Duplication cyst of the stomach with respiratory epithelium in adult: an uncommon finding.

Report of case and review of literature

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Duplication cyst of the stomach with respiratory epithelium in adult: an uncommon finding. Report of case and review of literature

Gastrointestinal duplication is a congenital rare disease entity. Duplication cyst of the stomach with pseudo stratified columnar ciliated epithelium is extremely rare. The very appearance of a gastric duplication cyst in an adult can present a diagnostic dilemma. In majority of reported cases, the diagnosis is established during surgical exploration. We report on a 34 year-old female patient suffering from repeated episodes of epigastric pain and gastroesophageal reflux. Abdominal computed tomography and endoscopic ultrasound demonstrated a intramural lesion attached to the gastric fundus, suggestive of gastrointestinal stromal tumor (GIST). At exploratory laparotomy a non-communicating cyst, was found along the greater curvature of the stomach in the esophagogastric transition. The lesion was excised along with an adjacent sleeve of the stomach and esophagus wall because shared muscular layer with the stomach and esophagus. The final pathologic examination revealed that the inner wall of the cyst was lined by a pseudostratified columnar ciliated epithelium (respiratory type) and, in part, columnar and gastric foveolar epithelium. Even though a panel of imaging modalities is available, it is still difficult to obtain a preoperative diagnosis. Duplication cyst can be mistaken for a soft tissue tumor of the gastrointestinal tract. There is no therapeutic algorithm. Surgical treatment is recommended for symptomatic cases.

KEY WORDS: Duplication cyst, Gastric duplication, Respiratory epithelium.

Introduction

Gastric duplication cysts (GDC) are very rare in adults[1]. The majority of gastric duplication cases also tend to have the gastrointestinal mucosa inside. The term "foregut duplication" is preferred when pseudostratified ciliated epithelium predominates [2-4]. The very appearance of a gastric duplication cyst in an adult can present a diagnostic dilemma [5]. Duplication cyst of the stomach with pseudo stratified columnar ciliated epithelium is extremely rare [6]. We report the case of a foregut duplication cyst of the stomach in an adult patient, which clinically and radiologically mimicked a gastrointestinal stromal tumor (GIST), discuss problems in the differential diagnosis, and review the pertinent literature.

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Case report

A 34 year-old female patient presented with repeated episodes of epigastric pain and gastroesophageal reflux;

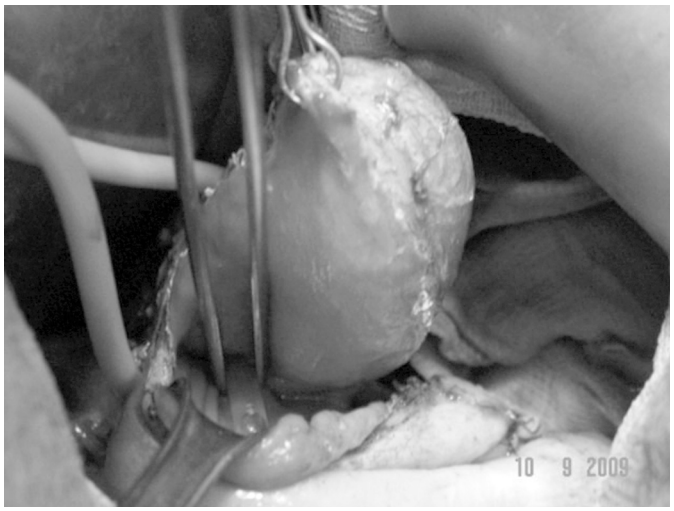

Fig. 1: The lesion was excised along with an adjacent sleeve of the stomach and esophagus. There was no communication between the lumen of the stomach and the cyst.

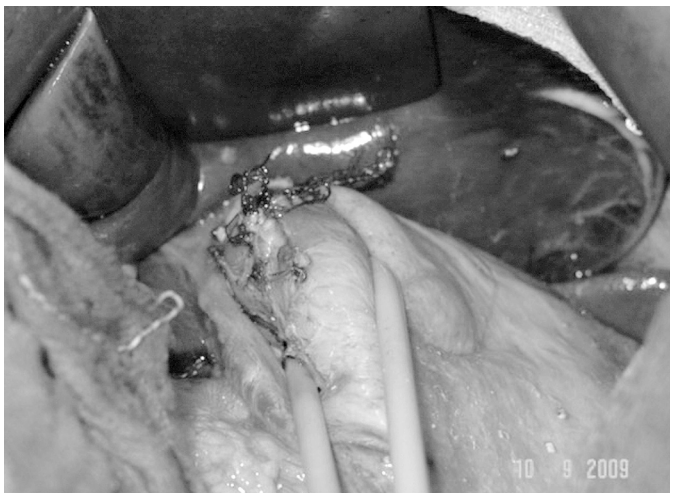

Fig. 2: The primary defect was repaired with esophago-gastric dual-layer manual suturing, combined with Dor fundoplication.

