

Case Records of the Massachusetts General Hospital



Weekly Clinicopathological Exercises

FOUNDED BY RICHARD C. CABOT

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Case 8-2000

PRESENTATION OF CASE

An 8½-year-old girl was admitted to the hospital because of an abdominal mass and pain.

The child had had recurrent abdominal pain during the preceding two years. Her mother believed that the symptoms had begun at about the time that an elevated coliform count was discovered in the local water supply. Seventeen months before admission, the girl's blood count and sedimentation rate were normal, and stool examination showed no ova or parasites. Seven weeks before admission, intermittent fevers began, with anorexia, loss of about 4.5 kg in weight, and a reduction in the frequency of bowel movements. Six weeks before admission, a radiograph of the abdomen showed a nonspecific pattern of bowel gas and stool throughout the colon; there was a calcified nodule in the right upper quadrant that was consistent with the presence of a gallstone. Treatment with mineral oil was recommended, but it provoked vomiting and was discontinued. Five days before admission, the child began to vomit all ingested foods and liquids. She had fever and diarrhea, with dark stools but no obvious hematochezia. Two days later, her temperature rose to 39.9°C.

Two days before her admission to this hospital, the girl was admitted to another hospital because of increasing abdominal pain and persistent vomiting; there was little oral intake and no vomiting at that time. Examination revealed dehydration and diffuse abdominal tenderness without signs of peritoneal irritation, organomegaly, or a mass. The urine was normal. Laboratory tests were performed (Tables 1 and 2). An abdominal radiograph showed a moderate amount of stool in the right side of the colon and in the rectosigmoid. There was little or no bowel gas in the

TABLE 1. HEMATOLOGIC LABORATORY VALUES.

VARIABLE	TWO DAYS BEFORE	
	ADMISSION	ON ADMISSION
Hematocrit (%)	35.5	27.1
Mean corpuscular volume ( $\mu\text{m}^3$ )	69	71
Erythrocyte sedimentation rate (mm/hr)	11	
White-cell count (per $\text{mm}^3$ )	14,300	17,900
Differential count (%)		
Neutrophils	44	80
Band forms	39	7
Metamyelocytes	5	0
Myelocytes	1	0
Lymphocytes	6	7
Monocytes	5	6
Platelet count	481,000	332,000

TABLE 2. BLOOD CHEMICAL VALUES.

VARIABLE	TWO DAYS BEFORE	
	ADMISSION	ON ADMISSION
Urea nitrogen (mg/dl)*	32	
Sodium (mmol/liter)	134	134
Potassium (mmol/liter)	3.5	3.2
Chloride (mmol/liter)	102	
Carbon dioxide (mmol/liter)	19	

\*To convert the value for urea nitrogen to millimoles per liter, multiply by 0.357.

left portion of the abdomen. No distended bowel loops or masses were detected, and the psoas shadows were normal.

Fluids and electrolytes were given. On the evening of the first hospital day, the patient passed a bloody stool, and ceftriaxone and metronidazole were administered. On the next evening, she vomited dark liquid, and piperacillin-tazobactam and gentamicin were substituted. There was no further vomiting. On the third hospital day, the hematocrit was 27.9 percent. An abdominal radiograph (Fig. 1) showed a gallstone in the right upper quadrant and a large mass with an air-fluid level in the left upper quadrant. The distal portions of the small intestine were not dilated. A computed tomographic (CT) scan of the abdomen and pelvis (Fig. 2), obtained after the oral and intravenous administration of contrast material, showed a large mass containing contrast material and an air-fluid level in the left upper quadrant. The mass had a



**Figure 1.** Plain Film of the Abdomen.

A large gas-filled structure in the left upper quadrant has caused cephalad displacement of the transverse colon. Distended loops of bowel are visible in the midabdomen. A dense gallstone is present in the right upper quadrant.



**Figure 2.** CT Scan of the Abdomen.

A large structure in the left side of the midabdomen that is filled with fluid and gas displaces nondistended loops of bowel. The structure appears to originate near the transverse duodenum and extends into the pelvis.

thin wall and lay just below the stomach and slightly below the pancreas. The stomach and first and second portions of the duodenum contained contrast material and appeared normal. No contrast material or distention was observed in the small bowel distal to the mass. The third portion of the duodenum appeared edematous and thickened. Gas was present in the transverse colon. A gallstone, 8 mm in diameter, was present within the gallbladder, the wall of which was not thickened. No gas was detected in the biliary tract. The child was transferred to this hospital.

