Case Report

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Congenital Hypertrophic Pyloric Stenosis

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Abstract

Hypertrophic pyloric stenosis (HPS) causes a functional gastric outlet obstruction as a result of hypertrophy and hyperplasia of the muscular layers of the pylorus. In infants, HPS is the most common cause of gastric outlet obstruction and the most common surgical cause of vomiting. Hypertrophic pyloric stenosis is a condition characterised by hypertrophy of two muscle layers of pylorus,l enghthening of canal, thickening of pylorus which becomes oedematous causing functional obstruction of gastric outlet. The gastric outlet obstruction due to the hypertrophic pylorus impairs emptying of gastric contents into the duodenum. As a consequence, all ingested food and gastric secretions can only exit via vomiting, which can be of a projectile nature. While the exact cause of the hypertrophy remains unknown, one study suggested that neonatal hyperacidity may be involved in the pathogenesis.

Keywords: Hypertrophic pyloric stenosis (HPS); Hyperplasia.

Introduction

Hypertrophic pyloric stenosis is a condition characterised by hypertrophy of two muscle layers of pylorus,lenghthening of canal, thickening of pylorus which becomes oedematous causing functional obstruction of gastric outlet [1]. The first description of pyloric stenosis was given by Hirschspung in 1888 [2]. It occurs in 2-4/1000 live births, more in males (4:1) and more so in the first born child. Cause is unknown, but genetic factor (IHPS1) is implicated, where there is deficient production of neuronal nitric oxide synthase causing impaired relaxation of pyloric smooth muscle. Use of macrolide antibiotics such as erythromycin in the first two weeks of life, during late pregnancy and through breastmilk in the first two days of life increases the risk. The child presents at 3-5 weeks of age with nonbilious, projectile vomitting immediately after feeding. On palpation of abdomen, olive like mass is felt in the right upper quadrant. The child presents with clinical signs of dehydration, sunken fontanelle, malnutrition and poor weight gain.

Congenital hypertrophic pyloric stenosis is diagnosed with clinical findings, hypochloraemic hypokalemic metabolic acidosis and ultrasonography.

Case History

One and half months male with a birth weight of 2.25kg had come to the outpatient department weighing 1.5kg. The mother gave a history of lactation failure. She was feeding him with saline orally for the first 15days of life. Later he was started on cows milk. After 10-15 minutes of feeding,he would vomit the same. Since the past one week the number of episodes of vomitting had increased and the baby had lost weight and so he was brought here. On examination, baby was irritable with cry, tone, activity decreased. He had an old mans look with severe muscle wasting, Grade IV PEM according to IAP classification. He had sunken eyes,with skin pinch going very slowly. When the baby was kept NBM no swelling was visible and even the sonography was unremarkable. As soon as the feed was started a swelling appeared in midepigastric region with visible peristalsis from left to right.

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His hemoglobin was 6.7g%, Sr. Sodium:132mmol/L, Sr. Potassium: 3.5mmol/L, Sr. Urea:51mg%. Abdominal ultrasonography showed pyloric canal with length of 1.7mm and muscle thickness of 5mm and doughnut sign,features suggestive of

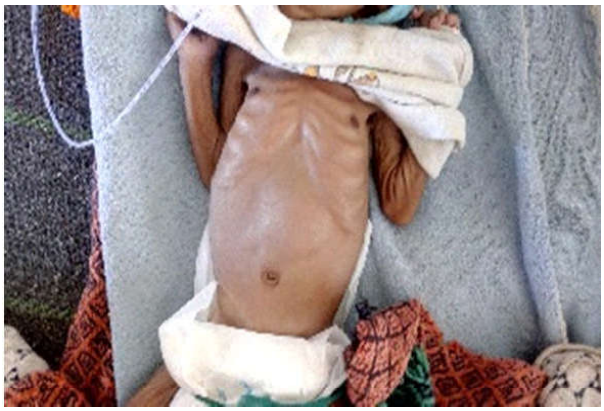

Fig. 1: Olive shaped swelling

hypertrophic pyloric stenosis. He was started on iv fluids,iv antibiotics and blood was transfused. Once he was haemodynamically stable, pyloromyotomy was done.

Discussion

Infantile hypertrophic pyloric stenosis was first described by Hirschsprung. It occurs in approximately 3 of every 1,000 livebirths, making it the most common indication for surgical intervention in infancy. It is characterized by hypertrophy and thickening of the circular and longitudinal muscle of the pylorus, leading to a gastric outlet obstruction.

Etiology

Hypertrophic pyloric stenosis is inherited by a multifactorial threshold model, and the generalized occurrence risk for siblings is 5-9%. Associated congenital anomalies are reported in 6-20% of patients with pyloric stenosis. A rare association with developmental delay has also been reported.