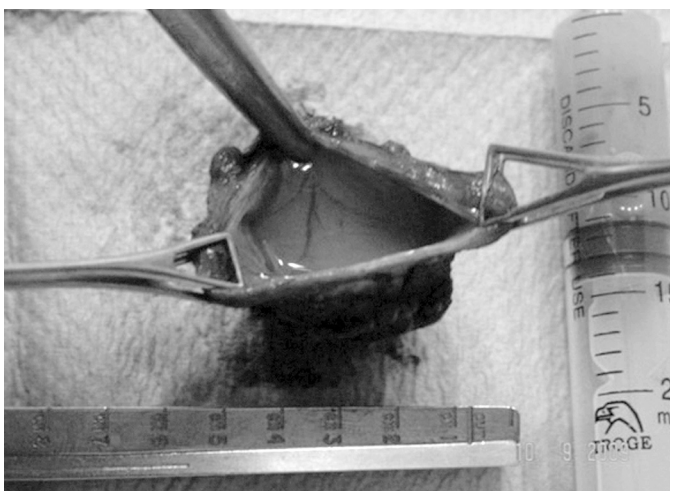

Fig. 3: A unilocular cyst 45 mm in maximum diameter, contained a yellow mucoid material.

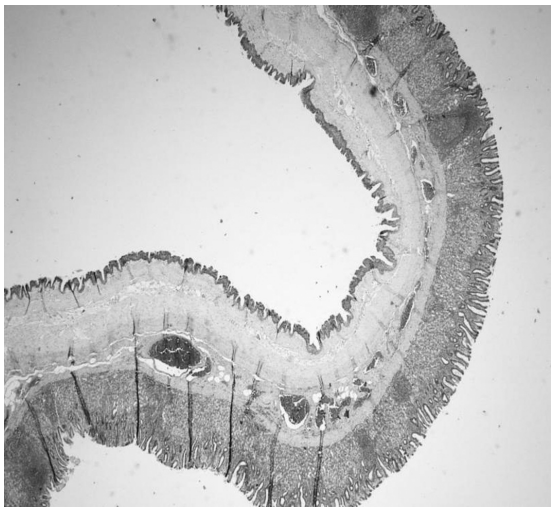

Fig. 4: Wall of the gastric body where it was recognized on the external side wall of the cysts.

she had a past medical history of nephrolithiasis and vertebral abnormalities. Her clinical examination and laboratory tests were normal, including tumor markers (carcinoembryonic antigen and CA19-9). A gastroscopy was performed, showing a 40 mm submucosal process on the gastric fundus, with normal overlying mucosa and tender to pressure with forceps biopsy. Endoscopic ultrasound revealed a intramural mass of the muscularis propria measuring 45 x 33 mm, with ecopattern frankly and homogeneously hypoechoic. The abdominal computed tomography (CT) revealed a low-density mass on the gastric wall, maximum diameter of 50 mm, well circumscribed and with hyperdense margins at the contrast enhancement. Preoperative diagnosis was a GIST. At exploration through an upper midline incision, a cystic mass was found along the greater curvature of the stomach in the esophagogastric transition. While stripping it from gastric wall the cyst was perforated, and fluid leaked from the cyst. Therefore the lesion was excised along with an adjacent sleeve of the stomach and esophagus wall because shared muscular layer with the stomach and esophagus. The gastric and esophageal lumen was exposed (Fig. 1). The primary defect was repaired with esophago-gastric dual-layer manual suturing, combined with Dor fundoplication (Fig 2). Intraoperative histopathological examination gives evidence of a gastric duplication.

The gross observation of the resected specimen showed a unilocular cyst 45 mm in maximum diameter with a clear margin. It contained a yellow mucoid material and there was no communication between the cyst lumen and the stomach (Fig. 3). The final pathologic examination revealed that the inner wall of the cyst was lined by a pseudostratified columnar ciliated epithelium (respiratory type) and, in part, columnar and gastric foveolar epithelium (Fig. 4-5). The cartilaginous tissue was not identified. The recovery was uneventful and the patient was discharged on the seventh postoperative day.

Fig. 5: Detail of the cystic wall covered with a pseudostratified columnar ciliated epithelium.

Discussion

Duplication of the alimentary tract is an unusual congenital anomaly that may occur at any level from the oral cavity to rectum. Ileal duplication are most common, followed by those of the esophagus, colon, jejunum, and stomach [7]. Gastric duplications account for between 3% and 20% of gastrointestinal duplications and occur twice as frequent in females as in males [4].

