

## Case Report

# Gastric duplication concurrent with congenital pulmonary airway malformation in a newborn

Bin Xu<sup>1</sup>, Wangfang Xie<sup>2</sup>

Departments of <sup>1</sup>Ultrasound, <sup>2</sup>PICU, The Children's Hospital of Zhejiang University School of Medicine, Hangzhou 310003, China

Received January 5, 2017; Accepted January 29, 2017; Epub March 15, 2017; Published March 30, 2017

**Abstract:** Gastric duplication (GD) is a rare gastrointestinal tract malformation, while congenital pulmonary airway malformation (CPAM) is also a rare congenital malformation. We reported a newborn whose transabdominal ultrasonography suggested GD and the subsequent computed tomography (CT) examination indicated concurrent type II CPAM in left lower lobe. The patient received open surgery to remove the gastric mass and the left lower lobe mass at the same time, which were confirmed pathologically to be GD and type II CPAM of left lung. To the best of our knowledge, this was the first report on the special case of GD combined with CPAM in English literature.

**Keywords:** Gastric duplication, congenital pulmonary airway malformation, newborn, malformation, congenital cystic adenomatoid malformation

## Introduction

Gastrointestinal duplication is a rare congenital malformation, among which, gastric duplication (GD) accounts for 2 to 9%, and the pathogenesis remains unclear at present. GD is not associated with specific clinical manifestations, and preoperative diagnosis mostly depends on imaging examination. Congenital pulmonary airway malformation (CPAM), which is also referred to as congenital cystic adenomatoid malformation (CCAM) of lung, is a rare congenital bronchial-pulmonary dysplasia that takes up about 25% of congenital lung malformation, and it is frequently seen in dyspneic newborns. GD may combine with duplication in other sites of the gastrointestinal system or congenital malformation of other systems; while CPAM can combine with congenital malformations such as spine and congenital heart disease. However, as far as we know, there has been no report on GD combined with CPAM. Therefore, we presented a rare case of a newborn boy who was found to have GD combined with type II CPAM.

## Case report

A two-day-old newborn boy went to see a doctor in the outpatient of our hospital as a result of

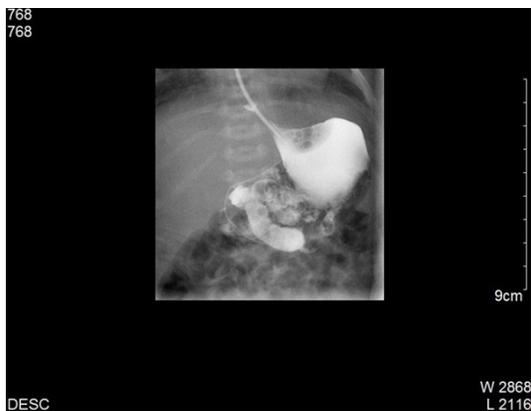
little milk intake after birth. The affected child was the first gestation and first production, and he was born 39 weeks + 6 days after pregnancy through cesarean section, with the birth weight of 3500 g and clear amniotic fluid; the Apgar score at birth was 9-10-10 points, and no asphyxia was seen after birth. Prenatal ultrasonography was completed in the local hospital, which did not reveal congenital malformation. The affected child had normal crying in the visit, with no fever, no convulsion, no cough, no shortness of breath, no dyspnea, no cyanosis, no vomiting or diarrhea, and quiet sleep, and the meconium had been evacuated. The patient had little milk intake after birth, with about 90 ml of milk being consumed within about 24 h.

