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Duplication cyst of the pylorus: A rare cause of gastric outlet obstruction

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CASE REPORT

Duplication cyst of the pylorus: a rare cause of gastric outlet obstruction

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SUMMARY

Alimentary tract duplications are a rare congenital malformation. They can present with varied symptoms owing to the locality of the duplication, along the gastrointestinal tract. Out of these duplications, the ones along the pylorus are the most rare. These are usually only diagnosed intraoperatively, as it is not a common differential on imaging due to its rarity. In lieu of the literature currently available, pyloric duplication cyst can present anytime from 1 week of age to 5 years, with some cases being detected antenatally due to the prevalence of regular antenatal scanning. Surgery remains the main stay of treatment with the goal of complete excision of the cyst and complete removal of the cyst mucosal lining. We report the case of a 5-year-old girl, which to our knowledge is the first ever reported case from Karachi, Pakistan.

BACKGROUND

Congenital pyloric duplication cyst is one of the exceedingly rare forms of alimentary tract duplications, with only 22 cases reported in English literature.[1] Majority of these are extraluminal masses more often arising from the pyloroduodenal junction. Due to its rarity, diagnosis is almost always never made before surgery, as the threshold for suspicion of a duplication cyst arising from the pylorus remains relatively high. We present a case of pyloric duplication cyst presenting as progressive gastric outlet obstruction in a 5-year-old girl, to report our experience with its surgical management.

CASE PRESENTATION

A 5-year-old girl presented to our outpatient clinic. She had been experiencing incoercible vomiting and abdominal pain for the past 2 months. The abdominal pain was the first symptom to manifest. Progressive vomiting, which was non-bilious and non-projectile in nature, then followed. The frequency of vomiting increased over the course of a month; thus on presentation, she was unable to consume solids or liquids orally. The child's previous history was unremarkable. Her prenatal and birth history were unremarkable. She had no siblings and the parents did not recall any children being born with any congenital malformations within the family. At the time of presentation, her clinical examination was unremarkable, with no mass appreciated on palpation.

INVESTIGATIONS

On admission, her laboratory investigations revealed: haemoglobin level of 15.20 g/dL, total leucocyte count of 9.90×10^9/L, prothrombin time of 12 s and partial thromboplastin time of 25 s. Liver function test was also within normal limits. An ultrasound of the abdomen revealed a thick-walled, round cystic mass approximately measuring 4.6×4.0 cm in the region of the pylorus and antrum, which was protruding into the lumen of the stomach causing gastric outlet narrowing. The cyst appeared to be dual walled with an echogenic component and minimal septae. Along with this, few of the mesenteric lymph nodes were enlarged (figure 1). A CT scan revealed a cystic mass compressing the antrum and pyloric channel causing hold-up of barium meal. The mass itself took up no contrast (figure 2).

DIFFERENTIAL DIAGNOSIS

We made the preliminary differentials of a gastric duplication cyst and choledochal cyst.

TREATMENT

An open laparotomy was undertaken for assessment and cyst removal. At laparotomy, a cystic mass was identified anterior and lateral to the pyloric channel (figure 3).

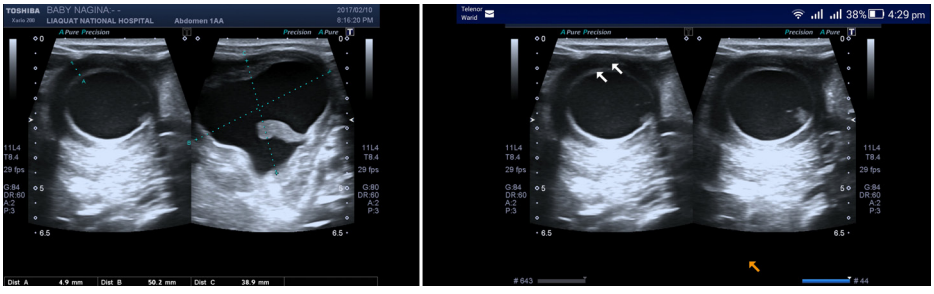

Figure 1 Ultrasound showing duplication cyst. Dual wall of the cyst (white arrows) can also be appreciated.

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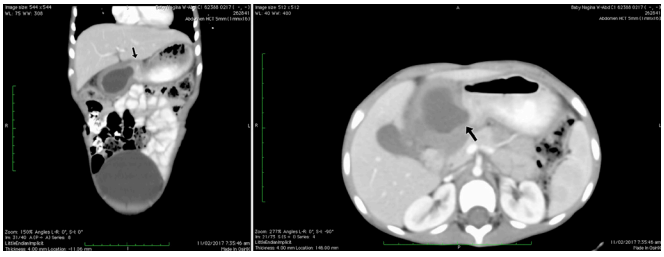

Figure 2 CT scan of the abdomen with cystic lesions in close proximity to pylorus sharing a common wall (arrow). Both coronal and transverse planes show no follow-through of barium within cyst.

The cystic mass appeared to compress the pyloric channel, resulting in gastric outlet obstruction. The duplication cyst did not communicate with the pyloric channel but did share a muscular wall with the antimesenteric border of the pylorus. Majority of the cyst was excised and the entire mucosal lining was carefully stripped away. The shared muscular wall of the cystic remnant was sutured (figure 4). No aberrant tissue was identified.

OUTCOME AND FOLLOW-UP

After the resection, the patient had an uneventful recovery and remained asymptomatic on subsequent follow-ups. Histological evaluation of the resected specimen confirmed the presence of a cyst containing fibromuscular wall with gastric type mucosa and pancreatic heterotopia.

