

A case of midgut volvulus due to mesenteric lymphangioma, not associated with malrotation in a 13-year-old boy

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= Abstract =

A 13-year-old boy, complained of an intermittent suddenly aggravated severe abdominal pain and diarrhea, was diagnosed as a small bowel volvulus without an intestinal malrotation, due to mesenteric lymphangioma. He took abdominal ultrasonography, abdominal CT scanning, upper gastrointestinal study and got an operation. The small bowel volvulus with cystic lymphangioma was confirmed by gross and pathologic findings. (**Korean J Pediatr 2008;51:431-434**)

Key Words : Midgut volvulus, Cytic lymphangioma, Malrotation

Introduction

A small bowel volvulus is an uncommon condition. It usually occurs within the first year of life. Infants with volvulus show symptoms of duodenal obstruction, but older children or adults may have vague clinical manifestations^{1,2)}. A small bowel volvulus may be associated with intestinal malrotation²⁾ however, cases without malrotation have been reported^{3,4)}. A mesenteric lymphatic malformation can cause an abdominal mass, which rarely has been associated with a small bowel volvulus^{5,6)}. We report a very rare case of a small bowel volvulus due to a mesenteric lymphangioma, not associated with malrotation, in a 13-year-old boy.

Case report

A 13-year-old boy was taken to the pediatric emergency department with intermittent colicky abdominal pain for 7 days and watery diarrhea for 2 days. Over the past four years, he had a history of multiple similar episodes of abdominal pain, but improved with a glycerine enema. The vital signs were a blood pressure 120/60 mmHg, pulse rate 96/min, respiration rate 22/min, and body temperature 36.5 °C. He looked acutely ill.

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The abdomen was soft and flat, but specific abdominal tenderness was detected and the bowel sounds were increased. A standard flat and upright view x-ray of the abdomen were normal. Ultrasound and computed tomography (CT) of the abdomen demonstrated a whirl-like appearance of the superior mesenteric vein and mesentery around the superior mesenteric artery. In addition, multicystic masses were seen at the right lower abdomen (Fig. 1A). An upper gastrointestinal contrast study showed a dilated proximal duodenum and a corkscrew or spiral appearance of the duodenum and jejunum (Fig. 1B).

At surgery a clockwise volvulus of the small bowel loops on its mesentery, about 60 cm distal to Treitz's ligament, was present and diffuse mesenteric edema and enlarged mesenteric lymph nodes were also present (Fig. 2A). The duodenojejunal flexure and ileocecal junction were in the normal position. The intestine demonstrated passive congestion probably due to stretched mesentery and traction on the root of the mesentery. A counterclockwise de-rotation was performed. serous fluid of 100 to 150 ml, and a soft, milky white mass measuring 7×5 cm superior to the volvulus along with enlarged mesenteric lymph nodes were found (Fig. 2B). The mass was found in the small bowel mesentery about 15 cm distal to Treitz's ligament. The mass was resected along with 15 cm segment of small intestine.

The mucosa of the resected small intestine showed multiple conglomerated submucosal nodules. On section, the intestinal wall showed a poorly demarcated multicystic ma