Physical examination: the patient had fair reaction, flat anterior fontanel, mildly yellowish systemic skin, no chest malformation, symmetric and rough bilateral breath sounds, no obvious rales in both lungs, and regular heart sounds with no obvious murmurs; soft abdominal wall, slightly swollen abdomen, and no epigastric vein could be seen; no obvious enlargement of liver and spleen could be palpated, with fair muscle strength and muscle tone of four extremities, and physiological reflex could be elicited. Laboratory examination: the abnormal indexes in blood routine included: white blood

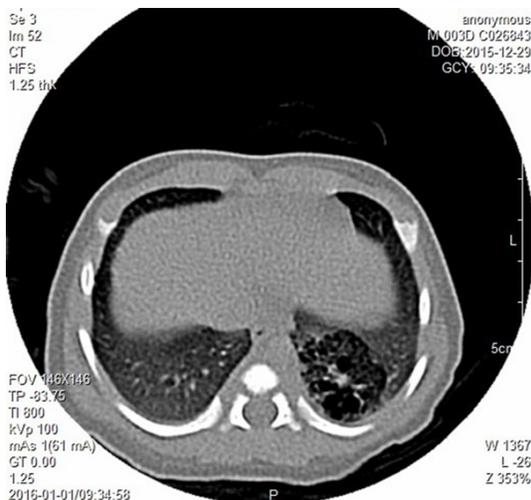
## Gastric duplication concurrent with congenital pulmonary airway malformation



**Figure 1.** Cystic mass (white arrow) in the left upper abdomen, which had the typical “double wall” and “muscular rim” signs.



**Figure 2.** GI suggested a local bright shadow and a quasi-circular filling defect in fundus of stomach.



**Figure 3.** Chest CT revealed a polycystic mass in the left lower lobe, with unequal cyst cavity size.

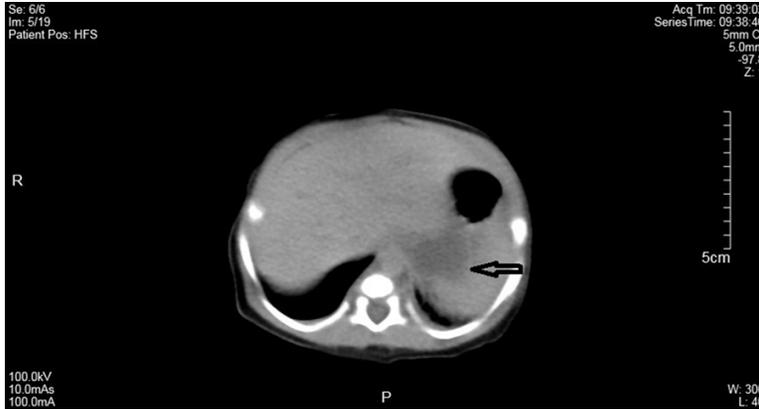
cell count:  $14.92 \times 10^9/L$  (normal value: 15 to  $20 \times 10^9/L$ ), red blood cell count  $4.59 \times 10^{12}/L$

(normal value: 6.0 to  $7.0 \times 10^{12}/L$ ), and hemoglobin 166 g/L (normal value: 170 to 200 g/L). Hypersensitive C-reactive protein was 10 mg/L (normal value: 0 to 8 mg/L).

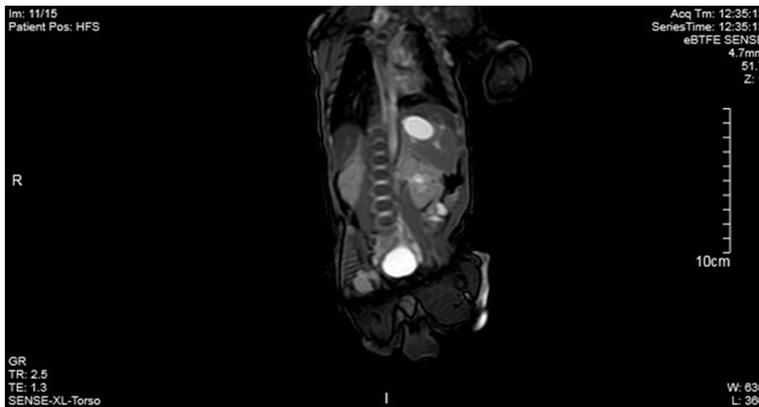
Abdomen ultrasonography was conducted on the day of visit: a cystic mass of about  $1.94 \text{ cm} \times 1.91 \text{ cm} \times 1.87 \text{ cm}$  in size could be seen in the left upper abdomen, the cyst wall of which showed hierarchical structure, and signs of “double wall” and “muscular rim” could be seen (**Figure 1**). Multi-section scanning revealed that boundary could not be drawn between one side of the mass cyst wall and the gastric wall, therefore, the ultrasonic diagnosis was gastric duplication (GD).