The patient's developmental history was normal, and her immunizations were up to date. There was no history of foreign travel and no family history of inflammatory bowel disease or liver disease.

The temperature was 38.1°C, the pulse was 134, and the respirations were 22. The blood pressure was 100/70 mm Hg.

On examination, the patient appeared pale but flushed and not acutely ill. Abdominal examination showed marked distention, without abnormal bowel sounds, tenderness, or other signs of peritoneal irritation.

Laboratory tests were repeated (Tables 1 and 2).

A diagnostic procedure was performed.

#### DIFFERENTIAL DIAGNOSIS

DR. MICHAEL R. CURCI\*: May we review the x-ray film and CT scan?

DR. ROBERT T. BRAMSON: A plain film of the abdomen (Fig. 1), obtained at the other hospital on the day the patient was transferred to this hospital, shows a mass in the left upper quadrant containing air and fluid that appears to be viscous. The distal portions of the small bowel are not dilated.

On the CT scan (Fig. 2), orally administered contrast material is visible in the stomach, and just below it a cystic mass contains contrast material and air, with an air–fluid level. The mass has pushed small-bowel loops and mesentery to the right. Several loops of bowel appear to have edematous folds. Scattered small collections of air medial to the mass suggest that the bowel may be perforated.

DR. CURCI: The salient findings that must be explained by the diagnosis of this child's illness include a history of partial intestinal obstruction and chronic abdominal pain, a cystic mass, anemia, and an enteric fistula. The mass appears to contain thick, viscous fluid, with a mixture of orally administered contrast material and air, and is surrounded by an enhanced wall — a finding consistent with the presence of an inflammatory reaction. The retroperitoneum on the left is not involved, since the kidney and psoas shadows are normal.

I am not aware of any tumor with this radiologic

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appearance. The chronicity of the patient's symptoms does not suggest a lesion of traumatic origin such as a pancreatic pseudocyst. The pancreas appears normal on the CT scan, and no abnormalities of the bile duct are evident, even though a solitary gallstone is present in the gallbladder, which does not have a thickened wall. The first and second portions of the duodenum also appear normal. No liver-function tests or pancreatic studies were performed.

Inflammatory lesions should be included in the differential diagnosis. The patient had a white-cell count of 14,300 per cubic millimeter, with 44 percent neutrophils and 39 percent band forms. Appendicitis with perforation is always a consideration in a child with abdominal pain. The radiographs do not show the location of the cecum. The calcification in the right upper quadrant could be a fecalith in a malrotated colon, but this diagnosis is ruled out by the CT scan. Also, the chronicity of the patient's symptoms is evidence against this diagnosis. An enteric fistula rarely occurs in association with a fecalith, and the communication appears to involve the proximal portion of the intestine. Inflammatory bowel disease is unlikely. Crohn's disease does not cause large abscesses, because the inflammatory process develops slowly and results in a localized stricture, an enteric fistula, or both.

A number of congenital lesions merit consideration: lymphangioma, malrotation, and intestinal duplications. Lymphangioma can appear as a multiloculated cyst and is often manifested by abdominal pain, fever, vomiting, ascites, and a palpable mass. It may become infected and have an enhanced wall on abdominal CT scanning, as in this case, but it is not associated with anemia or an enteric fistula.<sup>1,2</sup> A chronic malrotation may be manifested by abdominal pain, intermittent obstruction, anemia, and an abnormally positioned jejunum, as seen on the CT scan in this case. Malrotation, however, would not explain the multicystic mass or enteric fistula. The CT scan shows a normally positioned proximal duodenum. The fourth portion of the duodenum appears to be to the left of the midline, with the ligament of Treitz in its normal anatomical position. The superior mesenteric artery and vein are anterior to the pancreas and in their normal positions. The proximal jejunum is displaced in the right upper quadrant but is not obstructed, a finding that rules out the presence of a volvulus.

