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Duodenal duplication cyst in a 52-year-old man: A challenging diagnosis and management

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ABSTRACT

INTRODUCTION: Duodenal duplication is a rare congenital malformation. Although more frequent in childhood, it may rarely be observed in adulthood. Pre-operative diagnosis can be difficult.
PRESENTATION OF CASE: We report the case of a 52-year-old man with a duodenal duplication cyst, who was misdiagnosed even after a primary surgery. Definitive treatment needed an extensive diagnostic workup and a second delicate operation.
DISCUSSION: This article discusses the incidence of duodenal duplications, their types, their clinical presentations, the radiologic and diagnostic features along with different therapeutic options.
CONCLUSION: Duodenal duplication should always be one of the differential diagnoses proposed when approaching upper abdominal cystic formations.

1. Introduction

Duplications of the gastrointestinal system are rare congenital malformations observed in 1 out of 25,000 deliveries. Most duplications are detected when children present with gastrointestinal (GI) bleeding or with symptoms related to obstruction. Less than 30% of all cases of intestinal duplications are diagnosed in adults.[1] Intestinal duplications in the duodenum are extremely rare and comprise less than 5% of all intestinal duplications. Because of their rarity, duodenum duplications can represent a diagnostic challenge. Treatment is mainly surgical and total excision, if possible, is the procedure of choice. However, in some cases, because of extensive size or delicate location, alternative procedures, such as subtotal removal or digestive derivation, are required.[2] In this article, we report the rare case of an incidentally discovered duodenal duplication in a 52-year-old male. The diagnosis was confirmed by the operative findings and subsequent histopathologic examinations. We present thereafter a literature review of this rare and challenging clinical entity.

2. Presentation of case

A 52-year-old man, known to suffer from type 2 diabetes mellitus for the last 6 years, presented to our care 3 months after being operated for a laparotomy at another institution. On that occasion and one month prior to the laparotomy, he complained of mild vague left flank pain with no other associated symptoms. Upon seeking medical advice, an ultrasound scan (USS) of the abdomen was ordered to rule out kidney stones.

The USS showed a large cyst (15 cm × 10 cm) with no signs of hemorrhage or calcification and the diagnosis of a "pancreatic cyst" was suggested. A computed tomography (CT) scan of the abdomen was done supporting the presumed diagnosis, so he was scheduled for open internal drainage of the presumed "pseudo-cyst".

During the laparotomy, his surgeon noticed that the preoperative diagnosis was not accurate so he terminated the procedure after taking several biopsies, the results of which were 'non-specific' and hence inconclusive. However, the patient's course was insignificant till 1 month after the operation, when he started to complaint of melena, hematochezia and fever. He was hospitalized several times during which he received 12 units of packed red blood cells (PRBCs) over a 4-week interval.

Meanwhile, the gentleman presented to our care with fever, general weakness, and lower limb edema. Total parenteral nutrition (TPN) and broad spectrum antibiotics were started. A magnetic resonance imaging (MRI) scan of the abdomen showed a cystic lesion (12.2 cm × 8.9 cm) presenting a thick wall enclosing debris and air; this mass is located between the liver, duodenum and head of the pancreas causing severe compression to the 3rd and 4th parts of the duodenum (Fig. 1).

A gastroscopy was done and showed that the 2nd part of the duodenum contained a wide opening at the anterior duodenal wall distal to the ampulla of Vater, creating thereby a lumen through which food is passing (Fig. 2). So, the diagnosis of a pancreatic pseudocyst that was surgically drained (during the operation presented above) and thereafter complicated by bleeding and super-infection was suggested. Therefore, the decision to operate on the patient was undertaken in view of the patient's septic condition and worsening status. Upon surgical exploration, a large sub-duodenal mass

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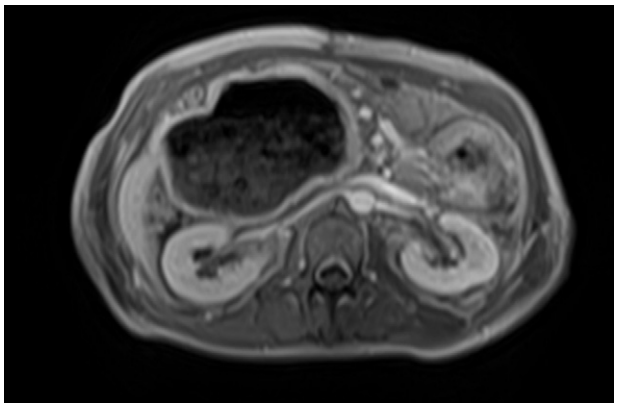

Fig. 1. MRI of the abdomen with the cyst in the R upper quadrant.