The patient was admitted with GD on the day of visit. Upper abdominal gastrointestinal angiography (GI) was performed after admission, which suggested a local bright shadow in fundus of stomach (**Figure 2**), the fundus of stomach showed quasi-circular filling defect after switching all kinds of positions, no contrast was seen in the bright shadow at all time, the pylorus was past smoothly, homogenous contrast distribution could be seen in duodenum, and no morphological or positional abnormality was seen. GI suggested a space-occupying lesion in fundus of stomach. The patient developed tachypnea gradually on the 3<sup>rd</sup> day of admission, thus chest and abdomen conventional CT examination was conducted as well. CT examination suggested that a mass which was constituted by gas-bearing cavities of various sizes could be seen in the left lower lobe (**Figure 3**), which was honeycomb with the cyst cavity of less than 2 cm. A cystic mass with the size of about  $2.2 \text{ cm} \times 1.8 \text{ cm}$  could be seen behind the fundus of stomach on abdomen CT, which was well-defined, with homogenous density in the cavity, the CT value was about 10 HU, the cyst wall was thick and adhered closely to the gastric wall (**Figure 4**). CT suggested type II CPAM in left lower lobe and GD behind the fundus of stomach. The patient received abdomen Magnetic resonance imaging (MRI) examination on the same day. A cystic mass in fundus of stomach could be seen on MRI images, with thick cyst wall and homogenous signal in the cyst cavity; therefore, MRI also suggested GD (**Figure 5**).

Open surgery was performed on the 6<sup>th</sup> day after admission after excluding surgical contraindications. The abdomen was accessed throu-



**Figure 4.** Abdomen CT suggested a homogenous low-density mass behind fundus of stomach, with thick wall that adhered closely to the gastric wall (black arrow).



**Figure 5.** MRI T2 image suggested a cystic mass in fundus of stomach, with smooth border, thick cyst wall and homogenous signal within the cyst cavity.

gh the oblique incision below the costal border in left upper abdomen, a mass with the diameter of about 2 cm could be seen in fundus of stomach, which located near the cardia, and clear boundaries with esophagus could be seen. The gastric body was raised for incision, the mass and gastric wall were separated along mass border, and it could be seen that muscular layer of gastric body was below the mass; the cyst was separated from muscular layer of gastric body carefully, and the mass was removed after being dissociated. The left lateral diaphragm was cut open above the stomach after putting the stomach back into the abdominal cavity, a lesion could be seen in the SX lung segment of left lower lobe after entering the left thoracic cavity, which was well-defined from the surrounding lung tissues; the diseased lung segment was resected along

intersegmental fissure, and the diaphragm was closed with continuous suture, followed by closing the peritoneum, abdominal muscular layers, and skin with layer-by-layer suture. The lesions were confirmed through postoperative pathology to be GD and type II CPAM in the left lower lung (Figures 6 and 7), and inflammatory cell infiltration could already be seen in the GD cyst. The patient had an uneventful recovery postoperatively, and was discharged on the 7<sup>th</sup> day after operation. He pays follow-up visits in the outpatient of our hospital every two months after being discharged, and he has been followed up for 5 times. At present, he has normal food intake, digestion and respiration, as well as height and weight.

