**Giant Gastro-duodenal Duplication Cyst with Juxta-Pancreatic Communication**

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**ABSTRACT:** Enteric duplication cysts are rare congenital malformations with a low incidence and there are only a few reports in the literature. Their clinical presentation varies according to the location and the type of duplication. Their overall prognosis is good if early surgical intervention is provided. We report a 2-month-old boy who presented with a case of a giant gastro-duodenal duplication cyst with a juxta-pancreatic communication and was successfully treated surgically. It is imperative to be aware of this rare congenital malformation that can present clinically with a wide range of non-specific symptoms that can cause significant morbidity and mortality if the treatment is delayed.

**Keywords:** Pancreatic Duct; Congenital Abnormalities; Intestinal Diseases; Newborn; Case Report; Colombia.

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**Case Report**

A 2-month-old boy arrived at the Paediatric Emergency Department (ED) of a tertiary care centre in Bucaramanga, Colombia, in 2016, with vomiting after feeds, abdominal distention and an abdominal mass found by his mother. The patient had a history of suspected duodenal atresia by antenatal ultrasounds (double bubble sign found). However, this congenital malformation was ruled-out postnatally after a normal upper gastrointestinal series test and in the absence of vomiting or gastric aspirates in the neonatal period.

Physical examination at the ED revealed a giant abdominal mass located in the hypogastrium [Figure 1A]. Given the clinical context, a contrasted computed tomography (CT) scan of the abdomen was done. A huge hypodense cystic mass with the dimensions of 11 × 12.8 × 6 cm and an approximate volume of 320 cc was documented [Figure 1B].

The patient underwent surgical exploration through a midline vertical incision after considering a mesenteric cyst versus an enteric duplication cyst as a possible diagnosis. During surgery, a giant boot-shaped cystic mass with a diameter of 15 × 12 cm occupying approximately 70% of the abdomen was found [Figure 2A]. The mass was adherent to the greater curvature of the stomach, sharing the serosa and muscular layers. It was hyper-vascularised, with blood supply from the left gastric artery, gastroepiploic artery, splenic artery, short gastric arteries and the pancreatic artery. Although a normal pancreatic duct was found, an aberrant duct communicating the cystic mass with the pancreas was also identified. The juxta-pancreatic communication was confirmed by intraoperative cholangiogram. Intraoperative process included the dissection of the giant enteric duplication cyst by layers and the ligation of contiguous vessels related to the cyst. The mass was disconnected from the greater curvature without compromise of the muscular and mucosa layers of the stomach. After removing this giant mass, no evidence of loss to the greater curvature of the stomach or the pancreas was identified.

Physiological examination of the stomach revealed normal perfusion. Additionally, ligation of the aberrant duct was performed using a 4-0 polypropylene suture after a normal intraoperative cholangiography that verified a normal transit of the contrast to the duodenum [Figures 2B and C]. Gross examination revealed a mass with an approximate dimension of 10 × 10 × 5...
A final diagnosis of a gastroduodenal duplication with a juxta-pancreatic communication was made. Histopathological examination showed the wall of the cyst composed of the muscular layers with extensive atrophic changes. Although the mucosa architecture was distorted, Paneth cells and enteric cells were identified. The submucosa and the muscularis layer were also documented [Figure 3]. The patient was evaluated in postoperative follow-up visits at one, two and six months. His recovery was uneventful.

Prenatal ultrasounds were also reviewed while writing this report. The authors did not find evidence of images suggesting an enteric duplication cyst. No maternal polyhydramnios was documented during antenatal follow-ups. Informed consent from the patient’s guardian was obtained for publication purposes.

