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Segmental dilatation of the ileum and intestinal malrotation*

Hüseyin ÖZBEY, Tansu SALMAN, Cem BONEVAL, Selim AKSÖYEK, Alaaddin ÇELİK
Department of Pediatric Surgery, University of Istanbul Medical Faculty, Istanbul, Turkey

Özet

İleumun segmental dilatasyonu ve intestinal malrotasyon

Segmental intestinal dilatasyon (SDI), nadir bir fonksiyonel intestinal obstrüksiyon nedenidir. Burada, aralıklı karın ağrısı, safrasız kusma ve gelişme geriliği şikayetleri ile kliniğimize başvuran, eksplorasyonda segmental ileum dilatasyonu ve intestinal malrotasyon saptanan 4 yaşındaki bir kız çocuğu sunulmuştur. Dilate ileum segmenti rezeke edilerek uç-uca anastomoz uygulanmıştır. Segmental intestinal dilatasyon ve malrotasyon birlikteliğinin barsağın "lokal gelişme aktivitesi"ndeki düzensizliği yansıtan bir bulgu olabileceği düşünülmüştür.

Anahtar kelimeler: *İleum, malrotasyon, intestinal obstrüksiyon*

Summary

Segmental dilatation of the intestine (SDI) is an uncommon cause of functional intestinal obstruction. Segmental dilatation of the ileum with malrotation was found in a 4-year-old girl presented with recurrent episodes of abdominal pain, nonbilious vomiting and severe growth retardation. She was treated successfully with resection of the dilated segment and end-to-end anastomosis. It is proposed that the association of SDI with malrotation may be a presentation of the failure in "localized growth activity" of the bowel.

Key words: *Ileum, malrotation, intestinal obstruction*

Introduction

Among the causes of intestinal obstruction observed in childhood, congenital segmental dilatation of the intestine is unusual and remains as a diagnostic dilemma [4,8]. The affected segment may be at jejunum, ileum or colon [3,6,8,11,16,17]. Segmental dilatation of the ileum has also been referred to as "ileal dysgenesis" or "giant Meckel's diverticulum" [2,5,13]. With the reported case, the relevant literature is reviewed to highlight the pathogenesis of this malformation.

Case Report

A, 4-year old girl presented with 3 days history of crampy abdominal pain associated with nonbilious vomiting. According to her parents these episodes were occuring repeatedly and resolving spontaneously every 3 or 4 weeks, since birth.

On physical examination her height and weight were found at the 3rd percentile. The abdomen was soft and no distention was observed. Hypoactive bowel sounds were detected on lower quadrants but no obstructive sign was present on erect radiographs. Laboratory values were within normal limits except leucocytosis. Nonbilious aspirate was observed through the nasogastric tube.

In upper gastrointestinal contrast studies, the gastroduodenal and jejunal passage was normal but a gas filled intestinal segment (which was later interpreted as "stomach like shadow") was observed behind the contrast filled stomach (Figure 1). A barium enema revealed an upper left quadrant localized cecum. On the second day of her admission, persistence of the abdominal pain and observation of bilestained gastric aspirate leod to exploration. The preoperative diagnosis was incomplete gastric

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Adress: Dr. Hüseyin Özbey, İ.Ü. İstanbul Tıp Fakültesi Çocuk Cerrahisi Anabilim Dalı, 34390 Çapa-İstanbul

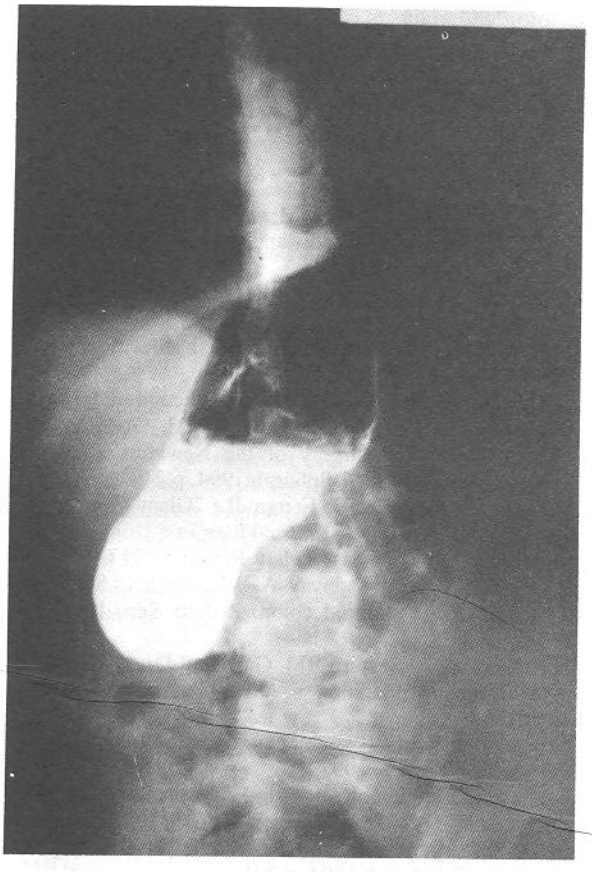

Figure 1. Contrast filled stomach and a "stomach like shadow".

volvulus and/or malrotation with mild obstructive bands. A laparotomy was performed via a supraumbilical median incision.

