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Retroperitoneal isolated enteric duplication cyst

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ABSTRACT

Enteric duplication cysts are rare congenital malformations of the gastrointestinal tract with an estimated incidence of 1:100,000 live births. Fewer still are isolated enteric duplication cysts (IEDC). Accurate diagnosis and timely excision of IEDCs can help in avoiding possible complications including bleeding from gastric mucosa and malignant transformation later in life. Currently, in the paediatric population, there are twenty reported cases in the literature worldwide. Of these, only three have been described in the retro peritoneum. We present a retroperitoneal IEDC, which was juxtaposed to the inferior vena cava. To the best of our knowledge, this has not been reported before in literature.

1. Introduction

Duplication cysts of the alimentary tract usually occur in communication with the gastrointestinal tract and typically share a common wall and blood supply. They are tubular or spherical structures with an epithelium similar to the intestinal tract. Rarely, they occur completely isolated from the gastrointestinal tract with an independent blood supply. The later are referred to as either non-communicating or isolated enteric duplication cysts (IEDC) [1,2]. There are approximately three dozen reported cases of IEDC altogether, with a slightly higher frequency in the paediatric population [3]. Clinically they present in a variety of ways from asymptomatic masses to abdominal pain, abdominal distension and bowel obstruction. This case report is about an eight-year-old child who presented following intestinal obstruction secondary to an IEDC.

2. Case report

A previously healthy eight years old male child, presented to the paediatric surgical clinic with a recent history of severe abdominal pain and vomiting for eleven days. According to the child and his parents, the abdominal pain started insidiously two weeks prior, which was then accompanied by vomiting. The vomiting was initially non-bilious, but later progressed to bilious. With these complaints, he was admitted to an external facility where he was investigated for intestinal obstruction. His plain abdominal radiographs were reported to be normal. However, an ultrasound of the abdomen done showed a cystic abdominal mass. The child was treated conservatively with bowel rest, analgesia and antibiotics. He subsequently opened his bowels and was discharged home. However, he continued to have abdominal pain.

On examination in our clinic, the child was able to point at the sight of his pain in the right upper quadrant of the abdomen. Palpation was however not able to reveal any mass. The rest of the examination was unremarkable.

A repeat ultrasound scan at our facility confirmed a poorly defined, 3.7 cm multiloculated cyst, which was retroperitoneal and abutting on the duodenum superiorly and the inferior vena cava medially [Fig. 1]. With a possible differential diagnosis of either a mesenteric cyst or an enteric duplication cyst, a contrast enhanced CT scan of the abdomen was done. A 2.2 × 3.9 × 2.7 cm (AP x Trans x CC) cyst with contrast enhancing walls was seen closely abutting the inferior vena cava and the junction of the second and third part of the duodenum in the retroperitoneum (Figs. 2 and 3). However, it did not seem to have any communication with the gastrointestinal tract.

Based on the preoperative imaging, the differential diagnosis at the time included mesenteric cyst, lymphangioma, or a cystic mass of unknown origin. Given the persistent pain and the unusual location of the cyst, a laparotomy was planned for excisional biopsy.

At laparotomy, findings included a palpable cystic mass in the root of the mesentery with multiple enlarged lymph nodes. The mesentery however, was freely mobile over the cyst. Therefore, an incision was

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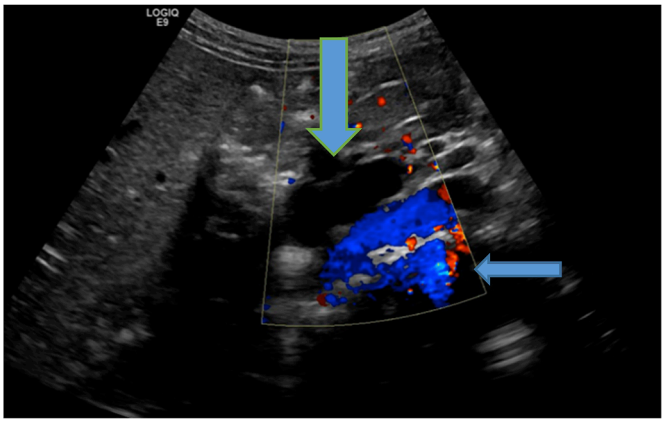

Fig. 1. A poorly defined, multilocular cyst (wide arrow) is demonstrated lateral to the IVC (narrow arrow) in the retroperitoneum measuring 3.7 cm × 1.5 cm.