I favor the diagnosis of a bowel duplication, a rare lesion that is diagnosed before the age of two years in 60 to 83 percent of affected patients.<sup>3-6</sup> Bowel duplications are found less often in older children and only occasionally in adults.<sup>7</sup> They can occur throughout the gastrointestinal tract, causing symptoms that vary depending on their location. The small intestine is the most common site of duplications; most of them involve the ileum. They occur along the mesenteric border of the adjacent intestine and share blood supply with it. Jejunal duplications account for 8 to

16 percent of duplications. In several large series, ectopic gastric mucosa was present in 20 to 48 percent of patients with duplications<sup>3-6,8</sup> and was responsible for ulceration, hemorrhage, perforation, or a combination of these complications involving either the duplication or the adjacent tissue.<sup>9-11</sup>

Duplications can be either spherical or tubular. Spherical lesions vary in size and usually do not communicate with the adjacent intestine. They have thick walls and are filled with mucus. The initial manifestations are obstruction, intussusception, and a volvulus. Tubular duplications vary in length and can involve long segments of intestine. They are difficult to diagnose if they have both a proximal and a distal opening and if there is no obstruction, unless gastric mucosa is present in the duplication, with hemorrhage, ulceration, perforation, or a combination of these complications. If a duplication has only a proximal communication, dilatation, stagnation of the contents, and infection occur in the obstructed distal portion, which may enlarge to such an extent that it mimics chylous ascites. Also, bleeding may occur within the duplication or the adjacent intestine, where a marginal ulcer may develop. Perforation of the duplication into the adjacent intestine, peritoneum, or other nearby structures can also occur.<sup>12,13</sup>

A tubular duplication that involved the proximal jejunum, contained ectopic gastric mucosa, and lacked a distal communication would explain many of the findings in this case. It would account for the abdominal mass, chronic abdominal pain, anemia due to gastrointestinal bleeding, and evidence of an inflammatory reaction on the CT scan. The diagnosis must also explain the different findings on the two abdominal films that were obtained 48 hours apart before the patient's transfer to this hospital. Of particular note on the second radiograph is evidence in the left upper quadrant of a perforation into a large mass that contains both air and liquid. During the two-day interval, the patient's condition remained stable, without evidence of a progressive septic course, suggesting that the perforation was contained within a walled-off structure. On the CT scan, as one follows the proximal jejunum into the right side of the abdomen, extraluminal air, which may correspond to a limited perforation, becomes apparent. I believe that the explanation for these radiologic changes was the perforation of a tubular duplication into the adjacent jejunum<sup>10</sup> and that the perforation was due to ectopic gastric mucosa that caused ulceration within an ectopic duplication that lacked a distal opening. The perforation would account for the mixture of oral dye and air from the gastrointestinal tract.

My choice of a diagnostic procedure would have been a laparotomy to resect the congenital lesion. Technetium pertechnetate scanning might have identified ectopic gastric mucosa but would not have eliminated the need for an operative procedure.

DR. R. ALAN B. EZEKOWITZ: What about an infection, particularly an amebic one?

DR. CURCI: Since the patient had not traveled out of the country, I think an infection is an improbable diagnosis. In addition, I do not believe that the sequence of events is consistent with the diagnosis of amebiasis.

DR. DANIEL P. RYAN: The preoperative diagnosis was an infected enteric duplication cyst.

### CLINICAL DIAGNOSIS

Infected enteric duplication cyst.

### DR. MICHAEL R. CURCI'S DIAGNOSIS

Enteric duplication cyst with intraluminal perforation and an enteric fistula.

### PATHOLOGICAL DISCUSSION

DR. RYAN: An exploratory laparotomy revealed fibrinous and fibrous adhesions between loops of bowel. The mass was a cyst in the mesentery of the jejunum, about 1.5 cm below the ligament of Treitz. We removed the involved jejunal segment and the attached cyst and performed an end-to-end anastomosis.

DR. CAROLYN C. COMPTON: On pathological examination, the round-to-ovoid, fluid-filled cyst, which was 15 cm in diameter, was firmly attached to the adjacent jejunum (Fig. 3). The serosal surface of the cyst was tan-brown, appeared scabrous, and had areas of hemorrhage. The lumen of the cyst contained watery brown fluid and grumous material. An open communication, 1 cm in diameter, was identified between the cyst and the jejunal lumen. The wall of the cyst was leathery and irregular in thickness, and its interior surface was shaggy. No mucosal lining was recognizable on gross examination.

