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Pyloric Stenosis Complicating Esophageal Atresia.

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Abstract:

Background: Post surgical complications of esophageal atresia (EA) include anastomotic leak, anastomotic stricture, gastroesophageal reflux, tracheomalacia, and recurrent tracheoesophageal fistula (TEF). However, hypertrophic pyloric stenosis (HPS) is complicating EA which seems to be rare. The aim of this report is to emphasize on HPS as a possible complicating post operative course of EA, the diagnosis of this complicating disorder may be delayed.

Case presentation: A 3000 gram male infant was born at 38 weeks gestation to a 21-year-old, gravid mother by cesarean section. He presented vomiting with profuse foamy discharge and salivation. Esophageal atresia was confirmed by looped orogastric tube in the upper pouch of esophagus in chest X-ray. He underwent surgical correction and was discharged with a good condition on 7th post operation day. Nevertheless three weeks later, he developed recurrent vomiting. The diagnosis of HPS was confirmed by ultrasonography and barium upper gastrointestinal (GI) series. Pyloromyotomy was done on him and He was discharged without any complication. At 3.5 months old, he developed regurgitation and choking cyanosis. At this time esophageal stenosis was diagnosed by barium swallow. The stricture was treated by gastroduodenoscopy.

Conclusion: Infantile HPS should be considered in any case of persistent vomiting and feeding intolerance after surgery for esophageal atresia. A high index of suspicion is required for diagnosis to avoid complication arising from a delayed diagnosis.

Keywords: Esophageal Artesia, Tracheoesophageal fistula, Hypertrophic pyloric stenosis

Introduction:

Hypertrophic pyloric stenosis complicating the post operative course of EA/ TEF is less well known. There have been 35 cases of this complicating disorder have been reported. The incidence of EA/tracheoesophageal fistula (TEF) is roughly 1 to 2 per 5000 live births.^(1, 2)

Esophageal atresia was first reported by William Durson in 1670, and Thomas Gibson described EA with TEF in 1697. Associated congenital malformations are seen in 50% of the patients; other gastrointestinal malformations are in 25% of the cases, and VACTERL (vertebral/ anal atresia / Cardiac/ TEF/ Limb deformity) associations occur in 20% of patients who have EA. The complications following operation for EA that are relatively common include anastomotic leak, anastomotic stricture, gastroesophageal reflux, tracheomalacia, and recurrent TEF.⁽³⁾

Hypertrophic pyloric stenosis is an acquired disease in which the circumferential muscle layer of the pyloric sphincter becomes thickened, resulting in the narrowing of the pyloric channel. The etiology of HPS is unknown, nevertheless, understanding of the condition and effective management have undergone a remarkable evolution in the 20th century, reducing the mortality rate from over 50% to nearly 0%.^(2, 4) The typical patient is a term male infant between 3-6 weeks of age who has progressive non-bilious projectile vomiting.

In any patient, presence of non-bilious projectile vomiting following EA repair should attract the attention of the HPS; although is not a common cause of vomiting after EA surgery. Anastomotic stenosis is the most common cause of further surgery after repair of EA. Infants

with a stricture have feeding difficulty and dysphagia with regurgitation and choking cyanosis, and the diagnosis can be confirmed by Barium Swallow or Esophagoscopy. The therapy includes balloon dilatation under fluoroscopic control.⁽⁵⁾

In this report, we emphasize on the diagnosis of HPS in any patient with recurrent vomiting and feeding intolerance after operation of EA and also on dilatation of esophageal stricture by Savary bougie gastroduodenoscopy instead of balloon dilatation.

