was inflamed but not perforated. Other pathological causes were sought after. On further inspection, we found a perforated duplication cyst at the mesenteric border of the intestine, about 90cm from the ileocaecal junction (Fig 1, Fig 2). Segmental bowel resection and primary anastomosis was performed.

The patient's post-operative recovery was uneventful, with the white cell count returning to normal. Patient was discharged three days after the operation. The histopathological report described all the characteristics of enteric duplication with heterotopic gastric mucosa. Patient is still under follow-up in our surgical outpatient clinic.

DISCUSSION

Enteric duplication is a rare cause of an acute abdomen. It may be asymptomatic, or present with vague non-specific symptoms or symptoms of complications. It is difficult to diagnose pre-operatively as symptoms are non-specific and usually mimic other more common causes of acute abdomen. It should be considered especially in the young patient presenting with vague symptoms of acute abdomen.

Enteric duplication can arise at any site along the entire length of the alimentary tract. It can be divided into 2 types; a) communicating and b) non-communicating. The exact etiology of enteric duplication is unknown, however, various theories have been postulated. Abortive attempts of twinning, phylogenetic reversion, adhesions between endoderm and neuroectoderm, persistence of embryonic diverticula, and recanalisation and fusion of longitudinal folds have all been blamed as the origin of this rare congenital anomaly.

Enteric duplication can present with vague abdominal pain such as epigastric pain which usually leads to the diagnosis of gastritis, peptic ulcer disease or GERD by most clinicians. This may be due to co-existing diseases or may be due to the inflammation of heterotropic mucosa which is a common association and can be present in up to 30% of the cases[2]. However, most patients present with symptoms and signs of complications such as gastro-intestinal bleeding, obstruction or perforation.

Radiological imaging such as transabdominal or endoscopic ultrasonography and computed tomography may help in the diagnosis, especially in cases with non-acute presentation.

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Corresponding Author: Islah Bin Ab Rashid, Department of Surgery, Hospital Universiti Kebangsaan Malaysia, Jalan Yaakub Latif, 56000 Cheras, Kuala Lumpur Email: islah75@hotmail.com

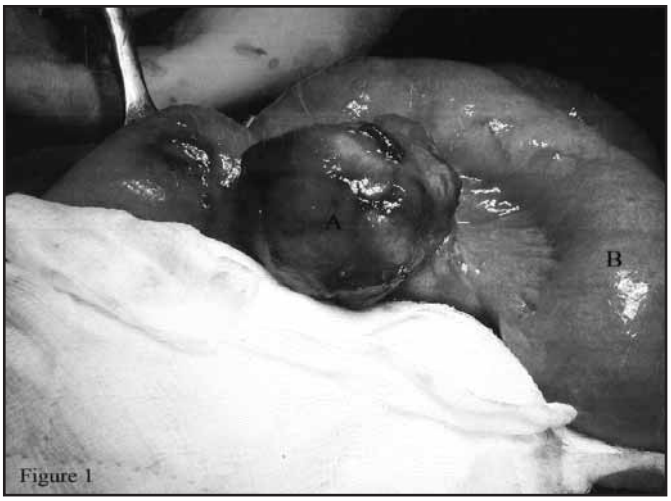

Fig. 1: Intraoperative finding of the duplication cyst
A) Duplication cyst.
B) Ileum

Fig. 2: Cut section of the ileum showing the communication between the duplication and the ileum
A) Duplication cyst.
B) Communication between the duplication cyst and the ileum

Endoscopic ultrasonography has the advantage of providing additional diagnostic information which is extremely helpful in the proper management of the disease. Unfortunately, it was not done in our case as the patient already had acute complications which required emergency surgical intervention.

Enteric duplication presenting as acute appendicitis is even rarer. There are only five cases reported in the literature up till the year 2000 including only one adult case. In the case described, the patient had been under follow up for gastroesophageal reflux disease. He was prescribed omeprazole after OGDS was performed. The duplication cyst in this case may have been the source of the patient's on going epigastric pain.

Considering the past history and patient's presentation, which was burning epigastric pain and fever with signs of peritonitis, a diagnosis of perforated gastric ulcer was made. However, the migratory pain was due to the perforated cyst with abscess formation. Appendicectomy through a Lanz's incision was performed.

Enteric duplication can be treated surgically by simple excision or by dissecting the common wall between the intestine and the cyst. Alternatively, selective mucosal resection will be helpful in patient with a long segment of enteric duplication as it will preserve the common shared blood supply to the native bowel[3]. Other methods such as marsupialization has also been reported[3]. In our patient, a segmental resection of the bowel with primary anastomosis was performed as the cyst had already ruptured and the surrounding bowel was unhealthy. The patient recovered well post operatively without any complications.

In conclusion, enteric duplication is a rarely encountered condition. It usually presents with vague symptoms suggestive of other pathology, making diagnosis difficult. It should be a differential diagnosis in patients presenting with right iliac fossa pain.

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