

Case Report of Late Onset Hypertrophic Pyloric Stenosis in Lebanese Infant

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Abstract: Described as the most common cause of gastric outlet obstruction and the most common surgical cause of vomiting in infants, Hypertrophic Pyloric Stenosis (HPS) is defined as a hyperplasia of smooth muscle fibers of the pylorus that is responsible for the narrowing of the pyloric canal and obstruction of gastric emptying. It occurs at a rate of 1–4 per 1000 live births with a male to female ratio 4:1. The classic presentation of HPS is nonbilious, projectile vomiting in a full-term neonate with a classic presentation at 3 to 4 weeks of age. We report in here a delayed presentation of HPS in an infant aged 4 months who presented with projectile nonbilious recurrent episodes of post prandial vomiting, where history, examination and investigations supported the diagnosis of HPS and Ramstedt's pyloromyotomy was curative.

Keywords: Hypertrophic Pyloric Stenosis (HPS), nonbilious vomiting, Ramstedt's pyloromyotomy.

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INTRODUCTION

In infants, hypertrophic pyloric stenosis (HPS) is the most common cause of gastric outlet obstruction characterized by a partial blockage that interferes with the passage of stomach contents into the small intestine. This disease is caused by diffuse hypertrophy and hyperplasia of the smooth muscle cells of the antrum of the stomach and pylorus, resulting in narrowing and elongation of pyloric channel. Infants feed well but vomit forcefully (projectile vomiting) shortly after eating and can become dehydrated and undernourished. An “olive” representing the enlarged pylorus can be palpated in the right upper quadrant in 60-80% of infants [1-3]. Hypertrophic pyloric stenosis usually presents at 3 to 5 weeks of age and very rarely after 12 weeks of age [4, 5] with a prevalence ranging from 1 to 4 per 1000 live births with geographic variations [6, 7]. The late presentation of HPS in childhood is a rare condition with a prevalence of 1:100,000 in the literature [8, 9]. In this article we report a late presentation of HPS in a 4 months old Lebanese baby, presented with projectile non bilious

post prandial vomiting, confirmed using ultrasonic examination of the abdomen and treated by Ramstedt's pyloromyotomy [10].

CASE DESCRIPTION

A 4 month old boy presented with complaints of non-bilious vomiting after feeds, projectile in nature. He was born to a 36 years old mother at 39 weeks of gestation by normal delivery; she was G4P2A2 mother with a non-consanguineous marriage. The baby doesn't have a history of previous admission to neonatal care unit with birth weight of 3 kilograms and current weight 6 kilograms, exclusively bottle fed. At 1 month of age the patient started to have vomiting mainly post prandial, upon which parents sought medical advice; and the baby was considered as having a gastroesophageal reflux and was maintained on Omeprazole, Domperidone and thickened formula. Mother noted improvement of symptoms for 1 month after initiation of treatment. Subsequently, at 3 months of age the vomiting persisted despite treatment and became projectile in nature, more

frequent and blood tinged especially last 4 days. The mother mentioned also a change in bowel habits with absent stools during last 4 days. On physical examination: vitals were within normal limits, yet the baby was irritable, not in distress, and looks mildly dehydrated with non-depressed open anterior fontanel with good peripheral perfusion, his abdomen was soft but mildly distended with positive bowel sounds and without evidence of any organomegaly or hernia. Initial laboratory investigations including CBCD and electrolytes showed moderate elevation in WBC (14300 cell/hpf) with lymphocytic predominance (62.7%), hypochloremia (Cl 85 mmol/l) and alkalosis (bicarbonate 29 mmol/l). KUB was done urgently revealed distended stomach, absence of air fluid levels. IV fluid infusion started. Differential diagnosis consisted of lactose intolerance, cow's milk protein allergy, severe gastroesophageal reflux disease, duodenal atresia and pyloric stenosis. Upper GI series was done and showed delayed passage of contrast from stomach to duodenum with severely distended stomach (**Error! Reference source not found.**).



Fig-1: Radiography of upper gastrointestinal series showing distended stomach, poor passage of contrast from stomach to duodenum

Pyloric stenosis was suspected, and diagnosis was confirmed by ultrasound of the abdomen that proved a hypertrophied, 6 mm thick pyloric muscle (**Error! Reference source not found.**).



Fig-2: Abdomen ultrasound showing pyloric muscle of 6 mm thickness

Nasogastric tube was inserted for gastric decompression; baby kept NPO and was prepared for pyloromyotomy (**Error! Reference source not found.**).

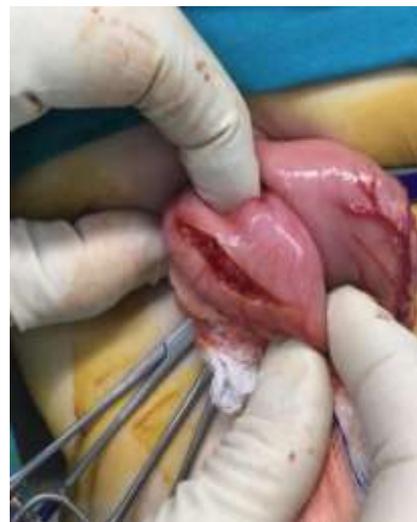


Fig-3: Pyloromyotomy

Oral intake was resumed 6 hours post-operative with oral rehydration solution for the first 8 hours followed by progressive introduction of formula and the patient discharged in the next day without any complication. Baby was followed after two months and had normal development and weight gain.

DISCUSSION

As described for the first time by Hirschsprung in 1888, Hypertrophic Pyloric Stenosis (HPS) is defined as the most common surgical cause of vomiting in infants [11]. The etiology of HPS is

most likely multifactorial with both genetic and environmental sides implicated [12]. To begin with, the assumption that vomiting is a minor symptom by many families and physicians may lead to lack of awareness of that symptom and probably misdiagnosis of the underlying causative pathology [11]. Vomiting which is typically characterized by its acute postprandial onset that is non bilious and forceful "projectile", with demanding of the infant to be re-fed shortly afterwards described as "the hungry vomiter"[13], remains the main finding of HPS. HPS constitutes the most common cause of non-bilious projectile vomiting in infants; however this presentation has also been reported with gastroenteritis, gastroesophageal reflux disease, hiatal hernia, adrenal insufficiency, and inborn errors of metabolism [14]. We discuss in this article the case of a 4 month old boy admitted for projectile non-bilious vomiting that started at 15 weeks of age which was considered as an uncommon age of onset for hypertrophic pyloric stenosis. At 1 month of age, the baby experienced typical effortless postprandial vomiting, diagnosed as gastroesophageal reflux disease and started on medical treatment with improvement of symptoms for one month. Furthermore, the worsening of vomiting at 12 weeks of age that became more frequent and projectile in nature, and at 15 weeks of age, increased our index of suspicion for HPS. The diagnosis was confirmed by an upper GI series and an abdominal ultrasound. The patient was treated by a pyloromyotomy with significant clinical improvement in the follow-up visit. This case can be added to the few reported cases in the literature with delayed presentation of HPS. In addition, this delay in presentation raises the possibility of an acquired cause of the disease rather than a congenital one [15].

CONCLUSION

Vomiting as an indicator of many serious etiologies during infancy and it should be kept in mind as it can be an important symptom for many surgical emergencies. Pay attention to the history of a repetitive, non-bilious vomiting and failure to gain weight for the possibility of Hypertrophic Pyloric Stenosis as a causative etiology, regardless of the age of presentation.

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