ORIGINAL ARTICLE

VARIETY OF GASTROINTESTINAL DUPLICATIONS IN CHILDREN: EXPERIENCE AT TERTIARY CARE HOSPITAL

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Background: Gastrointestinal duplication is a rare developmental anomaly that can be present anywhere along the GI tract, most often being found in ileum. The purpose of this study is to share our experience in evaluation of the presentation, investigations, management challenges and complications of patients with this very rare condition. **Methods:** This descriptive case series was conducted at the Department of Paediatric Surgery, National Institute of Child Health Karachi, Pakistan, from April 2018 to October 2019. Data was analysed with regard to age, clinical presentation, investigations, surgical procedures, site and type of lesion, histopathology, complications and outcomes. **Results:** A total of five patients were managed in one and half year. The patients' ages ranged from antenatally diagnosed foetus to 12 years old child. New-born who presented with antenatal diagnosis of abdominal cyst turned out to have duodenal duplication cyst. Among other four were thoracoabdominal duplication cyst, gastric duplication, jejunal duplication and ileal duplication, last two presented with perforation. Other presentations were abdominal pain, swelling and vomiting. Diagnosis was made on clinical ground, x-ray of abdomen, ultrasound and computed tomography. All cysts were resected successfully and patients remained asymptomatic till one year follow up except one patient who expired postoperatively due to liver failure. **Conclusion:** Enteric duplication can present in variety of ways depending on anatomical location. Prompt diagnosis and complete excision of cyst is the aim of treatment. However, these rare types of duplication are a challenge to operating surgeons.

Keywords: Gastric Duplication; Duodenal duplication; Ileal duplication; Thoracoabdominal Duplication; Perforation; Postoperative intussusception

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INTRODUCTION

Gastrointestinal (GI) duplication anomalies are rare and usually present within the first 2 years of life.[1] Duplication can be found anywhere but is most common in the Ileum (30%) followed by duodenum, stomach, jejunum, colon and rectum.[2] It can present in variety of ways, abdominal pain, mass, bleeding per rectum, or it may remain silent and incidentally diagnosed during surgery being performed for other reasons. It may also present acutely with peritonitis, intussusception, volvulus or intestinal obstruction. It can be occasionally non-communicating or communicates with the gut. There are two forms of duplications: Cystic duplications (75%) are more common than tubular duplications (25%).[3]

GI duplications pose diagnostic challenge due to varying presentation related to anatomical region of involvement and hence require high index of suspicion during diagnostic evaluation. Treatment options depend on clinical presentation, type of cyst and location of cyst. Aim of treatment is complete excision of the cyst with minimal resection of the involved segment of the gut, however partial excision and stripping of the residual mucosa are alternatives, especially when there is tubular variety or when cyst cannot be excised completely.[4]

As the disease is very rare, most of the published literature on enteric duplications is in the form of case reports, with only a few case series.[2,5,6] and two of these case series are from Pakistan[7,8]. This study was done to highlight the varied presentation of gastrointestinal duplications, their diagnostic challenges and outcomes.

MATERIAL AND METHODS

This descriptive case series was conducted at the Department of Paediatric Surgery, National Institute of Child Health Karachi, Pakistan, from April 2018 to October 2019. All the patients with diagnosis of gastrointestinal duplication who were managed during this period were included in the study prospectively. Written informed consent, both in English and native language Urdu, were obtained from parents for the use of patients' pictures and relevant data. Institutional ethical review board approval was taken. Data that was gathered consisted of variables including age, clinical presentation, diagnostic modalities used,

surgical procedures performed, types and site of duplication, histological findings and complications. Data was analysed using descriptive statistics.

RESULTS

A total of five patients with diagnosis of gastrointestinal duplication were managed during this one and half year and all were included in study prospectively that spanned from April 2018 to October 2019. All five patients had different sites of GI origin including gastric (n=1), duodenal (n=1), jejunal (n=1), ileal (n=1) and thoracoabdominal (n=1) duplication. Three patients were males and two were females (Male: Female = 1.5: 1). Age ranged from newborn to 12 years at presentation. Three out of five patients (60%) were less than one year old, one of them presented at birth with antenatally diagnosed cyst. Table-1 summarizes the age, sex, clinical presentation, site and type of duplication, surgical procedure, complications and outcomes.

In cases of jejunal and ileal duplication who presented with acute abdomen, Xray abdomen was done which showed pneumoperitoneum. In both the cases final diagnosis was made intraoperatively. The newborn with abdominal (duodenal) duplication was explored with initial impression of infantile choledochal cyst with obstructive symptoms as suggested by radiology whereas CT scan was diagnostic in other two cases (gastric and thoracoabdominal).

Two out of five (40%) duplications were cystic while thoracoabdominal tubular duplication had involvement of oesophagus and few centimetres of jejunum sparing proximal jejunal segment near DJ. In this case resection anastomosis of jejunal part was done in first stage due to initial presentation with abdominal symptoms and later investigated for oesophageal component due to intraoperative finding of presence of duplication at GEJ for which resection and mucosal stripping of common wall was done. The ileal duplication that presented with pneumoperitoneum also had tubular variety with communication at distal end of the segment where perforation was found, which underwent resection anastomosis as was done for jejunal tubular duplication.