DISCUSSION

Alimentary tract duplications are extremely rare and occur in approximately 1 out of 4500 births.[2] These duplications can occur anywhere along the gastrointestinal (GI) tract, but are most commonly found in the ileum. Perhaps, the most rare type is found along the pyloric channel, representing 2.2% of all gastric duplications.[3] The female to male ratio is 2:1.[4] The first report on duplication cyst was by Calder in 1733. However, it was not until 1937 when William E. Ladd tried to simplify the nomenclature by suggesting the use of the term 'duplications of the alimentary tract'.[5] A recent review on alimentary duplications indicates that they are mostly diagnosed in the neonatal period.[6] Solitary pyloric duplication cyst, however, is exceedingly rare with only a few scattered case reports.

Of the digestive tract duplications, 85% do not communicate with the GI lumen.[7] The exact aetiology of enteric duplication is still under much debate, with two theories being the most prevalent[5] : (1) Bremer suggested that duplications arise as a result of incomplete or defective vacuolisation of the intestine. (2) Bentley and Smith postulated the theory of split notochord syndrome. According to this theory, in the third week of gestation, the notochord grows cephalad and normally separates from the endodermal cells. Formation of a diverticulum in between this separation subsequently leads to cyst formation.

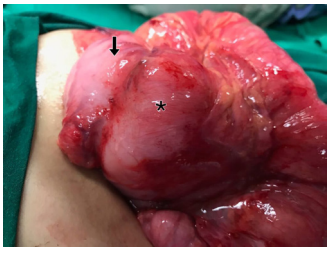

Figure 3 Cystic dilation (*) observed intraoperatively, in close proximity to the normal pylorus (arrow).

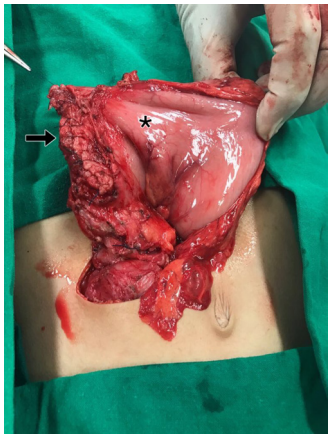

Figure 4 Normal gastric outlet continuity achieved with intact pylorus (*). Duplication cyst remnant (arrow) was sutured securely.

What is interesting to note about our case was the delay in presentation. Our patient presented at 5 years of life. Such a late presentation was also encountered only once in the literature by Ahmed *et al*.[1] Moreover, the patient became symptomatic over a period of 2 months prior to presentation, which was not consistent with other series, who reported such symptoms occurring in the initial weeks of life and thus mimicking infantile hypertrophic pyloric stenosis.[3 4 8]

Although preoperative diagnosis of pyloric duplication is quite challenging, ultrasonography, contrast CT scanning, MRI and oesophagoduodenoscopy can facilitate in differential diagnosis. An Endoscopic Retrograde Cholangio-Pancreatography (ERCP) should also be attempted in such cases, which was deferred in our patient, as we did not have a high suspicion of a communicating duplication cyst.

The use of endoscopic ultrasound (EUS) along with fine needle aspiration cytology (FNAC) in pyloric duplication have not been well documented in the paediatric population as far as our literature search reveals, as suspicion for the anomaly remains low.

On EUS, appearance of a duplication cyst arising from any part of the stomach usually appears as a hypoechoic lesion with a heterogeneous internal echo texture and regular margins.[9] A recent review on the use of EUS in duplication cyst in adults revealed that EUS-FNAC has been adopted as a diagnostic tool to rule out other ominous lesions from duplication cyst. However, the use of FNAC is still not universal as it poses a risk of infection of the cyst.[10]

We did not use EUS-FNAC as the facility was not available at our institution. In our opinion, although FNAC may prove diagnostic, this technique does not decrease the risk of malignant transformation of the aberrant gastric mucosa of the cyst; thus, surgical management would still be required.

The primary surgical management is the complete resection of the duplication cyst without undue injury to the lumen. If complete resection is not possible due to excessive adherence with the bowel wall, complete mucosal stripping or cauterisation of the mucosa should be achieved to decrease the risk of bleeding and neoplastic changes within the mucosa. Alternatively, the raw area may be patched with omentum or colon, or maintenance of a seromuscular gap has also been described.[11]

Some cases also call for complete excision of the cyst along with the common muscular wall with pyloroplasty to maintain

continuity. In our case, it was not possible to dissect the common wall between the duplication cyst and the pylorus, resulting in an almost complete resection of the cyst.

Learning points

- Gastric duplication cyst can present anywhere from 1 week of age up to 5 years of age and is thus not limited to the neonatal period alone.
- A high index of suspicion is required for diagnosis of gastric duplication in paediatric patients presenting with a cystic mass associated with the alimentary canal on imaging.
- Surgical exploration with either laparotomy or laparoscopy remains the mainstay of diagnosis, with identification of fistulous communications or aberrant pancreatic tissue in mind.
- Complete mucosal stripping and/or cauterisation of the cyst's mucosa is mandatory to decrease future chances of mucosal bleeding and malignancy.

Contributors MA: identified the rarity of the case and the need to report it, providing his expertise and intra-op pictures, edited and framed the final draft of the report itself and obtain consent from patient's parents. SMJ: gathered details about the case and formed and structured it into a report, also gathered follow-up information from patient's subsequent visits and provided the literature search. AS: contributed to the case report discussion section and in-patient details. Performed discussions with the pathology department for the case's interpretation. RR: contributed the radiological images for the case and reviewed the case with his senior colleagues due to the initial difficulty faced with making a diagnosis.

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Patient consent Parental/guardian consent obtained.

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