·病例报告·

Esophageal atresia/tracheoesophageal fistula complicated by hypertrophic pyloric stenosis: a case report

ZHANG Zhi-Bo

(*Department of Pediatric Surgery, Shengjing Hospital, China Medical University, Shenyang* 110003, *China*)

食道闭锁气管食管瘘合并肥厚性幽门狭窄 1 例报道

张志波 中国医科大学附属盛京医院小儿外科,辽宁沈阳 110004

[摘 要] 患儿,女,2 d。因出生后喂养困难,口鼻持续有泡沫样唾液流出 2 d 入院。患儿胎龄 36 周$^{+2}$,出生体重 2 510 g,身长 46 cm,Apgar 评分生后 1 min 和 5 min 均为 10 分,系剖腹产娩出。母健康,妊娠期间无异常,产前超声检查未提示有羊水异常。出生后发现喂养困难,口鼻持续有泡沫样唾液流出,其他未见异常。生后 2 d 就诊。就诊时发现口鼻内有泡沫样液体,上腹饱满,胃肠减压管插入受阻,疑诊为食道闭锁(esophageal atresia, EA),行胸腹平片检查诊断为 EA。同时发现胃高度扩张,提示存在气管食管瘘(tracheo-esophageal fistula, TEF),而远端肠气缺失则提示高位肠梗阻;食道 CT 证实了 EA 合并 TEF,CT 同时提示了食道盲端距离。入院后 2 d 行右侧开胸胸膜外食道吻合术,同时结扎切断气管食管瘘。随后行开腹探查术,旨在探查、治疗高位肠梗阻。经仔细探查除了发现幽门肥厚水肿外并未发现其他高位肠梗阻,幽门管长 2 cm,术中诊断为肥厚性幽门狭窄(hypertrophic pyloric stenosis, HPS),行幽门肌切开术,证实幽门肌厚达 4 mm。术后恢复过程顺利。经 10 d 全胃肠道外营养治疗后,食管造影证实吻合口通畅良好,无吻合口狭窄或吻合口瘘,随后给予母乳喂养,耐受良好。术后 14 d 出院。染色体核型为正常 46, XY (GTG 500 bands)。随访至今患儿已 14 个月,体重增长良好,进食正常,未出现术后并发症。

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Although additional congenital anomalies in association with esophageal atresia (EA) and tracheoesophageal fistula (TEF) are well known, infantile hypertrophic pyloric stenosis (HPS) complicated by EA/TEF is rare. Here a case of neonate suffered from EA /TEF and HPS is reported.

Presentation of case

A 2-day-old baby girl was born from healthy parents by cesarean section at 36 weeks 2 days of gestation. Her birth weight was 2 510 g, and the Apgar score at birth was 10. The pregnancy had been normal without polyhydramnios on the prenatal US scan. The baby was otherwise well, except for feeding intolerable and foamy saliva kept running from the mouth and nose. Two days after birth, she was carried to the hospital. On presentation, foamy saliva was observed coming from her mouth and nose and she had a distended upper abdomen. The naso-gastric tube could not be introduced. EA was suspected and so chest radiography was performed. Radiography found the obstructed tube and the dilated proximal pouch of the esophagus, so the diagnosis of EA was confirmed. A highly distended stomach and lack of gas from the outlet of stomach indicated a TEF of the distal tract and high intestinal obstruction (Figure 1). A later esophageal computed tomography (CT) study confirmed EA as well as TEF (Figure 2). CT also indicated the distance of the two ends. Two days after admission, the baby underwent reconstructive surgery with primary anastomosis of the two esophageal parts through a right extrapleural thoracotomy, with ligation and section of the fistula. A laparotomy was then performed in order to detect and repair the high intestinal atresia. After careful exploration, no other lesions but an edematous and hypertrophic pylorus were found, with the length of pyloric channel reaching nearly 2 cm. HPS was confirmed, and pyloromyotomy was performed which revealed the muscle thickness of about 4 mm. The baby was well in the post-operative period. She underwent total parenteral nutrition (TPN) for the first 10 days after surgery, and then a

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[Biography] ZHANG Zhi-Bo, Female, MD., Associated professor. Email: cathyzbzhang@sina.com

contrast esophagogram showed a patent anastomosis without leaks or stenosis. Breast milk was progressively introduced with a good tolerance. The baby was discharged from hospital 14 days postoperatively. The karyotype revealed a normal pattern (46, XY). She is now 14 months old, is tolerating a normal diet and has a satisfactory weight gain. No postoperative complications were found.

