AN UNCOMMON ASSOCIATION OF H – TYPE TRACHEOESOPHAGEAL FISTULA WITH INFANTILE HYPERTROPHIC PYLORIC STENOSIS*

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SUMMARY: Oğuzkurt P, Tanyel FC, Haliloğlu M, Hiçsönmez A. (Departments of Pediatric Surgery and Radiology, Hacettepe University Faculty of Medicine, Ankara, Turkey). An uncommon association of H-type tracheoesophageal fistula with infantile hypertrophic pyloric stenosis. Turk J Pediatr 1999; 41: 143-146.

Although infantile hypertrophic pyloric stenosis following esophageal atresia repair is known, infantile hypertrophic pyloric stenosis following H-type tracheoesophageal fistula has not been encountered previously. A case of H-type tracheoesophageal fistula and infantile hypertrophic pyloric stenosis is presented. The patient, operated on for H-type fistula, a rare congenital anomaly of the esophagus, on the tenth day of life was readmitted 19 days later because of continous vomiting after every feeding. The clinical findings and physical and radiological examinations revealed infantile hypertrophic pyloric stenosis which required surgical treatment. It is suggested that the association of H-type tracheoesophageal fistula with infantile hypertrophic pyloric stenosis is coincidental, given the estimated incidence of one in every 84,375,000 males and 337,500,000 females. *Key words: hypertrophic pyloric stenosis, H-type tracheoesophageal fistula.*

Esophageal atresia (EA) with tracheoesophageal fistulas (TEF) and infantile hypertrophic pyloric stenosis (IHPS) are among surgical problems frequently encountered in the newborn period. Although the occurrence of infantile hypertrophic pyloric stenosis is reported in patients with esophageal atresia and tracheoesophageal fistula, this entity has not been previously reported in patients with H-type tracheoesophageal fistula in English language literature. To the best of our knowledge this is the first described case.

Case Report

A four-day-old male was admitted to the hospital because of coughing, choking and cyanosis during and after feeding. He was the product of the fifth pregnancy of a 32-year-old mother and had a birth weight of 3000 g. Physical examination revealed severe icterus and coarse rales in both hemithoraces. Laboratory examinations including complete blood count, urinalysis and blood chemistry

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were within normal limits except for indirect hyperbilirubinemia with 24.2 mg/dl. After exchange transfusions were repeated twice, his indirect bilirubin level returned to normal limits. On posteroanterior chest x-ray there was consolidation at the right upper lobe. Cineesophagography was performed and revealed H-type tracheoesophageal fistula at the level of the second thoracic vertebra (Fig. 1). On his tenth day of life, he was operated on through a left cervical transverse incision, and a large fistula was isolated, transected and sutured. He was fed orally on the fourth day following the operation and was discharged without any complaints. When he was 29-days-old, he was readmitted because of vomiting and coughing for 15 to 30 minutes following feeding. Barium meal performed with the suspicion of refistulization revealed the previous fistula region to be normal. Gastroesophageal reflux was observed on fluoroscopic examination. On erect abdominal x-ray taken the next day, the stomach air was dilated (Fig. 2). On physical examination a small mass (olive) was palpable above the umbilicus and the abdomen was scaphoid. Laboratory investigations revealed alkalosis and mild hypochloremia. Abdominal ultrasonography was performed and supported the clinical diagnosis of hypertrophic pyloric stenosis (Fig. 3). The patient was operated on through a small right upper quadrant transverse incision. A hypertrophic pyloric muscle was found and Ramstedt pyloromyotomy was performed. The patient was fed on the first day of the operation and he had an uneventful recovery. He had no problems during the six month follow-up period.

Fig. 1: Cineesophagography of the patient demonstrating the H-type tracheoesophageal fistula at the level of the second thoracic vertebra.

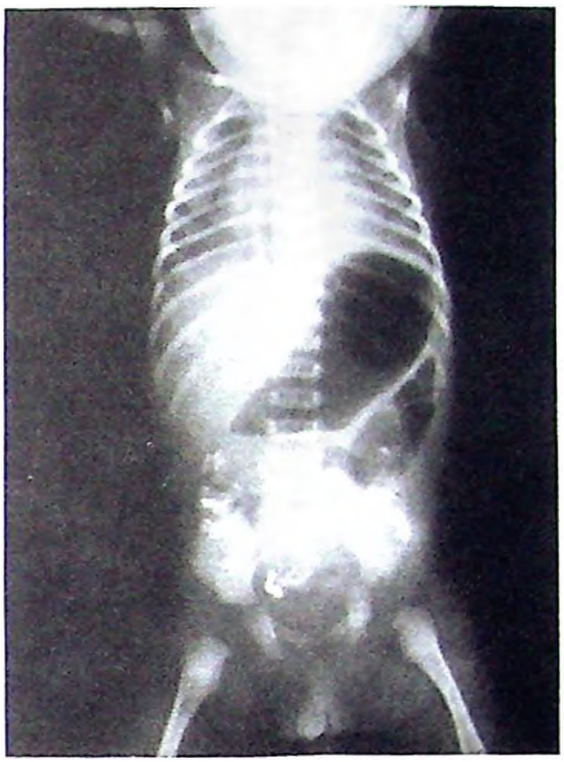

Fig. 2. Plain abdominal x-ray with the dilated gastric air.