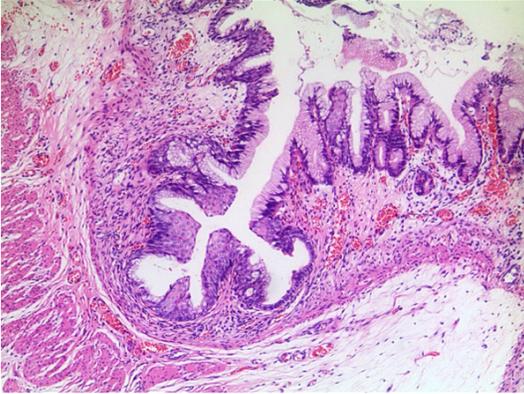
### Discussion

Gastrointestinal duplication is a kind of rare congenital malformation, which is more common in ileum (35%), stomach (9%) and colon (7%) [1]. Gastric duplication (GD) frequently

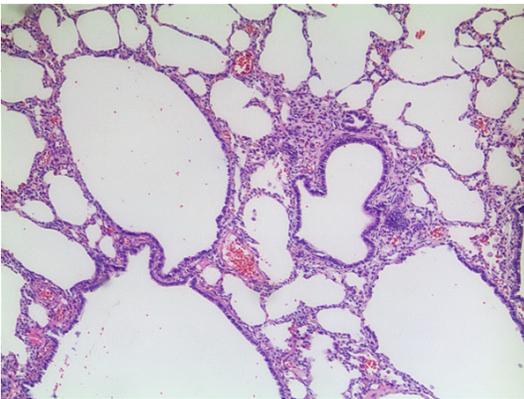
occurs in female, with the male-to-female morbidity ratio of about 1:2-8. GD can be divided into two types, namely, cyst type and tubular type, with the former being more common; most cyst type GD does not connect with the gastric cavity, and mostly locates in greater curvature; while the latter can be connected with the gastric cavity.

GD shows no specific clinical manifestations, and the common clinical manifestations are nausea, vomiting, difficulty in feeding, vague stomachache, abdominal mass, and weight loss; therefore, clinical diagnosis depends greatly on imaging examination. At present, there are different opinions on which kind of imaging examination leads to a better diagnosis. Some scholars consider that CT is the optimal imaging examination method [2], which

## Gastric duplication concurrent with congenital pulmonary airway malformation



**Figure 6.** A smooth muscle cyst wall lining the gastric mucosa could be seen through hematoxylin and eosin (HE) staining under the 100-fold light microscope.



**Figure 7.** Type II CPAM could be seen under the 100-fold light microscope. Multiple cyst cavities could be seen on the section, with cyst wall lining with metaplastic squamous epithelia.

mostly manifests as a mass with smooth border, fluid attenuation in the cavity, thick cyst wall with one side adhering closely to the gastric wall, and intensified border [3]; but it is believed in some research that CT can hardly diagnose the nature of mass in an accurate way [4]. Our case has typical manifestations on CT, including a cystic mass, with smooth border, and thick cyst wall, which adheres closely to gastric wall; consequently, we can make an accurate diagnosis. Our case reveals a typical cystic mass in gastric area on MRI, which is well-defined, with homogenous signal in the cavity, and thick gastric wall, thus, GD is suggested. However, some scholars suggest [4] that MRI does not improve the accurate diagnosis rate of GD. GD can hardly be directly discovered in most gastrointestinal images, but

such images can reveal gastric cavity compression, allow for observing the gastric cavity and gastric wall, and can accurately show whether the mass is connected with gastric cavity. There are also studies treating gastrointestinal imaging as the examination method in postoperative follow-up [2]. The ultrasonography in this case clearly displays the characteristic “double wall” and “muscular rim” signs of the lesion cystic wall [5] and fluid sonolucent area in the cyst cavity; the boundary between one side of cyst wall and gastric wall can not be drawn, which forms the common gastric wall; therefore, ultrasonography directly makes a diagnosis of GD. The preliminary diagnosis of GD in this case is completed through ultrasonography. The diagnosis can be more accurate if ultrasonography can clearly distinguish the normal gastric wall structures, such as mucosal layer, submucosal layer, muscular layer and serosal layer in the cyst. In addition, endoscopic ultrasound (EUS) and EUS guided fine needle aspiration may aid in diagnosis [6].

