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Case Report

Split Notochord Syndrome: Ileal Duplication Causing Intermittent Episodes of Vomiting

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Tsuchida, S., Takahashi, T., Noguchi, A., Takahashi, I., Mikami, T., Hebiguchi, T., Morii, M., Yoshino, H., Nanjo, H. and Takada, G. *Split Notochord Syndrome: Ileal Duplication Causing Intermittent Episodes of Vomiting*. Tohoku J. Exp. Med., 2006, **209** (4), 379-382 —— Split notochord syndrome is a group of developmental abnormalities caused by abnormal splitting or deviation of the notochord, clinically resulting in the duplicated bowel associated with vertebral anomalies. In this syndrome, initial presentations due to duplicated bowel, vomiting, abdominal pain, and failure to thrive, usually occur before 1 year of age. We here report a 12-year-old boy with intermittent vomiting, previously diagnosed with cyclic vomiting syndrome. On abdominal x-ray examination, a defect in the closure of posterior vertebral arches was observed in the 5th lumbar vertebral body, indicating the complication of spina bifida occulta. This finding suggested the diagnosis of split notochord syndrome. A magnetic resonance imaging study revealed a cystic mass lesion in the pelvic cavity. ^{99m}Tc-pertechnetate scintigraphy, which is frequently used to detect ectopic gastric mucosa for the diagnosis of Meckel's diverticulum, showed a positive spot corresponding to the cystic mass lesion. Surgical resection of the cystic mass lesion demonstrated ileal duplicaion with ectopic gastric mucosa. Surgical findings suggest that symptoms of the patient were due to ulceration, inflammation, or bleeding caused by acid-peptic juice secreted from ectopic gastric mucosa. Duplication of the alimentary tract should be considered as a possible cause in patients with symptoms suggesting cyclic vomiting syndrome. ———— split notochord syndrome; duplication of alimentary tract; cyclic vomiting syndrome

Duplication of the alimentary tract is a rare cause of vomiting and abdominal pain in childhood (Ildstad et al. 1988; Iyer and Mahour 1995). The duplicated bowel is rarely associated with vertebral anomalies, known as split notochord syndrome (Bentley and Smith 1960). This syndrome is a group of abnormalities caused by abnormal splitting or deviation of the notochord leading to a persistent connection between the gut and dorsal skin (Bentley and Smith 1960). In this syndrome, initial presentations usually occur before 1 year of age (Faris and Crowe 1975;

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Singh and Singh 1982; Gupta and Deodhar 1987). We here report a 12-year-old boy with intermittent vomiting, previously diagnosed with cyclic vomiting syndrome, caused by ileal duplication with ectopic gastric mucosa. Lumbar spina bifida occulta demonstrated on an abdominal x-ray examination suggested split notochord syndrome, resulting in the diagnosis of ileal duplication.

Case Report

The patient was a full term 3,860 g boy uneventfully born to healthy Japanese parents. There were no problems noted during infancy. From the age of 5 years, he started to present with intermittent episodes of vomiting and abdominal pain. Each episode lasted 2 days and occurred about every 3 months. At the age of 10 years, he was medicated with phenytoin as a treatment for cyclic vomiting syndrome. However, the treatment did not decrease the frequency of the episodes and the patient was finally referred to our hospital because of the intractable attacks at the age of 12 years. The attack consisted of abdominal pain and vomiting, leading to admission to hospitals for the treatment of complicated dehydration. The symptoms lasted 2 days and resolved without any specific treatment.

On admission during an attack, his height and weight were 153.1 cm (+0.53 s.d.) and 41.3 kg (–0.11 s.d.), respectively. Heart rate and blood pressure were 84/min and 110/58 mmHg, respectively. Physical examinations did not show any abnormal findings, including specific abdominal signs. There was nothing remarkable in the laboratory findings. Blood gas analysis showed the following: pH, 7.44; pCO_2, 40.1 mmHg, HCO_3, 26.7 mmol/L; base excess, 2.5 mmol/L. Organic acid analysis of urine did not demonstrate any specific findings. On abdominal x-ray examination, a defect in the closure of posterior vertebral arches was observed in the 5th lumbar vertebral body, showing the complication of spina bifida occulta (Fig. 1). The patient was not associated with other developmental disorders of the sinal cord, examined by a magnetic resonance imaging (MRI) study. MRI studies of the brain and abdomen were also performed. Although there was no abnormality in the brain or pituitary gland, a cystic mass lesion was found in the pelvic cavity (Fig. 2). ^{99m}Tc-pertechnetate scintigraphy showed a