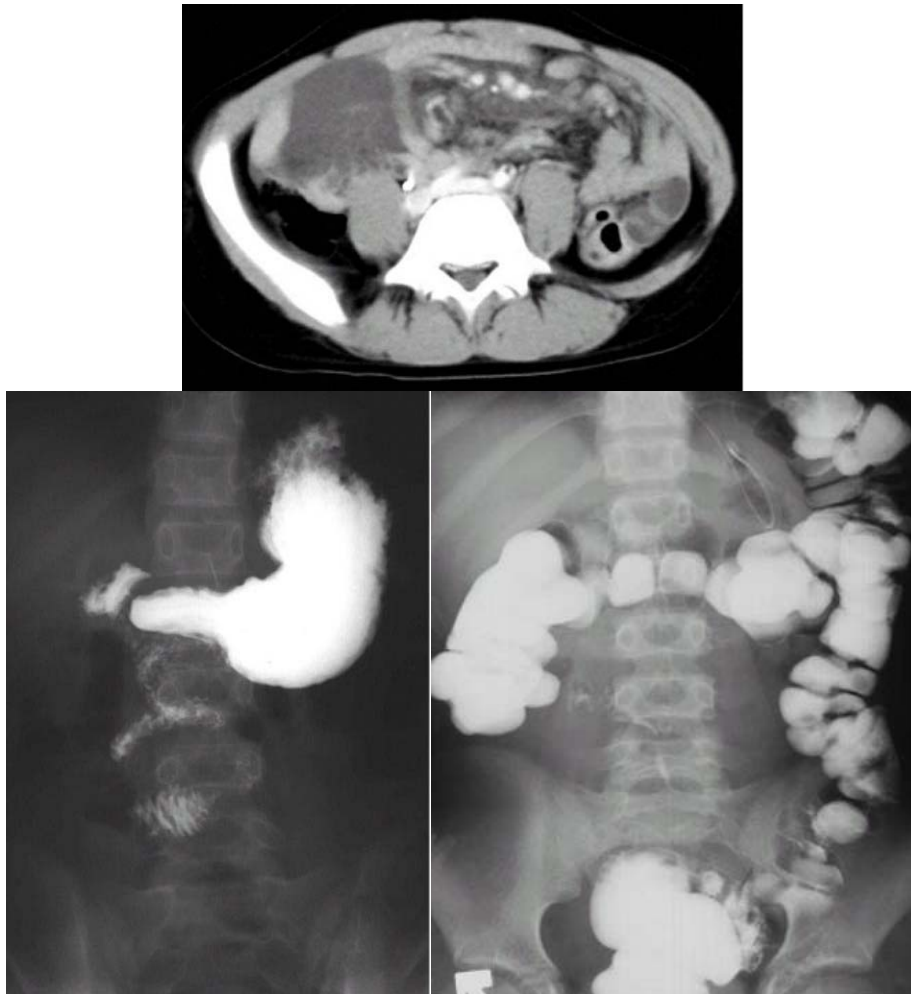


Fig. 1. A) CT scans show multicystic masses on the right lower abdomen. B) Upper gastrointestinal study shows dilated proximal duodenum and corkscrew or spiral appearance of duodenum and jejunum. C) Simple abdomen obtained on next day shows no evidence of rotation anomaly of the entire colon.

ss from the mucosa to the subserosa of the mesentery, measuring 16 cm at the largest dimension. The cysts were round and of variable size, measuring up to 7 cm in diameter. They contained yellow clear serous fluid. Microscopically, the cysts were lined by flat endothelial cells with no muscular coat. The post-operative period was uneventful and the patient was discharged 12 days later and is followed as an outpatient.

Discussion

Midgut volvulus is an intestinal obstruction caused by abnormal twisting of the bowel and usually presents in the first year of life. It is less common thereafter. It is a potentially life-threatening cause of obstruction and infarction

of the bowel. The most common cause of pediatric midgut volvulus is bowel malrotation. A midgut volvulus has been reported in 41–67% of cases with a bowel malrotation⁸⁾. However segmental volvulus without a malrotation, duplication cyst, retroperitoneal band, meconium plug, mesenteric defect and no causative abnormality have also been reported as causes of a midgut volvulus^{3,4)}.

The primary symptoms are a sudden onset of bilious emesis and abdominal pain in neonates and infants. However, the symptoms may be vague and nonspecific including chronic, intermittent vomiting and cramping abdominal pain, failure to thrive, constipation, bloody diarrhea and hematemesis in older infants and children^{2,7)}.

The spiral and corkscrew appearance of the duodenum and jejunum is diagnostic of a midgut volvulus and the

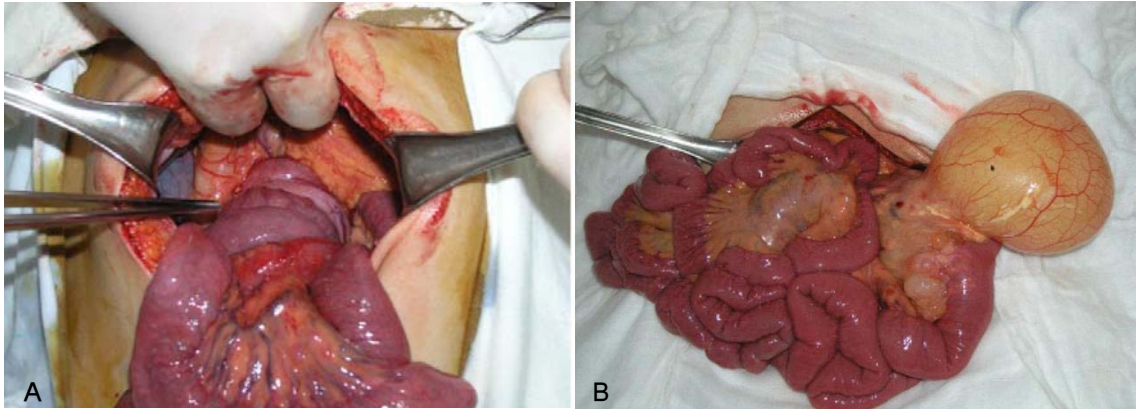


Fig. 2. A) operative finding shows a clockwise rotation of small bowel around the axis of superior mesenteric vessels. B) A soft, milky white mass arises from the small bowel mesentery.

