Idiopathic localized dilatation of the ileum: CT findings


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Abstract
We report computed tomographic and pathologic findings of an adult case of idiopathic localized dilatation of the ileum presenting as hematochezia and bowel perforation. If a cyst-like structure that has narrow communications with proximal and distal bowel loops and a layered enhancement pattern similar to those of adjacent bowels on the computed tomogram of a patient with gastrointestinal bleeding, idiopathic localized dilatation of the ileum should be suspected.

Key words: Intestines, abnormalities—Intestines, hemorrhage—Intestines, perforations—Intestines, CT—Intestines, biopsy.

Idiopathic localized dilatation of the ileum (ILD1) is a rare condition in which one or more segments of the ileum are dilated with more or less abrupt transitions to normal segments proximally and distally. There should be no evidence of deficient innervation. This condition has many synonyms such as segmental dilatation of the small bowel, ileal dysgenesis, segmental megaileum, and even giant Meckel’s diverticulum [1].

To the best of our knowledge, fewer than 50 cases have been reported in the world literature since the first description in 1962 [2]. However, computed tomographic (CT) findings for ILDI have not been previously reported. We describe the CT appearance of a case of ILDI causing bowel perforation and panperitonitis, which was confirmed pathologically.

Case report
A 69-year-old man visited the emergency room because of a sudden onset of severe abdominal pain and three episodes of hematochezia. Laboratory studies were within normal ranges except that hemoglobin (10.4 g/dL) and hematocrit (30.4%) levels were low. Physical examination showed a rigid and distended abdomen with generalized and rebound tenderness. The impression was panperitonitis.

Plain radiography of the abdomen showed an air–fluid level within a mildly distended bowel loop in the lower abdominal area, but we thought that a nonspecific finding. Helical CT with dynamic contrast enhancement showed multiple dilated bowel loops due to paralytic ileus caused by probable panperitonitis. In the pelvic cavity, there was a large cyst-like structure that was 9.5 cm in its largest diameter. This structure had no valvulae conniventes, which were present in adjacent dilated loops. However, the enhancing wall of the cyst-like lesion had the same layering pattern as that of adjacent bowel loops. Two abrupt narrow transitions to proximal and distal normal bowel loops were found (Fig. 1). CT scans did not show evidence of pneumoperitoneum.

Although a definite focus of bowel perforation was not verified, the clinical presentation of panperitonitis was so evident that exploratory laparotomy was performed. There was a 1-cm perforation of the ileum approximately 50 cm proximal to the ileocecal valve within a segment of
saccular bowel dilatation demarcated by two narrow points proximal and distal to the perforation site. Segmental resection of the involved loop and end-to-end anastomosis were done.

The pathologic specimen showed a focal dilated ileal loop without valvulae coniventes. This was distinct from the adjacent normal bowel in which the folds were present and normal in appearance. Two abrupt transitional points were found proximally and distally between normal and abnormal segments of the ileum. Small perforated and nonperforated ulcerations were present in the center of the dilated segment of the ileum (Fig. 2). Microscopic examination showed no abnormality in the bowel wall.

Discussion

Most reported cases of ILDI occur in childhood [3–5]. However, a small number become manifested in adulthood, as in our case [1, 6, 7]. Usual manifestations consist of abdominal pain, growth failure in childhood, and gastrointestinal bleeding with iron-deficiency anemia. Functional obstructive symptoms due to the atonic ileal segment are often noted. Panperitonitis is a relatively rare manifestation [1–3, 5–7].

The cause of ILDI is unclear but may be congenital. The usual manifestation during childhood and the presence of related congenital anomalies in reported cases support a congenital basis of this abnormality. There have been a few hypotheses about the pathogenesis of ILDI; neuromuscular dysfunction of the bowel [1, 7], prolonged intrauterine bowel obstruction by extrinsic compression [1, 3, 4], physiologic herniation through the abdominal wall as an obstructing lesion [8], and an early insult of heterotopic tissues to the developing gut [5].

Pathologically, the involved segment is composed of a thin but otherwise normal bowel wall with normal ganglion cells and nerve plexuses in most cases. The segment usually contains ulceration and, less commonly, heterotopic gastric mucosa. Decreased or absent valvulae coniventes are noted in most cases despite only minimal to moderate distention [1]. Grossly, the involved segment is tubular, bilobated, or multilobated and measures 6–21 cm [1].

The radiographic appearance of ILDI is easy to recognize and quite specific if some studies are combined. The characteristic plain film finding of the abdomen is that of a single distended bowel loop, which may have a