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Enteric duplication in children: A case series

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ABSTRACT

Background: Enteric duplication (ED) cysts include a wide variety of cystic lesions, which can involve any part of the gastrointestinal tract (GIT). They can be referred to foregut, midgut, hindgut derived, depending upon the portion of GIT involved. The main purpose of this study was to document the variety of presentation, investigations, and treatment options employed. **Patients and Methods:** This was a retrospective study at Paediatric Surgery Department of Services Hospital, Lahore from August, 2011 to August, 2013. The details of all the patients, including gender, and age, presenting complaint, abdominal examination findings, diagnostic modality, site, type, associated malformations, surgical option, and outcome were analysed. **Results:** A total of eight patients with histopathological diagnosis of EDs managed were included in the study. Of these eight patients, six were males and two were females, with an average age of 2.4 years. The main diagnostic tool used was ultrasound in almost all patients. The most commonly involved site was ileum in 5 of 8 (62.5%) patients. Two cases had gastric involvement, while one patient had involvement of the descending colon. Six of eight were of tubular type (75%), while remaining 2 (25%) were of cystic type. Excision was possible in all these patients. **Conclusion:** ED can present with a wide spectrum of symptomatology. It can present as mass abdomen, intestinal obstruction or even can mimic as hydrocoele. High index of suspicion is therefore required. Ultimate aim of treatment is excision of cyst with preservation of vascularity of native gut.

Key words: Duplication cyst, enteric, hydrocoele, intestine, mass abdomen

INTRODUCTION

Enteric duplications (EDs) are uncommon congenital anomalies, which can involve any part of the gastrointestinal tract (GIT). The most common site encountered is the small intestine, which is involved in 50% of cases, two-third of which are present in ileum.[1] EDs of all parts of GIT have been reported in the literature, but interestingly its presentation varies from case to case. It may present with abdominal pain, mass, bleeding per rectum, haematuria or it may remain silent and incidentally diagnosed during surgery being performed for other medical reasons.[2] It may also present acutely with intussusception, volvulus or intestinal obstruction. Abdominal examination may give a clue for duplication cyst, but it may not be palpable in 50% of cases. The main diagnostic tools for its diagnosis are ultrasound, computed tomography (CT) and magnetic resonance imaging.[3]

This variability in its presentation, pathology, diagnostic investigations and treatment options led us to perform this study and look for EDs in our population of patients.

PATIENTS AND METHODS

This retrospective descriptive study was conducted at Paediatric Surgery Department of Services Hospital, Lahore, from August, 2011 to August, 2013 in which all patients with histological diagnosis of EDs were included after getting Ethical Committee approval. The demographic details of all the patients including gender and age at presentation were noted. The chief complaint, abdominal examination findings, diagnostic modality, site of EDs, type, associated malformations, surgical option, and outcome were noted and mentioned in the proforma. Results were analysed by descriptive statistics.

RESULTS

A total of eight patients with histopathological diagnosis of EDs were included in the study. Of these eight patients, six were male and two were female; their average age was 2.4 years. All the patients except one

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were below 4 years of age and two were infants. Two of these patients presented with intestinal obstruction and two with abdominal mass. One of the patients presented with intussusception, while one patient presented as hydrocoele. All the details are summarised in Table 1.

Associated malformations were present in only one of these patients who had high variety of imperforate anus. Main diagnostic tool used was ultrasonography (USG) in almost all the patients in this study, while CT was also used in two of the patients. The most commonly involved site was ileum in 5 of 8 (62.5%) patients. Two cases had gastric involvement, while one patient had involvement of the descending colon. Six of eight were of tubular type (75%), while remaining 2 (25%) were of cystic type. All the patients underwent surgical excision, while partial excision and mucosal stripping was done in one of them. In 3 of 8 patients, it was communicating with native gut (37.5%), while in remaining, it was present as an isolated cyst (62.5%). In 3 of 8 patients, native gut had to be excised along with the duplication cyst. Outcome was satisfactory in all the patients except one patient who developed adhesive bowel disease and had to be re-explored.

DISCUSSION

Enteric duplications are uncommon congenital anomalies with incidence of 1 in 4500 autopsy series. They mostly present in paediatric age group.[4] The first case of duplication cyst was reported by Calder in 1733.[5] Previously, EDs had been called as giant diverticula, enterogenous cysts, giant thoracis cysts, duplication and unusual Meckle's diverticula.[6] According to Ladd, term 'ED' will be applied if a congenital lesion has (1) coat of smooth muscle, (2) GI type epithelial lining and (3) intimate anatomical location with some part of GIT. EDs can arise from any part of GIT from oropharynx to anus. They can be classified into foregut, midgut and hindgut, depending on the site of origin.[7]

Enteric duplications are present in both genders with slight male predominance.[8] Same findings were noted in this study where 6 out of 8 patients (75%) were male. The majority of EDs are diagnosed before 2 years of life. In our study, two patients were infants and 5 of 8 (62.5%) were of age <2 years, but all were below 7 years of age.

