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Proximal Jejunal Atresia Consist Of Two Membranous Septa

İki Membranöz Septa ile Birlikte Proksimal Jejunal Atrezi

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ABSTRACT: Intestinal atresia may show many operative findings whereas they have almost same clinical presentation. Careful intestinal exploration and proper surgical intervention effect postoperative mortality and morbidity. Here we presented prenatally diagnosed proximal jejunal atresia that was composed of two membranous septa.

Keywords: intestinal atresia, newborn

ÖZET: İntestinal obstrüksiyonların benzer klinik bulguları olmakla birlikte ameliyat sırasında çok değişik şekillerde ortaya çıkabilirler. Dikkatli bir intestinal eksplorasyon ve uygun cerrahi girişimler postoperatif mortalite ve morbidite üzerinde etkilidir. Biz burada prenatal olarak tanısı konulmuş, iki adet membranöz septa ile birliktelik gösteren bir proksimal jejunal atrezi olgusu sunduk.

Anahtar Kelimeler: İntestinal atrezi, yenidoğan

INTRODUCTION

Intestinal atresia accounts for one third of all cases of neonatal intestinal obstruction and the incidence has been reported as approximately one in 4000 to 5000 live births in most series (1-3). Atresia occurs mostly on jejunoileal segment, whereas colonic location is seen rarely. On operative exploration many types of intestinal anomalies can be detected but few theories have been focused on embryologic etiopathogenesis. Failure of recanalization and vascular accident or any other intestinal injuries are main approaches that explain formation of atresia. Recently, in a mouse model, studies have shown that some forms of atresia were hereditary and resulted from dysregulation of proliferation and apoptosis of the developing intestine through the fibroblast growth factor pathways (4). Epithelial plugging was considered mostly related to duodenal atresia but as in our case, rarely cause septal obstructions in the jejunoileum and if distal intestine is not checked, multiple septa will be challenging problem that deteriorates morbidity.

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CASE REPORT

Prenatal ultrasound screening of a pregnant woman presented with polyhydramnios revealed dilated gastric and proximal intestinal segments in the fetus. This finding was suggestive of gastrointestinal tract obstruction and elective caesarean section was performed at term of gestation. After delivery, the female newborn was admitted to neonatal intensive care unit with abdominal distention. Gastric decompression was provided by 8 F nasogastric tube and bilious content was drained. Physical and radiological examinations of the patient have proven proximal intestinal obstruction and no additional anomaly was detected. Laparotomy was performed and intestinal exploration has shown an obstruction on proximal jejunum with dilated proximal and unused narrow distal segment. Intact intestinal continuity without mesenteric defect has indicated an intraluminal obstruction that is related to type 1 jejunal atresia (Figure 1). The bowel was opened at the antimesenteric site and a membranous septum was seen and while checking distal lumen opening by feeding tube, we found out another septum that was placed at 5 cm distally (Figure 2).

