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CONGENITAL SEGMENTAL DILATATION OF THE COLON WITH HETEROTOPIC ESOPHAGEAL MUCOSA*

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Swenson and Rathauser[1] first reported segmental dilatation of the colon in three patients as a distinct entity. Following this description, six more cases appeared in the English literature[2-7]. Among these, heterotopic esophageal mucosa in the dilated segment was encountered on only one occasion[3].

In this report, a newborn infant with segmental dilatation of the colon, containing heterotopic esophageal mucosa, is described and the various aspects of this rare anomaly are discussed.

Case Report

A full-term, five-day-old newborn girl weighing 2500 grams was admitted to the Hacettepe Children's Hospital with a history of abdominal distention, bilious vomiting and failure to pass meconium after birth. The course of the pregnancy and the delivery had been normal.

On physical examination, the baby was found to have a very distended abdomen with no tenderness or muscular rigidity on palpation. No bowel sounds were present and no meconium was forthcoming on rectal examination. The other systems were normal.

Abdominal roentgenograms showed an enormously dilated intestinal segment with an air-fluid level (Fig. 1). A barium enema demonstrated a narrow rectum with dilatation of the remaining colon (Fig. 2). CBC, serum electrolytes and BUN were found to be normal.

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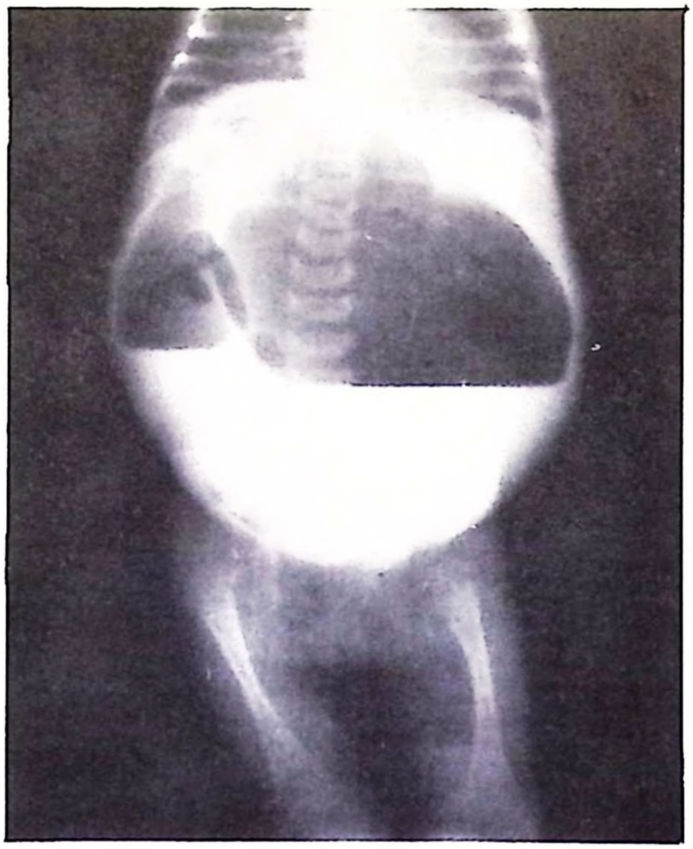

Fig. 1: Plain erect abdominal X-ray showing enormously dilated intestinal segment.