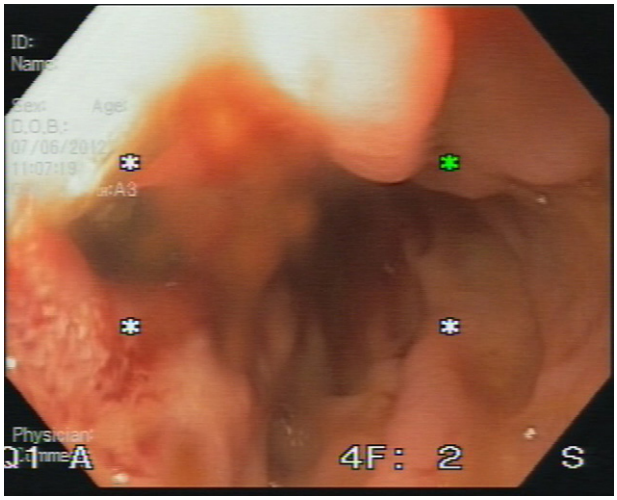

Fig. 2. An endoscopic view. The opening is shown on the L side.

pushing the duodenum ventrally and the ascending colon medially was noticed (Fig. 3). Upon incising the mass, food debris spilt out and a communication between the distal end of the mass and the duodenum was identified.

So, a full thickness biopsy from the wall was taken. An anterectomy with a Billroth II-type gastrojejunal anastomosis and a Roux-en-Y cysto-jejunostomy were done (Fig. 4). The patient had a smooth post-operative course and was discharged 10 days later. The final pathology report showed pieces of benign small bowel

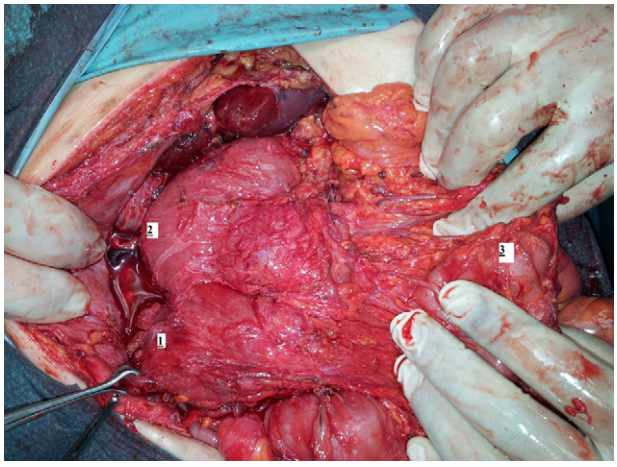

Fig. 3. An intra-op view: (1) cyst, (2) duodenum, (3) ascending colon.

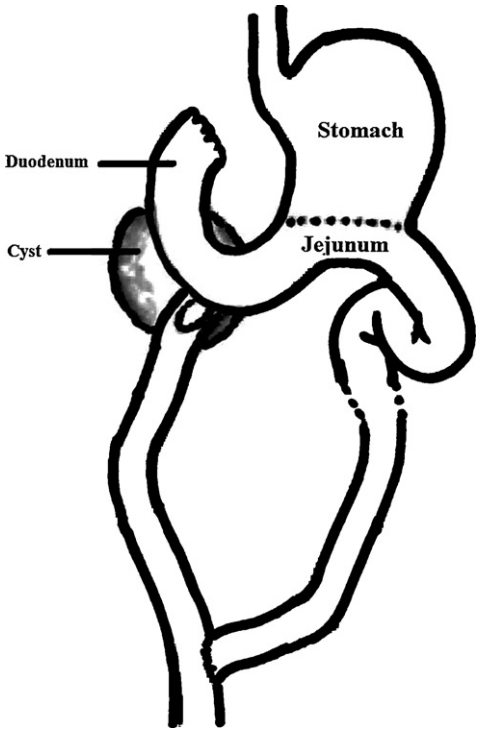

Fig. 4. Schematic diagram of the bowels at the end.

intestinal duodenal wall with marked ulceration reaching the serosa.

