Pediatrik Cerrahi Dergisi, 7:44-45, 1993

Hypertrophic pyloric stenosis with esophageal atresia : An etiologic dilemma of hypertrophic pyloric stenosis

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Summary

We present a case of hypertrophic pyloric stenosis (HPS) with isolated esophageal atresia. The diagnosis was established peroperatively during the gastrostomy procedure, when the child is twenty six days old. The operation had been delayed for unpredicted reasons over a period of three weeks. During this interval she was maintained only on total parenteral nutrition. The presented case is unique in developing HPS without oral or gastrostomy feeding and some of the related theories on the etiology of HPS are discussed.

Key word: *Esophageal atresia, hypertrophic pyloric stenosis*

Introduction

Additional congenital anomalies in association with esophageal atresia show an incidence up to 50 percent [5,7,10,11]. Within this scope, the appearence of esophageal atresia in combination with hypertophic pyloric stenosis (HPS) is very rare [5-7,8,10]. About 119 reported cases are encountered in the literature [1,2,4-7,8-10]. We present a case of isolated esophageal atresia associated with pyloric stenosis.

Case Report

A female baby, 1500 gm of body weight was born in a rural district health center on July 9, 1991. Because of prematurity, jaundice and respiratory distress; she was referred to Department of Pediatrics of Ege University Hospital on the 16 th of July, 1991. During her treatment she was put on total parenteral nutrition. When she was reverted to oral feeding, regurgitation was encountered and the diagnosis of esophageal atresia was established. Then, she was referred to the Department of Pediatric Surgery on the 2 nd of August, 1991.

Family history revealed no abnormalities and there was neither mention of X-ray nor medication during gestation. The 4 siblings, were all alive and healthy. On physical examination, there was undue secretion in nasopharynx. Abdomen was scaphoid and soft. Neither cardiac nor nephrologic or any other gastrointestinal system pathology were encountered. Since the plain abdominal roentgenogram revealed a gassless abdomen, the case was diagnosed as isolated esophageal atresia.

A gastrostomy was planned when she was twenty-six days old. At laparotomy the stomach was too small (4 X 2 cm) and the pylorus was hypertrophic (2.5 X 1.5 cm). A pyloromyotomy was performed followed by a gastrostomy.

Discussion

The etiology of HPS is still to be eluciated [3,9,11,12]. There are a number of theories on its etiology. Formerly, Lynn proposed a simple physiologic explanation that milk curds propelled by gastric musculature against a pyloric canal in spasm produce edema of the pyloric mucosa and submucosa with subsequent narrowing of the pyloric canal [9]. He postulated a responsive work-hypertrophy of the pyloric and gastric musculature, setting up a vicious cycle that progresses to high grade obstruction of the pyloric canal.

Nevertheless, it has been postulated that the presence of a gastrostomy may set the above mentioned

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factors into motion by local irritation and overdistention of the stomach at feeding time [1]. Inversely, another theory of continuous gastric decompression reducing intraluminal pressure to a le-vel insufficient to keep the pyloric canal open is also suggested [11]. Gastrostomy has also been incriminated as the cause of reccurrent pyloric stenosis following pyloromyotomy [12].

In contrast to our case in all the 119 reported cases encountered in the English literature, HPS had been developed after oral or gastrostomy feedings. In our case, esophageal atresia did not permit oral feedings for over a period of three weeks, that is the well known scope of 3-6 weeks, when the HPS is encountered most frequently.

During this 3 weeks period the patient hadn't been fed at all. Consequently, any jeopardizing effect to produce edema leading to either narrowing or work-hypertrophy of the pylorus was not present. Since HPS had developed before gastrostomy, the gastrostomy related theories are invalid to reveal the etiology in our case.

In conclusion, our case clearly contradicts both the irritation theory of propelled milk curd, and the theories incriminating gastrostomy.

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