A familial pattern exists, although HPS does not follow classic Mendelian genetics. The risk for developing HPS is about 7% if the father was previously affected and about 10-20% if the mother was affected.It is associated with B and O blood groups.

It is uncommon in premature babies. The incidence of pyloric stenosis in premature babies ranges from 1% to 16% of all cases.

1. The exact etiology of HPS is not known.
2. One theory proposes the lack of pyloric inhibitory innervation leading to reduced levels of nitric oxide, a smooth muscle relaxant. As a result, the pylorus experiences unopposed contraction following muscarinic stimulation.
3. Elevated levels of prostaglandins have also been implicated owing to the increased incidence of pyloric stenosis in infants who have received PGE to maintain a patent ductus arteriosus.
4. HPS has also been associated with other GI anomalies, such as tracheoesophageal fistula, pyloric atresia, antral webs, gastric duplications, and gastric volvulus.
5. Furthermore, HPS has been linked to other

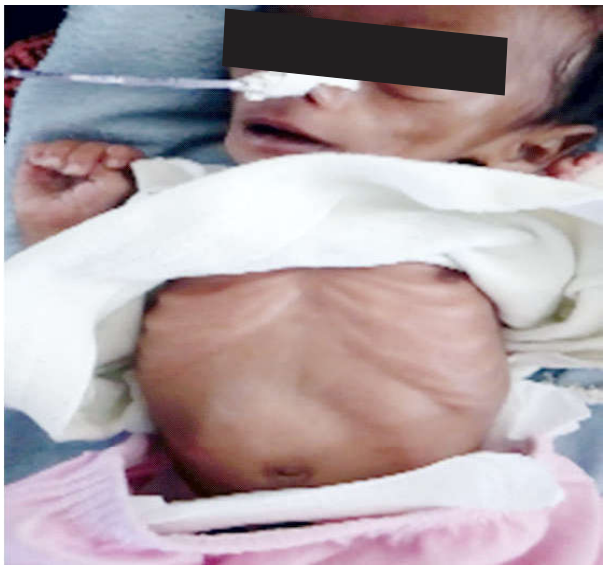

Fig. 2: Olive shaped swelling appeared in midepigastric region with visible peristalsis

disease states, such as eosinophilic gastroenteritis, epidermolysis bullosa, trisomy 18, and Turner Syndrome.

Clinical Presentation

Typical presentation of an infant with hypertrophic pyloric stenosis (HPS) is onset of initially nonbloody, always nonbilious vomiting at 4-8 weeks.

Although vomiting may initially be infrequent, over several days it becomes more predictable, occurring at nearly every feeding. Vomiting intensity also increases until pathognomonic projectile vomiting ensues. Slight hematemesis of either bright red flecks or a coffee-ground appearance is sometimes observed.

Patients are usually not ill-looking or febrile; the baby in the early stage of the disease remains hungry and sucks vigorously after episodes of vomiting

Prolonged delay in diagnosis can lead to dehydration, poor weight gain, malnutrition, metabolic alterations, and lethargy. Parents often report trying several different baby formulas because they (or their physicians) assume vomiting is due to intolerance. Hyperbilirubinemia is the most common association of pyloric stenosis known as icteropyloric stenosis On physical examination, the infant may exhibit poor weight gain or even weight loss.

Marasmus, however, or severe protein-calorie malnutrition, is rarely seen today. Jaundice may be seen in approximately 5% of infants. After feeding, a wave of gastric peristalsis may be seen traversing the abdomen from left to right, representing intense contractions against an obstruction. Abdominal distention may be a late finding, as is usually the case with proximal GI obstructions. The hypertrophied pylorus may be palpable. The pylorus is firm, mobile, and olive-shaped. It is located in the right upper quadrant of the abdomen, beneath the liver edge. It is best palpated from the left side while the infant is feeding since the abdominal muscles are relaxed. A palpable "olive" is pathognomonic of pyloric stenosis.

Diagnosis

Ultrasonography

- The criterion standard imaging technique for diagnosing HPS
- Muscle wall thickness 3 mm or greater and pyloric channel length 14 mm or greater and pyloric diameter of 10-14mm are considered abnormal in infants younger than 30 days