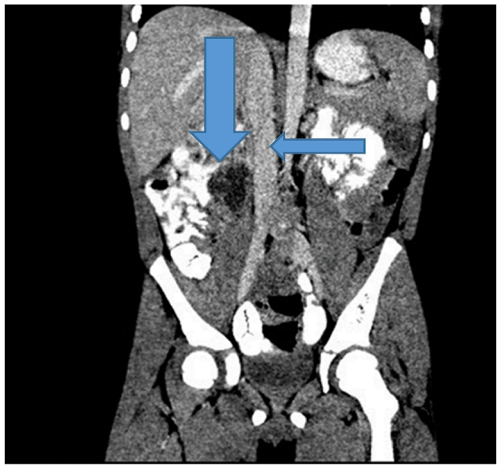

Fig. 2. A 2.2 × 3.9 × 2.7 cm (AP x Trans x CC) cyst with contrast enhancing walls (wide arrow) is seen closely abutting the inferior vena cava medially (narrow arrow).

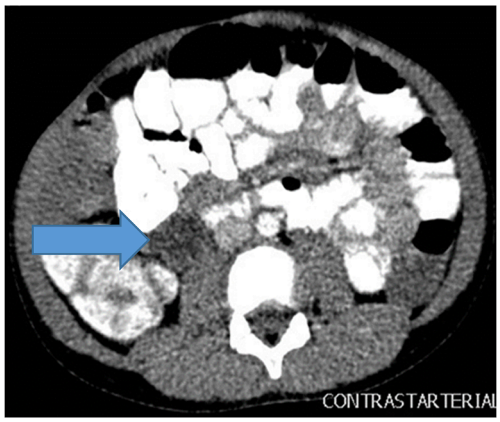

Fig. 3. wide blue arrow is the IEDC with the Inferior vena cava on its medial wall.Superio-medially is the juction of 2nd and 3rd part of the duodenum.

made lateral to the reflection of the ascending colon and the colon was reflected medially. The cyst was then separated meticulously from the Inferior vena cava medially, the psoas muscle posteriorly and the third part of the duodenum superiorly. [Figs. 4 and 5 below].

The specimen was then submitted for histology. Gross examination revealed a cystic mass measuring 3.5 × 2.6 × 2.3 cm, which was

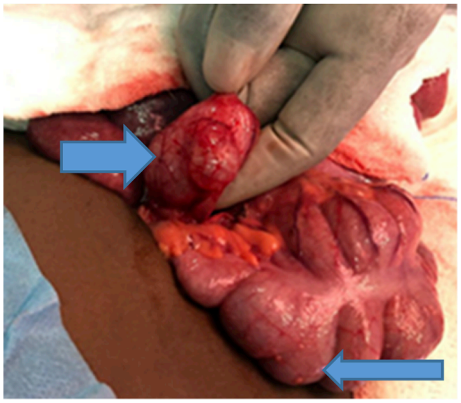

Fig. 4. Intraoperative images of IEDC (wide arrow) Wide blue arrow :IEDC. Narrow blue arrow is the ascending colon reflected medially.