The presentation of EDs varies greatly and usually is confused with other GIT pathologies. Sometimes its presentation is so dramatic and really confusing as in one of our patients who presented with hydrocele, which was explored for herniotomy and during surgery the contents of sac gave us a suspicion. The patient was investigated and re-explored for excision of duplication cyst of ileum [Figure 1]. It is one of very rare presentation of EDs and only two cases have been reported in literature of this type of presentation.[9] In two of our patients, it presented in emergency department as intestinal obstruction and one patient presented with intussusception [Figure 2]. One of our patient with gastric duplication, presented with hematemesis and that patient had been under treatment of paediatric gastroenterologist for gastroesophageal reflux disease, which later on turned to be ED. In another patient in our series, duplication of the descending colon was accidentally discovered, while performing colostomy for anorectal malformation. During colostomy, incidentally ED of the descending colon was found. Two of eight patients in our series were of foregut origin who presented with upper GI symptoms. Remaining six patients were midgut derived who presented with abdominal, inguinal and lower GI symptoms.

Abdominal examination in case of EDs may be unremarkable, but a mobile mass may be palpable in

Table 1: Details of all cases

Patient	Gender	Age	Presentation	Abdominal examination	Associated malformation	Investigation	Site	Type	Communication with native gut	Surgery	Outcome
1	Female	8 months	Intestinal obstruction	Peritonitis		USG	Ileum	Tubular	Yes	Excision	Adhesive bowel disease
2	Male	3 years	Hydrocele	Non-specific		USG, CT	Ileum	Cystic	No	Excision	None
3	Male	18 months	Mass abdomen	Palpable mass		USG, CT	Ileum	Tubular	Yes	Excision	None
4	Male	2 years	Intussusception	Peritonitis		USG	Ileum	Tubular	No	Excision	None
5	Male	2 years	Mass abdomen	Palpable mass		USG	Gastric	Cystic	No	Excision	None
6	Female	3 years	Vomitting	Non-specific		USG	Pylorus	Tubular	No	Excision	None
7	Male	3 months	Rectovestibular fistula	Non-specific	Anorectal malformation	USG	Ascending colon	Tubular	Yes	Excision	None
8	Male	7 years	Intestinal Obstruction	Peritonitis		USG	Ileum	Tubular	No	Excision	None

USG: Ultrasonography; CT: Computed tomography

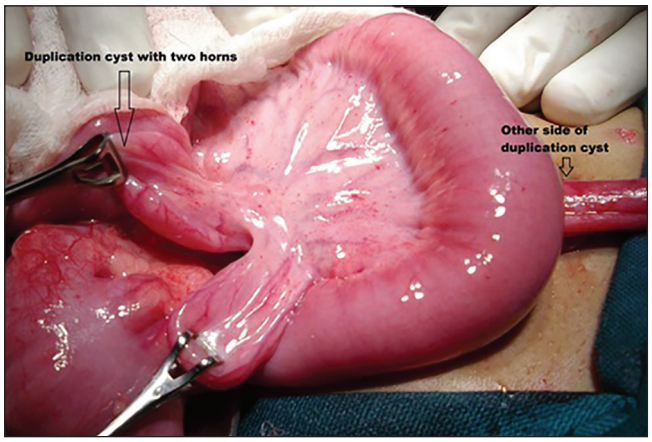


Figure 1: Duplication cyst of ileum presented as hydrocoele

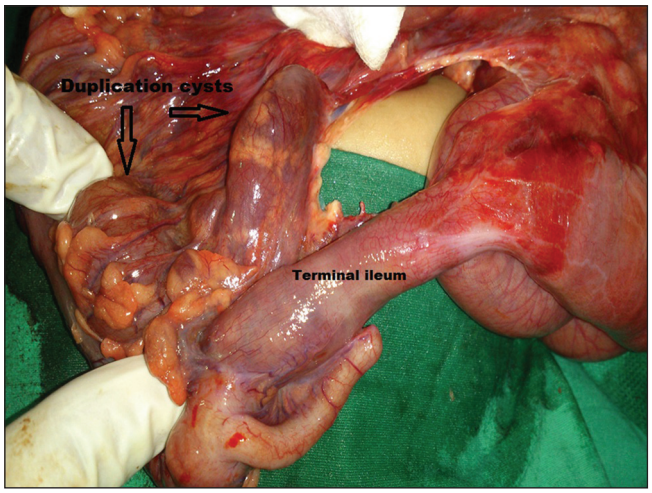


Figure 2: Two duplication cysts of ileum with mesenteric defect who presented with intussusception

50% of cases.[10] However, examination findings depend upon the presentation of patients. Abdominal mass was palpable in two patients in this series, while three patients had signs of acute abdomen, in three of eight patients abdominal examination was unremarkable. Associated malformations are reported in 50% of patients with EDs, most common being vertebral defects.[11] However, in this study, only one patient had associated anorectal malformation.

