Pediatrik Cerrahi Dergisi, 10:101-103, 1996

Midgut atresia with partial situs inversus

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Özet

Parsiyel situs inversus ile birlikte orta barsak atrezisi

Abdominal situs inversus (Sİ) ile birlikte orta barsak atrezisi olan bir olgu sunulmuştur. Ameliyat öncesi parsiyel Sİ ve proksimal barsak tıkanıklığı tanısı radyolojik olarak konabilmiş ve laparotomide de bu tanı konfirme edilerek tıkanıklık sebebi olarak orta barsak atrezisi saptanmıştır. Ameliyatta tapering jejunoplasti sonrası jejunum ve kolon anastomozu yapıldı. Ameliyat sonrası dönemde fonksiyonel tıkanıklığa bağlı olarak ağızdan beslenmeye geçilemeyen ve total parenteral beslenme tedavisi uygulanan bebek, ameliyat sonrası 18. gün kateter sepsisi ile kaybedilmiştir. Olgumuz oldukça nadir olan abdominal Sİ ve orta barsak atrezi birlikteliğini sergilemektedir.

Anahtar kelimeler: *Situs inversus, barsak atrezisi*

Summary

A female newborn with abdominal situs inversus (SI) in association with midgut atresia is presented. The initial diagnosis of partial SI and proximal intestinal obstruction was easily made by radiologic modalities. An emergency laparotomy confirmed the diagnosis of SI and the intestinal obstruction was found to be due to midgut atresia. Tapering jejunoplasty and jejunocolic anastomosis with resection of atretic segmens were performed. Postoperatively, the patient needed to receive total parenteral nutrition via central catheter since oral feedings could not be instituted because of functional obstruction at the anastomosis. She died of catheter related sepsis at the 18th postoperative day. It appears that this case represent a very rare association of abdominal SI with midgut atresia.

Key words: *Situs inversus, intestinal atresia*

Introduction

Various types of intestinal atresia with abdominal SI have been reported in the literature [1,2,8]. However, the relationship between these anomalies still remains obscure. It is hard to say that such an association can occur by chance, but it can be suggested that these anomalies represent simultaneous developmental events.

Case Report

A one day-old female infant whose grandmother was known to have the diagnosis of partial situs inversus presented with bilious vomiting. Physical examination revealed mild jaundice and epigastric fullness. The abdominal radiograms showed a few dilated loops of bowel with air fluid levels with no other gas in the rest of the abdomen and a right-sided fundic gas. The heart was placed on the left side on chest radiograms (Fig. 1). A barium enema showed a microcolon. Partial situs inversus associated with jejunal atresia was clinically suspected. Following the preoperative resuscitation a laparotomy through a transverse incision was performed.

The stomach and the spleen were found to be located in the right hypocondrium, the liver and the duodenum were in the left. Proximal jejunum was found to be markedly dilated to a point 15 cm distal to the ligament of Treitz where the bowel became a fibrous cord with an avascular fibrous mesentery (Fig. 2,3). The entire midgut, distal jejunum, cecum, appendix and ascending colon were found to be atretic. The extenal appearance of the spleen was normal.

Tapering jejunoplasty and jejunocolic anastomosis with resection of atretic segments were performed. Postoperatively, the patient had few stools but oral feedings could not be instituted because of the functional obstruction at the anastomosis. A contrast study of gastrointestinal tract performed on the 12th postoperative day showed delayed passage through

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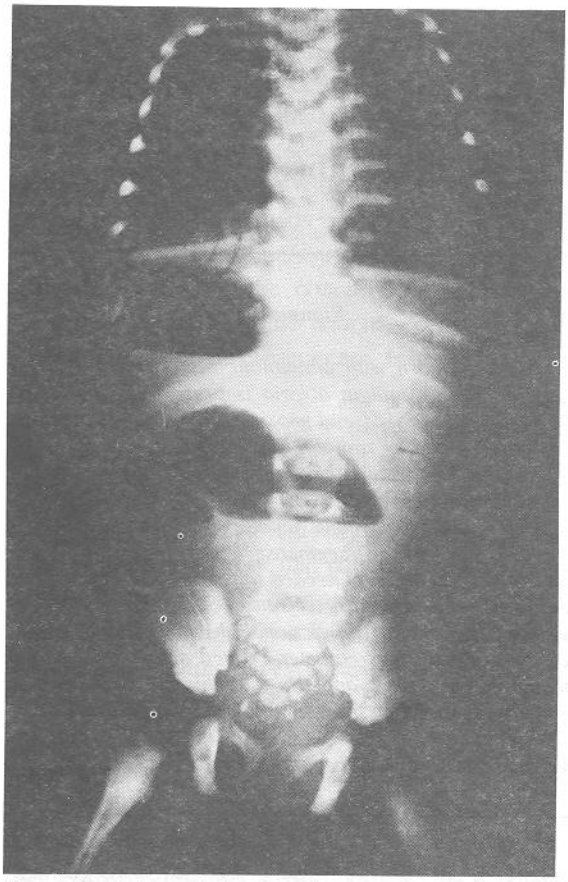

Figure 1. Plain upright abdominal and chest X-ray confirming proximal intestinal obstruction and partial situs inversus.