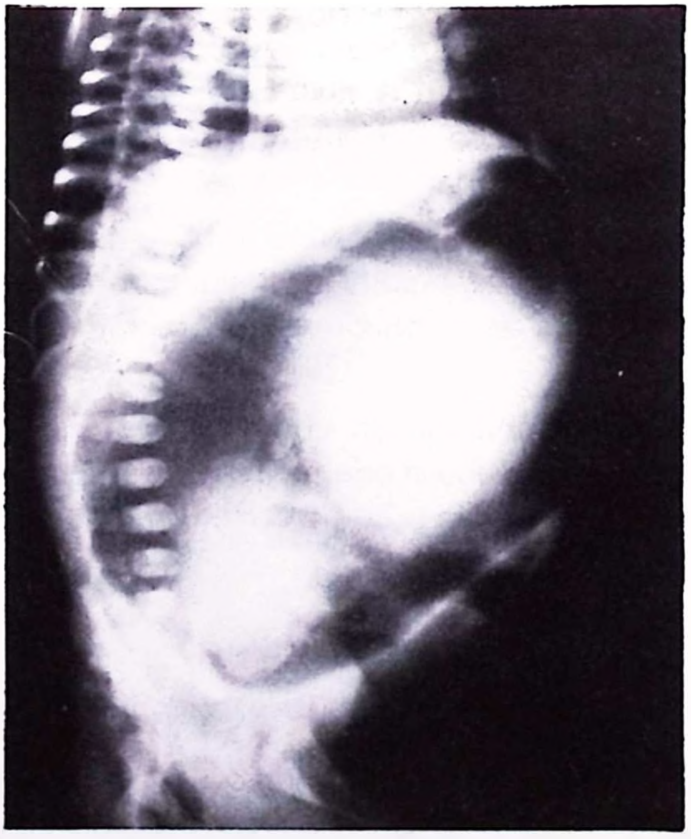

Fig. 2: Severely dilated colon and narrow rectum on barium enema.

Following the fourth fluid therapy and nasogastric decompression, a laparotomy was performed. There was a preoperative diagnosis of Hirschsprung's disease. On exploration, the colon was found to be dilated and thick-walled from the hepatic flexura to the rectosigmoid junction, in the absence of an extrinsic obstructive lesion. Teniae coli were not noted in this dilated segment. There was no pathological finding in the vascularization pattern. The caliber and thickness of the wall of the rectum and ascending colon were normal.

Frozen section examinations showed normal plexuses with ganglion cells in the dilated segment, the ascending colon and the rectum as well. The dilated segment was resected and proximal terminal colostomy and closure of the rectal stump was carried out. The postoperative course was uneventful except for a wound infection.

Histological examination showed normal ganglion cells and nerve bundles in submucous and myenteric plexus throughout the resected segment. The muscular layers of the resected segment were found to be normal. A well-formed esophageal mucosa was detected in some areas (Fig. 3).

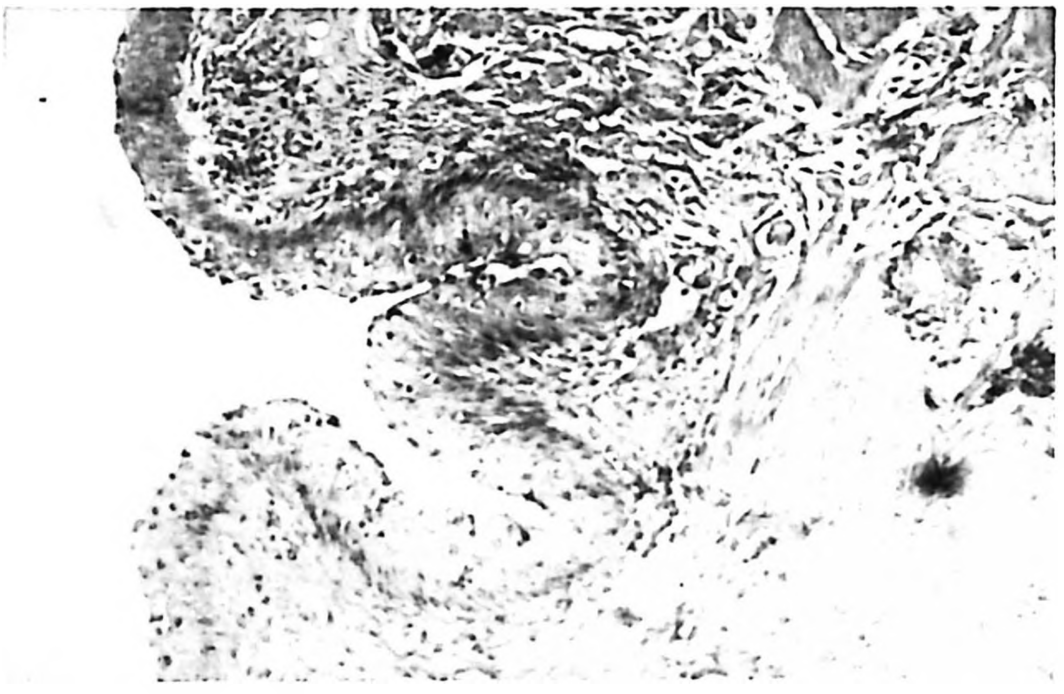

Fig. 3: Photomicrograph of the heterotopic esophageal mucosa (HE x 10).

