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## Giant Gastroduodenal Duplication Cyst with Juxta-Pancreatic

### Communication

#### *A rare intraoperative finding*

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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 present a case of a giant gastroduodenal duplication cyst with a juxta-pancreatic communication in a 2-month-old boy who 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.

#### Introduction

Gastrointestinal tract duplications or enteric duplication cysts are rare congenital malformations formed during the embryonic development of the digestive system with an incidence of 1:4,500 births. These malformations most frequently occur in the esophagus, jejunum, ileum, colon, and

stomach.<sup>1</sup> The involvement of the pancreas is very rare.<sup>2</sup> The clinical presentation of these malformations is with a wide diversity of signs and symptoms, including abdominal pain and distension, gastrointestinal bleeding, obstruction, or intussusceptions.<sup>3-5</sup> Urgent surgical intervention is needed when the severity of symptoms and signs involve significant complications such as massive bleeding or perforation of the intestine.<sup>3</sup> We are presenting a case of a giant gastroduodenal duplication cyst with a juxta-pancreatic communication, a rare intraoperative finding that was successfully treated surgically.

### **Case Report**

A 2-month-old boy arrived at the pediatric emergency department (ED) 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 (GI) 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 computerized tomography (CT) scan of the abdomen was ordered. A huge hypodense cystic mass with a diameter of 11x12.8x6 cm and an approximated volume of 320cc was documented (Figure 1B).

The patient underwent surgical exploration through a midline vertical incision after considering a mesenteric cyst vs an enteric duplication cyst as a possible diagnosis. During surgery, a giant boot-shaped cystic mass with a diameter of 15x12cm 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-vascularized, 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 the mucosa layer of the stomach. After removing this giant mass, no evidence of loss to the greater

curvature of the stomach or the pancreas was identified. Normal perfusion of the stomach was also confirmed. Additionally, ligation of the aberrant communication was performed using a 4-0 polypropylene suture after a normal intraoperative cholangiography that verified a normal transit of the contrast to the duodenum. (Figure 2B and 2C). Gross examination revealed a mass with an approximate dimension of 10x10x5cm. 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 1, 2, and 6 months. His recovery was uneventful.

Informed consent from the patient's guardian was obtained for publication purposes. 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.

## **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 hypogastrium. Gastrointestinal tract duplications are rare anomalies that are usually found in early childhood but can remain asymptomatic till 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.<sup>2, 6</sup> 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.<sup>2</sup> The embryogenesis of the enteric duplications remains unclear. However, several hypotheses including canalization impairment, intrauterine vascular accident, cellular migration defects, and diverticulization have been postulated.<sup>7</sup> Furthermore, in those cases with an associated pancreatic communication, the mechanism of embryonic maldevelopment is thought to be more complicated.<sup>8</sup>

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 our case, the enteric duplication was sharing a blood supply of arteries with a common embryonic origin, there is no clear literature about this relationship.<sup>7</sup> Regarding location, the gastroduodenal region is a rare site for enteric duplication to occur (10% of cases), and only around 20% of cases at this level have an associated pancreatic communication. From those cases involving the stomach, about 66% of gastroduodenal duplications occur at the greater curvature.<sup>9,10</sup> In a case series by Lopez-Fernandez et al., the authors 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.<sup>11</sup> Therefore our case corresponds to a rare entity with a very low incidence and reports a *giant* enteric duplication cyst with an aberrant pancreatic duct.

Radiologic studies, including abdominal ultrasonography, magnetic resonance imaging (MRI), or abdominal CT scan, are usually needed for antenatal or postnatal diagnosis.<sup>12</sup> An abdominal CT scan is recommended for postnatal diagnosis due to its ability to document the anatomical relationship with the surrounding structures.<sup>2</sup> As mentioned before, our patient had an antenatal diagnosis of duodenal atresia by ultrasound that was ruled out postnatally. Interestingly, in a case reported by Okamoto et al., a gastrointestinal mass found antenatally was diagnosed as a duplication cyst postnatally.<sup>13</sup> Noteworthy, the case reported by Okamoto et al. and our case reinforce the statement that in patients with antenatal images suggesting intraabdominal cystic masses or congenital abnormalities of the intestines, a complete evaluation of the newborn and radiologic studies in the postnatal period must be completed to achieve 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.<sup>4</sup> Some authors suggest that enteric duplications should be excised not only for the symptoms but also for the risk of developing adenocarcinoma within the cyst.<sup>2</sup> In those cases with a precise preoperative diagnosis, a laparoscopic approach is recommended.<sup>14</sup> 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.<sup>15</sup> In our case, adequate diagnosis and surgical treatment led to a satisfactory outcome. This case describes an uncommon condition that pediatricians and pediatric surgeons should be aware of as part of differential diagnosis that must be ruled out during postnatal evaluations when intraabdominal 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. Our 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 intraabdominal 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 the treatment is delayed.

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### **Authors' Contribution**