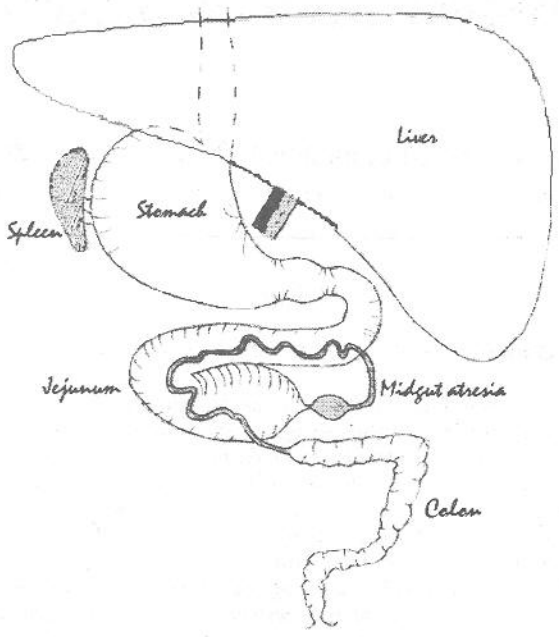

Figure 2. Line diagram showing midgut atresia and abdominal situs inversus.

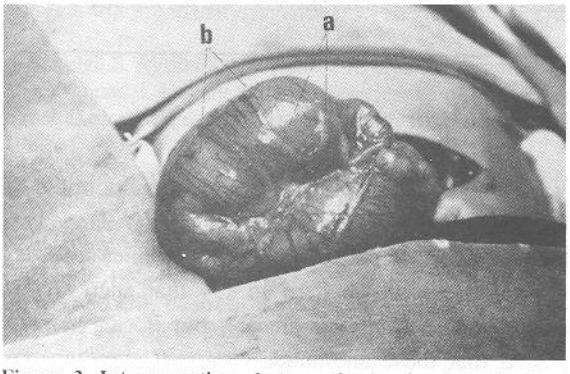

Figure 3. Intraoperative photograph showing intestinal atresia (a) and proximal dilated jejunum (b).

the anastomosis. The patient received total parenteral nutrition via a central catheter during the postoperative period and died of catheter related sepsis at the 18th postoperative day. Her parents refused permission for autopsy.

Discussion

Situs inversus viscerum is a condition in which the transposition of the abdomino-thoracic viscera occurs in a manner to present a mirror image of the normal. Transposition of the viscera may be complete or partial (situs inversus totalis or partials). In the latter, transposition may be confined only to the thoracic or to the abdomial viscera [5].

The transposition of the viscera, whether total or partial, is commonly associated with various anomalies of cardiovascular, urinary or alimentary systems. Among the alimentary tract anomalies, annular pancreas, duodenal atresia, duodenal web, jejunoileal atresia and malrotation have been most frequently reported. Situs inversus is also one of the major components of various syndromes associated with splenic anomalies [2,4,8].

Although it was recognized hundreds of years ago, the embryologic cause of situs inversus still remains an enigma. Cockayne in 1938 proposed that the complete transposition of the viscera was genetically determined, involving an autosomal recessive gene [6]. The rare reports of siblings with situs inversus supported this hypothesis but there have been many other workers who concluded that there was in-

sufficient evidence to confirm such a conclusion. It is also suggested that altered mechanical influences rather than genetic factors on the developing embryo results in visceral transposition [3]. In general, it is accepted (speculated) that the change which leads to the transposition of the viscera occurs as an early event in utero life, whether its cause is genetic or mechanical [4,8].

The embryogenesis of different types of intestinal atresia is better understood. For many years the most convincing explanation was a lack of revascuolization of the solid-cord stage of intestinal development. However, the clinical observations that bile pigments, squames, and lanugo hairs often found distal to atretic segments and postmortem observations of vascular abnormalities suggested that other etiologic factors were involved in the pathogenesis of intestinal atresia.

Spriggs in 1912 first suggested that the mechanical accidents, including vascular occlusions, might have a role in the development of intestinal atresia. Since that time, the so-called vascular theory, which postulated that failure of the blood supply to the gut may cause intestinal atresia, has been confirmed by many clinical and experimental observations [7].

According to the vascular theory, if the superior mesenteric artery (SMA) is occluded proximal to the middle colic artery (or just distal to the level where it leaves aorta), the whole midgut derivates supplied by the SMA and associated mesentery will then undergo ischemic necrosis. This kind of intestinal atresia and fibrous avascular mesentery were seen in this case.

The exact relationship between intestinal ab normalities and situs inversus remains obscure. The gut fixation and rotation anomalies may not be dif ficult to embryologically understand in abdomina SI since the association of these anomalies can b explained on the basis of multiple organ malrotatio involving the intestine and other abdominal viscer together. However, the association of various kind of intestinal atresia are less easy to understand, but i is hard to say that the association of these relativel rare anomalies may be due to chance. It seems likel that they represent simultaneous developmenta events. One can speculate that a teratogenic insul may be a common cause which contributes both ab dominal situs inversus and intestinal atresia or situ inversus is the initial anomaly which leads to th other malformations such as midgut atresia.

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