

The Delayed Diagnoses of Hypertrophic Pyloric Stenosis due to Hospitalization in Neonatal Intensive Care Unit: A Report of 5 Cases

Nuriye Tarakci*, Murat Konak, Hüseyin Altunhan and Rahmi Ors

Department of Neonatology, Meram Medical Faculty, Necmettin Erbakan University, Turkey

*Corresponding author: Nuriye Tarakci, MD (Research Fellow), Department of Neonatology, Meram Medical Faculty, Necmettin Erbakan University, Konya, 42080, Turkey, Tel: 00903322236697; Fax: 00903322236585; E-mail: nuriyetarakci@hotmail.com

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Abstract

Hypertrophic pyloric stenosis (HPS) is the most common cause of gastric outlet obstruction in infants. It commonly requires surgery. Its incidence is 0.17-4.4 cases per 1,000 live births. The clinical findings of pyloric stenosis typically appear within three to five weeks after birth. Its most important clinical finding is non-bilious projectile vomiting. If its diagnosis is missed in early period, the most common finding is dehydration (with hypochloremic hypokalemic metabolic alkalosis). However the findings of HPS might be frequently masked in infants hospitalized for longer periods in neonatal intensive care unit. Because vomiting is the one of most common symptoms, it may be related to the different etiological factors of vomiting (congenital or genetic causes, NEC, sepsis, nutritional intolerance), blockage of projectile vomiting with gastric drainage by previously inserted indwelling orogastric/nasogastric catheter which prevents development of excessive gastric dilatation. The accurate diagnosis delays with elimination of severe alkalosis and electrolyte disorder and prevention of malnutrition with administered parenteral nutrition. Herein we would like to draw attention the delayed diagnosis of HPS in five neonatal cases who were hospitalized in the neonatal intensive care unit (nicu) for longer periods.

Keywords: Hypertrophic pyloric stenosis; Delayed diagnosis; Newborn

diagnosis of HPS due to hospitalization for longer periods in NICU (Table 1).

Introduction

Hypertrophic pyloric stenosis (HPS) is characterized by diffuse hypertrophy and hyperplasia of the muscular layers of the pylorus. The pyloric muscle hypertrophy results in narrowing of the pyloric canal. The obstruction blocking gastric emptying and the increased peristaltic movements lead to the hypertrophy of gastric muscles and gastric dilation [1]. Though its incidence changes among different ethnic groups, it ranges between 0.17 and 4.4 cases in 1000 live births [2].

Case	Gestational week	Birth weight (gr)	Gender	Underlying condition	Age at diagnosis (day)
1	37	2800	Female	Esophageal atresia	36
2	34	2600	Male	Williams syndrome	43
3	35	2800	Female	Down syndrome	39
4	39	2840	Female	Multiple congenital anomalies	49
5	29	1300	Female	Prematurity	38

Table 1: The patients were diagnosed as HPS.

Clinical symptoms manifest from the 2-6 weeks of life and peak at 3-5 weeks [1,3]. Herein we presented 5 neonatal cases with delayed

Case Reports

Case 1

The first case weighing 2800 g was delivered by caesarean section at 37 weeks of gestation. She was operated on the postnatal 2nd day because of diagnosis with the esophageal atresia. After seven day of her successful operation, she was started on orogastric feeding. The time of full oral feeding was on the postoperative 18th day. After 5 days, non-bilious vomiting episodes were observed. Free drainage was applied. Total parenteral nutrition was administered. Serum electrolytes, blood gases were normal. Direct abdominal graphs were unremarkable. Any evidence of obstruction was not detected on contrast-enhanced upper gastrointestinal radiograms. On abdominal US, length and diameter of the pyloric canal were 17 mm and 6 mm, respectively. Her symptoms persisted pyloromyotomy was performed at postnatal 36 days of her life. On the postoperative 7th day, she reached to full enteral nutrition.

