

## CASE REPORT

# Duodenal Duplication Cyst in Close Proximity of Ampulla of Vater: A Surgical Challenge

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### Abstract

Prompt preoperative diagnosis of duodenal duplication cyst is uncommon owing to its extreme rarity. The ideal treatment of intestinal duplication cyst is complete surgical excision, though in few cases, due to the proximity to the bilio-pancreatic duct, the complete excision is not possible. We herein present an infant presenting with bilious vomiting starting few days after birth. Ultrasonography and CT scan provided the diagnosis of the duplication cyst in relation to duodenum. A successful surgical management by a subtotal excision was done. Although duodenal duplication is seldom seen, it should be considered in differential diagnoses of upper gastrointestinal tract (GIT) occlusion.

**Key words:** Bilious vomiting; Duodenal duplication; Subtotal excision.

### INTRODUCTION

Duodenal duplication cysts are rare congenital malformations of the intestinal tract (1 per 100,000 births) and represent only 2-12% of all GIT duplications [1,2]. Because of its rarity, duodenum duplication can represent a diagnostic and management challenge. Surgery and complete excision is the preferred method of treatment however, surgical procedure occasionally tailored depending upon its relationship with important surrounding structures [3]. We report a case of duodenal duplication cyst having communication in proximity to ampulla of Vater precluding its complete excision.

### CASE REPORT

A 1-month-old female infant, born at term with spontaneous vaginal delivery having 3200g birth weight, presented with progressively frequent bilious vomiting since birth. Prenatal ultrasounds were normal. One week before the admission, the mother observed progressive increased bilious vomiting. General physical examination was normal; her abdomen was soft non-tender and non-distended. There was no palpable mass. Laboratory investigations yielded normal results. Ultrasonography showed a cystic mass measuring 50mm adjacent to the second portion of the

duodenum. The mass had a thick wall consisting of two layers (outer hypoechoic, inner echogenic). Computed tomography scan of the abdomen showed a well-defined ovoid cystic structure, located in the right upper abdomen adjacent to the second portion of the duodenum and head of the pancreas (Figure 1). On the basis of this “two-layer” or the “gut signature” sign, the diagnosis of duodenal duplication was established. Barium examination revealed a communication between the duplication and the normal lumen.

After optimization, under general anaesthesia, laparotomy was performed. After releasing multiple intra-abdominal adhesions especially with gallbladder, we found a tubular duodenal duplication in close approximation to the second and third portions of the duodenum, measuring 10 cm in length (Figure 2). The contrast material was administration into the gallbladder, in order to identify the cystic duct, the common bile duct, and the duodenum. The duodenal duplication was opened in longitudinal direction, which showed the internal common wall between the cyst and the duodenum. A communication between D2 and duodenal duplication was noted; measuring 3 cm in length, in proximity to the major Papilla, which was recognized by methylene blue injection into the gallbladder (Figure 3). The cystic wall was excised up to common wall with stripping of lining

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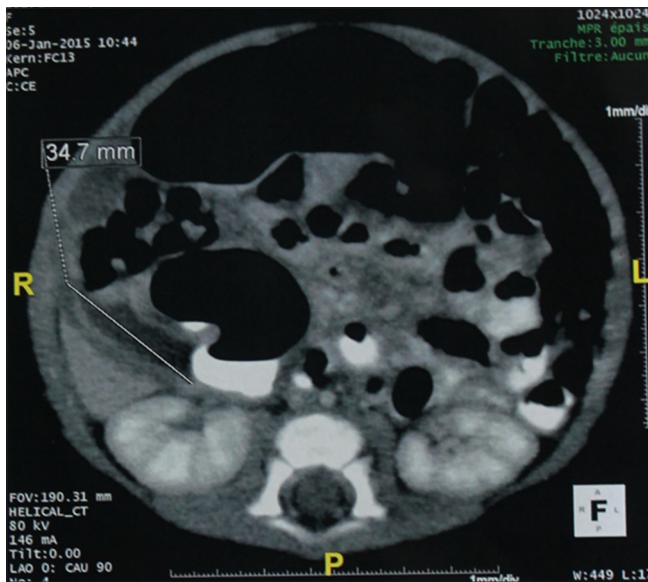


Figure 1: Computed tomography scan of the abdomen showed a 35 mm well-defined ovoid cystic structure, located in the right upper abdomen and adjacent to the second portion of the duodenum and to the head of the pancreas

