

Foregut Duplication Cyst as a Rare Cause of Hematemesis: A Case Report

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ABSTRACT

Duplications of the gastrointestinal tract are rare congenital malformations that usually present in the first two years of life with symptoms of poor weight gain and palpable abdominal mass. We report here a case of a 9-year-old boy who was evaluated for massive upper gastrointestinal bleeding. Upper endoscopy revealed a submucosal large mass in fundus of the stomach with a

3-centimeter diameter ulcer. Our provisional diagnosis was gastrointestinal stromal tumor (GIST) which was later revised to gastric duplication cyst. Gastric duplication cyst is misdiagnosed as leiomyoma or GIST endoscopically and it should be considered in the differential diagnosis of gastrointestinal mass in young children.

Keywords: Duplication Cyst; Stomach; Foregut; Hematemesis

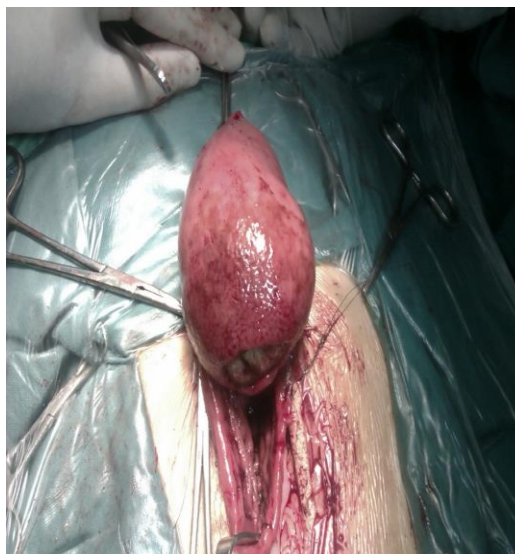
INTRODUCTION

Duplications of the alimentary tract are rare congenital anomalies [1]. Among alimentary duplication cysts, the gastric duplication cyst (GDC) is an uncommon congenital anomaly and is often lined by gastrointestinal mucosa [1]. It accounts for 2-8% of all gastrointestinal duplications [1]. GDC lined by pseudostratified columnar ciliated epithelium (PCCE) is extremely uncommon, and only 17 cases have been reported so far [1]. They can occur anywhere throughout the gastrointestinal tract [2]. Symptoms and signs can include vomiting, abdominal pain, and a palpable mass; in some cases, symptoms related to its complications may also be present [3]. The etiology has not been well characterized yet [2]. Radiographic studies such as ultrasonography and computerized axial tomography may help in making a preoperative diagnosis but it is usually confirmed by surgery [2]. We report here case of a 9-year-old boy with massive gastrointestinal bleeding with histopathological diagnosis of gastric duplication. Our patient presented with hematemesis and melena whereas the duplication cyst was of non-communicating form. In this article, we report a case of foregut duplication cyst, arising from the stomach, which clinically mimicked leiomyoma.

CASE REPORT

A previously healthy 9-year-old boy was admitted in our hospital with complaints of hematemesis and melena. He had loose bloody stool for two days and abdominal pain without nausea and vomiting for one day before presentation. Furthermore, he mentioned symptoms of common cold and coryza. He had no medical history of weight loss. At admission, patient was pale and appeared to be in hypovol-

Figure 1: Cystic lesion in stomach with ulcer and clot on significant artery



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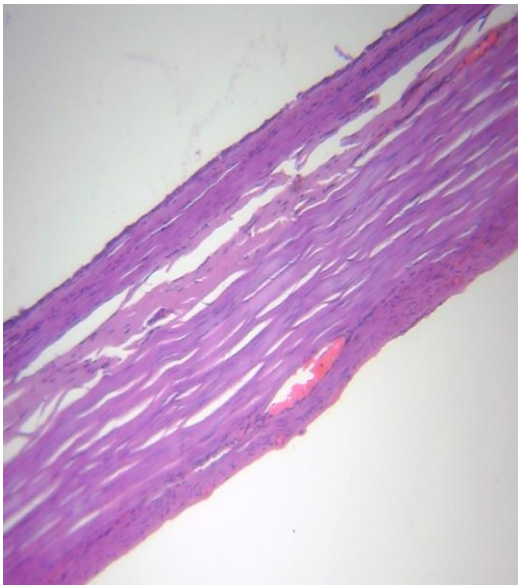
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Figure 2: Cystic lesion removed from the lesser curvature (without mucosa)