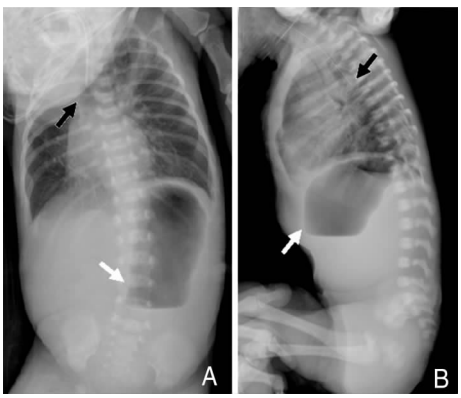

Figure 1 Plain X-ray study of the chest and abdomen (A: Anterior-posterior position; B: Lateral position) Black arrows indicate the blocked nasoesophageal tube as well as the dilated proximal esophageal pouch, white arrows indicate the dilated gastric bubble, and the other parts of the abdomen are blank.

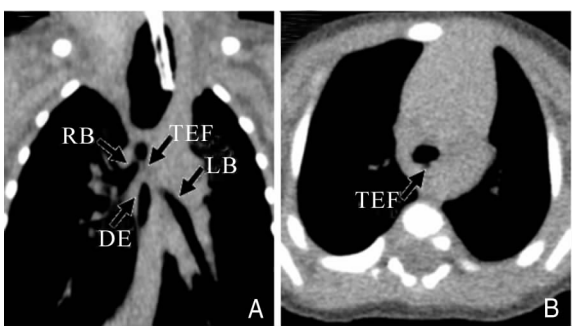

Figure 2 Esophageal CT study A: Coronal section; B: Transverse section. CT scan shows abnormal developments of esophagus and trachea.

LB: left bronchus; RB: right bronchus; TEF: trachoesophageal fistula; DE: distal esophagus.

Discussion

There is a well-known incidence of additional congenital anomalies in infants with EA. EA has been divided into two different forms: isolated EA (IEA, 50%) and syndromic EA (SEA, 50%). The most frequent associated malformations are cardiac (13%-34%), limb (5%-19%), renal (5%-14%) and vertebral malformations (6%-21%), as well as anal atresia (10%-16%)[1-4]. Patients with EA show signs of onset immediately after birth, while HPS often happens in infants at ages of 3-6 weeks. Although it is also a very common pediatric condition, it is rarely associated with TEF and EA (a reported frequency of 1%-10%). In 1986, Magilner[5] reported 2 cases of this combination and reviewed other 35 reported cases. Since then, less than 10 cases have been reported about TEF/EA associated with HPS [6-8]. In these reports, HPS always happened in the postoperative course of EA complicated by TEF or not. It is now believed that HPS is an acquired entity, and has an interesting age- and tissue-specific nature; and mostly happens at ages of 3-6 weeks. So, the delayed onset of HPS in all these reports was comprehensible. It is believed that the concurrence of the two conditions in a newborn has never been described previously.

The management of an infant with such a complex constellation of anomalies requires logical definition of surgical priorities. Pyloromyotomy was performed rightly after repair of the EA in order to allow proper placing of the nasogastric tube and normal gastric emptying. This minimized the risk of gastroesophageal reflux and decreased the outlet pressure of the esophagus to guarantee the healing of the anastomosis. Following the optimal repair of the esophagus, this infant is doing well now at the age of 14 months.

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