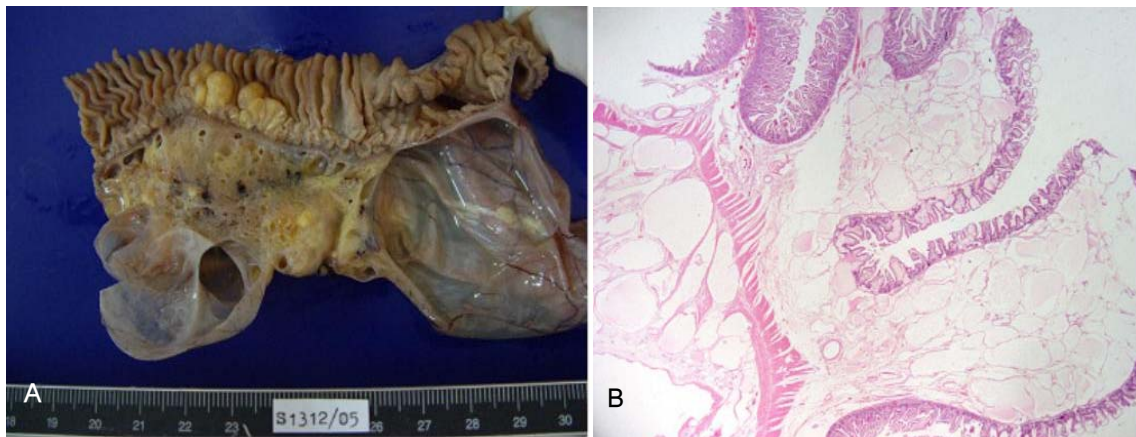


Fig. 3. A) Gross photograph of the cystic lymphangioma. A poorly demarcated cystic mass involves from the mucosa to the subserosa. The cysts have variable sizes and contained clear yellow serous fluid. B) Microphotograph of the cystic lymphangioma. The cysts occupy whole layers of the intestine, from mucosa to subserosa. The cysts have very thin wall with no muscular coat (H&E stain, $\times 200$).

bowel is usually twisted in a clockwise direction. Other signs of a small bowel volvulus include abnormal orientation of the superior mesenteric artery and superior mesenteric vein, which creates a whirl-like appearance on ultrasound and CT^{2, 6, 7}.

An abdominal lymphangioma is also an uncommon lymphangiomatous malformation occurring mainly in childhood⁹. There is lack of communication of the small bowel or retroperitoneal lymphatic tissue with the main lymphatic vessels, resulting in cystic mass formation¹⁰. Abdominal lymphangiomas may develop in the mesentery of the intestine, in the omentum or in the retroperitoneum¹¹. The clinical presentation is diverse and can range from an incidentally discovered abdominal mass to symptoms of an acute abdomen¹². Cystic lymphangioma is rarely associated with a small bowel volvulus^{5, 6}. In cases where cystic lymphan-

gioma and midgut volvulus were reported to occur together intestinal malrotation was always an associated finding^{5, 6, 13}, except for one case. The clinical presentation of the mesenteric cyst is usually dependent on the size, location and complications such as torsion, hemorrhage, infection or rupture of the cysts.

Three recent case reports of a lymphatic cyst in association with midgut volvulus have been reported as distinct from torsion of the cyst itself^{5, 6, 13}. While some authors have reported that the cyst in the mesentery is the primary pathology⁵, which can secondarily cause volvulus, others postulate the presence of a longstanding or intermittent volvulus as the cause of lymphatic cyst^{6, 13}. For the latter theory the history of intermittent abdominal pain and diarrhea was thought to support this theory. The thin walled lymphatics are probably the first to be occluded, followed

by the veins and arteries subsequently¹⁾. Chronic lymphedema develops due to lymphatic obstruction, which can cause a protein losing enteropathy and diarrhea. In our patient, there was a history of intermittent abdominal pain and diarrhea.

Because other mesenteric cysts can cause a volvulus^{3, 4)}, the mass effect is usually associated with a volvulus. Additional study is needed to understand the pathology of the cystic lymphangioma and volvulus.

Another unusual case presented in adolescence had enlarged mesenteric lymph nodes and anti-clockwise rotation of the volvulus¹³⁾. Our case had a multicystic lymphangioma (Fig. 3A) in cross section and was not associated with intestinal malrotation on the standard abdominal x-ray obtained on the next day (Fig. 1C) following the upper gastrointestinal study.

The treatment for a midgut volvulus is surgery. Ladd's procedure is most commonly performed for the intestinal malrotation. This procedure consists of untwisting of the midgut and division of adhesions obstructing the duodenum. The small bowel is placed on the right side of the abdomen and the colon on the left side. The last procedure was not needed in our case because of the absence of an intestinal malrotation. After corrective surgery, there is about a 7% risk of recurrence of a volvulus in children¹⁴⁾. Early diagnosis in these cases is important because delayed diagnosis can result in bowel necrosis and death.

한 글 요약

13세남아에서 발견된 창자 회전 이상이 동반되지 않은 남성 림프관종에 의한 창자꼬임 1례

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13세 남자 환자가 반복적이고, 발작적으로 발생하는 복통과 설사를 주소로 내원하였다. 복부 초음파검사, 복부 전산화 단층 촬영검사와 상부 위장관 검사를 통하여 창자 회전 이상은 동반되지 않은 작은 창자 꼬임을 진단하였다. 수술을 통해서 치료를 하였으며 병리, 조직 검사를 통하여 남성 림프관종에 의한 작은 창자 꼬임으로 밝혀졌기에 보고하는 바이다.

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