Figure 3: Microscopic view of gastric duplication cyst lining



emic shock. His blood pressure, pulse rate, respiratory rate and axillary temperature were 70/50, 120, 24, and 37°C, respectively. On physical examination, we found a 7 x 4 centimeters mobile mass in the epigastric zone. Peripheral pulses were weak and capillary refill was increased (four seconds). On complete blood count, hemoglobin and hematocrit were low (10.6 g/dl and 30.9%, respectively). The coagulation tests revealed no abnormality (platelet count=241×10⁹ per liter, PTT=30 seconds, and PT=12 seconds). Black color of

stools was noted and there were many red blood cells on microscopic examination of stool. Other tests, such as electrolytes, were in normal range. After resuscitation and blood transfusion, endoscopy was performed. During endoscopy, a large submucosal mass in the fundus of stomach with a large 3 centimeter in diameter ulcer was noted with a clot in a significant artery in the crater of the ulcer (Figure 1). Primary impressions for this patient were leiomyoma or GIST. The risk of re-bleeding was high and we were not able to perform other imaging in our center. Therefore, without any further investigations, the child underwent laparotomy. In laparotomy, we found a large mass in the gastric fundus. Most of the mass was palpated inside the stomach wall. Fundus of the stomach was explored, the mass was evaluated and it was found as a 7.5 x 4.5 centimeter duplication cyst. Duplication cyst was non-communicating with the stomach and was associated with large ulcer (three centimeters). Because of operative findings, after ligation of the short gastric vessel, patient underwent segmental resection of the stomach. Following which the stomach was repaired in two layers. Pathological examination of the resected specimen revealed a gastric duplication cyst lined by PCCE (Figures 2, 3 and 4). After operation, the patient had one episode of melena and his vital signs remained stable (blood pressure= 90/55, pulse rate=85, respiratory rate=20, and axillary temperature=37°C). One week after surgery, the patient was doing well.

DISCUSSION

Gastrointestinal tract duplications are rare congenital malformations that can occur anywhere along the gastrointestinal tract from the tongue to the anus. Ileum is the most common site for duplications [2, 4, 5]. Gastric duplication cyst is uncommon and accounts for 2-8% of all gastrointestinal duplications [6].

When there are gastrointestinal mucosa in duplications (usually gastric, but may be small intestinal or colonic), the correct term is "gastric duplication" whereas the term "foregut duplication" is indicated when pseudostratified ciliated epithelium predominates. The last form of duplication cyst of the stomach is extremely rare [7]. A review of 80 cases with duplication cysts showed a 2:1 female to male ratio [2, 7].

Gastric duplications are usually single, non-communicating and are less than 12 centimeter in diameter. The most common sites of occurrence are greater curvature or anterior/posterior gastric

wall [3]. It has been shown that gastric duplication cysts are in no communication with the gastric lumen [8].

Although no evidence of gastric duplication cysts in the first degree persons of a family have been seen in the literature, Paul et al reported gastric duplication cysts in siblings and they recommended evaluation of these cysts in relatives [9]. Gastric duplication cysts have also been called as enterocytoma, heterotrophic gastrointestinal cystic choriostoma [10]. Different kinds of tissue types can be seen in these cysts, such as esophageal, gastric and enteric mucosa and even respiratory type-epithelium [9].

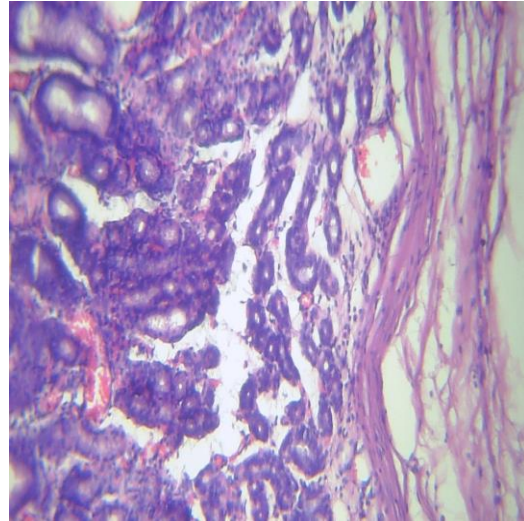
Gastric duplication cysts lined by pseudostratified columnar ciliated epithelium can present late (25-76 years in age) and are seen in females more often than in male. GDC lined by PCCE is easily misdiagnosed as a gastrointestinal stromal tumor. Malignant change in these cysts is rare, but because of persistent exposure to carcinogenic materials, GDC must be considered as a lesion at risk for malignant change in patients who are more than 50 years of age [11]. There is no definite embryologic process that can explain the formation of these cysts, but some theories have been proposed. One hypothesis suggests that some epithelial inclusions cause fusion between primordial structures [10].

In another hypothesis, duplication cysts are a result of an abnormality in the notochord development. In addition to this, abnormal canalization of the alimentary tract may be a probable process for foregut duplications. Most alimentary tract duplications are cystic whereas minority are tubular [12].

Presentation of GDC depends on their size and location. They can present with abdominal pain, vomiting and occasionally a palpable abdominal mass but are usually asymptomatic. Significant hematemesis, melena and anemia are some of the complications of this disease and may be the only presenting symptoms, as in the case of our patient. Other complications are malignant transformation, pancreatitis and perforation with associated acute abdomen [2, 7, 13-16]. A definite association is reported between gastric duplication cysts and other congenital malformations such as intestinal atresia, spinal malformation, intestinal malrotation, and urinary tract anomalies [17].