At laparotomy, an isolated (25 cm) dilatation of the terminal ileum was found just behind the stomach. Malrotation was present with an upper left quadrant localized cecum (Figure 2). No obstructive bands were observed in association to malrotation. Resection of the dilated segment, end-to-end anastomosis and appendicectomy were performed.

The cecum was left in upper left quadrant. The postoperative course was uneventful and she was discharged on the eight postoperative day. Histologically, focal increases in the thickness of submucosa with lymphoid hyperplasia and cystic changes, indicating chronic inflammation was observed. The muscle layer was normal along with normal ganglion cells.

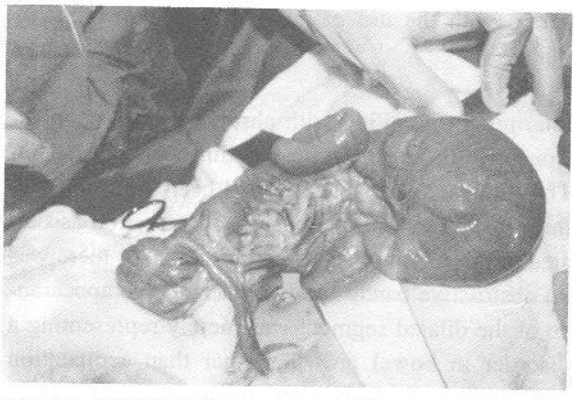

Figure 2. Intraoperative appearance of segmental dilatation of the terminal ileum.

Discussion

Segmental dilatation of the intestine (SDI) is a rare malformation which was first described by Swenson and Rathauser in 1959 [14]. The etiology of SDI is obscure. Primary dsplasia of the intestinal segment, vascular insufficiency during intussusceptions (hydrostatically reduced) resulting in hypoplasia of the intestinal muscle, volvulus or strangulation of the intestine in the umblical ring during the early stage of development were some of the etiologic theories [1, 8,17].

Heterotopic tissues have been reported in various cases [8,15]. Generally, there is no evidence of deficient innervation but functionally it may act as a stagnant loop, usually with a very thin or absent muscular layer. Segmental dilatation of the colon was reported in association with anorectal malformations, when it has to be differentiated from rectal ectasia and the pouch colon [4].

The age at onset of the symptoms may be from shortly after birth to 1-year of age and the clinical presentation may extend up to adolescence [7,12,14]. Although SDI may be the cause of acute intestinal obstruction and/or perforation in the neonatal period, frequent episodes of abdominal pain, vomiting and chronic growth suppression may be the clinical findings in older patients [12,13].

The radiographic diagnosis of SDI is difficult. The "stomach like shadow" is reported to be a diagnostic radiological finding, as found in our case [4]. Preoperative demonstration of the anatomy has been re-

ported with the use of duodenal infusion with barium [13].

Intermittent vomiting with chronic growth suppression were the main clinical findings of our patient. The "stomach like shadow" had been interpreted as intestinal dilatation secondary to obstruction associated to malrotation. As shown in Figure 1, there was no obstructive bands and the macroscopic appearance of the dilated segment was merely representing a disorder in bowel growth, rather than a dilatation proximal to an obstruction. Hence, the entity localized to the terminal ileum should be recognized as an intrinsic ileal dysgenesis. Its definitive treatment consist of resection of the dilated segment with end-to-end anastomosis, as in other localizations.

In contrast to the "general force", "localized forces" are though to be responsible for the movement of the various parts (duodenal and umblical loop) of the midgut. The growth activity of the umbilical loop leads to the development of several loops with passive movement and push of the cecum up to a cranial position. It is proposed that localized growth activity may be responsible from the rotation anomalies rather than the rotation of the whole gut [9,10].

The most common site for SDI is the terminal ileum, which may be due to embryological hyperactivity of this part of the gut. The common association of SDI with rotation anomalies may be a presentation of the developmental behaviour of the bowel. In our opinion, continuing localized growth activity with pause in lengthening may result with segmental dilatation of the terminal ileum and unusual localization of the cecum. Similarly, the neonates with congenital short bowel (constantly associated with rotation anomalies with thickened bowel wall and/or dilated bowel) should be another clinical presentation of the failure of the localized growth activity of the bowel. Further studies including the immunecytological analyses of the affected part of the intestine, in order to reveal the disturbances in its growth pattern is needed.

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