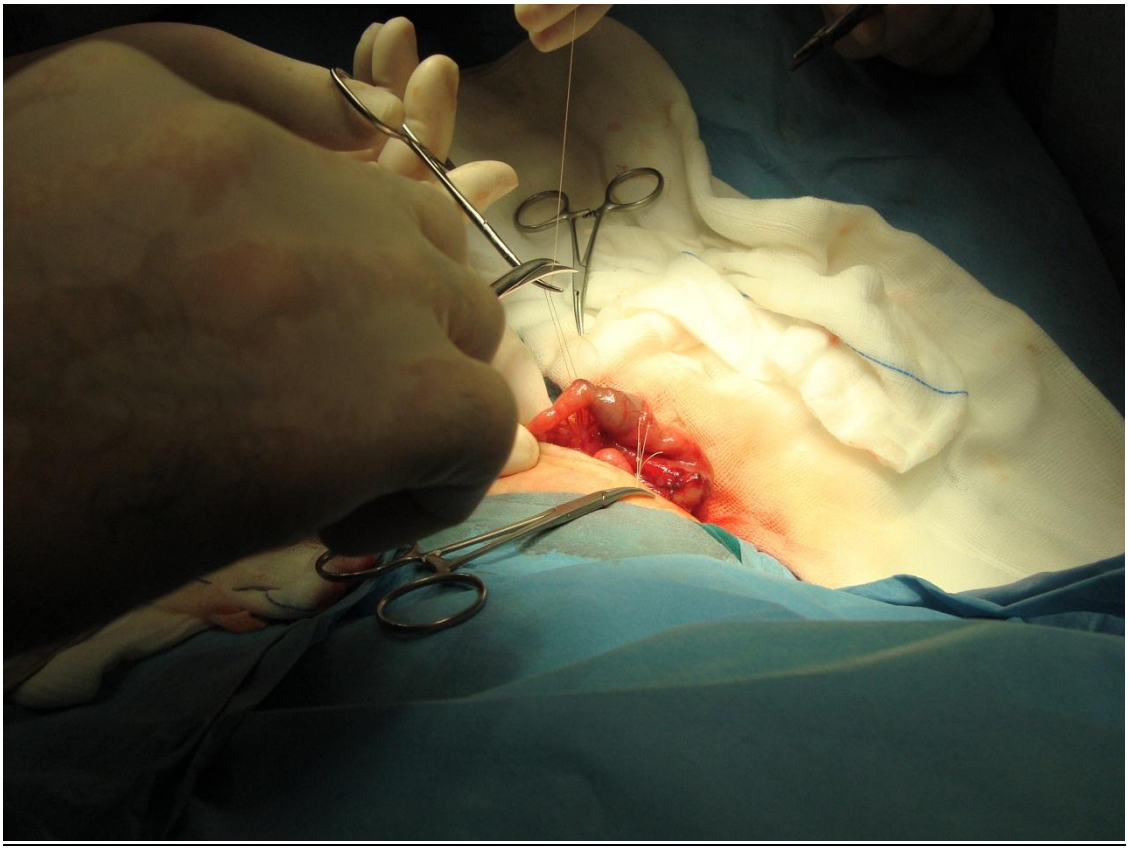

Figure 1. Gross appearance of atretic intestinal segment: intact intestinal continuity and proximal dilatation.

Between two septa minimal meconium was trapped. Atretic intestinal segment was resected and end-to-end oblique anastomosis was carried out to provide adaptation of large proximal lumen to unused narrow distal lumen. Postoperative course was uneventful and the patient was discharged on postoperative 8th day.

DISCUSSION

Abdominal distention and bilious vomiting are cardinal clinical findings of neonatal intestinal obstruction that may necessitate urgent operative intervention. However antenatal diagnosis may be possible by meticulous ultrasound screening of fetus. In this way, proper and rapid management of newborn can be provided and delayed diagnosis and related complications are also prevented. The accuracy rate of detecting small bowel atresia was reported between 23 to 31% by Basu et al (12). Dilated stomach and intestinal segments are reported as most common prenatal diagnostic findings. Here, ultrasound findings of patient were suspicious for intestinal obstruction and rapid postnatal investigation proven jejunum atresia and then early surgical intervention was performed.

Operative findings of intestinal atresia vary in a large scale and multiple theories have been studied experimentally to give an explanation to embryologic pathogenesis of these anomalies (5, 6). Tandler's theory included the lack of recanalization of solid cord stage of intestine. As proliferation and recanalization occurs prior to the 11th week of intrauterine life, in the duodenal atresia, associated congenital malformations have a higher prevalence (2, 3). Mostly, jejunoileal atresia was separated with a cordlike segment or a V-shaped mesenteric gap defect.