**JFMP** and **AP** were involved in conceptualization, 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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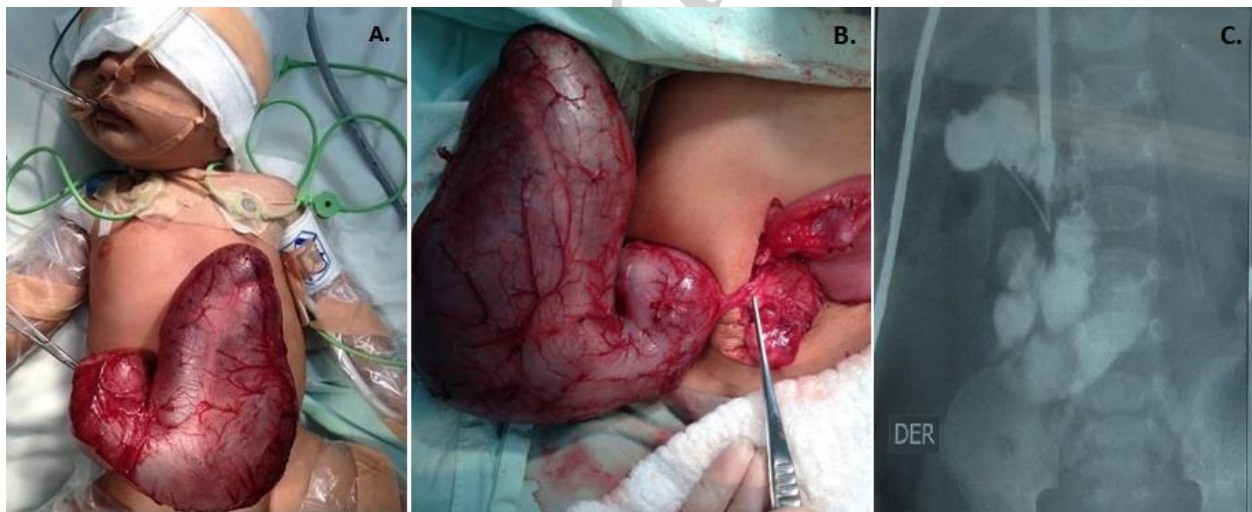
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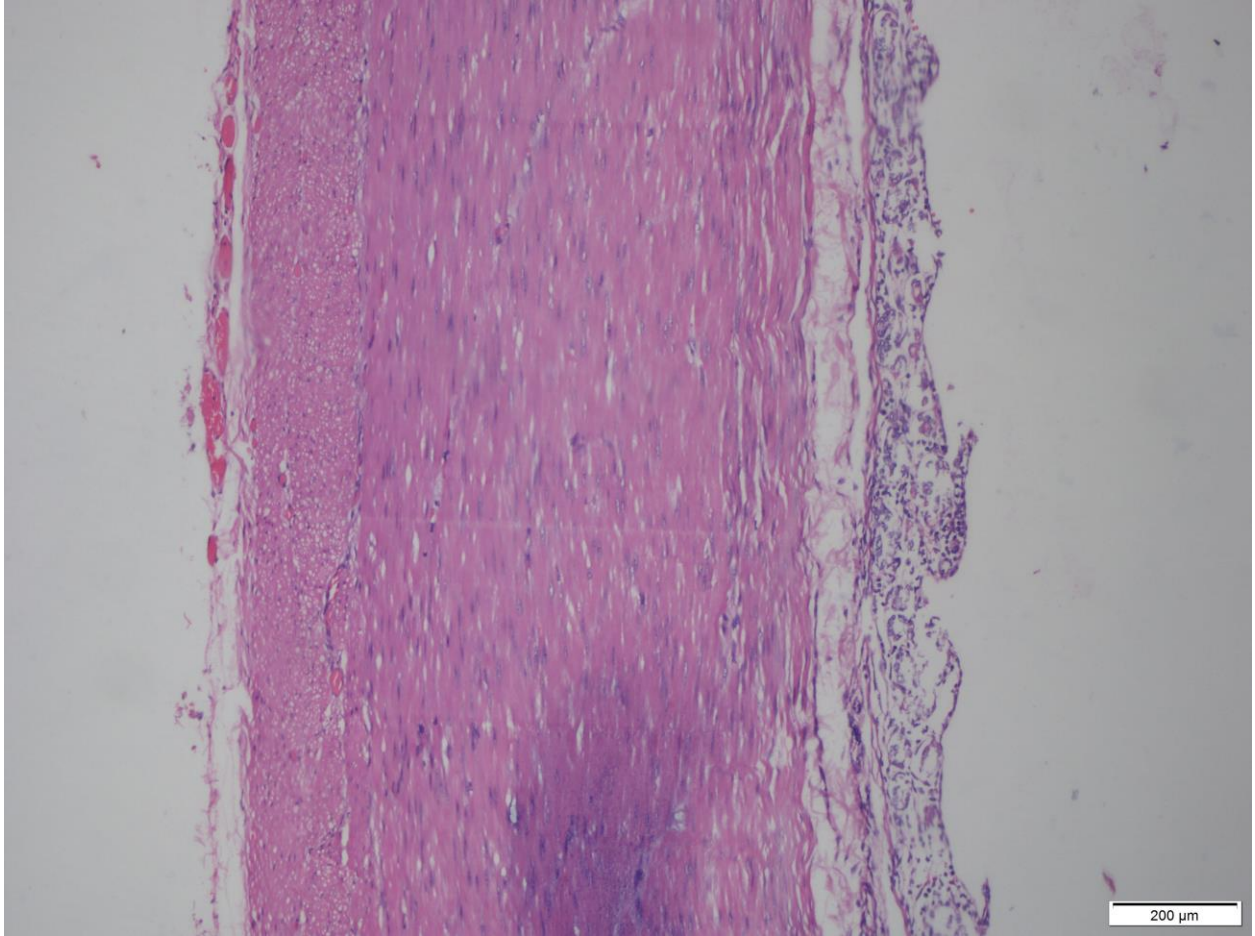


**Figure 1:** A. Findings on examination at the pediatric emergency department B. Contrast-enhanced computed tomography (CT) scan of the abdomen: 11 x 12.8 x 6cm mass with an approximated volume of 320cc.



**Figure 2:** A. Boot-shaped giant mass occupying 70% of the abdomen. B. Juxta-Pancreatic communication. C. Normal intraoperative cholangiography.





**Figure 3:** Microscopic examination of the excised cyst wall showed gastric and duodenal mucosa, submucosa, and muscle coats consistent with an enteric duplication cyst.

Accepted