Case History:

A 3000 gram male infant was born to a 21-year-old primigravida mother at a 38 week gestation by caesarean section due to decrease fetal activity with Apgar score 8 and 10 at one and five minutes after birth, respectively. His mother didn't take any medications during pregnancy. Polyhydraminous was detected by prenatal ultrasound. Otherwise she was healthy. The parents were not first cousins. The newborn's clinical condition after birth was good. He was put on his mother's chest for the first feeding. The baby developed vomiting and choking cyanosis after the first feed, and experienced profuse foamy discharge and salivation. He was suspected to have proximal esophageal atresia which was confirmed by looped orogastric tube in the upper pouch of esophagus in chest X-ray. Abdominal x-ray showed gaseous distension (due to TEF). A systolic murmur was

detected by cardiac examination. The echocardiography demonstrated tricuspid regurgitation and patent foramen ovale. The patient was operated with diagnosis of EA/TEF. The post operative course was uncomplicated and the patient was discharged with a good condition on 7th post operative day.

Nevertheless, when he was 27 days old, he was presented with repeated non-bilious vomiting, that gradually progressed to projectile vomiting. He was admitted in neonatology ward, arterial blood gas and electrolyte determination, abdominal ultrasonography and contrast study (Fig-1) were done. Ultrasonography and upper GI series were in favour of HPS. After pyloromyotomy under general anaesthesia his condition improved and stopped vomiting. The patient had a regular outpatient follow up without any problem. When he was 3.5 months old, he presented with regurgitation and gradually had choking cyanosis after meal. Esophageal stricture was detected in the middle third part of esophagus. Dilatation of stenosis by Savary bougie gastroduodenoscopy (PENTAX –EG2540) was effective. (Fig-2)



Figure 1



Figure 2

Discussion:

Infantile hypertrophic pyloric stenosis (IHPS), that is the most common surgical disorder causing vomiting, presents at 2-8 weeks of age. The etiology of IHPS remains a mystery. Some suspected risk factors include birth rank, maternal age, male sex, family history and monozygosity in twins. Various theories have attempted to explain its etiology.

It is suggested that enteric neuronal damage and nitric oxide synthases dysfunction may be implicated, but environmental modification may exist to account for the variability in its occurrence.⁽⁶⁾ IHPS cause functional gastric outlet obstruction leading to non-bilious projectile vomiting.

The frequency of pyloric stenosis is approximately one in 400 births.⁽¹⁾ Most of the literatures suggest that pyloric stenosis is an acquired entity and its onset usually occur after birth. Imaging techniques particularly ultrasounds provide non-invasive and accurate techniques for identification of HPS. Pyloromyotomy is a common operative procedure performed on these infants.^(5, 7)

Laparoscopic pyloromyotomy is a new treatment modality. Peter et al. com-

pared two techniques and concluded that there were no significant differences in operating time and time to full feeding and also less postoperative pain and emesis and more rapid recovery after laparoscopic pyloromyotomy.^(8, 9) The occurrence of HPS as a possible complicating factor after EA repair is rare. HPS is different from most of the post operative complications of EA/ TEF that are apparent in the neonatal period. Magilner in 1986 reported two cases of HPS after repair for EA and TEF, their diagnosis was confirmed by Barium swallow, and feeding difficulty has improved after surgical correction of HPS.⁽¹⁾

Zhi-Bo reported one case of hypertrophic pyloric stenosis after operation of esophageal atresia and Karmakar reported a newborn baby with EA/TEF who expired due to anastomotic leak and mediastinitis. HPS was confirmed by autopsy examination.^(10, 11)

One of the common complications following EA/TEF repair is esophageal stenosis which is late complication after anastomotic. These strictures may result from various causes including fibrosis, ischemia, leakage, acid peptic injury, and excessive tension. An anastomotic stenosis should be suspected whenever dysphasia or respiratory symptoms in an infant who had previous tolerated feedings. Under fluoroscopic guidance, the stenosis segment is dilated and there may be a need for repeating the procedure. Finally, a refractory stenosis may be managed with segmental resection.^(2, 5) But in our case esophageal stricture was dilated by Savary bougie gastroduodenoscopy with a good result and his symptoms improved after the treatment.

Conclusion:

HPS should be considered in the evaluation of recurrent vomiting in an infant who had had recent surgery for EA/ TEF, and also gastroduodenoscopy dilatation might be a good modality of treatment of esophageal stenosis instead of balloon dilatation under fluoroscopic control.

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