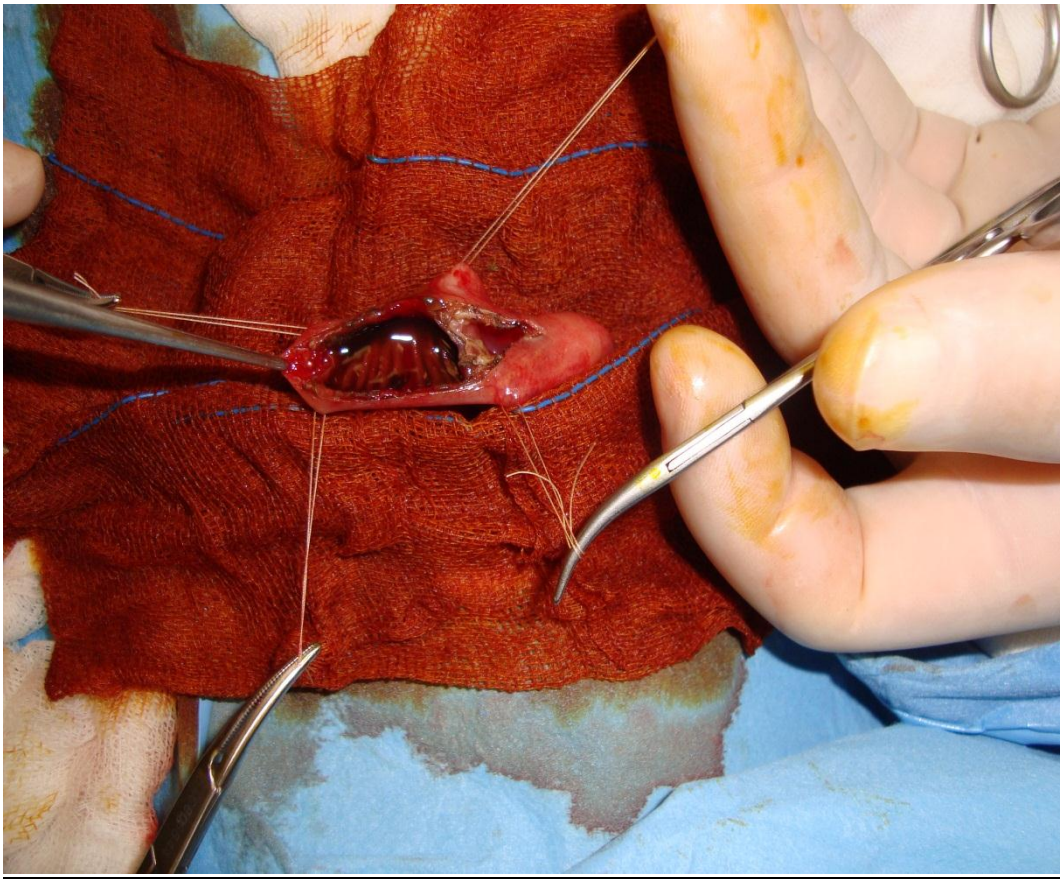

Figure 2. Lumen was opened and septa were detected.

Louw and Barnard demonstrated the role of late intrauterine mesenteric vascular accidents as the likely cause of jejunoileal atresia (7). Intrauterine volvulus and intussusceptions were commonly observed in single mid- and low jejunoileal atresia resulting in gap and cord type defects. Volvulus may not only cause jejunoileal atresia but also result from anatomic changes after the development of jejunoileal atresia in some cases. Intestinal atresia can also be coexisted with gastroschisis and meconium ileus (8, 9). The *Fgf10-Fgfr2b* signaling pathway is known to regulate mesenchyme-epithelial interactions, and its loss of function results in gastrointestinal tract atresia without vascular malformation. Fairbanks et al suggested that an accident in fibroblast growth factor pathway disturb proliferation and apoptosis of developing intestine and account for hereditary type of the intestinal atresia (4).

Epithelial plugging is uncommon in pathogenesis of jejunoileal atresia, but type 1 intestinal atresia that refers septal obstructions without mesenteric gap or defect, may explain these anomalies. These types of atresia are consisted 19% of all cases of jejunoileal atresia (10, 11). In our case, careful prenatal screening provided antenatal diagnosis of atesia and proper postnatal management of newborn patient. Laparotomy and exploration showed intact intestinal continuity and no mesenteric gap. After opening first narrowed antimesenteric site a septum was detected and another one was found out while checking distal lumen. We want to emphasize confirming whole intestinal luminal continuity. Trapped meconium between septa may show initial opening of proximal septum.

Jejunal atresia composed of septa is a rare type of atresia and we conclude that etiopathogenesis can be elucidated by recanalization theory or a disturbed proliferation and apoptosis through fibroblast growth factor pathway. Checking distal intestine is also important because multiple septa can be observed.

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