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Colonic perforation due to congenital segmental dilatation: A case report

Haluk SARIHAN, Musa ABEŞ, Kadriye YILDIZ

Karadeniz Technical University Faculty of Medicine, Departments of Pediatric Surgery and Pathology, Trabzon, Turkey

Summary

Congenital segmental dilatation of the colon is an uncommon surgical pathology that deserves attention particularly with respect to the differential diagnosis of Hirschsprung's disease. We report a male infant with congenital segmental dilatation involving the transverse colon. In addition the patient had microcephaly, renal agenesis and inguinal hernia on the right side.

Key words: *Segmental colonic dilatation, microcephaly, renal agenesia*

Introduction

Segmental dilatation of intestine is a rare condition and was first described by Swenson and Rathauser in 1959 [1,4,6]. Recently, 32 cases have been reviewed collectively [1]. Segmental dilatation may involve any part of gastrointestinal tract; most common site is ileum [1,5]. Exact cause of colonic segmental dilatation (SCD) is unknown. Innervation of the colon is normal but constipation and obstruction occur from the lack of peristaltisis [1,2,5,6].

Clinical course and radiological appearence of patients with SCD may resemble Hirschsprung's disease [1,4,6]. It may be associated with various congenital anomalies [5]. We report a case of congenital segmental dilatation of the transverse colon associated with microcephaly, right renal agenesis and right inguinal hernia. This association has not been reported previously.

Address: Dr. Haluk Sarıhan, KTÜ Tıp Fakültesi Farabi Hastanesi, Çocuk Cerrahisi Anabilim Dalı, 61080, Trabzon

Case report

A 5 month-old boy was admitted to our clinic for abdominal distension and bilious vomiting. The patient had a history of constipation since birth. The pregnancy was unremarkable. Peritonitis was evident; there was no palpable mass. The patient had developed gastrointestinal obstruction prior to the present findings. Microcephaly and right inguinal hernia were also observed. Plain abdominal x-ray demonstrated a large dilated transverse colon with air/fluid levels (Fig. 1). The patient was resuscitated and operated as an emergency case. At laparatomy the transverse colon was found to be dilated from the middle to the hepatic flexura.

The dilatation appeared to be at least four times larger than normal diameter. Other parts of colon were normal. There was a perforation in the center of the affected colon which penetrated into the liver. The dilated part of colon was resected. Full thickness biopsy of descending colon for Hirschsprung's disease was obtained and a covering colostomy raised. Postoperative course was uneventful. Intravenus pyelography demonstrated right renal agenesis. Colostomy was closed after 2 months. Histopathological examination of specimens showed normal mucosal and submucosal layers but hypoplastic and/or absent muscular layers in the dilated segment (Fig. 2). A normal arrangement of ganglion cells and nerve bundles were present in the dilated and distal segments of the colon.

Discussion

SCD is a rare surgical condition which is often dealt with as an emergency [1]. The sigmoid colon is the most commonly involved colonic segment [6]. Pa-

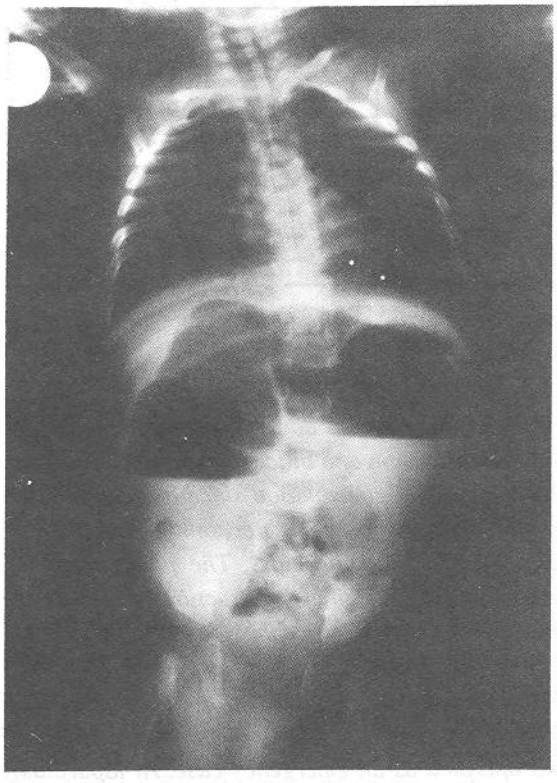

Figure 1. Plain abdominal x-ray with massive large dilatation of transvers colon.