Microscopical examination of the cyst confirmed the absence of a mucosal lining, which was replaced by acutely inflamed granulation tissue and collections of foamy histiocytes. The wall of the cyst contained smooth muscle, the presence of which distinguished the lesion from a pseudocyst and identified it as an enteric duplication cyst. The muscle was markedly fibrotic (Fig. 4). The cyst and adjacent jejunum shared a muscular wall (Fig. 5) at their juncture. The communication with the intestine was lined entirely by inflamed granulation tissue, a finding consistent with the presence of an inflammatory fistulous tract. The cyst contained gram-negative bacilli and gram-positive cocci owing to the communication with the adjacent bowel. No gastric mucosa or peptic-type ulceration was seen.

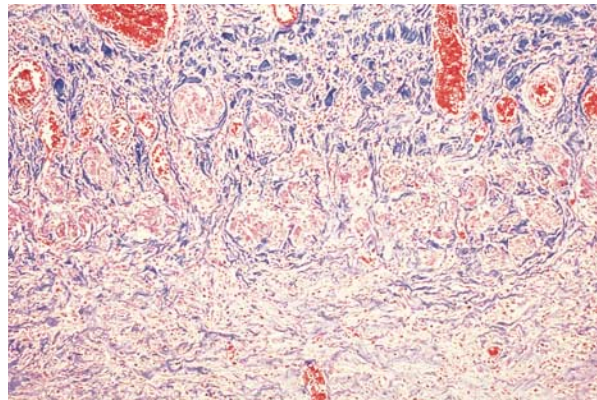
A cholecystectomy was also performed. The lumen of the gallbladder contained yellow calculi. Microscopical examination showed mild chronic cholecystitis.

Duplication cysts can occur anywhere in the gastrointestinal tract, from the tongue to the anus.<sup>3,14-18</sup> They may be single or multiple and vary widely in size.<sup>6</sup>



**Figure 3.** Enteric Duplication Cyst.

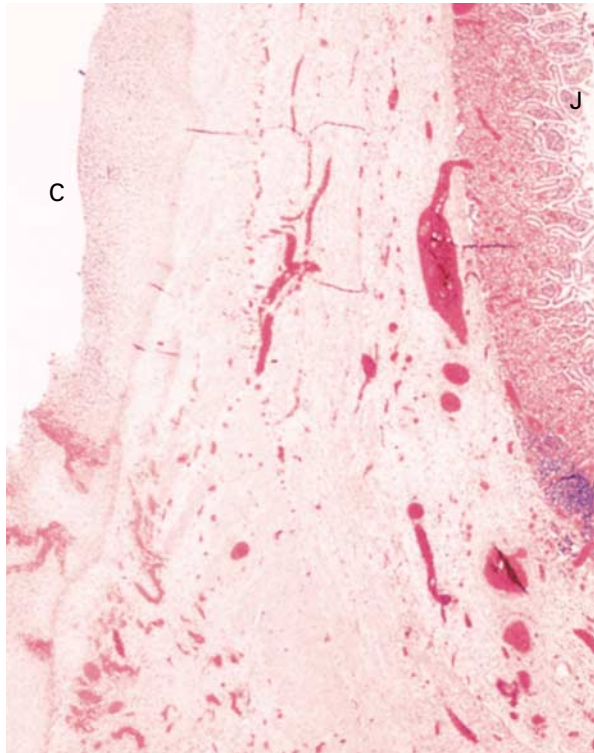
The adjacent jejunum has been opened longitudinally to show a communication (arrow) with the cyst. The outer surface of the cyst is shown to the right and left of the opened jejunum.



**Figure 4.** Bundles of Atrophic, Pink-Tan Muscle Cells Encased by Blue Collagen Fibers (Masson Trichrome,  $\times 125$ ).

Spherical duplications usually do not communicate with the intestine and are usually filled with clear secretions. However, as seen in this case, fistulae may form as a consequence of inflammation and necrosis. In contrast, tubular duplications typically open at one or both ends into the adjacent bowel lumen and may become filled with bowel contents, leading to the blind-loop syndrome.