Postoperatively the patient with gastric duplication developed small bowel intussusception on second postoperative day which was manually reduced. Patient operated for jejunal duplication developed anastomotic leakage for which redo anastomosis was done. Patient with ileal tubular duplication expired due to liver failure whose exact cause could not be known as this case was reported earlier.[18] Histopathology confirmed the diagnosis and all cases showed presence of ectopic gastric mucosa (100%). All four patients remained asymptomatic during 1 year follow up.

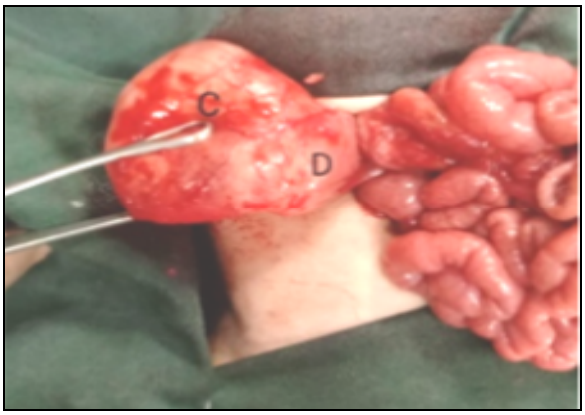

Figure-1: Intraoperative picture of duodenal duplication; c= cyst, d= duodenum

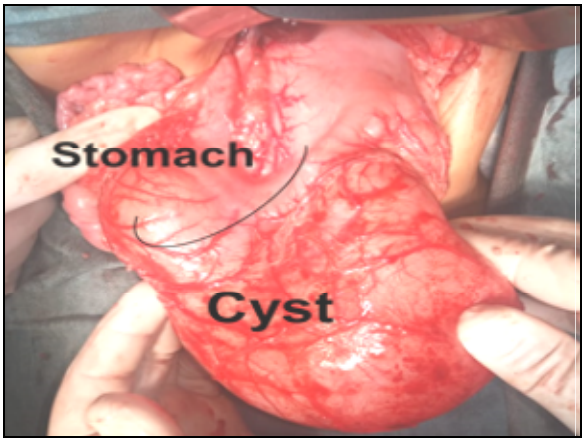

Figure-2: Intraoperative picture of gastric duplication

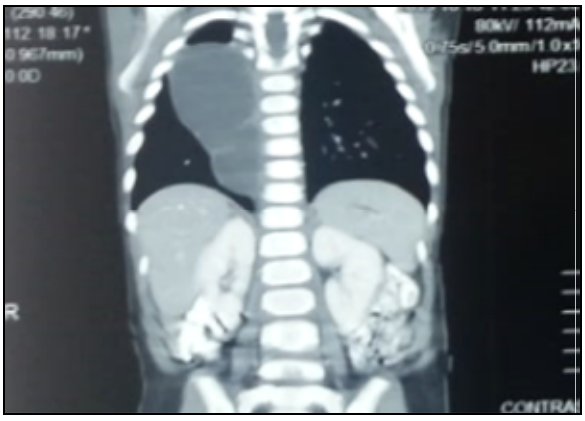

Figure-3: CT scan of patient with thoracoabdominal duplication

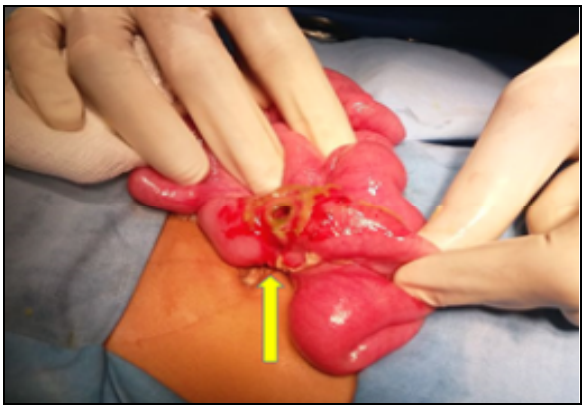

Figure-4: Intraoperative picture of ileal duplication showing perforation

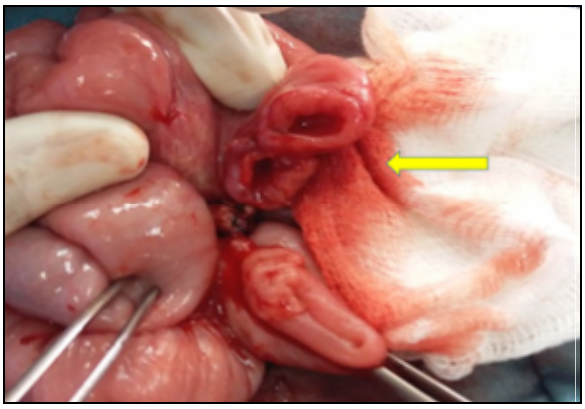

Figure-5: Intraoperative picture of ileal duplication showing two lumens upon resection