tients ages have ranged from 1 day to 22 years [1,6]. The males are affected twice as more than females [4]. Symptomatology of this entity resembles Hirschsprung's disease and clinically it is difficult to differentiate these disease [4]. The clinical presentations are variable. The neonate may present with low intestinal obstruction. The common complaint in the elder patients is persistent constipation [6]. Physical examination may reveal a smooth non-tender mass. Plain abdominal x-ray findings include large, sometimes massively dilated segment of intestine and air/fluid levels [6]. Barium enema shows a normal appearence at the unaffected colon and dilatation at affected segment [4,6]. The diagnosis of SCD is extremely difficult and so far it has rarely been identified before operation.

According to histopathological findings there are two groups of SCD [1]: Hypertrophied muscular layers within the dilated segment are generally encountered in elder patients [4]. In younger patients a very thin or nonmuscular layer have been described [3]. Submucosa and mucosa are almost always normal both the dilated segment and the rest of the gastrointestinal tract carry normal ganglion cells [3]. Tenia coli are generally absent in the dilated segment but serosal vessels may be prominent [1]. In our case the mucosa and submucosa layers of affected colon segment were normal but the muscular layer was hypoplastic or completely absent.

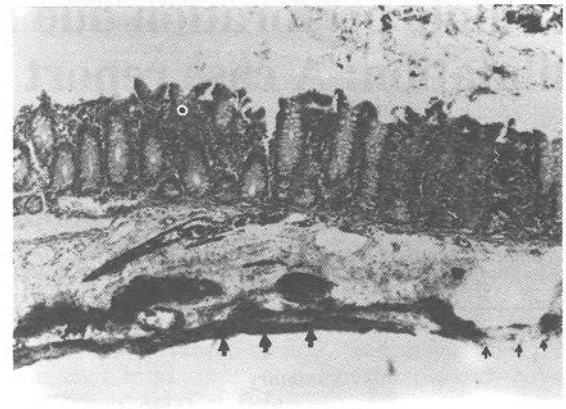

Figure 2. Histopathological examination demonstrating areas of hypoplastic muscular layer (large arrows) and absent of muscular layer (small arrows) with normal mucosal and submucosal layers (HEX30).

The pathogenesis of absent intestinal musculature is obscure. It is hypothesized to be congenital by most the authors: An embryological etiology of absent intestinal musculature is based on the fact that at 7 to 40 mm the embryo has multiple diverticula in the intestine which normally regress. After this process the sorrounding mesenchyme condenses to form the muscular coats of the bowel. Failure of this process would lead to segmental absence of the intestinal musculature [3]. This condition produces either segmental dilatation of intestine or leads to intestinal perforations. Our case presented with both symptoms due to segmental hypoplasia or aplasia.

Segmental dilatation is associated different congenital anomalies both intestinal and extraintestinal in nearly 50 % of cases. Malrotation, short bowel syndromes, anomalous portal vein, Meckel's diverticulum, pancreatic malformation, bladder exstrophy, meningoencephalocele with hydrocephalus, omphalocele, covered anus, split sacrum, dorsal hemivertebra with deformity of ribs, esophageal and duodenal atresia, pulmonary and genito urinary malformations have been reported as associated anomalies [1,5].

We have found microcephaly, inguinal hernia and renal agenesia in our case. The etiology of these combined anomalies is unknown. The mother had not receive any medication during pregnancy. A single fetal insult resulting hypoxia may have affected kidney, brain and intestine at the same time. It is known that critical development of these organs occur in the same embryological week.

Surgical treatment consists of resection of dilated segment and reconstruction of the tract [2,6]. Differential diagnosis may require frozen section pathological investigations intraoperatively. Colostomy may be advisable when frozen section is not available.

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