

Case Report

PERFORATION OF LONG SEGMENT ILEAL DUPLICATION WITH GASTRIC HETEROTOPIA - A CASE REPORT

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ABSTRACT

Alimentary tract duplication cysts are rare congenital malformations, found primarily in children under the age of 15 years. It is becoming increasingly recognized as a cause of abdominal pain, obstruction and hemorrhage, but few instances have been published where perforation has occurred. Majority of the duplications present as spherical cysts and usually range from a few millimeters to less than ten centimeters in size. Although very few patients present with peritonitis as the initial manifestation, the possibility should be borne in mind when diagnosing and planning therapy for such emergencies, particularly in children. We describe herein the case of a 2.5-year-old boy in whom generalized peritonitis was caused by perforation of a long segment tubular communicating ileal duplication cyst.

Key Words: Small Intestine, Ileal Duplication, Perforation

INTRODUCTION

Alimentary tract duplication cysts are rare congenital malformations with an incidence of one in 4500 birth (Puligandla *et al.*, 2003) that can occur at any site in the digestive tract, but most frequently in the ileum. A duplication of the alimentary canal has three characteristic features; it is lined by alimentary tract epithelial mucosa, has a well developed smooth- muscle layer, and is fixed to at least one point in the gastrointestinal tract (Lister, 1990). Most patients with duplication present with an abdominal tumor, bowel obstruction, intussusception, and/or melena (Ildstad *et al.*, 1988; and Stringer *et al.*, 1995) but peritonitis caused by perforation of the duplication is rarely seen. We report one such case of a child presenting with perforation of an ileal tubular duplication.

CASES

A 2.5-year-old boy was referred to our hospital with distension and pain abdomen, vomiting, fever since two days. On admission, he had blood pressure of 80/65 mmHg, a heart rate of 120/min, and a temperature of 100 F. Physical examination revealed distension abdomen with generalized tenderness and guarding. Rectal examination revealed an empty rectum. Bowel sounds were absent upon auscultation. Laboratory data showed an elevation in the white blood cell count to 15900/mm³. Abdominal X-ray showed a small intestinal bowel gas pattern, but no pneumoperitoneum, and abdominal ultrasonography revealed a small amount of fluid in the rectovesical pouch; however, the ileocaecal wall was normal and the appendix was not visualized. Subacute intestinal obstruction was suspected and conservative management involving the passage of a nasogastric tube, IV rehydration, and antibiotics was initiated. The patient failed to respond to conservative treatment. An emergency laparotomy was performed the next day, about 50ml of purulent ascites was found in the pelvis. A 50 x 2-cm tubular duplication was observed, 50 cm from the ileocecal valve, adjacent to the ileum on the mesenteric side, and a 5-mm perforation was noted in the middle of the duplicated segment. Therefore, his condition was diagnosed as generalized peritonitis caused by perforation of an ileal tubular duplication. The duplication and adjacent normal intestine were removed, and an end-to-end reanastomosis was performed.

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Macroscopic examination showed a 50 x 2-cm duplication on the mesenteric side of the ileum. On cutting the normal ileal (antimesenteric) segment showed normal mucosal folds while duplicated segment was of different colour (grey white). Examination of the resected duplication also revealed a 0.5-cm orifice in the middle of it. The proximal end of the duplication was not connected to the normal intestine, while the distal end was communicating to the normal intestine.

Histological findings revealed that the duplication was lined by heterotopic gastric mucosa with fundic glands, and the area around the perforation consisted of intestinal mucosa, with edema and bleeding. These findings indicated that the heterotopic gastric mucosa had caused peptic ulceration on the intestinal mucosa of the duplication, resulting in perforation. The patient was discharged from hospital on postoperative day 10 and has remained well and symptom-free.

DISCUSSION

Alimentary tract duplication cysts are congenital malformations that can occur anywhere from the lingual root to the anus. By definition, they are covered by smooth muscle, lined by alimentary tract mucosa, and located adjacent to some segment of the alimentary tract (Ladd and Gross, 1940). Morphologically, they are divided into spherical and tubular types, the tubular type being less commonly found in the small intestine than the spherical type (Wrenn, 1993). Each type can be further subdivided into a communicating type, which connects with the normal intestine, and a noncommunicating type. In this regard, tubular duplications often communicate with the normal intestine at the proximal and/or distal end (Wrenn, 1993). In our patient, the tubular duplication communicated with the normal intestine at the distal end.

Although duplications can be encountered in patients of any age, majority of them are found in infants and children, with a slight preponderance in males (Puligandla *et al.*, 2003). The ileum is most frequently involved, followed by such sites as the colon, esophagus, and jejunum (Macpherson, 1993).

Most cases of duplication present as an abdominal tumor, bowel obstruction, intussusception, and/or melena (Ildstad *et al.*, 1988; and Stringer *et al.*, 1995) however, there have been a few reports of patients presenting with peritonitis caused by perforation of the duplication (Stringer *et al.*, 1995; Niesche, 1973; Collins, 1972; and Waterston *et al.*, 1980). A review of these cases indicates the following as conditions giving rise to perforation: increased pressure within the duplication, Wrenn (1993) ischemia, Wrenn (1993) peptic ulcer, Niesche (1973); Collins (1972) and Waterston *et al.*, (1980) and calculary formation. Marks and Stunt (1984) In our patient, the perforation was caused by peptic ulceration in the intestinal mucosa within the heterotopic gastric mucosa. The presence of such heterotopic gastric mucosa was also reported by Vaage and Knutrud (1974) and Stringer *et al.*, (1995) in 5 of 31 patients (16%) and 7 of 18 (39%) patients with small intestine duplication, respectively. Perforation due to heterotopic gastric mucosa can involve the heterotopic gastric mucosal segment or the communicating orifice to the normal intestine. Patients with heterotopic gastric mucosa frequently develop melena as a result of ulceration, (Ildstad *et al.*, 1988) but asymptomatic progression to sudden perforation is rare. Our patient suffered ulcerative perforation in the heterotopic gastric mucosa of the duplication.

Peptic ulceration of ectopic tissue can account for unusual and often misleading symptoms, making diagnosis difficult. Although ^{99m}Tc-pertechnetate is useful for detecting heterotopic gastric mucosa (Waterston *et al.*, 1980) it is difficult to apply this procedure for distinguishing between heterotopic gastric mucosa and Meckel's diverticulum. It is also virtually impossible to employ this procedure for patients with acute abdomen, due to limitations in time. Although abdominal ultrasonography is an alternative diagnostic tool for detecting spherical-type duplications, it cannot be used for diagnosing the tubular type, as in the present case. In the surgical treatment of duplication cysts, resection of the

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duplication alone is difficult since the duplication and adjacent intestines are fed by a common vessel. Therefore, the recommended procedure is removal of the duplication along with a short segment of the adjacent normal intestine.

In conclusion, the case described in this report serves to illustrate that perforation of a long segment duplication which exhibited extensive gastric heterotopia, although rare, should be considered in the differential diagnosis of acute peritonitis, especially in children.

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