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Case Report

Paediatrics Section

A Rare Coincidence of Infantile Hypertrophic Pyloric Stenosis and Esophageal Atresia with Tracheo-Esophageal Fistula: A Case Report

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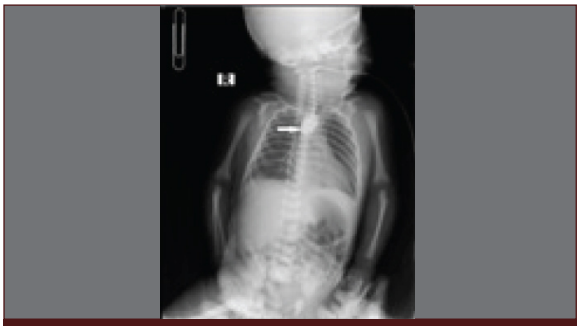
ABSTRACT

The coincidence of Infantile Hypertrophic Pyloric Stenosis (IHPS) and Esophageal Atresia (EA) with Tracheo-Esophageal Fistula (TEF) is rare. Although, vomiting and regurgitation in operated cases of EA and TEF are attributed to Gastroesophageal Reflux (GER) and the stricture of the anastomosis, it may be also associated with IHPS as well. We report a case of 3-hour-old female infant, who had EA and TEF operation and diagnosed to have IHPS at 9th week of age. Early diagnosis and treatment can prevent complications.

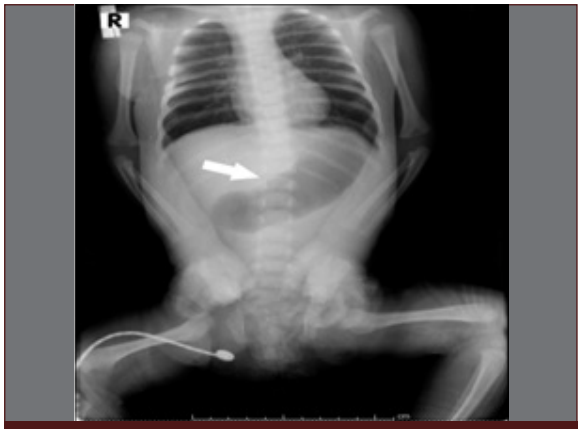
Keywords: Newborn, Gastroesophageal reflux, Olive sign, Salivation

CASE REPORT

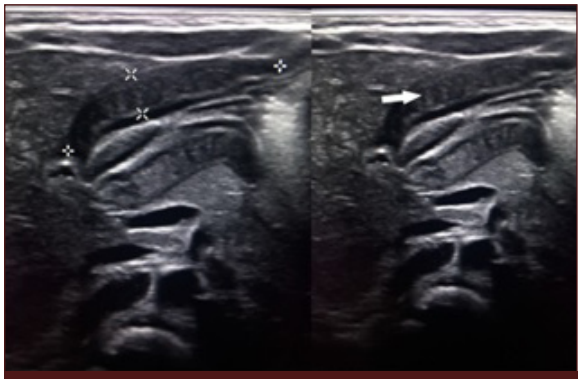
A 3-hour-old female newborn was admitted to our Neonatal Intensive Care Unit (NICU) with a complaint of excessive salivation and failure to pass a nasogastric tube into the stomach. Baby was born at 36 week gestational, 2690 grams, by normal vaginal delivery as the second child of 33-year-old mother. Her mother had no history of radiation exposure, drug ingestion, alcohol use, smoking during pregnancy, family history was unremarkable for congenital anomalies and there was no consanguinity. At presentation, her body temperature was 36.8°C, with blood pressure of 84/48 mmHg, heart rate of 144 beats per minute and respiratory rate of 72 breaths per minute. Her breath sounds were equal, she had grunting, nasal flaring, tachypnoea, subcostal retraction, central cyanosis. She was frothing from the mouth. The attempt of putting a no. 10 catheter into the stomach was not successful. Pulmonary consolidation on the right lung and decreased gas was seen in her chest and abdominal X-ray. The diagnosis of EA and TEF was confirmed, as the radio contrast agent didn't pass through the esophagus and the catheter rolled at the distended upper esophageal pouch [Table/Fig-1]. Her hemogram and blood biochemistry were normal. Echocardiogram and abdominal Ultrasonography (USG) were normal. Upon admission, the patient was started on antibiotics and she had assisted ventilation. On 2nd day of her life, primary esophageal anastomosis and ligation-division of fistula were performed via right thoracotomy. After the operation she had GER, treated with Histamine-2 receptor antagonist, domperidon and sodium alginate. The baby was extubated on her 7th day of life and mode of the mechanical ventilation was swithced to Continuous Positive Airway Pressure (CPAP). Since, she had laryngomalacia and tracheomalacia, she could not be weaned from CPAP untill she was 40 days old. She was reached to full enteral feeding on her 21st day of life. On 48th day of her life, after feeding, she had vomiting and had respiratory distress. Pneumonia due to aspiration of the milk was detected. Broad spectrum antiobiotic treatment was given and she had assisted ventilation. During the follow-up, she had feeding difficulties because of vomiting episodes. Regurgitation and vomiting were attributed to GER. The contrast esophagogram revealed GER and ruled