The majority of duplications are diagnosed in the first two years of life. Before an operation, the diagnosis of gastric duplication cysts can be difficult. An abdominal CT scan with contrast is

Figure 4: Microscopic view of gastric duplication cyst



gold standard for diagnosis of GDC, because it shows the exact margin of cystic lesion and can show the adjacent structures for evaluation of other anomalies. Magnetic resonance imaging (MRI) is another diagnostic method that can determine the cyst and its borders clearly [17]. Definitive management of gastric duplication cysts is to remove them surgically as these are nonfunctional and there is a risk of development of adenocarcinoma in future. The suggested treatment of symptomatic GDC is resection of the cyst without violation of the gastric lumen and an alternative procedure is segmental gastrectomy [18, 19]. Management of asymptomatic patients is controversial but some surgeons believe that duplication cysts should be excised to prevent GI bleeding [1].

CONCLUSION

We report the case of a gastric duplication cyst in a 9-year-old boy. Our patient presented with hematemesis and melena and he did not have any other gastrointestinal symptom. Gastric duplication cyst is misdiagnosed as leiomyoma or GIST endoscopically and it should be considered in the differential diagnosis of gastrointestinal mass in young patients.

REFERENCES

1. Jiang W, Zhang B, Fu YB, Wang JW, Gao SL, Zhang SZ, et al. Gastric duplication cyst lined by pseudostratified columnar ciliated epithelium: a case report and literature review. *J Zhejiang Univ Sci B* 2011; 12:28-31.
2. Faerber EN, Balsara R, Vinocur CD, De Chadareveian

- JP. Gastric duplication with hemoptysis: CT findings. *AJR Am J Roentgenol* 1993; 161:1245–1246.
3. Bartels RJ. Duplication of the stomach. Case report and review of the literature. *Am Surg* 1967; 33:747–752.
 4. Bond SJ, Groff DB. Gastrointestinal duplications. In: O'Neill JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG (eds) *Pediatric Surgery*, 5th edn. Mosby, St. Louis 1998. p. 1257–1265.
 5. Ladd WE. Duplications of the alimentary tract. *South Med J* 1937; 30:363–371.
 6. Kim DH, Kim JS, Nam ES, Shin HS. Foregut duplication cyst of the stomach. *Pathol Int* 2000; 50:142–145.
 7. Stephen TC, Bendon RW, Nagaraj HS, Sachdeva R. Antral duplication cyst: a cause of hypergastrinemia, recurrent peptic ulceration and hemorrhage. *J Pediatr Gastroenterol Nutr* 1998; 26:216–218.
 8. Skandalakis JE, Gray SW, Ricketts RR. The stomach. In: Skandalakis JE, Gray SW, eds. *Embryology for Surgeons*. Baltimore: Williams and Wilkins 1994. p. 150–183.
 9. Davis PL 3rd, Gibson KG, Evans AK. Foregut duplication cysts in siblings: a case report. *Int J Pediatr Otorhinolaryngol* 2010; 74:1331–1334.
 10. Gorlin RJ, Jirasek JE. Oral cysts containing gastric or intestinal mucosa. An unusual embryological accident or heterotopias. *Arch Otolaryngol* 1970; 91:594–597.
 11. Fukumoto K, Suzuki S, Sakaguchi T, Morita Y, Oishi K, Suzuki A, et al. Adenocarcinoma arising from gastric duplication: a case report with literature review. *Clin J Gastroenterol* 2008; 1:148–152.
 12. Veeneklaas GM. Pathogenesis of intrathoracic gastrogenic cysts. *AMA Am J Dis Child* 1952; 80:500–507.
 13. Bonacci JL, Schlatter MG. Gastric duplication cyst: a unique presentation. *J Pediatr Surg* 2008; 43:1203–1205.
 14. Shinde T, Lindner J, Dilverman J, Agrawal R, Dhawan M. Gastric-duplication cyst with an aberrant pancreatic-ductal system: an unusual cause of recurrent abdominal pain. *Gastrointest Endosc* 2009; 69:377–379.
 15. Daher P, Karam L, Riachy E. Prenatal diagnosis of an intrathoracic gastric duplication: a case report. *J Pediatr Surg* 2008; 43:1401–1404.
 16. Takahara T, Torigoe T, Haga H, Yoshida H, Takeshima S, Sano S, et al. Gastric duplication cyst: evaluation by endoscopic ultrasonography and magnetic resonance imaging. *J Gastroenterol* 1996; 31:420–424.
 17. Zhang J, Zhang KR, Bai YZ, Wang W. Thoracoabdominal foregut duplication cyst with respiratory epithelium and alimentary epithelium. *Am J Surg* 2010; 199:54–56.
 18. Oslen JB, Lemmens O, Anderson K. Adenocarcinoma arising in a foregut cyst of the mediastinum. *Ann Thorac Surg* 1991; 51:497–499.
 19. Holcomb GW 3rd, Gheissari A, O'Neill J, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. *Ann Surg* 1989; 209:167–174.