3. Discussion

Duodenal duplication cysts constitute a rare congenital anomaly of the gastrointestinal tract. A recent meta-analysis of the literature between 1999 and 2009 reported a total of 47 cases of duodenal duplication cysts.[3]

They form during the embryonic development of the human digestive organs. The definition includes a smooth-muscle coat, an alimentary epithelial lining and an intimate attachment to the GI tract.[4]

Jejunal duplications are the most common, followed by gastric, colonic then duodenal duplications that are the rarest, constituting less than 5% of all intestinal duplications.

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.[2,5,6] In our case, the duodenal duplication was cystic and located in the third and fourth parts of the duodenum, but it was of the communicating type and located on the lateral border and extending to the posterior side.

Like other intestinal duplications, most duodenal duplications are diagnosed in childhood, and for such a lesion to detected later in life is an extremely rare finding. A variety of clinical manifestations have been reported that are determined by the type, site and size of the duplication. Generally, patients present with a palpable mass in the abdomen, signs of intestinal obstruction, or abdominal pain. In a few cases, ectopic gastric mucosa can appear and cause ulceration, bleeding and even perforation.[2,7]

Our patient was asymptomatic and the cyst was firstly discovered while performing an ultrasonography for mild left flank pain. After the first laparotomy, he complained of gastrointestinal bleeding probably because of erosion and ulceration at the biopsy site and the final pathology report did not mentioned gastric mucosa.

The preoperative diagnosis of intestinal duplications is rarely accurate. Duodenal duplication is differentiated from other cystic lesions by the "gut signature" of its wall observed by abdominal or endoscopic US. Gut signature refers to the layered pattern of the wall, with the hyperechoic inner layer representing the submucosa and the hypoechoic outer layer representing the smooth muscle.[8,9] Peristalsis of the cyst wall noted upon real-time US is strongly suggestive of a duplication cyst.[10] Barium studies of gastrointestinal tract help to demonstrate the mass effect and displacement of normal alignment,[11] whereas a technetium scan can aid in the detection of heterotopic gastric mucosa in cases complicated with bleeding.[12] CT is valuable in identifying the type, location and the size of the duplication cyst.[13] In our case, despite the extensive workup the diagnosis was only made during the operation, and was only confirmed by histological examination.

The ideal treatment of duodenal duplication cysts is complete surgical resection. Due to proximity to the bilio-pancreatic duct, total resection sometimes requires pancreaticoduodenectomy. This major surgical procedure entails the disadvantages of high morbidity and mortality with poor quality of life. In our opinion, this procedure should remain an ultimate option. Less invasive approaches have been proposed, including partial resection or internal derivation.[14]

Recently, advances in therapeutic endoscopy, such as endoscopic mucosal resection and endoscopic submucosal dissection, provide a viable option for resecting neoplastic lesions of the alimentary tract with a low invasive potential.[15] They have also been widely introduced to submucosal lesions, and occasionally succeed in the removal of duplication cysts in older children and adults.[16,17]

Back to our case, the close relation of the cyst with the duodenum makes its complete resection impossible and, pancreaticoduodenectomy was very difficult because of severe adhesions and distorted anatomy. So to reduce morbidity and to decrease the risk of the surgery, we favored to do an internal derivation. The antrum was resected and a Billroth II-type gastro-jejunostomy was done in order to exclude the duodenum entirely from the passage of food, then a Roux-en-Y cysto-jejunostomy was done to drain the cyst and prevent bile from stagnation and infection.

4. Conclusion

Although extremely rare and rarely diagnosed in adults, duplications in the duodenum should always be a part of the differential diagnosis when approaching upper abdominal cystic formations. The management plan should be kept in mind in order to avoid non-indicated and potentially risky operations.

Conflict of interest statement

No conflicts of interest.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Ali al-Harake has operated and managed the patient.

Ahmad Basal has put the design of the paper and analyzed the data.

Mohamad Ramadan has helped in editing and figures arrangement.

Mohomad Chour has written the article and collected relevant literature.

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