Case 2

A male infant (birth weight, 2600 g) was born at 34 gestational weeks as the first child of a woman, who delivered by spontaneous vaginal route. He was on ventilator because of respiratory distress. He has depressed nasal root, lateral thinning of his eye brows, long philtrum and nose with anteverted nostrils. Hypercalcemia developed during monitorization of the patient having subvalvular pulmonary stenosis detected by echocardiographically. The poor nutritional status of the patient was attributed to the presence of Williams syndrome. However, his nutritional status improved with time. From the postnatal 26th day on, his non-bilious vomiting episodes were observed and

gastric drainage was applied. Then total parenteral feeding was initiated. His contrasted abdominal radiograms were unremarkable. However, on the 43rd day of his life, (because of attribution of his vomiting episodes to sepsis and NEC etc.) diagnosis of HPS was made. Following pyloromyotomy, enteral feeding was started and returned to normal. He was discharged from the hospital.

Case 3

A 34-year-old mother, gravida 3, para 3 was delivered a baby weighing 2800 g by cesarean section at 35 weeks of gestation. The newborn was admitted to NICU because of persistent non-bilious projectile vomiting episodes starting after its birth and being born with Down's syndrome. He has ASD and VSD diagnosed by echocardiography. His diagnosis of Down syndrome was confirmed by chromosomal analysis reported as 47 XX. The patient was partly fed through orogastric route supported by total parenteral nutrition. His laboratory tests and blood gases were normal. Contrast-enhanced abdominal radiograms were unremarkable. On abdominal ultrasonograms, thickness of the pyloric muscle and length of the pyloric canal were 5 mm and 18 mm, respectively. With these findings, on the 39th day of its hospitalization, delayed diagnosis of pyloric stenosis was made, and pyloromyotomy was performed. After surgery, nutritional intolerance was not observed. The patient was discharged.

Case 4

A term girl infant weighing 2840 g was born to mother 38-year-old, gravida 5, para 5 delivered by C/S. Feeding of the baby with multiple congenital anomalies detected on physical examination was already challenging. Beginning from the 30th postnatal day, her gastric residuals increased and diagnosed as feeding intolerance. Gastric drainage was started and total parenteral nutrition was re-administered again. Contrast-enhanced abdominal radiograms were unremarkable. There were no abnormalities in blood gases and serum electrolytes. On abdominal US, thickness of pyloric muscle and length of the pyloric canal were 7 mm and 16 mm, respectively. The patient was diagnosed as HPS and operated on the 49th day of her hospitalization. Her nutritional intolerance improved during the postoperative period. However, the patient died 2 months later, because of other associated pathologies.

Case 5

A baby girl with a birth weight of 1300 g who was one of the twins delivered by a 25-year old mother at 29th gestational weeks of her first pregnancy, was transferred into NICU because of her prematurity. Despite improvement of her complications of prematurity, her nutritional problems persisted. Her gastric residuals and non-bilious vomiting episodes increased. Total parenteral nutrition was initiated. In addition, gastric contents were drained. Nutritional intolerance was thought to be related to inadequate intestinal maturation due to prematurity. Electrolyte imbalance and alkalosis were not detected. Her abdominal physical examination was normal. Her gastric residuals and non-bilious vomiting episodes were persisted. On contrast-enhanced radiograms, gastric dilation and delayed gastric emptying were observed. On abdominal US, thickness of pyloric muscle and length of the pyloric canal were 6 mm and 17 mm, respectively. On her 38th day of hospitalization, pyloromyotomy was performed. During the postoperative period, her vomiting episodes disappeared. The patient who tolerated full feeding was discharged from the hospital.

Discussion

In infants, HPS is one of the lesions which most frequently require surgical interventions [4]. Clinical symptoms start to manifest from 3-5 weeks of life on and generally result in electrolyte, acid-base imbalance and malnutrition [4,5].

Etiology of HPS is not known exactly. However, involvement of genetic, maternal, neuronal, humoral and environmental factors has been reported. Male infants have 5-fold higher incidence than female infants. This phenomenon has been associated with hypertrophic effects of testosterone hormone [6]. Lower plasma nitrite levels, abnormalities in various components of pyloric muscle like smooth muscle cells, growth factors, extracellular matrix elements, nerve and ganglion cells play important roles in its etiology. Pyloric stenosis has been reported during very early phase of life, especially in patients treated with erythromycin within the first weeks of their lives [7]. The role played by maternal fluoxetine use, viral respiratory infections [8] and bottle-feeding [9] and other macrolides has been demonstrated in the development of HPS. HPS can be associated with other malformations including malrotation, esophageal atresia, diaphragmatic hernia [4]. One of the cases presented by us was a patient with Down syndrome who had been operated with the indication of esophageal atresia and during follow-up period signs of HPS became apparent. Similarly, our two cases followed up with diagnosis of premature RDS, received claritromycin during early phase and during their follow-ups they were operated with the indication of HPS.

