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Case Report

Double Jeopardy- Hypertrophic Pyloric Stenosis and Pyloroduodenal Duplication Cyst in A Neonate

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Abstract

Duplication cysts of the pylorus are the rarest of all gastrointestinal duplications and the co-existence with infantile hypertrophic pyloric stenosis (IHPS) is anecdotal at best. Surgical excision with minimum collateral damage is the mainstay of treatment. Palpation of the pyloroduodenal region with adequate pyloromyotomy is essential for good results. Here we chronicle the case of a neonate with this unusual pathology.

Keywords: *Duplication Cyst; Infantile Hypertrophic Pyloric Stenosis; Pyloromyotomy*

Abbreviations

IHPS: Infantile Hypertrophic Pyloric Stenosis

Introduction

The first description of a pyloric cyst simulating IHPS was by Ramsay., *et al*. in 1957 [1]. In Grosfeld's series, a pyloric location was most uncommon and seen in only 4.3% [2]. Others have quoted a much lower incidence and it remains one of the rarest of duplications with only a handful of reports found in literature. Further, a pyloroduodenal cyst has been reported by only three groups till now [3-5].

Case Report

A 22-day old boy presented with a history of non-bilious, projectile vomiting since birth and refusal of feeds for the past one day. Delayed passage of meconium or stooling issues were not reported. Antenatal imaging had been non-contributory. The history was significant for the presence of a palpable lump in the abdomen especially after feeds (as noticed by the mother). The child was dehydrated and icteric but otherwise hemodynamically stable at presentation with a venous blood gas suggestive of metabolic alkalosis.

Abdominal examination revealed a fullness in the upper abdomen with visible peristalsis from the left hypochondrium towards the right. A 3 x 2 cm firm, globular lump (the "olive") was palpable in the periumbilical region towards the right. Ultrasonography revealed a thick-walled cyst with a diameter of 15 mm, supero-lateral to the pylorus (Figure 1a). The pyloric length was 14.1 mm with a thickness of 16.7 mm. An oral contrast study was corroborative and showed a grossly dilated stomach with pyloric stenosis (Figure 1b).

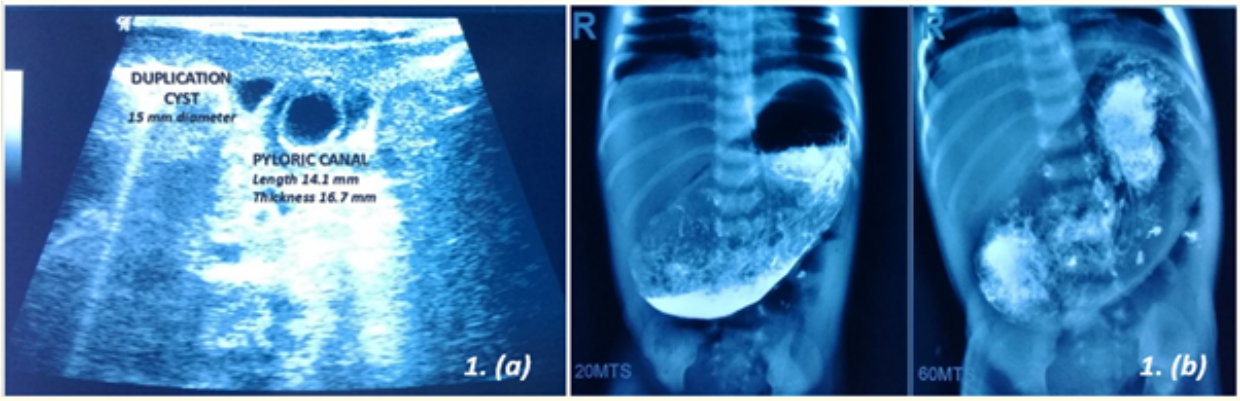

Figure 1 (a): *Cyst and hypertrophic pyloric muscle seen on USG* ***(b):*** *Oral contrast study and retention in delayed film.*

Laparotomy after appropriate fluid resuscitation showed a dilated stomach with a 2 x 2 cm globular cyst within the antero-superior aspect of the pyloric channel extending till the first part of the duodenum (Figure 2a). Partial Kocherization of the duodenum revealed the complete extent of the cyst. The layers of thick muscle and the cyst wall were divided till the mucin filled core. There was no apparent communication with the stomach. The mucosa was lifted between stays and lifted off till the submucosa of the actual pylorus pouted out. A wedge of muscle was also resected (Figure 2b). Further palpation guided the extension of this incision till the distal antrum. Gastric contents were pushed with ease across the pylorus suggesting an adequate pyloromyotomy. A negative saline leak test assured us of the integrity of the mucosa.