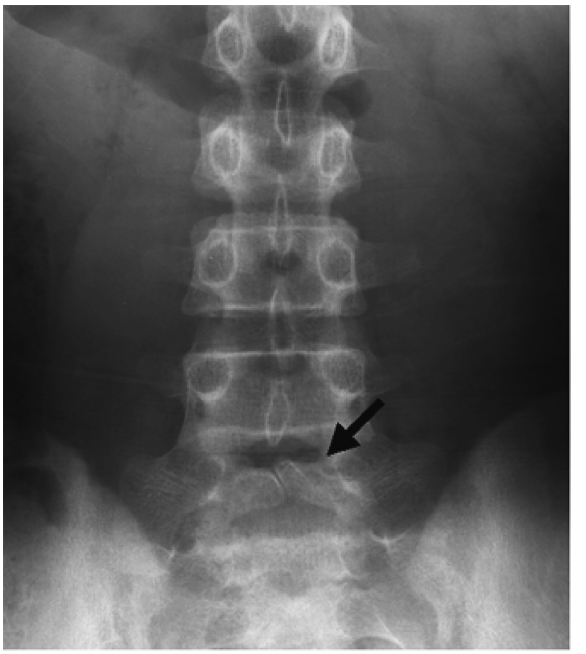

Fig. 1. Abdominal x-ray. A defect in the closure of posterior vertebral arches was observed in the 5th lumbar vertebral body (arrow).

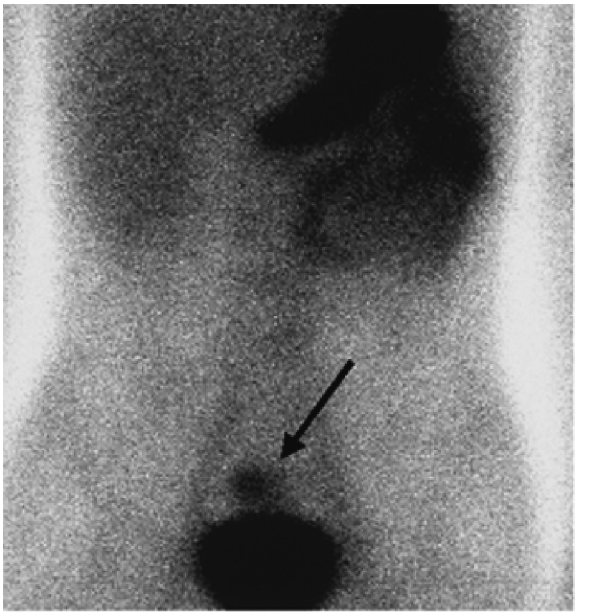

Fig. 2. Image on ^{99m}Tc-pertechnetate scintigraphy. The arrow shows the presence of ectopic gastric mucosa.

positive spot corresponding to the cystic mass lesion, suggesting the presence of ectopic gastric mucosa (Fig. 3). Surgical resection of the cystic lesion was performed for the treatment. The cystic lesion (about 3.0 × 3.6 cm) was ileal duplication, occurred at about 20 cm from the ileocecal junction, and located on the mesenteric border, making it difficult to excise it without removing attached bowel. Histologically, the duplication contained ectopic gastric mucosa that caused bleeding in the adjacent intestine (Fig. 4).

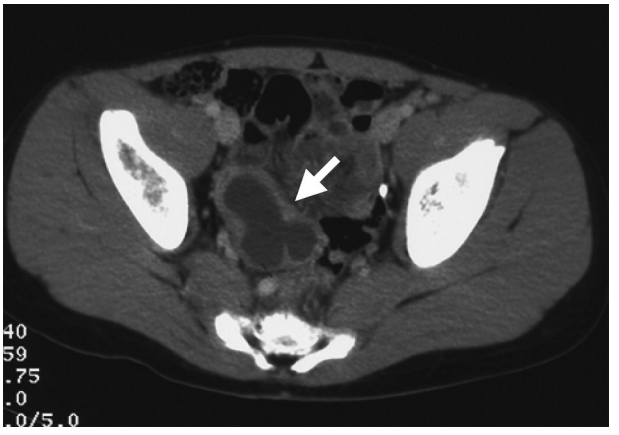

Fig. 3. CT study of the pelvic cavity. The arrow shows a cystic mass lesion in the pelvic cavity.