Table-1: Characteristics of patients (n=5)

Age/ Gender	Presentation	Investigation	Site	Type	Surgical procedure	Complication	Outcome
03 month/ male	Abdominal mass/ non-bilious vomiting	Ultrasound CT Scan	Stomach	Cystic	Complete cyst excision with mucosal stripping of common wall.	Postoperative intussusception (manually reduced)	Discharged
Newborn/ female	Antenatally diagnosed cyst in RUQ	Ultrasound CT Scan	Duodenum	Cystic	Partial excision and mucosal stripping of common wall	None	Discharged
12 year/ female	Pneumoperitoneum	Xray abdomen	Jejunum	Tubular	Resection & end to end anastomosis	Anastomotic leak /redo-anastomosis	Discharged
8 month/ male	Pneumoperitoneum	Xray abdomen	Ileum	Tubular	Resection & end to end anastomosis	liver failure	Expired
2 year/ male	Abdominal pain and recurrent RTI	CT scan	Thoraco-abdominal	Tubular	Staged excision	None	Discharged

DISCUSSION

Gastrointestinal duplications are rare anomalies of the alimentary canal presenting in childhood with an incidence of 1 in 100,000 cases per year.[9] Numerous theories have been developed however no single theory has been able to account for all variants. Split notochord theory is widely accepted as embryological origin of GI duplication. Other theories include partial twinning, Embryonic Diverticula and Recanalization Defect.[10]

Enteric duplication is defined by 3 Rowling's criteria: (1) the wall of the duplication is in continuity with one of the duplicated organs; (2) the duplication is surrounded by a smooth muscular layer; and (3) a layer of digestive mucosa is present, more often typical or heterotopic like gastric mucosa.[11,12] These may remain silent and incidentally discovered during a surgical procedure or can present at any time with GI symptoms related to site of origin. Similarly, our cases had broad range of presentation from antenatally diagnosed cyst with obstructive symptoms to abdominal pain and vomiting to acute abdomen with peritonitis.

Duplications are now more frequently discovered on antenatal scans owing to better antenatal care[13], one of our cases also had an antenatal diagnosis which caused obstructive symptoms postnatally and, with initial impression of infantile choledochal cyst, the patient underwent surgery where it was found to be a duodenal duplication confirmed on histopathology.

Histologically, most duplications have mucosal lining native to the lesion, but 25–30% specimens contain ectopic tissue[14] however all of our cases showed presence of ectopic mucosa. Heterotopic gastric mucosa secretes acid which often causes peptic ulcer disease, leading to perforation as seen in two of our case. Tubular duplication often communicates with the main intestine proximally or distally or at both ends as reported by G. Ekbote *et.al.*[15] Similarly, two of our cases, jejunal and ileal duplication, had communication with native gut distally. Perforation was found in mid segment of jejunal tubular duplication, while in case of ileal duplication perforation was found in distal end. Patient with Thoracoabdominal duplication had no communication with native gut. Ileum is found to be the most frequent site of duplication in literature.[16] However, we came across and managed duplications involving almost all the regions during this study period time and there are very scantly reported cases of duplication involving stomach and duodenum. Cases of small bowel duplication (jejunal and ileal)

were managed with resection & anastomosis of the gut, however near total cyst excision along with partial stripping of mucosa was done in cases of oesophageal, gastric and duodenal duplication to avoid injury to or severing the common blood supply of native bowel, this treatment option was consistent with the treatment strategies opted in literature worldwide.[16]

Postoperative intussusception is reported in literature after surgeries in retroperitoneal region, also there are reported cases of duplications presenting with intussusception[17] however there is no reported case of intussusception developing after surgery for gastric duplication which was found in one of our patients who underwent manual reduction of intussusception during same hospital admission. The patient with ileal duplication developed jaundice and altered sensorium on 4th Postoperative day with acute liver dysfunction. Despite aggressive ICU care and ventilator support, on 7th postoperative day, patient expired. No definitive cause could be identified.[18]

After discharge from hospital no recurrence of symptoms or other complications were found in our patients at follow ups conducted at 1 month, 3-month, 6 month and 1 year. Malignancy has also been reported in literature with a duplicated gut for which these patients need long term follow up till adulthood[19] and therefore we plan to follow these patients at regular interval of 3–5 years keeping high index of suspicion even in case of vague symptoms.

CONCLUSION

Enteric duplication can present in variety of ways depending on anatomical location. Prompt diagnosis and complete excision of cyst is the aim of treatment. However, these rare types of duplications pose diagnostic and therapeutic challenge to operating surgeons. They may be unexpectedly encountered intraoperatively, and appropriate surgical management requires that the attending surgeon be familiar with the pathology and clinical characteristics of GID hence one should be sharp enough to tailor procedure on merit of the case.

AUTHORS' CONTRIBUTION

SP: Study conception and design, analysis and data interpretation. SP, SA, SI: Data acquisition, critical revision. SA: Drafting of manuscript.

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