Diagnosis of HPS is mostly based on clinical findings in 60-80 % of the cases. Vomiting generally starts after the 3rd postnatal week. However symptoms may manifest within the first week of life or the infants may be asymptomatic up to the 5th month. Following a vomiting episode, the infant is hungry and wants to be fed again. With persistent vomiting, hypochloremic metabolic alkalosis develops due to loss of fluid, hydrogen ion and chloride from the body. With palpation during physical examination "olive sign" can be detected on the pyloric region. Since abdominal muscles relax after a vomiting episode, "olive sign" can be better detected on palpation. After feeding of the baby, peristaltic waves of the pyloric region can be seen. On ultrasonographic evaluation, suspected cases can be easily clarified. Thickness of the pyloric muscle (>4 mm) and length of the pyloric canal (>14 mm) reinforce the diagnosis. Ultrasonographic examination has a diagnostic sensitivity of nearly 95 percent. Using contrast-enhanced imaging techniques, elongation of the pyloric canal, intra-antral protrusion of the pyloric muscle (shoulder sign) can be visualized [10]. However, contrasted radiograms are not routinely used for the imaging of the upper gastrointestinal system. In cases with pediatric HPS, if the infant is not considered to be in the suggestive age for HPS or clinical suspicion of HPS is not strong, then contrast-enhanced radiograms can be obtained to rule out malrotation and gastroesophageal reflux [1]. Since some of our patients had congenital anomalies and all of them had clinical symptoms suggesting HPS, we obtained contrast-enhanced upper gastrointestinal system radiograms in all of them. Because of lack of any electrolyte disorders and gastric dilation, even it was the appropriate time to diagnose HPS, at first, HPS was not firstly taken into consideration. Despite expected time of onset of symptoms of HPS, in infants hospitalized for longer periods in neonatal intensive care units, the interval between the emergence of symptoms and time of diagnosis may be delayed. Nutritional intolerance and vomiting episodes of the patients are frequently associated with problems of gastroesophageal reflux, necrotizing

enterocolitis, sepsis and prematurity. Feeding through orogastric/nasogastric catheters and gastric drainage in case of development of intolerance prevent gastric dilation and formation of olive sign. Compensatory replacement of non-bilious residuals by drained, and administration of total parenteral nutrition prevents development of acid-base and electrolyte disorders in these patients. In the literature, cases with delayed diagnosis of HPS have been reported. In an investigation performed by Boybeyi et al. cases with clinical complaints diagnosed as HPS later than the period of infancy have been also reported [3]. However, in the literature, we could encounter only one case with delayed diagnosis who had manifested clinical symptoms of HPS during treatment of their different complaints [11]. Our two cases having genetic and congenital anomalies and three premature patients had been hospitalized from birth. Nutritional intolerance and vomiting episodes of our patients were frequently associated with problems related to gastroesophageal reflux, necrotizing enterocolitis, sepsis and prematurity. In all of our cases, indwelling orogastric/nasogastric catheters prevented projectile vomiting episodes and gastric dilation, while total parenteral nutrition inhibited weight loss and development of hypochloremic metabolic alkalosis. All of these factors contributed to delayed diagnosis.

Patients diagnosed as HPS are treated with surgery performing routinely pyloromyotomy. This treatment modality was curative in all of our patients and their nutritional intolerance improved.

As a conclusion, we can say that, in patients treated in neonatal intensive care units for various indications, nutritional intolerance can be seen with or without any association with the underlying disease. These superimposed and concomitant conditions may delay the diagnosis of pyloric stenosis. In this article, we would like to emphasize on high index of suspicion to prevent any delay in diagnosis of HPS.

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