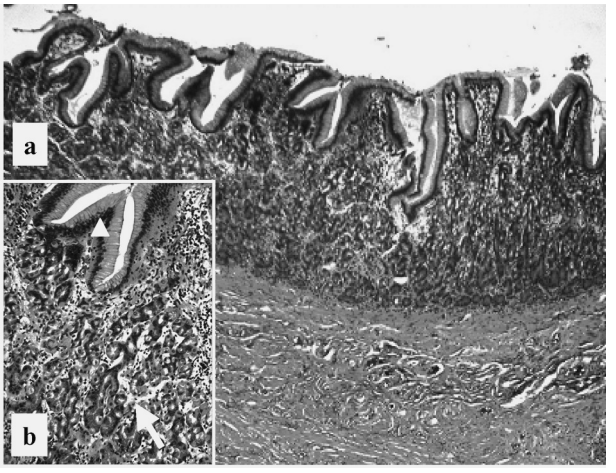

Fig. 4. Microscopic findings of the ileal duplication. The ileal duplication contained ectopic gastric mucosa (hematoxylin and eosin, × 40) (a). The mucosa showed typical structure of gastric mucosa containing foveolar (arrow head) and glandular (arrow) compartments (× 200) (b).

One year postoperatively, the patient has been doing well with no episodes of vomiting or abdominal pain.

Discussion

From the age of 5 years, the patient started to present with intermittent vomiting and abdominal pain without any abnormal findings of the abdomen or on abdominal x-ray. Since treatment was not effective, the patient was referred to our hospital because of intractable attacks. Lumbar spina bifida occulta, which happened to be found in an abdominal x-ray examination, suggested split notochord syndrome, resulting in the diagnosis of colonic duplication.

Duplication of the alimentary tract includes a variety of cysts, diverticulae, and tubular malformations (Ildstad et al. 1988). Gastric mucosa is frequently observed in the walls of such duplications, irrespective of their site of origin in the alimentary tract (Bajpai and Mathur 1994). Vertebral anomalies have not been associated consistently with duplications at any site (Bajpai and Mathur 1994). Split notochord syndrome is a group of abnormalities thought to be caused by abnormal splitting or deviation of the notochord leading to a persistent connection between the gut and dorsal skin (Bentley and Smith 1960). Clinically, the syndrome is diagnosed whenever there is an association of mediastinal or abdominal cyst with malformation of the spinal cord and column. In this syndrome, the ventrally situated yolk sac, or primordial gut endoderm, may become either herniated or adherent to the neural tube through the split notochord, forming enteric fistula to the skin, diverticulum, enteric cyst, and alimentary tract duplication in various combinations.

Split notochord syndrome can occur at any level in the spine, but the majority of reported cases have involved the cervical or thoracic region (Alrabeeah et al. 1988). The cases involving the lumbar region are very rare. There has been a case report of tubular colonic duplication, sigmoid colon diverticulum, and lumbar anterior spina bifida in a 28-year-old man diagnosed with split notochord syndrome (Kurisu et al. 1992).

The patient was exceptional in having reached the age of 28 years without complaints. Our case is also a rare case of split notochord syndrome without specific symptoms in infancy or early childhood. Those two cases suggested that some cases of split notochord syndrome with lower vertebral abnormalities could be asymptomatic during early childhood. In addition, our case had been treated as cyclic vomiting syndrome for several years before establishment of the diagnosis. Surgical findings suggest that the attacks were due to ulceration, inflammation, or bleeding caused by acid-peptic juice secreted from ectopic gastric mucosa. Duplication of the alimentary tract should be considered as a possible cause in patients with symptoms suggesting cyclic vomiting syndrome (Li and Misiewicz 2003).

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