The manifestations of intestinal duplications vary. Their size, location, and complications determine the signs and symptoms. Common clinical manifestations of small-intestine duplications include a palpable abdominal mass, abdominal pain, melena, vomiting, and



**Figure 5.** Wall Shared by the Cyst and the Adjacent Jejunum (Hematoxylin and Eosin,  $\times 35$ ).

The cyst (C) is lined by inflamed granulation tissue, and the jejunum (J) by normal mucosa.

an acute abdominal presentation with intestinal obstruction, which may be complicated by a volvulus or intussusception.<sup>18,19</sup>

Two of the most characteristic features of an intraabdominal duplication are its firm attachment to the adjacent gut and the presence of smooth muscle in its wall.<sup>20</sup> The lesion is typically lined by glandular mucosa that contains one or more cell types normally present in the gastrointestinal tract.<sup>21</sup> The most common type of mucosa is gastric; it is found in approximately 50 percent of duplications.<sup>22</sup> Resultant peptic injury often causes mucosal erosion or ulceration and hemorrhage.<sup>18,19</sup> Occasionally, as in this case, the entire mucosa becomes necrotic and inflamed.<sup>23</sup> In addition to peptic injury, increasing pressure and ischemia may have a role in mucosal necrosis,<sup>14</sup> especially in cysts as large as the one in this case. Ultimately, with transmural necrosis and the formation of fistulae, hemorrhage into the adjacent bowel may occur, and massive hemorrhage may be the initial manifestation of the duplication. Some authors have emphasized that duplications should always be considered in the differential diagnosis of lesions that cause massive gastrointestinal hemorrhage but that cannot be diagnosed by endoscopy.<sup>18</sup>

The formation of a fistula may lead to secondary infection of the cyst, as it did in this case. With either infection or hemorrhage, the contents of the cyst change from clear fluid to turbid fluid or a semisolid material, with a consequent change in the features of the cyst on imaging.

The first use of the term “duplications” to refer to these congenital lesions has been attributed to Reginald Fitz, who described the lesions in 1884 and thought they were remnants of the omphalomesenteric duct.<sup>6,24</sup> Although it is now clear that they are not, their pathogenesis has not been established. Several theories have been proposed to explain the pathogenesis of duplications, including the persistence of embryonic diverticula, epithelial sequestration, the sparing of a bowel segment as a result of embryonic vascular insufficiency or trauma, and anomalous development of a duplicate lumen due to an error in recanalization after the solid phase of enteric mucosal development in the fetus.<sup>6,17</sup> The last theory, proposed by Bremer in 1944,<sup>25</sup> is the most widely accepted. It is possible, however, that duplications at different anatomical sites develop through different mechanisms.<sup>16,23</sup> In particular, it has been suggested that the genesis of intrathoracic duplications, which are often associated with defects of the spinal cord, may be related to anomalous development of the notochord.

The treatment of an enteric duplication depends on the age and condition of the patient; the type, location, and extent of the lesion; and the presence or absence of associated abnormalities.<sup>6,14</sup> Complete surgical excision is recommended when it is feasible. Duplications cannot be removed by simple excision. A segmental resection of the involved intestine is required, because enteric duplication cysts are always mesenteric and share their blood supply and muscular wall with the adjacent bowel.<sup>14,18</sup> If the duplication is too extensive to excise completely (e.g., in the case of a complete duplication of the small bowel), partial excision can be performed with internal drainage or removal of the mucosal lining, in order to eliminate the possibility of acid secretion and consequent peptic injury. Complete excision of the cyst or its mucosa is also advisable because adenocarcinoma has been reported to develop in duplication cysts in adults.<sup>11</sup> In infants and children, however, duplications are uniformly benign.<sup>6</sup>

**DR. CURCI:** In patients with very long tubular defects, decompression is associated with a high risk of recurrent hemorrhage and ulceration, especially if there is ectopic gastric mucosa. In the surgical literature, as Dr. Compton mentioned, a mucosal-stripping operation is recommended for very long duplications. This operation can be difficult to perform, but it may be the only long-term solution in some cases.

#### ANATOMICAL DIAGNOSIS

Enteric duplication cyst communicating with the jejunum.

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35-MILLIMETER SLIDES FOR THE CASE RECORDS

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