

A rare case of late presentation of infantile hypertrophic pyloric stenosis in a 5-year old child

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ABSTRACT

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The case describes a late presentation of Infantile Hypertrophic Pyloric Stenosis in a 5 year old boy, and its management.

INTRODUCTION

Infantile Hypertrophic Pyloric Stenosis (IHPS- ICD₁₀: Q40.0) is one the most common gastro-intestinal diseases in the first few weeks of life. Pyloric hypertrophy causes a functional gastric outlet obstruction that usually gives symptomatology in the first weeks of the infancy¹. Atypical presentation and late manifestation of the symptoms are rare. In the present article, a late presentation of IHPS is described in a 5-year old boy.

CASE REPORT

A 5-year old male patient was admitted in the

pediatric ward to investigate recurrent episodes of projectile vomiting, especially after the meals, since 1 month. Parents reported 4 kilograms of weight loss during the last 20 days. Previous history of the child was unremarkable; but mainly due to lack of regular pediatric surveillance. Blood pressure, heart rate and arterial blood gases were within normal range for age. Laboratory exams showed WBC 5400c/mm³ (segments 8,3%, lymphocytes 80,1%), sodium 136 mmol/l , potassium 3,9 mmol/l , SGOT 23 u/l, SGPT 43 u/l ,while CRP was negative. Abdominal ultrasound displayed gaseous distension of the bowel with otherwise unremarkable findings and the X-Ray exam with barium meal revealed mild gastroesophageal reflux. Abdominal Computed tomography scan, with oral and I.V. contrast, showed prominent gastric dis-

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tension with average stomach wall and attenuated lumen caliber. Remarkable dilatation of the stomach and associated stenosis of the pyloric lumen with increased thickness of the pyloric mucosa was also found during the gastroscopy that followed.

Based on the findings, an explorative laparotomy was decided. During the surgery, typical pattern of pyloric stenosis that found and it was managed with the classic Ramsted pyloromyotomy. Enlarged scattered mesenteric lymph nodes were also found. Three of them were sent for histopathological specimen examination, which revealed marked sinusoidal and lymphoid hyperplasia (Figures. 1 to 3).

Figure 1. Intraoperative display of pyloric stenosis.



Intraoperative and postoperative course of the patient was unremarkable. Twenty-four hours postoperatively, the nasogastric tube was removed and the patient was fed with no complications. The boy was discharged 48 hours later.

Figure 2. Intraoperative photograph of surrounding lymphadenitis.

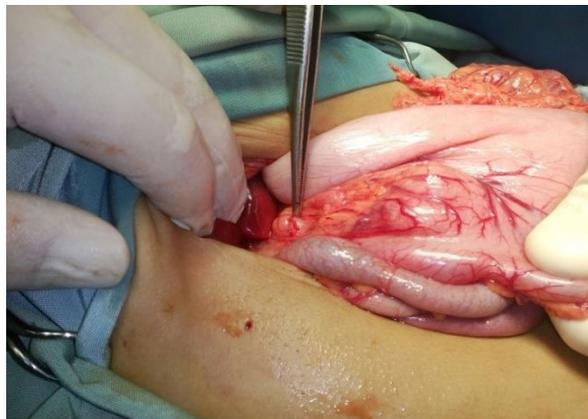


Figure 3. Intraoperative image of pyloromyotomy.



DISCUSSION

The incidence of hypertrophic pyloric stenosis is 2.4 per 1000 live births in Caucasians, 1.8 in Hispanics, 0.7 in Blacks, and 0.6 in Asians^{1,2}. It is also less common among children of mixed race parents. Rarely, infantile pyloric stenosis can occur as an autosomal dominant condition. It usually presents at 3 to 4 weeks of age, and late presentation up to 5 months has

been reported. It is four times more likely to occur in males and is also more common in the first born.

Delayed presentation of HPS beyond infancy is an extremely rare occurrence. Pyloric stenosis seems to be multifactorial, with some genetic and some environmental components. Numerous theories for the pathogenesis of hypertrophic pyloric stenosis (IHPS) have been proposed but none of them has merit general acceptance^{1,5}. All of them fall in to three categories 1) Compensatory work hypertrophy 2) neurologic degeneration or immaturity 3) abnormal endocrine signals. Some authors have suggested that milk curbs developing in the stomach can obstruct the pyloric channel leading to redundancy in the pyloric mucosa and compensatory pyloric muscle hypertrophy. Other investigators have theorized that various neural components in the pyloric antrum may be immature or defective leading to increased muscle size. Some of these theories include the deficiency of nitric oxide synthase decreased interstitial cells of Cajal (pacemaker cells of the gut) and diminished expression of neural cell adhesion molecule (NCAM).

Diagnosis of IHPS is based mostly on the history and physical exam of the patient. Patient's history includes recurrent episodes of projectile vomits; the pick of the symptoms to be presented between the 4th and 6th week after birth. On physical assessment peristaltic waves

are usually visible in the epigastrium and a characteristic mass named "olive" is often palpable. In case of absence of these physical findings the radiological study of choice to confirm the diagnosis is abdominal ultrasound. Also gastrointestinal contrast study and CT scan with contrast can be indicative. Never the less in most centers U/S is the key study, although not helpful in our case. The most current ultrasonographic criteria for HPS include pyloric channel length 15 mm pyloric muscle diameter 13 mm and pyloric muscle wall thickness 3 mm^{4,6}.

Premature infants are diagnosed with IHPS later than term or post-term infants. Apart from that, they often may present atypically, with less forceful vomiting and absence of visible gastric peristalsis⁷. En plus, IHPS in premature babies born at or before 28wk of gestation is extremely rare⁸.

Differential diagnosis include duodenal, jejunoileal and pyloric atresia, antral web, intestinal malrotation, Sandifer syndrome, mid-gut volvulus, food allergy and acute infections⁹. Definitive treatment is corrective surgery. The Ramsted pyloromyotomy, performed through a right upper quadrant transverse incision, currently is the procedure of choice. Yet, new reports suggest laparoscopic pyloromyotomy as a safer alternative¹⁰⁻¹¹.

Very few cases of pyloric stenosis in children more than five years old have been reported. The larger age reported ever was a 17-year old female¹². Most of rest were non hypertrophic pyloric stenosis cases caused by intraluminal webs and diaphragms mucosal valves or heterotopic pancreas^{2,3}. In all these cases, the obstruction was incomplete and functional; the stomach was dilated with an apparently normal pylorus and without any muscular hypertrophy. All the layers of pylorus were normal by biopsy, and Heineke-Mikulicz pyloroplasty procedure was curative.

CONCLUSION

IHPS is a really rare disease entity in children more than 6 to 8 months. Never the less the both Pediatrician and Pediatric Surgeon have to be alert in case of patient with suchlike history and symptoms.

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