The pathogenesis is controversial. Several etiopathogenic theories have been proposed to explain their formation: a) persistence of a vacuole formed in the solid phase of bowel embryogenesis or persistence of an embryonic diverticulum; b) failed fusion and recanalization of the longitudinal folds, which would allow an epithelial bridge; and c) formation of a traction diverticulum as a result of a failure in the normal development of the notochord and endoderm [8]. Rowling established several criteria for defining gastric duplication: 1) the cyst wall must be contiguous with the stomach wall, 2) the cyst is surrounded by smooth muscle, which is contiguous with that of the stomach, and 3) the cyst is lined by gastric epithelium or any other type of gut mucosa [9]. In addition, the lining of the cyst may contain pancreatic epithelium and rarely heterotopic mucosa (respiratory) [4,5], and may or may not be calcified [10]. The term "gastric duplication" implies the presence of gastrointestinal mucosa (usually gastric, but may be small intestinal or colonic), the term "foregut duplication" is preferred when pseudostratified ciliated epitheliumpredominates [2,3].The respiratory mucosa is usually found in the esophageal duplication cyst, but is extremely rare in duplication cyst of the stomach [6]. These cysts usually have mucosa and circular muscle layers. The muscular layer of the cyst and the stomach are in close contact and sometimes they share a common muscular layer[1]. Duplications of the stomach are usually single, less than 12 cm in diameter and located on the greater curvature or on the posterior or anterior gastric wall [4]; only 5.5% of them are in the smaller curvature [3].Wieczorek *et al.*[11] reported that gastric duplications can be tubular or cystic; the cystic type does not communicate with the gastric lumen (about 80% of gastric duplication cysts [12]). Review of the literature revealed that the first case of gastric duplication was reported in 1911 [13] and approximately 150 cases have been reported since then [4]. To the best of our knowledge, only 20 adult cases of duplication cyst of the stomach with respiratory lining epithelium, except for our case, have been reported in the English literature[6,14-16].

Approximately 67 per cent of GDC are identified within the first year of life [12]. The diagnosis is made more often in women than men at a ratio of 8:1 [10]. Because duplication cysts of the stomach are very rare in adults, usually asymptomatic and have no specific symptoms and signs, the diagnosis is usually confirmed at laparotomy [1]. A wide range of symptoms and signs have been reported and vary from asymptomatic to non disease-specific presentations, e.g. vague abdominal complaints, nausea, vomiting, epigastric fullness, weight loss, anemia, dysphagia, dyspepsia, etc [4]. Rarely the patients present with gastrointestinal hemorrhage, perforation, malignancy or gastric outlet obstruction [17]. Gastrointestinal hemorrhage can result from intussusception, pressure necrosis of the cystic mucosa due to the expanding cyst, or peptic ulceration of the cyst lining [18]. Communicating cyst may connect with the pancreatic ducts and cause recurrent pancreatitis [19]. Up to 10 per cent of gastric duplication may contain ectopic pancreatic tissue which may be prone to pancreatitis [2]. Physical and laboratory examinations usually reveal no abdominal finding. Ultrasonography, endoscopic ultrasonography, CT scans, and MRI may help the diagnosis to be established preoperatively[1].

Abdominal CT and MRI can identify duplication cysts, but misdiagnosis of solid masses can reach up to 70% depending on the series [8]. The fact that these cysts vary in their content explains this variation in the ability of CT scans to differentiate with accuracy water-attenuation cysts from soft-tissue attenuation cysts; the diagnosis of foregut duplication cyst was certain in only 30% of patients [20]. In the case of complex content, resulting in echogenicity, non-accentuation of the contrast is essential in the differential diagnosis with gastric lesions with predominant development sub-serous and particularly the leiomyoma [21]. Because the majority of duplication are noncommunicating, barium studies and upper endoscopy are often non diagnostic [7]. Woolfolk et al. described the use of EUS for diagnosis and endoscopic cystotomy of a large gastric duplication cyst. The reported cases of preoperative diagnosis of gastric duplication cysts by EUS

were based on the identification of a layered wall structure. However, reliance on this feature alone is not infallible, and errors have been reported [22]. During EUS cyst peristalsis should be considered as a diagnostic feature of this condition [23]. EUS-guided fine-needle aspiration also allows to obtain tissue for cytohistological examination, which is essential for the differential diagnosis with other entities, as well as to exclude malignant transformation of the cyst [8,24]. Gastric duplication cyst, particulary those that have have respiratory-type epithelium, can be diagnosed by EUS-guided FNAB [24]. The demonstration of detached ciliary tufts in cyst fluid by using electron microscopy has been reported [20].