An end-to-side anastomosis was performed between the ascending colon and the rectum when the patient was two years of age. A tube was inserted from the appendectomy stump as a cecostomy in order to prevent anastomosis. However, a low output caecal fistula occurred because of leakage from the tube cecostomy. This fistula was closed 10 months later. The patient is four-years-old now and thriving well with normal bowel movements.

Discussion

Segmental dilatation of the colon is a rare entity and nine such cases have so far been reported[1-7]. The characteristics of the previously reported cases are lack of radiologic motility of the affected segment, normally functioning colon proximal and distal to the dilated segment, absence of the teniae coli in the dilated part, presence of ganglion cells throughout the distended and undistended colon and hypertrophic or normal muscular layers in the dilated segment. Table I summarizes some other aspects of these cases..

The ages of the patients ranged from 1-day to 22 years. There were six boys and four girls. The localization and the extension of the dilated segment varied, and in some cases, a large part of the colon was involved as in the case present.

The cause of the colonic dilatation is unknown. In the reported cases, the ganglion cells and nerve bundles were found to be normal. Further there was no obstructive lesion. However, the clinical features were obviously related to the dilated segment, because resection of this segment provided normal bowel movements. Despite the normal ganglion cells, there may be a defect of the innervation system of the affected bowel which cannot be identified by current methods[2]. As a result of the faulty innervation, the normal peristaltic waves are absent and this may lead to progressive dilatation. The severity of the neurogenic defect results in varied symptomatology; there was intestinal obstruction in certain severe neonatal cases, while chronic constipation was recorded in older cases.

Although there was heterotopic mucosa in the dilated segment of only two out of the ten cases, including the one presented, this association may lead to some important questions in terms of etiology. Heterotopic mucosa in the gastrointestinal system is not an uncommon finding; gastric mucosa in Meckel's diverticulum, esophagus and duplications are well-known examples. However heterotopic esophageal mucosa in the intestinal tract has been reported only on three occasions; once in an appendix vermiformis[8], and twice in the segmental dilatation of the colon and ileum[3,9]. The association of very unusual heterotopic esophageal tissue with segmental dilatation of the colon indicates a congenital origin of the condition.

The symptomatology of the segmental dilatation of the colon resembles Hirschsprung's disease. The neonatal cases were admitted to the hospital with signs of low intestinal obstruction[3,5]. In the older patients, the common complaint was persistent constipation until infancy[6]. The typical roentgenologic finding is a large, sometimes very large, dilated and showing an air-fluid level segment on plain films[1]. Barium enema reveals a normal colon distal to the affected part[7]. Relaxation of the internal sphincter on rectal manometry was reported as a diagnostic finding on one occasion[6]. However, preoperatively, most of the

TABLE I: Summary of Ten Cases with Segmental Dilatation of the Colon

Case	Author	Age	Sex	Location	Heterotopic tissue	Associated anomalies
1	Swenson and Rathauser[1]	6 months	F	Proximal transverse colon	–	Mobile caecum.
2	"	$2^{1,2}$ years	F	Descending colon	–	Foreshortened colon with incomplete rotation.
3	"	22 years	M	Sigmoid colon	–	–
4	de Lorimier[2]	4 years	M	Caecum to sigmoid	–	–
5	Aterman and Abacı[3]	20 days	M	Caecum, ascending colon, transverse colon	Gastric and esophageal mucosa	Extrophy of the bladder, covered anus, split sacrum, dorsal hemivertebra with deformity of ribs.
6	Brawner and Shafer[4]	12 years	M	Splenic flexura to recto-sigmoid junction	–	–
7	Helikson et al[6]	1 day	M	Sigmoid colon	–	–
8	Etzioni et al[6]	5 years	F	Sigmoid colon	–	–
9	Nguyen and Shandling[7]	9 years	M	Sigmoid colon	–	–
10	Present case	5 days	F	Hepatic flexura to rectum	Esophageal mucosa	–

reported cases were diagnosed as Hirschsprung's disease and the differential diagnosis between Hirschsprung's disease and segmental dilatation of the colon has generally been made following pathological examination[1-7].

The treatment of the segmental dilatation of the colon is the surgical removal of the affected segment and all of the cases reported responded well to resection of the segment as did our patient.

Summary

A five-day-old newborn girl with congenital segmental dilatation of the colon containing heterotopic esophageal mucosa is described. The etiology, the clinico-pathological findings and treatment of the anomaly are briefly discussed.

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