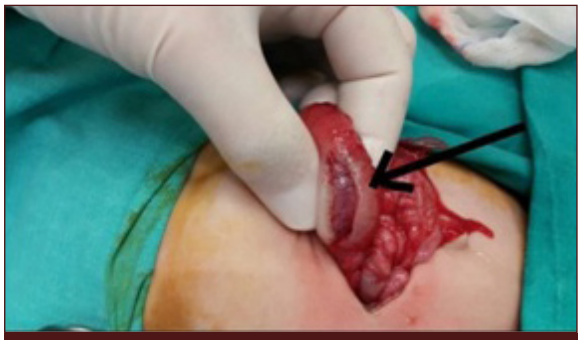
[Table/Fig-1]: Chest and abdominal X-ray. The arrow shows the dilated, blind-ending proximal esophageal pouch in the neck.



[Table/Fig-2]: Abdomian X-ray. The arrow shows dilated stomach and gastric outlet obstruction.



[Table/Fig-3]: Abdominal USG. The thickness of the pyloric muscle is 53 mm, the diameter of the pylorus is 4.6 mm and the canal length is 3 cm.



[Table/Fig-4]: Olive is seen before the pyloromyotomy.

out the anastomotic stricture. At the age of 9th week, abdominal X-ray showed grossly dilated stomach and gastric outlet obstruction was suspected [Table/Fig-2]. In the meantime, a projectile non-bilious vomiting was observed. GER, the stricture of the anastomosis, mechanical intestinal obstruction such as postoperative ileus and IHPS were the main pathologies in the differential diagnosis. Abdominal USG was performed. Pyloric muscle thickness measured 53 mm, pyloric diameter 4.6 mm and canal length 3 cm [Table/Fig-3]. The baby was diagnosed with IHPS when she was 65 days old. The deep palpation of the abdomen revealed the 'olive sign'. At the operation a transverse incision was made under general anesthesia. Hypertrophic pylorus (olive) was seen [Table/Fig-4]. Fredet-Ramstedt pyloromyotomy was done. On 5th day of the operation, she reached to full enteral feeding. At 76th day of her admission, she was discharged from the hospital with nasal oxygen therapy due to chronic lung disease, laryngomalacia and tracheomalacia.

DISCUSSION

IHPS following EA repair is rarely reported [1]. IHPS is characterized by postnatal hypertrophy of the longitudinal and circular muscles of the pylor causing gastric outlet obstruction and non bilious vomiting [2]. Although, there are some theories for the ethiology of IHPS, the most known theories are the genetic predisposition, the environmental theory and the neurogenic theory. According to Mahalik S et al., IHPS is an acquired condition in which the hypertrophy of the fibers of the mucosa, narrows the pyloric canal and results in the obstruction [3]. Paulozzi LJ et al., discussed the acquired theory for IHPS and claimed that *Helicobacter pylori* is the reason of IHPS [4]. Sorensen HT et al., suggested that maternal smoking during pregnancy or postnatally is a risk factor for IHPS [5]. Hoey reported seven neonatal cases of IHPS after erythromycin prophylaxis due to a pertussis epidemic [6]. Maternal risk factors that have been reported include hyperthyroidism, nalidixic acid use, intranasal decongestant use, raised pregnancy BMI. IHPS strongly aggregates in families even among distant relatives and there is a high concordance between monozygotic twins [7].