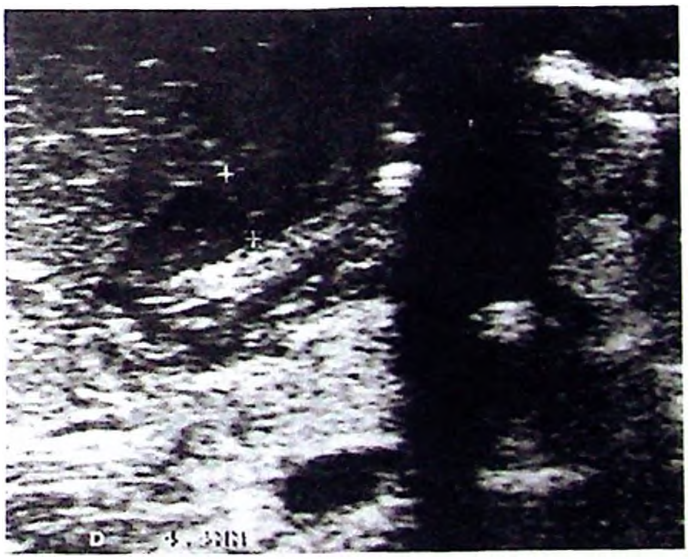

Fig. 3: Ultrasonography showing the thickness of the pyloric muscle.

Discussion

Nearly 50 percent of patients with EA have additional congenital anomalies[1,2]. The frequency of other gastrointestinal anomalies in patients with EA is 18 percent[2], the most commonly encountered being imperforate anus, duodenal atresia (with or without annular pancreas) and pyloric stenosis[1,3]. In patients with EA the frequency

of IHPS has been reported to be between one and 10 percent[4]. On the other hand, only six percent of patients with IHPS had additional anomalies, 13 percent of which are associated with the gastrointestinal system[4].

There are nearly 40 cases in the literature where IHPS developed following operations for EA[1]. However, IHPS associated with H-type fistula has not been reported perviously. Since their etiologies are different, it is suggested the occurrence is coincidental. Since EA is encountered once in 4,500 live births with males and females equally affected and since four percent of the cases are H-type TEF[5], the estimated incidence of H-type TEF becomes one in 112,500 males or females.

On the other hand, IHPS is encountered once in 300 live births, with a 4:1 male to female ratio[6]. These ratios make the estimated incidence of both IHPS and H-type TEF one in 84,375,000 males and 337,500,000 females.

Since no common etiologic factor is known for H-type fistula and IHPS, this suggests a coincidence. Although this is the first reported case of this association, it should be estimated as one in 84,375,000 males and 337,500,000 females.

REFERENCES

1. Magilner AD. Esophageal atresia and hypertrophic pyloric stenosis: sequential coexistence of disease. AJR 1986; 147: 329-330.
2. Ein SH, Shandling B, Wesson D, Filler RM. Esophageal atresia with distal tracheoesophageal fistula: associated anomalies and prognosis in the 1980s. J Pediatr Surg 1989; 24: 1055-1059.
3. Manning PB, Morgan RA, Coran AG, et al. Fifty years' experience with esophageal atresia and tracheoesophageal fistula. Beginning with Cameron Haight's first operation in 1935. Ann Surg 1986; 204: 446-453.
4. Bidair M, Kalota SJ, Kaplan GW. Infantile hypertrophic pyloric stenosis and hydronephrosis. is there an association? J Urol 1993; 150: 153-155.
5. Randolp GJ. Esophageal atresia and congenital stenosis. In: Welch KJ, Randolp JG, Ravitch MM, O'Neill JA, Rowe MI (eds). Pediatric Surgery (4th ed) Vol 1. Chicago: Year Book Medical Publishers, Inc; 1986: 684.
6. Benson CD. Infantile hypertrophic pyloric stenosis. In: Welch KJ, Randolph JG, Ravitch MM, O'Neill JA, Rowe MI (eds). Pediatric Surgery (4th ed) Vol. 2. Chicago: Year Book Medical Publishers, Inc; 1986: 811.