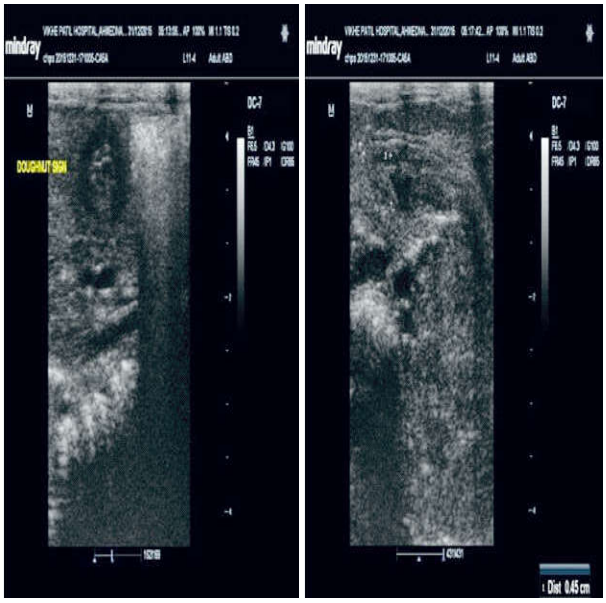

Fig. 3:

Barium upper GI study

- Effective when ultrasonography is not diagnostic
- Should demonstrate an elongated pylorus with antral indentation from the hypertrophied muscle
- May show the single "string sign" or "double track" sign when thin tracks of barium are compressed between thickened pyloric mucosa or the "shoulder" sign when barium collects in the dilated prepyloric antrum
- After upper GI barium study, irrigating and removing any residual barium from the stomach is advisable to avoid aspiration

Endoscopy

- Reserved for patients with atypical clinical signs when ultrasonography and UGI studies are nondiagnostic

The diagnostic test of choice is the ultrasound, which has approximately 90% sensitivity.

The "classic" laboratory finding in HPS is a hypochloremic, hypokalemic metabolic alkalosis. Repeated vomiting results in a loss of HCl, causing the hypochloremic metabolic alkalosis. The patient is likely dehydrated from repeated GI loss and poor oral intake. A metabolic acidosis (lactic acidosis) may result with severe dehydration.

Management

Surgical repair of HPS is fairly straightforward and without many complications. However, properly preparing the infant is vitally important.

Preoperative Management

- Directed at correcting the fluid deficiency and electrolyte imbalance
- Base fluid resuscitation on the infant's degree of dehydration
- Most infants can have their fluid status corrected within 24 hours; however, severely dehydrated children sometimes require several days for correction
- If necessary, administer an initial fluid bolus of

10 mL/kg with lactated Ringer solution or 0.45 isotonic sodium chloride solution

- Continue IV therapy at an initial rate of 1.25-2 times the normal maintenance rate until adequate fluid status is achieved
- Adequate amounts of both chloride and potassium are necessary to correct metabolic alkalosis
- Unless renal insufficiency is a concern, initially add 2-4 mEq of KCl per 100 mL of IV fluid
- Urine output and serial electrolyte determinations are performed during resuscitation
- Correction of serum chloride level to 90 mEq/L or greater is usually adequate to proceed with surgical intervention
- Before induction of anesthesia, aspirate the infant's stomach with a large-caliber suction tube to remove any residual gastric fluid or barium; saline irrigation is occasionally necessary to remove a large quantity of barium

Surgical Treatment

- Ramstedt pyloromyotomy remains the standard procedure of choice
- The usual approach is via a right upper quadrant transverse incision that splits the rectus muscle and fascia
- Laparoscopic pyloromyotomy may also be used
- Endoscopic pyloromyotomy is a simple procedure and can be performed as an outpatient procedure
- Endoscopic balloon dilatation of hypertrophic pyloric stenosis after failed pyloromyotomy can be used
- A supraumbilical curvilinear approach has gained popularity with good cosmetic results.

Postoperative Management

- Continue IV maintenance fluid until the infant is able to tolerate enteral feedings
- In most instances, feedings can begin within 8 hours following surgery
- Graded feedings can usually be initiated every 3 hours, starting with Pedialyte and progressing to full-strength formula
- Schedules that advance the volume of feeds more quickly or those that begin with ad lib feeds are associated with more frequent episodes of vomiting but do not increase morbidity and actually may decrease the time to hospital discharge
- Addition of an H2 receptor blocker sometimes can be beneficial
- Treat persistent vomiting expectantly because it usually resolves within 1-2 days
- Avoid the temptation to repeat ultrasonography or upper GI barium study; these invariably demonstrate a deformed pylorus, and results are difficult to interpret

Conclusion

Most important part of management is rehydration and correcting the acid-base imbalance [3]. Ramstedt Pyloromyotomy has given satisfactory results with the mortality rate being 0-0.5%. But preoperative preparation is the greatest factor in bringing the mortality to its present level [4].

The use of ultrasonography has nearly completely replaced the clinical skill of palpating the ''olive,'' a skill that is often only used after the diagnosis has been made and the child is under general anesthesia simply to demonstrate to students and house staff what is described in textbooks.

As clinicians in a highly technology–dependent health care system, it is important that we review the history of our craft to be reminded that astute clinical judgment and careful clinical examination must remain essential tools in the care of patient.

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