Laboratory investigations in case of EDs are usually non-specific however, they may show anaemia, which may be due to bleeding from heterotopic gastric mucosa present in ED cyst wall. One patient in this study was diagnosed prenatally and patient came to us for management opinion at 18 months age when he was operated. The main diagnostic tool used in all patients was USG abdomen, which showed a cystic mass in four of eight patients but in two of four patients it could not confirm the origin of the cyst. In one of the patient CT abdomen was obtained, which also failed to confirm its origin. In all patients, no further imaging modality was used and exploration was considered.

The most common site of EDs is ileum[12] as in our study where 5 out of 8 patients had ileal duplication cyst ($n = 5$). Gastric duplication is rare and accounts for 3.8% of all duplication of GIT.[2] In our study, one patient had gastric duplication ($n = 1$). EDs of the colon are rare and account for 13-18% of all duplications of GIT.[13] However, descending colon is a rare site in the colon to have ED. One of our patients had involved descending colon also ($n = 1$). Pyloric EDs are rare among duplication of GIT and are present in 2.2% of EDs. One of patients in our study also had pyloric ED ($n = 1$).[14,15]

Enteric duplications may be cystic, tubular or mixed and they are located on mesenteric side of intestine. EDs may share its blood supply with adjacent intestine by residing in leaves of its mesentery, posing a difficulty for safe resection.[16] In our series, six of eight cases were tubular type, while two patients had cystic EDs. In two of cases, duplication cyst was present in leaves of the mesentery of the intestine and we had to resect intestine along with ED. In two of eight patients, ED was communicating with a lumen to native intestine.

Li *et al*. have classified EDs of the small intestine on the basis of its blood supply into Type 1 (parallel type) and Type 2 (intramesenteric type).[17] In five of our cases involving small intestine, two were of Type 1 having duplication more to one side of the mesentery with a separate artery for duplication cyst, while three were of Type 2 with duplication cyst lying in the centre of mesentery and sharing blood supply with native gut.

The surgical approach varies with location and type of EDs. In all of our patients excision was performed; however in two cases, native gut had to be excised. In one patient with duplication cyst just proximal to ileocecal junction excision was done with mucosal stripping. Histopathological examination of specimens was obtained in all patients and it confirmed them to be EDs.

Surgery was uncomplicated in all of our patients except one who developed adhesive bowel disease and presented to us with intestinal obstruction. He was explored 8 months after initial surgery and was unexpectedly found to have two blind ending loops of small intestine lying close to each other with no signs of scarring or adhesions at initial anastomotic site at

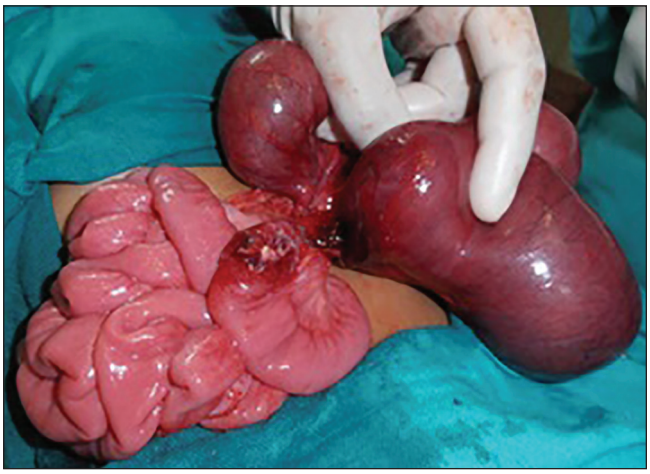


Figure 3: Transection of gut loops due to adhesive bowel disease (after permission from APSP J Case Rep)

ileum. It was an unusual complication of adhesive bowel disease, which had never been reported in literature (Figure 3).[18] Recurrence was found in none of our patients in 1-2 years follow-up.

CONCLUSION

Enteric duplications can present with a variety of symptoms and excision should be considered in all cases wherever possible.

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