IDIOPATHIC INTRINSIC PYLORIC STENOSIS – A VERY RARE CAUSE OF GASTRIC OUTLET OBSTRUCTION IN SMALL CHILDREN

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Abstract

This is the report on the case of a 2 year old boy, known from the age of 1 month with recurrent symptomatology indicative of an intermittent gastric outlet obstruction. The imaging studies confirmed the gastric outlet obstruction syndrome, in the presence of a dilated and plicated stomach suggesting an intermittent gastric volvulus. During the surgery a pyloric fibrotic stenosis was discovered, therefore a pylororomy was performed, followed by a Heineke-Mikulicz pyloroplasty and gastropexy. After the surgery, the patient’s condition improved fast, with complete remission of all symptoms.

The few cases of pyloric fibrous stenosis described in children were secondary, either of toxic etiology (ingestion of caustic or corrosive substances, nonsteroidal anti-inflammatory drugs) or associated with peptic ulcer disease. In our case no certain etiology could be

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Introduction

The gastric outlet obstruction (GOO) that occurs during infancy and childhood can have multiple causes; the most common is, by far, the hyperthrophic pyloric stenosis (HPS) with an incidence of 1 - 4 to 1000 newborns. Other causes of this syndrome, such as congenital (pyloric atresia and prepyloric gastric antral web) or acquired conditions (ingestion of caustic or corrosive agents, nonsteroidal anti-inflammatory drugs overdose, peptic ulcer disease, chronic granulomatous disease, eosinophilic gastroenteritis, Jodhpur disease) are very rare, about 1 in 100 000 live births (1).

The diagnosis of HPS is easy to make and is confirmed by the ultrasound measuring of the pylorus. The diagnosis of a non-HPS GOO can be established by upper gastrointestinal (GI) series, upper GI endoscopy or ultrasound exam; in most of the cases, the etiology is determined at surgery.

The treatment of GOO depends on its cause; in most cases it is surgical and it rarely is non-operative (in case of peptic ulcer disease or inflammatory diseases). The pyloroplasty has proved to be successful in the treatment of the pyloric fibrous stenosis, and its outcomes were excellent.

We report the case of a 2 years old boy who presented with an intermittent non – HPS GOO, with an early onset at 1 month of age. During the surgery, a pyloric fibrous stricture without any inflammatory component at the histopathological examination was detected.

Case presentation

A 2.6 year old boy was referred to our ward following recurrent episodes of forceful postprandial non-bilious vomiting occurring shortly (at about 10 minutes) after food intake, associated with diffuse abdominal pain and epigastric distention. The last vomiting episode dated 4 weeks before the presentation and was relieved after 2 weeks by drug treatment with antiemetics and antispastics.

The boy’s medical history showed numerous episodes of non-bilious postprandial vomiting since the early age of 1 month, which usually responded to symptomatic treatment with Esomeprazole und Trimebutine maleate; the ultrasound measurements of the pylorus showed no hypertrophy of the muscular wall or channel narrowing and therefore ruled out the diagnosis of HPS. There was no history of caustic or corrosive agent ingestion; oral nonsteroidal anti-inflammatory drugs (Ibuprofen) were given only after 3 months of age and only in the recommended doses.

When admitted in our service the boy’s condition was relatively good; the physical examination showed an epigastric distention with a mild tenderness during palpation; the rest of the systemic examination did not show any abnormality. The laboratory studies didn’t show electrolyte imbalance; serum urea and creatinine were normal.

The upper GI series showed a free passage of contrast agent through the esophagus and a subdiaphragmatic located cardia, without any gastroesophageal reflux in the Trendelenburg position. The stomach was grossly distended and plicated, with significant pyloric narrowing and delayed gastric emptying (Figures 1 and 2).

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The upper endoscopy performed in another institution did not display esophageal or gastric peptic lesions, or other intraluminal lesions.

The diagnosis of pyloric stenosis was made and the boy was planned for elective surgery. At the operation, the stomach was distended and thick-walled while its left side was redundant, creating a mediogastric plication; the pylorus was normal-sized. No extraluminal compression was noted. After performing a gastrotomy proximal to the pylorus, the pyloric canal was explored and subsequently an obvious intrinsic stenosis was detected at this level. A longitudinal pylorotomy was performed, cutting through a fibrous pyloric ring located in the muscle thickness. Heineke-Mikulicz pyloroplasty with an epiploic patch was done, followed by a left-side gastropexy. The pyloric biopsy showed a submucous fibrosis process at the gastroduodenal junction; a thickened muscularis mucosae was noted, containing collagen bundles intersecting with the muscle fibers and also entering the submucosa (Figures 3, 4 and 5). No inflammatory cells were detected.

Figure 1 – Frontal view from the upper GI series showing a grossly distended and plicated stomach, with significant pyloric narrowing and delayed gastric emptying.

Figure 2 – Lateral view from the upper GI series showing the mediogastric plication of the stomach.

