

Congenital Segmental Dilatation of the Colon

by

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Though many malformations of the intestine have been described up to the present, congenital segmental dilation is very rare. It is comparatively difficult to diagnose this disease prior to operation or autopsy. We have, recently, experienced a case in whom an autopsy was performed at Tohoku University Hospital and a segmental dilatation of the colon was found. This is a report of a case with pathological and histological findings.

Case Report

An 9-day-old female was admitted to the Second Department of Surgery, Tohoku University Hospital 4 days after birth because of abdominal distension and vomiting. Her sister died of severe infection on the 10th day of life with severe jaundice caused by leptomeningitis. Her parents had incompatible blood groups (mother : O, Rh(+), father : AB,Rh(-)). She was born by Caesarean section because of hydra-mnion and her birth weight was 2,980g. A plain film of her abdomen showed distended intestines, but intestinal atresia or Hirschsprung's disease was excluded by fluoroscopic and manometric examination. She was referred to Perinatal Ward for administration of severe jaundice. However, her general state became gradually aggravated and she died of severe jaundice on the 9th day of life. Serum total bilirubin was 27.9mg/100ml on her last day of life.

Autopsy disclosed that she suffered from nuclear icterus with yellow-stained hippocamps and pale globe of brain. Segmental marked dilatation of the transverse colon was disclosed at the same time. This dilated part was existed about 5 cm anal from the right colic flexure. The diameter of this dilated part was a little over 4 cm, while the other part of the transverse colon was about 1 cm in diameter. On the cut surface, the mucous membrane seemed to be normal and there was no obstruction in the colon and rectum (Fig. 1). Photomicroscopic findings of the dilated part disclosed that normal muscle layer was present and ganglion cells were seen in the myenteric plexus (Fig. 2). In the colon and rectum anal from this dilated part,

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Fig. 1 Gross findings of the colon.



Fig. 2 Photomicroscopic findings of the dilated Part of the colon.

ganglion cells were also seen in the myenteric plexus and Hirschsprung's disease was ruled out. From these findings, the diagnosis of congenital segmental dilatation of the colon with kernicterus was made.

Discussion

Several papers on the segmental dilatation of the intestine have been published up to the present. Dilated portions of the intestine were variable: jejunum by Rossi⁴

and KOMI³, ileum by SAWADA¹ and UEDA⁷, and colon by SWENSON⁶, de LORIMIER² and BRAWNER¹. The symptoms of this disease also differ, depending upon each case. Abdominal pain, abdominal distension, vomiting and diarrhea were commonly seen in the reported cases. The time of onset of the symptoms varies case by case and only two cases including the present case have been reported in the newborn period up to the present.

In gross findings, the dilated portion of the intestine looks like the abdomen of snake that swallowed the bait. In general, the intestinal wall seems to be thickened which was confirmed in the microscopic findings.^{2,4,6} On the other hand, UEDA⁷ described that atrophic muscle layer was seen in his case, while the dilated intestinal wall was thick in gross findings. Dilatation and meandering of the vessels were described in the serosal site of this wall by de LORIMIER² and SWENSON⁶. As concerns the ganglion cells in the myenteric plexus, almost all authors reported that there was no abnormal findings in these plexus.

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和文抄録

先天性限局性結腸拡張症の1例

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先天性限局性腸拡張症は, まれな疾患で, これまで空腸, 回腸および結腸のいずれからも発生が報告されているが, 術前に診断することは困難で, 開腹時, あるいは剖検時に発見される事が多い. 本症例は, 生後2日目より黄疸, 腹部膨満および嘔吐を主訴とし, 東北大学第2外科に入院した患児で, 検査により

Hirschsprung 氏病を否定され, 黄疸治療中に死亡した女児例であるが, 剖検により, 結腸に著明な拡張を認め, 組織学的な所見により Hirschsprung 氏病, 腸狭窄症などを否定し, 先天性限局性結腸拡張症であると診断された.