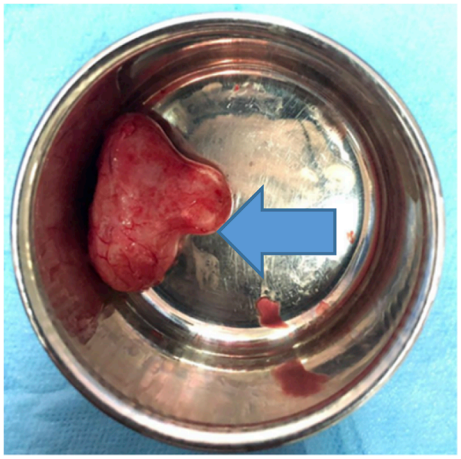

Fig. 5. gross specimen of the IEDC confirming complete excision.

multiloculated on cut surface and filled with mucoid fluid. On microscopy, the cyst wall showed organized smooth muscle [Fig. 6] and a surface simple mucinous columnar epithelium [Fig. 7], without goblet cells, suggestive of gastric epithelium.

Post operatively, the patient had a smooth recovery. He was discharged the next day and has been asymptomatic for over six months of follow up.

3. Discussion

According to Menon et al., [4] no discernible communication or connection with the adjacent alimentary tract, along with the presence of the typical histopathological features of a duplication cyst, qualify for the diagnosis of an IEDC. These cysts usually present either as an incidental abdominal mass on antenatal ultrasound scan or in

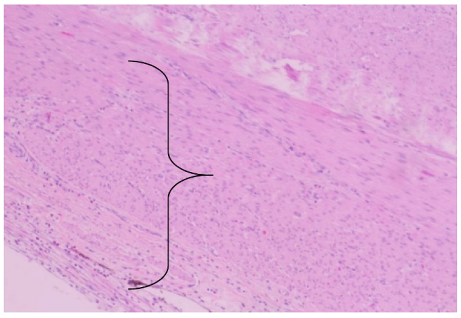

Fig. 6. Shows Smooth muscle wall (Bracket, x40).

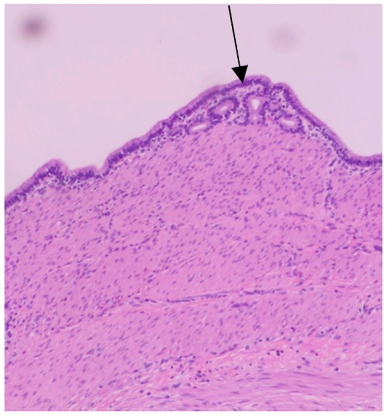

Fig. 7. Simple mucinous columnar lining (Arrow, x20).

the perinatal period. Presentation as a slowly growing abdominal mass with associated variable symptomatology from infancy to adulthood has also been reported [3].

In recent years, there have been a number of reported cases of IEDC from all geographical regions of the world. This raises the question regarding the etiology of IEDC. There are several theories regarding the etiology of duplication cyst communicating with adjacent bowel, however none unify any of the existing hypotheses. Furthermore, they do not explain the existence of IEDC. There is the aberrant luminal recanalization theory, which is based on a possible aberrancy in the canalization process leading to a parallel tract of normal bowel. Then there is the abortive twinning theory based on incomplete twinning during the fetal development. Other possible etiologies include Split Notochord theory, Persistent embryological diverticula theory and sequestration of part of the fetal gut [5,6].

There is a small but significant potential that enteric duplication cysts including IEDC can transform into malignant masses [7,8]. This is true for both symptomatic and asymptomatic masses. Secondly, because of the possibility of ectopic gastric and or pancreatic tissue, there is always a danger of an acute abdomen from an acute bleed [9,10].

Though children have the majority of the reported IEDC, the vast majority are peritoneal. There have been no report of IEDC next to the Inferior vena cava. Intestinal obstruction and persistent pain had led to the excisional biopsy and eventual diagnosis in our case.

Though rare, it is important to keep isolated enteric duplication cysts in the differentials of abdominal cystic masses in the both paediatric and adult population. They may have variable demographics, presentations and histology. However, due to the likelihood of both malignant transformation and acute bleeding, we recommend excision regardless of symptomology.

4. Conclusion

We felt the need to share our case to bring attention to this growing number of isolated enteric duplication cysts in the literature. This will help promote the recognition of this pathology as a separate entity in literature as well as research into the etiology, possible early diagnosis and treatment in the future to avoid the probable complications.

Patient consent

Consent to publish this case report was obtained from the parents.

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Declaration of competing interest

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