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GASTROINTESTINAL DUPLICATION CYST: A MASQUERADER

Pathology

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ABSTRACT

AIM: Gastrointestinal tract (GIT) duplications are rare congenital anomalies found in 0.2% of the children, most detected ante-natally or within first two years of life. We report three cases with an aim to study the clinical and histopathological findings.
METHODS: It is a retrospective analysis of the paediatric patients with gastrointestinal tract duplication over a period of five years. **RESULTS**: Two cases, 11 year and 4 month old males diagnosed as ileal duplication cyst presented with abdominal pain and respiratory distress respectively. Third case of a newborn female on prenatal ultrasonography showed CCAM, was eventually diagnosed as foregut duplication cyst on histopathology. All three cases were managed surgically. **CONCLUSION**: GIT duplications, although rare can arise anywhere along the GIT. Clinical scenario is diverse and mimics other intra-abdominal conditions thus presenting a great challenge to arrive at a diagnosis preoperatively. Therefore high index of suspicion and histopathology is must for the correct diagnosis and management.

KEYWORDS

Neonate, Gastrointestinal duplication cyst, Intestinal obstruction, Enteric, Esophageal, ileal duplication, foregut duplication, congenital anomaly, Cyst, Children

INTRODUCTION:
Incidence of gastrointestinal duplication is about 1 in 4500 live births and is found in 0.2% of the children. They can occur along the entire length of the gastrointestinal system and can present at any age but 80% of cases present within first two years and majority within first three months of life with antenatal diagnosis made in significant number of cases [1]. The duplications are classified according to their location, shape and presence of ectopic tissue within the duplication cysts. Most commonly in the ileum (33%), followed by the oesophagus (20%), colon (13%), jejunum (10%), stomach (7%) and duodenum (5%) [2]. Though there is no consensus on the exact embryological origin of enteric duplication cysts, the most widely accepted theory is the split notochord theory. As per Ladd, the term 'enteric duplication cyst' will be applied if a congenital lesion has (a) coat of smooth muscle, (b) Gastrointestinal type epithelial lining and (c) intimate anatomical location with some part of gastro-intestinal tract (GIT).

They can be classified into foregut, midgut and hindgut, depending on the site of origin [3]. During the first trimester, heterotopic rests of foregut-derived epithelium may persist, resulting in foregut duplication cysts, they are most frequently seen in the thorax or abdomen [4]. Esophageal duplications are the second cause of posterior mediastinal mass in children, after neuronal tumors, with a prevalence of approximately 1/22.500 live births [5]. The objective of this study is to report our 5 years of experience in histopathological diagnosis of gastrointestinal duplication cysts.

MATERIALS AND METHODS:
This study is a retrospective analysis of the paediatric patients who were diagnosed with gastrointestinal tract duplication over a period of five years. Data was retrieved from the medical records to obtain antenatal diagnosis, clinical details, diagnostic work-up, associated gut malformations, histopathological diagnosis and post-operative course of the cases.

Case summary:
Case 1 Eleven yrs male child with history of abdominal pain since 10 days. Past history of accidental pebble swallowing, USG suggestive of malrotation with impacted stone in meckel's diverticulum. *Gross* specimen showed 2 adhered loops of bowel measuring 9*4 and 5*3 cm with no communication. On opening, the ileal segment showed a 2cm impacted marble (Figure 1) with no evidence of mucosal ulceration or perforation. The adhered bowel loop was unremarkable. *Histopathological Diagnosis* : Ileal duplication cyst showing normal ileal mucosa (Figure 2).

Fig.1: Two adhered bowel loops with no communication and shows a impacted marble

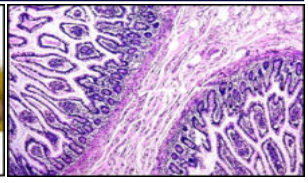

Fig.2: Ileal duplication cyst with normal ileal mucosa. (H&E,100x)

Case 2 Newborn female child, asymptomatic; antenatal USG suggestive of Congenital Cystic Adenomatoid Malformation (CCAM). *Gross* specimen showed 2 cystic structures, 3 cm & 4 cm in diameter with no communication (Figure 3). *Microscopy*: one cyst showed immature esophageal mucosa with underlying hypertrophied muscle and few ganglion cells while the other had lining of gastric mucosa with underlying longitudinal and circular muscle layers with nerve plexus (Figure 4&5). *Histopathological Diagnosis*: Foregut duplication cyst.

