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Unusual Case Of Jejunal Atresia: A Mixture Of Type I And Type IIIa

Alışılmadık Jejunal Atrezi Tipi: Tip 1 Ve Tip 3a Karışımı

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ÖZET

Intestinal atreziler yenidoğan barsak tıkanukluklarının en sık sebebidir. Bu atrezilerin %46 sını Jejunoileal tıkanıklıklar oluşturur. Jejunoileal atreziler 4 tipe ayrılır. Olgumuz bu tiplerden hiçbirine uymamaktadır. Tip 1 ve Tip3a karışımı olarak tespit edilmiştir. Tespit ettiğimiz bu atrezide mukosal web yırtılarak açılmıştır. Intestinal atrezilerde cerrahi tedavi, erken ve doğru teşhis mortaliteyi, komplikasyonları azaltmaktadır.

Sonuç olarak safralı kusma ile başvuran yenidoğanlarda intestinal atrezi hatırlanmalıdır. Bu atrezi normal tiplendirmeye uymayabilir.

Anahtar Kelimeler: Barsak atrezisi, doğumsal anomaliler, jejunoileal atrezi

ABSTRACT

Intestinal atresias are a most common cause of bowel obstruction in the newborn. Jejunoileal obstruction was found in 46% of the intestinal atresia. The classification of jejunoileal atresias defined 4 types of lesions. Our case does not conform these types. A mixture of type I and type IIIa. This atresia included fenestrated mucosal web with minimal mesenteric defect. Being a life threatening surgical emergency, prompt diagnosis and opportune treatment are required to prevent mortality and other complications in intestinal atresias.

In conclusion, intestinal atresias should be remembered in bilious vomiting of newborn. And intestinal atresia type may be unusual.

Key Words: Intestinal atresia; Congenital anomalies; Jejunoileal atresia.

Introduction

Intestinal atresia is a well recognized cause of bowel obstruction in the newborn. The management of neonates with intestinal atresia has improved in recent decades due to refinements in neonatal intensive care, operative technique, use of total parenteral nutrition (TPN), and neonatal anesthesia [1]. Jejunoileal obstruction was found in 46% of the intestinal atresia [1]. Being a life threatening surgical emergency, prompt diagnosis and opportune treatment are required to prevent mortality and other complications.

We presented an unusual case of jejunal atresia.

Case

We describe a case of 90 day old triplet, premature boy that was treated for necrotizing enterocolitis. A day after being discharged he presented with yellow-green vomiting to our hospital. Physical examination findings showed right inguinal hernia, weight was 1600 gr. and a plain abdominal radiograph showed double-bubble sign but an upper gastrointestinal contrast study showed filling in distal intestinal system (Figure 1).



Figure 1.

Upon persistent bilious vomiting and air-fluid levels in plain abdominal radiograph exploration was done by supraumbilical right transverse incision. Ladd bands, malrotation, Meckel diverticulum and jejunal atresia with 5 mm defect in the mesentery was found approximately 15 cm distal to Treitz ligament (Figure 2). Caecum was right lateral to duodenum. Ladd bands were excised. We have

not been able to catheterize the stenotic lumen with 6 Fr tube. Resection and end to end anastomosis and inguinal hernia repair was done. Primary repair was done to iatrogenic jejunal damage. Postoperative recovery was uneventful.



Figure 2.

Hystopathological finding: Fenestrated web of jejunal segment in specimen (Figure 3).

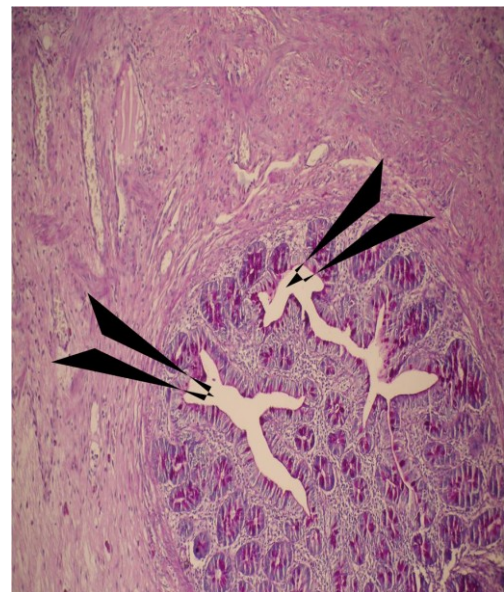


Figure 3.