Discussion

This is a case report of a giant gastroduodenal duplication cyst with juxta-pancreatic communication in a 2-month-old boy with vomiting after feeds, abdominal distention and an abdominal mass in the hypogastrum. Gastrointestinal tract duplications are rare anomalies that are usually found in early childhood but can remain asymptomatic until adulthood. Clinical presentation of this abnormality has a wide range of signs and symptoms from non-bilious vomiting to severe abdominal pain due to fluid leakage or rupture of the cyst.\(^2\) Most enteric duplications become symptomatic as a result of the obstruction of the gastrointestinal tract by external pressure, distention of the cystic mass or bleeding.\(^2\) The embryogenesis of the enteric duplications remains unclear. However, several hypotheses including canalisation impairment, intrauterine vascular accident, cellular migration defects and diverticulisation have been postulated.\(^7\) Furthermore, in those cases with an associated
pancreatic communication, the mechanism of embryonic maldevelopment is thought to be more complicated. The anatomy of gastrointestinal tract duplications usually includes a hollow, epithelial-lined and cystic or tubular mass attached to the wall of the gut and supplied by contiguous blood vessels. However, in the current case, the enteric duplication was sharing a blood supply of arteries with a common embryonic origin, there is no clear literature about this relationship. Regarding location, the gastroduodenal region is a rare site for enteric duplication to occur (10% of cases), and only approximately 20% of cases at this level have an associated pancreatic communication. From those cases involving the stomach, approximately 66% of gastroduodenal duplications occur at the greater curvature. A case series by Lopez-Fernandez et al. presented a total of 11 cases of pyloroduodenal duplication cysts treated over 26 years of practice. From those cases, only three corresponded to an enteric duplication with pancreatic communication. Therefore, the current case represents a rare entity with a very low incidence (a giant enteric duplication cyst with an aberrant pancreatic duct).

Radiologic studies, including abdominal ultrasonography, magnetic resonance imaging or abdominal CT scan, are usually needed for antenatal or postnatal diagnosis. An abdominal CT scan is recommended for postnatal diagnosis due to its ability to document the anatomical relationship with the surrounding structures. As previously mentioned, the current patient had an antenatal diagnosis of duodenal atresia by ultrasound that was ruled-out postnatally. Interestingly, in a case report by Okamoto et al., a gastrointestinal mass found antenatally was diagnosed as a duplication cyst postnatally. It should be noted that the case reported by Okamoto et al. and the current case reinforces the need for a complete evaluation of the newborn as well as radiologic studies in the postnatal period in patients with antenatal images suggesting intra-abdominal cystic masses or congenital abnormalities of the intestines; this would enable an early and accurate diagnosis and prevent complications.

The surgical approach of enteric duplications varies according to location and type of duplication. Surgical management for asymptomatic cases is controversial. It has been suggested that enteric duplications should be excised not only due to the symptoms but also for the risk of developing adenocarcinoma within the cyst. In those cases with a precise preoperative diagnosis, a laparoscopic approach is recommended. The presence of an aberrant pancreatic communication and its risk of postoperative pancreatitis makes a radiological evaluation imperative and an intraoperative endoscopic retrograde cholangiopancreatography is usually recommended to avoid morbidity. In the current case, adequate diagnosis and surgical treatment led to a satisfactory outcome. This case describes an uncommon condition that paediatricians and paediatric surgeons should be aware of as part of the differential diagnoses that must be ruled-out during postnatal evaluations when intra-abdominal cystic lesions are documented antenatally. This also shows how an abnormality in the embryogenesis of the gastrointestinal tract can manifest in an infant.

**Conclusion**

Gastrointestinal tract duplications are rare conditions. The current case describes a giant enteric duplication cyst that presented as an abdominal mass associated with abdominal distention and vomiting. Complete radiologic evaluation of patients with antenatally documented intra-abdominal cystic masses must be performed to pursue an early diagnosis and prevent complications of undiagnosed pathologies. Additionally, in the event of an abnormal pancreatic communication, the evaluation of the integrity of the pancreatic drainage during the procedure and the careful resection with preservation of adjacent structures is essential. It is imperative to be aware of this rare congenital malformation that can present clinically with a range of non-specific symptoms that can cause significant morbidity and mortality if treatment is delayed.

**AUTHORS’ CONTRIBUTION**

JFMP and AP were involved in conceptualisation, writing and editing of final manuscript. DGPD was involved in the histopathological examination as well as writing the manuscript. All authors approved the final version of this article.

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