The incidence of additional congenital anomalies associated with EA and TEF is approximately 30-50%, IHPS coincidence of postoperative EA and TEF is rarely reported in the literature [8]. However, Van Belen NW et al., have reported that the incidence of IHPS was 7.5% after EA repair. Postoperative vomiting occurs often in neonates, especially who have had abdomino-thoracic surgery such as EA and TEF repair, which is due to GER or anastomotic stricture [9]. Hassan RAA et al., reported a newborn who had EA and TEF operation in the 2nd day of life. The infant was diagnosed as IHPS at the 8th week of his life, although he had GER and anastomotic stricture proven by esophagogram [1]. Lipszyc ES et al., reported 2 cases who have had EA and TEF repair operations, in the 3th and 2nd day of life respectively. Both of the cases admitted to the hospital after the first discharge because of the persistant vomiting at the 5th and 7.5th week of life. These 2 cases also have other congenital anomalies such as hydronephrosis, cardiac defects or anus anomalies. Both of the patients underwent pyloromyotomy for the treatment of IHPS [8]. Our patient had an accomplished EAH and TEF repair operation in the 2nd day of her life. In the postoperative period, our patient had vomiting episodes which were attributed to GER. The enlarged pylorus, classically described as an "olive" which can be palpated in the right upper quadrant or epigastrium of the abdomen is called 'olive sign' which is seen approximately 60-80% of infants with IHPS. As our patient had persistant vomiting, so first we performed esophagogram and

ruled out the anastomotic stricture. As our patient had projectile non-bilious vomiting and had olive sign during deep palpitation of the abdomen, we suspected that of IHPS. We confirmed the diagnosis by USG at the 9th week of her life. Our case didn't have any other additional anomaly except IHPS. We have been following up our patient for the long term complications of the abdominal surgery such as postoperative ileus and stricture of the anastomosis, pulmonary and gastrointestinal complications. At 6 months of age, her weight was at the 10th percentile and height was 50th percentile.

USG is the best choice of imaging method for the diagnosis of IHPS. The diagnosis is confirmed using USG, which has 100% sensitivity and specificity in the experienced hands. Although, the pyloric muscle thickness of greater than 3 mm and pyloric canal length greater than 15 mm are considered diagnostic for IHPS, the diameter of the pyloric muscle is the most important criteria for the diagnosis however the measurements differ according to the patient's weight and age [10].

CONCLUSION

Infants having abdomino-thoracic surgery such as EA and TEF repair operation, IHPS should be kept on mind as a rare anomaly in the presence of persistant vomiting if GER or anastomotic stricture were ruled out. A high index of suspicion and diagnostic radioimaging may help earlier diagnosis and intervention.

REFERENCES

[1] Hassan RAA, Choo YU, Noraida R, Rosida I. Infantile hypertrophic pyloric stenosis in postoperative esophageal atresia with tracheoesophageal fistula. Journal of Neonatal Surgery. 2015;4(3):32-33.

[2] Khan AA, Yousaf MA, Ashraf M. Role of ultrasonography in eary diagnosis of infantile hypertrophic pyloric stenosis. J Ayub Med Coll Abbottabad. 2014;26(3):316-19

[3] Mahalik S, Prasad A, Sinha A, Kulshrestha R. Delayed presentation of hypertrophic pyloric stenosis: a rare case. J Pediatr Surg. 2010;45(2): e9-11.

[4] Paulozzi LJ. Is *Helicobacter pylori* a cause of infantile hypertrophic pyloric stenosis?. Med Hypotheses. 2000;55(2):119-25.

[5] Sørensen HT, Nørgård B, Pedersen L, Larsen H, Johnsen SP. Maternal smoking and risk of hypertrophic infantile pyloric stenosis: 10 year population based cohort study. BMJ . 2002;325(7371):1011-12.

[6] Hoey J. Hypertrophic pyloric stenosis caused by erythromycin. CMAJ. 2000;162(8):1198

[7] Mouafo Tambo FF, Kouna Tsala IN, Ngowe Ngowe M, Andze Ondobo G, Sosso MA. Hypertrophic pyloric stenosis in infants: is it a congenital or acquired disorder? Reflections on 2 cases. Springer Plus. 2014;3:555.

[8] Lipszyc ES, Klin B. Hypertrophic pyloric stenosis following repair of esophageal atresia and tracheo-esophageal fistula. Journal of Neonatal Surgery. 2014;3(3):30-31.

[9] Van Belen NW, Mous DS, Brosens E, de Klein A, van de Ven CP, et al. Increased incidence of hypertrophic pyloric stenosis in esophageal atresia patients. Eur J Pediatr Surg. 2014;24(1):20-24.

[10] Jobson M, Hall NJ, Bchir MB. Contemporary management of pyloric stenosis. Seminars in Pediatric Surgery. 2016;25: 219-24.

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