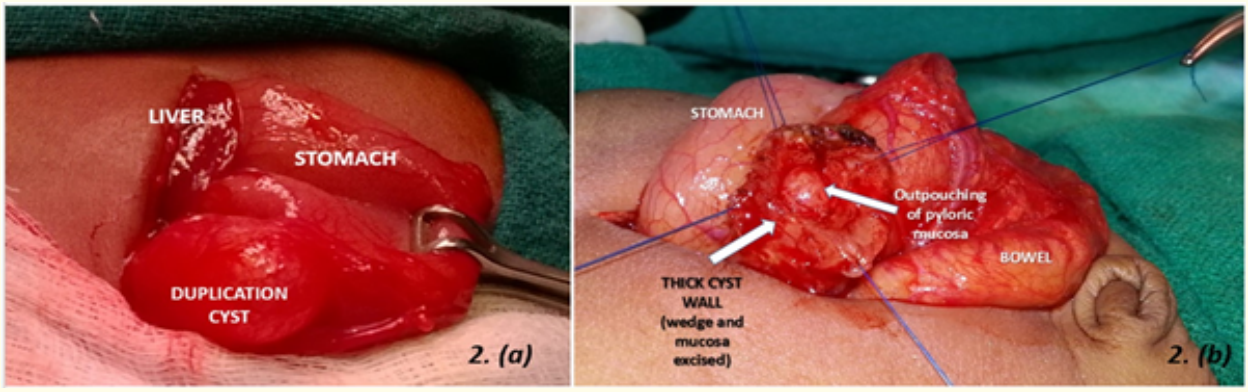

Figure 2 (a): *Duplication cyst in the pyloroduodenal region* ***(b):*** *After excision of cyst- pouting gastric mucosa is seen.*

Oral feeds were started on the first post-operative day. Histology was consistent with intestinal epithelium and mild subepithelial fibrosis. The child had an uneventful recovery.

Discussion

It is worthwhile to recapitulate Ladd's classic description of an alimentary duplication cyst here: a well-developed coat of smooth muscle, presence of epithelial lining resembling some part of the alimentary tract and an intimate attachment to at least one point of the alimentary tract. A multitude of theories have been put forward to explain the whole spectrum of GI duplications [6].

The symptoms of recurrent non-bilious vomiting, upper abdominal distension and a variable palpable mass in this age group suggest IHPS. The other differentials include pyloric atresia, antral web, gastric volvulus and of course, gastric duplication. This entity has also previously been confused with a choledochal cyst due to its subhepatic location.

Goyert., *et al.* were the first to diagnose it in the antenatal period in 1986 [7]. Despite a variety of ultrasonographic findings described in literature, a cystic appearance (often with echogenicity due to debris and mucus) is common. The "muscular rim sign" delineating the echogenic inner mucosa from the hypoechoic outer rim of muscle is indicative of a gastrointestinal origin. Conventional contrast studies give a general idea of the degree of obstruction and the extrinsic compression but are quite non-specific.

Only rarely is a communication with the cyst demonstrated. Upper GI endoscopy has also been utilized but feasibility issues in neonates limit its utility. Others have used multiplanar reformatted computed tomography of the abdomen with Gastrograffin as the contrast agent to confirm equivocal ultrasound findings [8].

Many options for surgical management exist but the core concept is a complete excision of the cyst or the mucosa at the least. The choice should be made on the basis of the anatomy of the cyst and the general condition of the child. If the common wall between the cyst and the stomach is amenable to dissection, a simple excision is often possible. In the event of an intimate relation of the cyst with the native bowel, the cyst and common wall should be removed with a reconstruction of the defect to avoid repeated inflammation from the remnants. Pyloroantrectomy with gastroduodenostomy has also been done [6].

To avoid the morbidity of an excision in the pyloroduodenal region, as in our case, the cyst may well be marsupialized, evacuated of contents and the epithelial lining excised. The additional hypertrophy of the pylorus in this child necessitated a formal pyloromyotomy after excising the mucosa of the cyst in entirety. Fisher., *et al*. [9] labelled this the "closed" approach and equated it with faster post-operative recovery.

As regards the co-existence of IHPS with gastroduplication, whether the hypertrophy is a consequence or coincidence remains in question. The clinical presentation and the hypertrophic muscles and mucosa of the pylorus were typical of IHPS. In the literature, there has been only one report that has described the coexistence of IHPS and gastroduplication [10].

Conflict of Interest: No financial interest or conflict of interest exists.

Bibliography

1. Ramsay GS. "Enterogenous cyst of the stomach simulating hypertrophic pyloric stenosis". *British Journal of Surgery* 44.188 (1957): 632-633.

2. Grosfeld JL., *et al*. "Enteric duplications in infancy and childhood: an 18-year review". *Annals of Surgery* 172.1 (1970): 83-90.

3. Tihansky DP., *et al*. "Pyloroduodenal duplication cyst". *American Journal of Gastroenterology* 81.3 (1986): 189-191.

4. Hamada Y., *et al*. "Pyloroduodenal duplication cyst: case report". *Pediatric Surgery International* 12.2-3 (1997): 194-195.

5. Upadhyaya VD., *et al*. "Duplication cyst of pyloroduodenal canal: a rare cause of neonatal gastric outlet obstruction: a case report". *Cases Journal* 2 (2009): 42.

6. Shah A., *et al*. "Pyloric duplication in a neonate: a rare entity". *Pediatric Surgery International* 21.3 (2005): 220-222.

7. Goyert GL., *et al*. "Prenatal diagnosis of duplication cyst of the pylorus". *Prenatal Diagnosis* 11.7 (1991): 483-486.

8. Tang XB., *et al*. "An intraluminal pyloric duplication cyst in an infant". *Journal of Pediatric Surgery* 43.12 (2008): 2305-2307.

9. Fisher R., *et al*. "Pyloric duplication in the newborn: A rare cause of gastric outlet obstruction". *Journal of Indian Association of Pediatric Surgeons* 12.1 (2007): 34-35.

10. Hishiki T., *et al*. "A rare presentation in a case of gastric duplication cyst communicating to the pancreatic duct: coincidental detection during pyloromyotomy for hypertrophic pyloric stenosis". *Journal of Pediatric Surgery* 43.9 (2008): e1-e3.

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