Duplication Cyst of Pyloric Canal: A Rare Cause of Pediatric Gastric Outlet Obstruction: Rare Case Report

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Abstract Duplication of the alimentary tract may occur in any site, from the mouth to the anus, and in recent years such anomalies have received wide notice. Pyloric duplication cyst is an extremely rare congenital anomaly of the alimentary tract, whose clinical presentation often mimics those of hypertrophic pyloric stenosis. Gastrointestinal duplications are observed in 1 of every 4500 autopsies, predominantly in white males. It represents 2.2 % of all gastric duplications, with only 18 have been reported up to 2011 (Table 1). In most cases preoperative diagnosis is not made. We report a case of a pyloric duplication cyst in a 3-year-old girl with progressive increased vomiting. The patient had an ultrasonography, upper gastrointestinal series, and computer tomography of the abdomen. The diagnosis was confirmed by surgery and histopathology examination. The patient was asymptomatic at 12-month follow-up. The clinical and radiological analysis can reveal configurational changes consistent with a large extrinsic mass rather than muscular hypertrophy and can lead to accurate preoperative diagnosis.

Keywords Pyloric duplication · Hypertrophic pyloric stenosis · Gastric outlet obstruction · Enucleation · Pyloroantrectomy

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Case Presentation

A 3-year-old girl, weight 10.2 kg, was presented to us with history of abdominal fullness, pain in abdomen, and recurrent nonbilious projectile vomiting after taking feeds for 2 months. Ultrasonography of the abdomen revealed a well-defined cystic lesion in epigastric region measuring 2.7 cm×2.9 cm in close proximity to inferior surface of the left lobe of the liver. Upper gastrointestinal barium study was suggestive of narrowing of pylorus due to soft tissue opacity causing external pressure effect. CT scan was suggestive of the congenital duplication cyst at pylorus. Clinical diagnosis was idiopathic congenital hypertrophic pyloric stenosis, but radiological investigations were suggestive of the pyloric duplication cyst. The patient was prepared for elective laparotomy via transverse supraumbilical incision and externalized the pylorus. There was a cystic mass of about 5 cm×4 cm at the greater curvature of pylorus, obstructing but not communicating with the patent lumen of the pyloroduodenal canal. The cystic mass was enucleated without damage to the underlying pyloric channel and primary repair was done. The excised cyst contained yellowish mucoid fluid. On histological examination, the common wall had pyloric mucosal lining at places showing necrosis with mild chronic inflammation with fibromuscular tissue, consistent with clinical diagnosis of the congenital duplication cyst of pylorus (Figs. 1, 2, 3, 4, 5 and 6) (Table 1).

Discussion

Pyloric duplication cysts are an extremely rare congenital anomaly, with incidence of 1 in 4500, predominantly in

ULTRASONOGRAPHY

Fig. 1 A well-defined cystic lesion

white females [1, 2]. They present with symptoms of gastric outlet obstruction and palpable mass which is clinically indistinguishable from IHPS. Enteric duplications can occur anywhere along the GI tract, but are most commonly found

BARIUM STUDY

Fig. 2 Narrowing of pylorus

C.T.SCAN

Fig. 3 A 2.7×2.1 cm well-defined cystic lesion

in the ileum and are perhaps the rarest along the pyloric channel and represent 2.2 % of all gastric duplications. The earliest case of a juxtapyloric cyst was reported by Ramsay in 1957. Till now only 18 cases have been reported so far in the English literature. In the 18 patients, excluding the present patient, with true duplication cysts, 15 presented

INTRAOPERATIVE FINDINGS

Fig. 4 Pyloric cyst at greater curvature of stomach

Fig. 5 Enucleation and excision of cyst

with persistent vomiting and 10 had a palpable abdominal mass. All duplications were noncommunicating. Upper gastrointestinal study may show pyloric narrowing but may not demonstrate the duplication. Ultrasound, however, will indicate the degree of pyloric hypertrophy and, if one is present, a duplication.

The etiology of duplications of this area is unknown, but the diverticulum theory of Lewis and Thyng and the Bremer theory of failure to recanalize are often mentioned; none completely explains the origin [1, 2]. Duplications of the alimentary tract are embryologic deviations differing from

vitello-intestinal remnants, or dorsal enteric remnants [3]. In this respect, the theory of failure to recanalize is often mentioned in the literature and could explain to some extent the origin of pyloric duplication [2]. The most widely accepted theory has been offered by Bremer [2]. Bremer has proposed that these anomalies represent errors of canalization between week 6 and week 8 of embryogenesis.

