Case Report

Hypertrophic pyloric stenosis: Early presentation with hyperbilirubinemia

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Abstract

Hypertrophic Pyloric Stenosis commonly presents with non-bilious progressive vomiting usually after a feed. The vomiting usually starts after third week with a range of symptoms onset from first week to fifth month of life. Less commonly it is associated with hyperbilirubinemia (unconjugated > conjugated) which subsides after surgical correction. We report a case of Hypertrophic Pyloric Stenosis who presented early in fifth day of life with repeated vomiting after each feed and unconjugated hyperbilirubinemia. Ultrasonography revealed Hypertrophic Pyloric Stenosis without any evidence of other cause of jaundice. Patient was treated conservatively, prepared for surgery, surgical correction done with Ramstedt's Pyloromyotomy following that vomiting and jaundice both subsided. Early presentation of HPS may suggest congenital etiology and association of unconjugated hyperbilirubinemia should alert clinician for future Gilbert Syndrome.

Key words: *Hypertrophic pyloric stenosis, Unconjugated hyperbilirubinemia, Vomiting*

Hypertrophic pyloric stenosis (HPS) commonly presents with non-bilious progressive vomiting, usually after a feed. The vomiting usually starts after 3rd week with symptom onset ranging from the first week to 5th month of life. Less commonly, it is associated with hyperbilirubinemia (unconjugated > conjugated), which subsides after surgical correction. Early presentation of HPS in the first week of life is relatively rare [1,2] and also, its association with unconjugated hyperbilirubinemia may complicate the clinical approach [3,4]. We present such a case of HPS, who presented with jaundice and repeated vomiting in 1st week of life, and was corrected by Ramstedt's pyloromyotomy.

CASE REPORT

A 22-day-old, weighing 2.2 kg, first born male baby was referred with the complaints of yellowish discoloration of the whole body and repeated non-bilious vomiting since 5th day of life. Baby was a product of full term, normal vaginal hospital delivery after an uneventful antenatal period with birth weight of 3 kg without any immediate postnatal intervention. On the 5th day of life, mother noticed increasing yellowish discoloration and repeated vomiting that was initially intermittent, gradually progressed to emesis soon after each feed. Vomitus was found to be non-bilious. After each episode of vomiting, baby used to cry again for the feed. From 9th day, onwards mother noticed that the baby passed very scanty stools for 3-4 days. There was no history of temperature instability, fast breathing, abdominal distension or mass, blood in vomiting or blood in the stool.

Initially, baby received supportive treatment for 6 days and finally, referred to our hospital when condition deteriorated. In our department, baby was admitted with findings of icterus up to sole, depressed anterior fontanel, temperature 36°C (axillary), respiratory rate - 48/min and heart rate - 160/min. His activity was depressed with poor cry decreased tone, but rooting and sucking was found to be good. On abdominal examination, no mass was palpable, and no

peristaltic wave was seen. His abdominal circumference and head circumference were found to be normal (23 cm and 33.5 cm respectively). Respiratory and cardiovascular findings were insignificant.

After emergency resuscitation, baby was put on nil per oral with the presumptive diagnosis of late onset sepsis with hyperbilirubinemia and conservative management was started. Initial investigations revealed no evidence of sepsis or hemolysis. The baby showed hyperbilirubinemia with serum bilirubin (total - 14.7 mg/dl, direct - 1.5 mg/dl), without any major blood group incompatibility (blood group of mother - B positive and of baby- O positive). Arterial blood gas analysis done on FiO_2 - 21% suggested hyponatremic and hypochloremic metabolic alkalosis (pH -7.52, pCO_2 - 35.2 mmHg, pO_2 - 58.5 mmHg, HCO_3 - 28.2 mmol/L, Na^+ - 122.4 mmol/L, K^+ - 4.07 mmol/L, Cl^- - 82.0 mmol/L, Ca^{++} - 0.488 mmol/L, OSM - 246 mosm/kg).

Ultrasonography with color Doppler revealed HPS with no evidence of peristalsis in pylorus (pyloric length 2.5 cm, pyloric thickness 3.7 cm, no evidence of malrotation, superior mesenteric artery and vein are in normal anatomical position).

Patient was conservatively managed with phototherapy, intravenous fluid; electrolyte and acid-base imbalance were corrected accordingly and referred to pediatric surgery department for surgical correction where patient was operated with Ramstedt's pyloromyotomy. The patient was put on breastfeeding on 3rd post-operative day and subsequent investigation on the same day revealed decreasing trend in serum bilirubin level and vomiting did not reappear on re-feeding. Patient was discharged on exclusive breast feeding on 10th post-operative day after removal of all stitches.

DISCUSSION

Nonbilious vomiting is the initial symptom of pyloric stenosis. The vomiting may or may not be projectile initially, but is usually progressive, occurring immediately after feeding. Emesis might follow each feeding, or it may be intermittent. The vomiting usually starts after 3rd week of age [1]. However in our patient, it started early in 5th day of life. Presentation in the 1st week of life is very rare. A Medline literary search revealed only 2 reported instances where the age of presentation was less than 1 week [5]. Apart from reports [1,2,5] of early presentation after birth and even cases reported to be diagnosed in intrauterine life. This suggests the congenital pattern of the disease [2].

Hyperbilirubinemia is one of the important clinical association of pyloric stenosis, and it is also known as icteropyloric syndrome [1]. Hyperbilirubinemia, mostly unconjugated, is associated with HPS only in 2-5% cases [3]. It usually subsides after surgical correction. It may be due to decreased level of glucronyl transferase [1] that results from a mutation in the bilirubin uridine diphosphate glucuronyl transferase UGT1A1 gene [1,4,6]. The clinical implication of hyperbilirubinemia in HPS with possible mutation in UGT1A1 gene is that it may be an early manifestation of Gilbert syndrome [4,6]. Increased levels of intestinal glucuronidase activity [2] or starvation [3] are the possible causes which contribute to hyperbilirubinemia. There are reports of Gilbert syndrome diagnosed in a patient who presented with jaundice and HPS, and they suggested that the association of jaundice with HPS is due to molecular defect within the gene promoter [4,6].

Thus, infantile hypertrophic pyloric stenosis may present early in 1st week of life, and probable congenital etiology could not be denied for the early presentation of HPS [2]. The association of unconjugated hyperbilirubinemia should alert clinicians for future consideration of Gilbert syndrome and testing for possible homozygous or heterozygous polymorphism at (TA) 7TAA [4,5].

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