About 50% GD can combine with congenital malformations in other sites, and duplication or atresia in other sites of gastrointestinal system, as well as spine malformation is commonly seen. There have been reports on esophageal duplication combined with type I CCAM [7, 8], but to the best of our knowledge, GD combined with CCAM (CPAM) has not been reported before in English literature.

CPAM, which was also called CCAM, was first proposed by Chin and Tang in 1949, and it was renamed as CPAM by Stocker afterwards. The morbidity of CPAM in live birth infants is 1:25000 to 1:35000 [9], which accounts for approximately 25% of congenital lung malformation. It can occur in all lobes of both lungs, with comparable incidence in both lungs, and it mostly occurs in a single lung and a single lobe. CPAM is a kind of lung dysplasia that is characterized by excessive growth of bronchiole, is associated with bronchial atresia, and thus gives rise to hamartoma-like developmental malformation; and it is more commonly seen in dyspneic newborns and children with recurrent pneumonia. According to Stocker classification [10], it can be divided into 5 types based on gross specimen and histological findings, which are type 0, I, II, III, and IV. Type II accounts for 15 to 30%, with the cyst diameter

## Gastric duplication concurrent with congenital pulmonary airway malformation

of <2 cm, the section shows honeycomb, with no cartilage or mucous glands in the cyst wall. The whole lesion shows dilated bronchus-like structure.

CPAM is mostly diagnosed through prenatal ultrasonography, but it is not suggested in prenatal ultrasonography in this case, as a result, we only conduct abdomen ultrasonography but not lung ultrasonography. It is suggested in numerous studies [9] that CT is more superior to ultrasonography in diagnosing newborn CPAM, and it is the preferred examination method that can clearly display the lesion morphology as well as cyst cavity size, in addition, it allows for classification; MRI can replace CT examination, but it has no more advantages over CT apart from free of radiation. CT examination is performed on this case on admission, and CPAM in left lower lobe can be found; CT can clearly display the CPAM lesion and gives out accurate classification, but MRI is mainly performed on abdomen, no detailed MRI examination is conducted on lung, and no definite CPAM is found on the only coronary lung T2 image.

There are numerous hypotheses attempting to explain the pathogenesis of gastrointestinal duplication, but none of the hypothesis can completely explain all GD so far [11]. There are theoretical studies on bronchopulmonary foregut malformations (BPFM) and lung-bud foregut malformation (LBFM) [7], which account for congenital malformations in respiratory system and digestive system deriving from the foregut. Approximately in the 4<sup>th</sup> week of embryogenesis, the foregut tissue is gradually divided into the dorsal digestive tract primordium and the ventral respiratory system primordium; afterwards, the foregut develops into the mouth of digestive system to the ampulla of duodenum, as well as respiratory system tissues like airway, lung and pleura. Therefore, we speculate that common origin of foregut during embryogenesis is the pathological foundation of GD combined with type II CPAM in this case, but it requires to be further investigated.

GD is likely to be concurrent with inflammation, hemorrhage, ulcer and perforation, and there are also reports on carcinogenesis in adults [1, 12] and even peritoneal metastasis [13]; therefore, it is suggested in numerous studies that timely resection should be performed on GD. Open surgery is frequently adopted previously,

and some scholars at present remove the lesion under laparoscope. There are controversies regarding whether surgery should be conducted on the asymptomatic CPAM, while surgery is mostly recommended for symptomatic CPAM. Early lesion resection may contribute to the further development of normal lung tissues in newborns [9]. Transabdominal resection of GD and CPAM lesions is performed in this case at the same time, which has reduced surgical incisions on body surface and the patient had an uneventful recovery. The affected child pays regular follow-up visits every two months after being discharged, and he has been followed up for 10 months. The boy shows no abnormalities in food intake, digestion and respiration, and the height and weight fall within normal range.