The usual location of pyloric duplication is along the greater curvature. More often, there is no communication with the stomach. Associated anomalies can occur in up to 50 % of cases and can include other alimentary tract duplications and vertebral defects [4].

The recommended criteria for diagnosis of duplication cyst include (i) proximity to the alimentary tract; (ii) common blood supply; (iii) outer smooth muscle coat sharing that is usually the gastric muscle coat; and (iv) alimentary epithelial lining (not necessarily of the adjacent gut).

Symptoms vary according to the location of the cyst and type of mucosal lining. The usual presentation is with abdominal mass, vomiting, weight loss, and failure to thrive [5]. In older children, it may present with abdominal pain, gastrointestinal bleeding, and fever [5]. These complications are attributed to inadequate drainage of the pent-up acid pepsin secretions, leading to ulceration, hemorrhage, and perforation. Although the usual age of presentation is early infancy, most are manifested by 1 year of age [5].

Surgery is the undisputed line of management but the surgical option varies. It is important that the surgery be individualized to each case. The surgeries performed have varied from simple excision to pyloroantrectomy. The stomach has blood supply on both sides unlike the mesenteric blood supply of the intestines [6]. Hence instead of resection of the involved segment of the bowel, it is sufficient to excise the cyst alone with the stripping of the mucosal lining; the raw surface being patched with transverse colon (for large defects) [6] or omentum [7], or even leaving a seromuscular gap, like following a Ramstad's pyloromyotomy [1]. This proposed "closed" approach allows for effective relief of obstruction, a rapid postoperative recovery, and the early reinstitution of feeding after 12 hrs. There are reports of excision procedures such as a limited pyloroantrectomy [8]. In all surgical procedures, it is essential to remove all the cyst lining [9]. Drainage procedures are only rarely advisable like when extensive resection would otherwise be necessary because of huge size or close proximity to the common duct etc. Procedures such as cystgastrostomy, cystenterostomy Roux-en-Y, and a "window" procedure have all been done in various situations. Marsupialization

HISTOPATHOLOGY

Fig. 6 Pyloric mucosal lining

Table 1 List of cases till now with management

Studies	Age at presentation	SEX	Size of cyst (cm)	Diagnostic study	Surgical treatment
Ramsay (1957)	8 days	F	3.0×2.0	-	Local excision
Abram & Dennison (1961)	7 days	F	2.0	UGI	Posterior gastrojejunostomy
Grosfeld et al. (1970)	3 days	M	1.8×1.0×0.4	-	Cyst excision
Anas & Miller (1971)	12 days	M	2.0×3.0	UGI	Cyst mucosal stripping
Bower et al. (1978)	3 weeks	?	1.5	?	Cyst excision
Keramidas et al. (1980)	20 days	F	3.0×2.0	UGI	Cyst excision
Teele et al. (1980)	5 months	M	Unknown	UGI AND US	Cyst excision
Galifer & Ferran (1980)	11 months	M	5.0	UGI	Pyloroantrectomy
Birenbaum et al. (1982)	2 days	F	2.0×3.5	UGI AND EGD	Cyst excision
Novak & Boeckman (1983)	2 months	M	3.0×1.5×1.5	UGI AND CT	Cyst excision
Bommen & Singh (1984)	1 day	M	3.0	UGI AND US	Cyst excision
Tihanski et al. (1986)	6 weeks	F	5.0	UGI	Cyst excision with Gastroduodenostomy
Goyert et al. (1991)	Perinatal	F	5.0×6.0	US	Cyst excision
Murty et al. (1992)	Newborn	M	5.0×8.0	UGI AND US	Cyst excision
Cooper et al. (1995)	5 weeks	M	0.8×0.5×0.3	UGI	Cyst excision
Manish et al. (1997)	3 days	M	8.0×5.0	US	Pyloroantrectomy
Shah et al. (2005)	3 days	F	6.0×5.0	US	Pyloroantrectomy
M Iwasaki et al.(2007)	0 months	F	1.4×2.2×1.2	US AND CT	Cyst excision and local excision
Present study	2 months	F	5×4	US AND CT	Cyst excision and local excision

has to be avoided because carcinoma has been found in adult duplications [4].

Asymptomatic cysts should possibly be removed because of possible late complications [5].

The results depend on the type of duplication, the presence of complications, and especially on the associated malformations [1].

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