Fig.3: Two adhered loops with no communication.

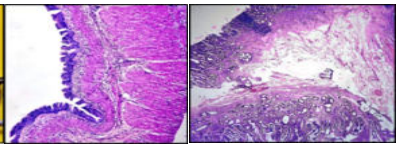

Fig. 4 & 5: Immature esophageal mucosa with hypertrophied muscle and gastric mucosa respectively. (H&E,40x)

Case 3 Four months male child presented with fever, cough and respiratory distress since birth. A right thoracic cyst was identified on USG and was resected. Gross specimen showed cyst measuring 7*5*1cm well defined, unilocular, smooth lining cyst. *Microscopy*: Cyst lined by small bowel mucosa followed by muscle and serosa. *Histopathological Diagnosis*: Ileal duplication cyst

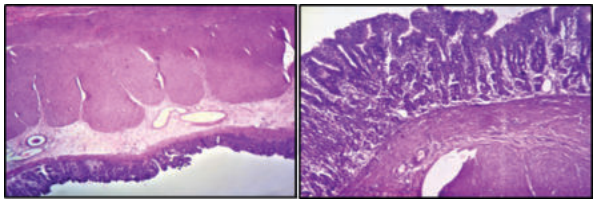

Fig. 6 & 7: Ileal duplication cyst with normal ileal mucosa. (H&E, 40x & 100x) respectively.

DISCUSSION:

Gastrointestinal duplications are the result of one or more congenital anomalies of uncertain etiology. They are generally saccular or tubular masses with presence of an intimate attachment to the GI tract, a layer of smooth muscle in the wall and an epithelial lining resembling some part of the GI tract. The cysts become incorporated into the bowel wall and share a common blood supply with the parent bowel [1]. Although most cases present within 2 years of life, our study had 1 case presenting at 11 years of age. The most common location reported in the literature is terminal ileum representing more than 70% of small GI duplications [1]. In our study 1 out of 3 cases the site was ileum while the other 2 cases presented as thoracic mass. The symptoms are non specific and vary from minor digestive problems to intestinal obstruction, gastrointestinal bleeding, or perforation [2]. In our study 2 out of 3 cases were symtomatic. Often they mimic other intra-abdominal conditions posing great diagnostic difficulty. Intestinal-type mucosa is found in most of the cases, and in 16–39% of the patients ectopic gastric or pancreatic cells are found. The mucosa should always be removed because of the malignant potential of ectopic tissue. Although no ectopic tissue was found in our cases [2]. Duplications can sometimes lead to complications which include perforation, intussusception, volvulus, and associated malignancy [1]. 23% of intestinal duplication cysts in adults were found to be ileal cancer [6]. No complications were found in our cases. Various congenital malformations are reported in 50% of patients, the most common being vertebral defects [3]. Although no such anomalies were found in our patients. Microscopically, gastrointestinal tract duplications contain smooth muscle in their walls and are lined with alimentary tract mucosa. The lining mucosa, however, is not necessarily that of the adjacent segment of the gastrointestinal tract [7]. Based on their epithelial type and other features, foregut duplication cysts may appear to closely resemble airway, esophagus, or small intestine. Therefore, the term *foregut duplication cyst* includes bronchogenic cyst, esophageal duplication cyst, and enteric duplication cyst [8]. USG is considered to be the first-choice imaging modality [9]. The aim of the intervention when possible should be total resection as partial excision is associated with high risk of recurrence [1]. All our cases had good prognosis with no recurrence. Thus apart from clinical history and radiology, histpathological examination is essential for definitive diagnosis.

CONCLUSION:

GIT duplications, albeit rare, are still not uncommon in the pediatric population. The clinical picture is quite diverse and often mimics other intra-abdominal conditions thus posing a great challenge to pediatric surgeons to arrive at clinical diagnosis preoperatively. Therefore high index of suspicion followed by histopathologic evaluation is imperative for accurate diagnosis and management.

Abbreviations:

GIT= Gastrointestinal tract, CCAM = Congenital Cystic Adenomatoid Malformation, etc = et cetera.

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