In the case of our patient, the preoperative CT and EUS findings were interpreted as being most consistent with a GIST and no performed EUS-guided FNAB. A misinterpretation of a gastric duplication cyst as a GIST [15,25,26] or leiomyoma [14] on CT scan has been previously reported. However, biopsy should rarely be used to confirm the diagnosis of a resectable GIST, as they can precipitate tumor rupture and lead to tumor dissemination or hemorrhage. The biopsy is indicated if the results will change the management [27].

When in close proximity to the pancreas, it is often difficult to discriminate GDC from congenital cysts or malignant pancreatic cystic tumours. There are many clinical and radiological similarities between gastric duplication cysts and pancreatic pseudocysts. But normal levels of pancreatic enzymes and no pancreatitis in personal medical history may suggest a duplication cyst [1]. A simultaneous increase in CEA and CA 19-9 in GDC in the absence of malignancy [7,28] or in fluid contained in the cyst was reported [28]. Associated anomalies are found in about 50% of gastric duplication cysts [3].

Detection of associated esophageal duplication and vertebral abnormalities in some patients may help the diagnosis [1,3,5,8]. Associated pathologies include pulmonary sequestration, multicystic kidney and gonadal dysgenesis, neoplasias, such as adenocarcinoma and carcinoid, originating in the duplication, and hypergastrinemia [29].

There is no therapeutic algorithm for duplication cysts. Surgical treatment is recommended for symptomatic cases. Management of asymptomatic cases remains controversial. Due to the potential complications and malignant transformation, (six cases reported in literature) [17], some authors considered surgical excision to be the best treatment, even in asymptomatic patients [1,8] or who are aged more than 50 years [15]. The production of oncofetal antigens suggests the possibility of a precancerous condition[28]. However malignant change of GDC lined by respiratory epithelium is rare. There has been no cancer in the previously reported nine duplication cysts of the stomach with pseudostratified columnar ciliated epithelium [6]. Complete resection of the cyst without violation of the gastric lumen whenever possible, is the ideal technique, achieved with both open and laparoscopic approaches [25,30]. Endoscopic resection may be an alternative to surgical intervention in selected patients in whom a GDC presents as a gastric polyp [31].

Marsupialization has been discouraged[5]. Small lesions can be totally excised, but the excision of larger cysts may require gastric resections. Stripping of the cystic mucosa from the common wall is the preferred treatment, so the gastric lumen remains intact [1,5]. However, the enucleation of the cystic mucosa can be very difficult (not a clear cleavage plane between the cyst and the stomach) and may result in a potentially unstable mucosa prone to complications. It was clear from our experience and that of others [30]. If a communication exists between the cyst and the gastric lumen, partial gastrectomy should be done[1]. Successful approaches including percutaneous or endoscopic aspiration of cystic fluid [10] have been reported, but are associated with complications, such as fistula formation and hemorrhage [1].

Conclusion

We encountered a adult patient with foregut duplication of the stomach. Even though a panel of imaging modalities is available, it is still difficult to obtain a preoperative diagnosis. GDC can easily be mistaken for a soft tissue tumor of the gastrointestinal tract. The diagnosis by EUS-guided FNAB can preclude surgery in asymptomatic patients, particularly those that have respiratory-type epithelium. The possibility of malignant transformation within these anomalies must also be considered, particularly in the context of a symptomatic patient.

Riassunto

La duplicazione gastrointestinale è una rara malattia congenita. La duplicazione cistica dello stomaco con epitelio colonnare ciliato pseudostratificato è estremamente rara. La duplicazione gastrica può presentare un dilemma diagnostico. Nella maggioranza dei casi riportati, la diagnosi è stabilita durante l'esplorazione chirurgica. Noi riportiamo il caso di una femmina di 34 anni sofferente di ripetuti episodi di epigastralgia e reflusso gastroesofageo. La TC dell'addome e l'EUS dimostravano una lesione intramurale del fondo gastrico suggestiva di un tumore stromale gastrointestinale (GIST). Alla laparotomia esplorativa veniva trovata una formazione cistica non comunicante con il lume gastrico, lungo la grande curvatura, a livello della giunzione esofagogastrica. La lesione veniva escissa insieme all'adiacente parete dello stomaco e dell'esofago, per la fusione dello strato muscolare. L'esame patologico finale rilevava che la parete della cisti era rivestita da un epitelio colonnare ciliato pseudo stratificato (di tipo respiratorio) e, in parte, da epitelio colonnare e foveolare gastrico. Anche se un pannello di tecniche di imaging è disponibile, è ancora difficile ottenere una diagnosi preoperatoria. Una duplicazione cisti-

ca può essere scambiata per un tumore dei tessuti molli del tratto gastrointestinale. Non esiste un algoritmo terapeutico. Il trattamento chirurgico è raccomandato per i casi sintomatici.

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