Figure 3 - Microscopic aspect of the pyloric biopsy, showing the gastroduodenal junction with duodenal mucosa on the left and gastric mucosa on the right. The muscularis mucosae contains collagen bundles intersecting with the muscle fibers, also entering the submucosa. Van-Gieson staining, 10×.

Figure 4 - Microscopic aspect of the pyloric biopsy, showing the yellowish muscularis mucosae fibers intermingled with thick red collagen bundles. The submucosa (on the left) also contains a large amount of collagen bundles. Van-Gieson staining, 4×.
The post-operative period was uneventful, the patient gradually resuming oral feeding beginning with the 4th day after surgery, without vomiting. At 3-month follow-up the condition of the child was good, with complete remission of previous symptoms.

**Discussions and conclusions**

The GOO presents with forceful, non-bilious, incoercible or recurrent vomiting, loss of weight, abdominal pain and epigastric distention, with preserved appetite. The most common cause of GOO in infancy is the hypertrophic pyloric stenosis (HPS), with an incidence of 1-4 to 1000 newborns, which typically manifests between 4 and 10 weeks of age. There may be other conditions which can produce the GOO symptoms in this age group, but they are quite rare, i.e. 1 in 100 000 children (1); they include either congenital diseases such as pyloric atresia, gastric antral web, gastric duplication, heterotopic pancreas (2), or acquired conditions such as peptic ulcer disease (3), chronic granulomatous disease (4), caustic or corrosive agents ingestion (5), nonsteroidal anti-inflammatory drugs overdose, gastric vulvulus, eosinophilic gastroenteritis and Jodhpur disease (6).

The onset of vomiting can be early, at birth, when the condition is congenital, or can be delayed in case of acquired diseases.

The diagnosis of GOO can be made by upper GI series, which reveal a dilated, often plicated stomach with a narrowed pyloric canal and a delayed gastric emptying. In case of HPS the ultrasound exam is the most appropriate since it allows for direct visualization and measurement of the pyloric channel and muscular wall, thus establishing the imaging diagnosis. The upper endoscopy may determine the etiology of a non-HPS GOO by visualising either lesions associated with a peptic ulcer disease, an antral web, polyps or other intraluminal masses / lesions.

It is essential to ascertain the cause of the pyloric stenosis because the therapeutic measures differ: medicamentous, with corticosteroids, for chronic granulomatous disease (4), endoscopic pneumatic dilatation for peptic stenosis (7), extramucosal pyloromyotomy for HPS, diaphragm excision with pyloroplasty for antral web, or pyloroplasty for peptic stenosis and Jodhpur disease respectively.

In our case the onset of symptoms was at the age of 1 month; at that time the diagnosis of HPS was ruled out by ultrasound imaging. Regarded as a gastroesophageal reflux disease, the illness was treated accordingly with Esomeprazole und Trimebutine maleate but the course of disease was oscillating, with periods of alleviation followed by recurrence of symptoms consisting in vomiting and colicky abdominal pain. Consequently additional imaging studies were performed, such as upper GI series which excluded the gastroesophageal reflux disease but displayed signs of GOO, and upper endoscopy which didn’t detect any esophageal or gastric lesions suggesting a peptic disease. An intermittent gastric volvulus was also taken into account, based on the atypical plicated appearance of the stomach during upper GI series.

As mentioned, the definitive diagnosis was made only by directly assessing the pyloric fibrous stricture during surgery and subsequently by histopathologically confirming it.

A very rare similar form of GOO was described by Sharma et al (1) under the name of Jodhpur disease, with 22 cases reported so far. The average age of presentation is 2.9 years with predilection of male sex (6). Its features include narrowing of the pyloric canal, increased gastric emptying time and a large-sized stomach; the US evaluation does not show pyloric muscle hypertrophy and the gastroscopy reveals no intraluminal pathology. Histopathologically there is a normal structure of the pylorus, without inflammatory, fibroproliferative or neoplastic cells. Regarding our patient, most of the above aspects were present except that the

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**Figure 5** - Red collagen bundles in the muscularis mucosae and submucosa, intersecting with the yellowish smooth muscle fibers. Absence of inflammatory cells. Van-Gieson staining, 40×.
pyloric biopsy displayed thickened muscularis mucosae, containing collagen bundles intersecting with the muscle fibers and also entering the submucosa. The complete absence of inflammatory cells excluded the acquired inflammatory trait, which distinguishes our case from the one reported by Ratan et al (8). As is the case with Jodhpur disease, pyloroplasty proved to be very efficient in treating this illness.

In those cases in which other known congenital or acquired causes of a non-HPS GOO are ruled out, the rare event of an idiopathic fibrous pyloric stenosis should also be considered; this pathologic entity has not been reported in literature until now.

As it happened in our case, the empiric antacid and symptomatic treatment only prolongs the child’s illness and delays the definitive surgical treatment, e.g., pyloroplasty. Therefore, we stress the importance of establishing the etiology of a GOO, even though this can be difficult to achieve (9). In this regard, upper GI series and endoscopy can be of great help.

References

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