Discussion

Intestinal atresia is a common cause of neonatal intestinal obstruction. Vascular accidents are thought to predispose to a majority of these lesions [2]. The two major theories regarding the pathogenetic mechanisms of intestinal atresia are Tandler's [1] concept of a lack of revacuolization of the solid cord stage of intestinal development and the classic study by Louw and Barnard [2] suggesting that a late intrauterine mesenteric vascular accident is the cause of most jejunoileal and colonic atresias. That can cause this vascular disruption of the blood supply include abnormalities in either the

formation or normal resorption of fetal blood vessels during development, compression or twisting of the mesenteric vessels as a result of a volvulus, or the release of emboli through placental vascular connections from a deceased monozygotic twin [3, 4]. While lack of revacuolization is the probable cause for most cases of duodenal atresia, compelling observations from other studies [5, 6] demonstrate that jejunoileal atresias occur as a result of intestinal volvulus, intussusception, internal hernia, or strangulation in a tight gastroschisis or omphalocele defect.

The classification of jejunoileal atresias varies somewhat based on the location of the obstruction. Duodenal lesions historically have been classified by Gray and Skandalakis, 14 with identification of 3 different types of lesions [7]. A type I defect represents a mucosal web with normal muscular wall (most common); type II, a short fibrous cord connecting the 2 atretic ends of the duodenum; and type III (least common), one in which there is complete separation of the atretic ends. The classification of jejunoileal atresias initially defined by Louw also recognized 3 types of lesions [8]. The classifications were later refined by Martin et al. and by Grosfeld et al. to include the apple peel deformity and multiple atresias [9, 10]. According to this method of classification, type I defect represents only mucosal or mucosa and submucosal web with an intact mesentery. Type II defects consist of a fibrous cord connecting the atretic bowel ends. A type IIIa lesion denotes an atretic segment with a V-shaped mesenteric gap defect, while type IIIb defines the apple peel deformity, in which there is a proximal jejunal atresia and the distal bowel is supplied by a single retrograde blood vessel. Type IV describes instances of multiple atresias ("string of sausage" effect). Type V or stenosis is defined as localized narrowing of the intestinal lumen without disruption of continuity or defect in the mesentery. Stenosis may also take the form of a type I atresia with a fenestrated web.

Our case does not conform these types. A mixture of type I and type IIIa. This atresia included fenestrated mucosal web with minimal mesenteric defect.

Babies with atresia or stenosis usually develop bilious vomiting on the 1 st day of life, but in 20% of newborn it may be delayed for 2 to 3 days. Abdominal distension is more pronounced with distal small bowel obstruction. 60-70% of these babies fail to pass meconium on the 1 st day of life [11]. Intestinal stenosis or

fenestrated web of type I atresia more likely to create diagnostic difficulty. Intermittent partial obstruction or malabsorption may subside without treatment. These babies usually fail to thrive and ultimately develop complete intestinal obstruction, which requires exploration [11]. This situation conforms to our case.

The treatment and management of neonates with bowel atresia has greatly improved. When the circumstances allow for prenatal diagnosis, delivery at or early transfer to a specialized center, and prompt surgical treatment, infants without critical associated anomalies have an excellent prognosis. Ongoing efforts to minimize delivery of low birth weight babies and optimize treatment of associated congenital anomalies through prenatal detection will hopefully contribute to further improvement in survival [12].

In conclusion, intestinal atresias should be remembered in bilious vomiting of newborn. And intestinal atresia type may be unusual.

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