This case suggests that GD may be concurrent with CPAM, therefore, they should be considered in imaging examination. In the discovery of any one of the two diseases, the scope of examination should be expanded to eliminate the possibility of another malformation, and imaging examination allows for accurate diagnosis of GD and type II CPAM, then open surgery could be remove them at the same time.

### Disclosure of conflict of interest

None.

**Address correspondence to:** Wangfang Xie, Department of PICU, The Children's Hospital of Zhejiang University School of Medicine, No. 57 Zhugan Lane, Xiacheng District, Hangzhou 310003, China. Tel: +86 57188873761; Fax: +86 57188873761; E-mail: xwfhz@hotmail.com

### References

- [1] Zheng J and Jing H. Adenocarcinoma arising from a gastric duplication cyst. *Surg Oncol* 2012; 21: e97-101.
- [2] Scatizzi M, Calistri M, Feroci F, Girardi LR, Moraldi L, Rubio CA, Moretti R and Nesi G. Gastric duplication cyst in an adult: case report. *In Vivo* 2005; 19: 975-978.
- [3] Hur J, Yoon CS, Kim MJ and Kim OH. Imaging features of gastrointestinal tract duplications in infants and children: from oesophagus to rectum. *Pediatr Radiol* 2007; 37: 691-699.
- [4] Laurent S, Vivario M and Lardinois F. Gastric duplication cyst with respiratory epithelium: a rare entity. *Acta Chir Belg* 2015; 115: 379-381.
- [5] Sutcliffe J and Munden M. Sonographic diagnosis of multiple gastric duplication cysts cau-

## Gastric duplication concurrent with congenital pulmonary airway malformation

- sing gastric outlet obstruction in a pediatric patient. *J Ultrasound Med* 2006; 25: 1223-1236.
- [6] Eloubeidi MA, Cohn M, Cerfolio RJ, Chhieng DC, Jhala N, Jhala D and Eltoun IA. Endoscopic ultrasound-guided fine-needle aspiration in the diagnosis of foregut duplication cysts: the value of demonstrating detached ciliary tufts in cyst fluid. *Cancer* 2004; 102: 253-258.
- [7] Kuga T, Inoue T, Sakano H, Zempo N, Oga A and Esato K. Congenital cystic adenomatoid malformation of the lung with an esophageal cyst: report of a case. *J Pediatr Surg* 2001; 36: E4.
- [8] Hasegawa S, Koga M, Matsubara T, Oga A and Furukawa S. Congenital cystic adenomatoid malformation complicated by esophageal duplication cyst in a 6-month-old girl. *Pediatr Pulmonol* 2002; 34: 398-401.
- [9] David M, Lamas-Pinheiro R, Henriques-Coelho T. Prenatal and postnatal management of congenital pulmonary airway malformation. *Neonatology* 2016; 110: 101-115.
- [10] Stocker JT. Congenital pulmonary airway malformation: a new name and an expanded classification of congenital cystic adenomatoid malformation of the lung. *Histopathol* 2002; 41 Suppl: 424-431.
- [11] Bonacci JL, Schlatter MG. Gastric duplication cyst: a unique presentation. *J Pediatr Surg* 2008; 43: 1203-1205.
- [12] Zhu Y, LV L, Pan W, Ren P, Han T and Xu X. Gastric duplication complicated by malignant transformation in adults: report of three cases. *J Gastrointest Dig Syst* 2015; 5: 374.
- [13] Liu K, Lin X, Wu J, Meng M, Su H, Tai W and Chang H. Peritoneal metastatic adenocarcinoma possibly due to a gastric duplication cyst: a case report and literature review. *BMC Gastroenterol* 2014; 14: 48.