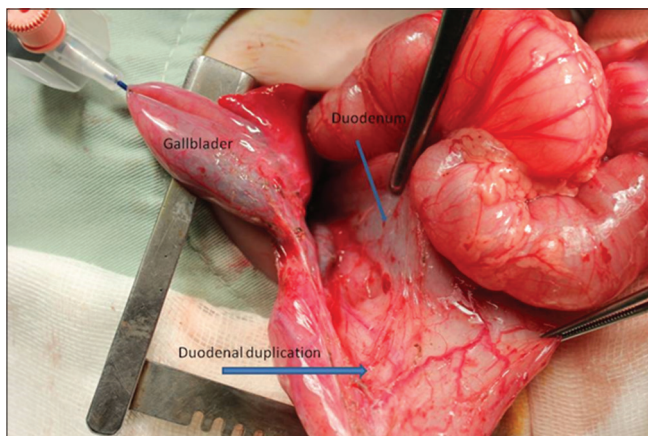


Figure 2: Duplication cyst sharing common wall with duodenum

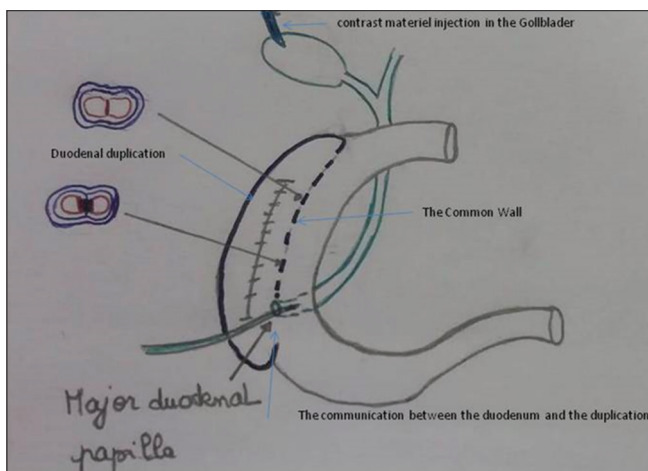


Figure 3: Schematic representation of operative finding

mucosa, followed by primary repair of the duodenum wall. The post-operative recovery was uneventful. Histologic examination revealed duodenal mucosal lining with fibromuscular tissue, with no ectopic tissue, consistent with the diagnosis of duplication cyst of the duodenum. At 3-year follow-up, she is doing fine with no recurrence.

## DISCUSSION

Ileal duplications are the most common; duodenal duplications constitute less than 5% of all GIT duplications [1]. Duodenal duplication cyst is a rare congenital anomaly. A recent meta-analysis (1999 and 2009) found a total of 47 cases of duodenal duplication [1].

Duodenal duplications can be cystic or tubular, communicating or non-communicating, but the most common type is cystic and non-communicating. These are generally located at the medial border of the first and second parts of the duodenum and extend to the anterior or posterior side [3]. In our case, the duodenal duplication was tubular and located in the second and third parts of the duodenum, but it was communicating type and located on the lateral border and extending to the posterior side.

Clinical presentations are usually non specific, such as abdominal pain, nausea, vomiting related to an upper gastrointestinal obstruction or specific including recurrent, or chronic pancreatitis. [1] In the index case, the baby had gradually frequent bilious vomiting as a presenting complaint.

Usually physical examination and plain abdominal radiographs are normal but a mass can be palpable although not detected in our case. On ultrasonography, duodenal duplication is seen as an anechoic, double-walled, bilobed cystic lesion containing debris in the pyloroduodenal region, or sometimes, as an echogenic mass [1]. It may have muscular peristalsis, which helps distinguish duplication cysts from other abdominal cysts [4, 1, 5]. However, on gastrointestinal tract (GIT) contrast radiographs, duodenal duplication cysts can present as smooth submucosal or extrinsic masses or oval filling defects in the duodenum [1, 4]. Computed tomography or MRI are valuable in identifying their location and size, their anatomy in relation to the pancreaticobiliary system, or any edema of the pancreas resulting from associated pancreatitis. On CT scan or MRI, duodenal duplication cysts are discrete, fluid-filled structures attached to the medial wall of the descending duodenum. Occasionally, cyst wall calcification is recognized [1].

In some cases, the histological examination, in duodenal duplication cysts can show ectopic gastric or pancreatic mucosa[6], which might be complicated by local

bleeding before or after the surgical treatment. Ectopic tissues and chronic inflammation of the surrounding intestinal mucosa can undergo malignant transformation [7]. For these reasons, complete excision of the cyst is the standard surgical procedure, but this is often not possible in the duodenum because of the proximity to the bile ducts and the pancreas [4].

In our case, the close relation of the cyst with the ampulla of Vater makes its complete resection impossible. We opted a subtotal removal of the cyst. The cystic wall was opened and excised up to the common wall with stripping of lining mucosa then the duodenum was gently closed. According to some authors: Removal of the free walls of the cyst, with mucosal stripping of the common muscular wall, is the preferred treatment for duplication of the duodenum [8]. Recently, duodenal duplication cysts are being treated endoscopically. Advances in therapeutic endoscopy, such as endoscopic mucosal resection and endoscopic sub mucosal dissection, provides a viable option for dealing such lesions. [4, 8].

In conclusion, duodenal duplication cysts are uncommon alimentary tract lesions. They most commonly present as abdominal pain and vomiting. Its diagnosis and management can be challenging as happened in the index case. The surgeon should be familiar with the anatomy and various surgical options for these lesions. Surgical complete excision is the preferred method of treatment; although, could not be achieved in the index case owing to its anatomical relationship with biliary and pancreatic structures.

#### Authors' contribution

All authors contributed equally in concept, design, literature review, drafting the manuscript, and approval of the final manuscript.

#### Consent statement

Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

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