Terminal Ileum Duplication
An Unusual Case of Small Bowel Obstruction
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ABSTRACT

Duplications of the alimentary tract are uncommon congenital anomalies that usually present during infancy and early childhood. The case of an adolescent presenting with small bowel obstruction secondary to a duplication cyst is presented and the challenges in the management described.

Keywords: Aetiology, ileum abnormalities, intestinal obstruction

INTRODUCTION

Duplications of the alimentary tract are uncommon congenital anomalies that usually present during infancy and early childhood. Eighty-five per cent are diagnosed before two years of age (1), and as a result, in patients presenting after this age, duplications are rarely considered as a differential diagnosis for small bowel obstruction. Recognition of this entity as a differential is not the only important consideration, but knowledge of the various anatomic, vascular and pathologic subtypes is also required. This will lessen the significant surgical challenge that may exist when the condition is diagnosed at operation, and theoretical or practical experience of a paediatric surgeon is unavailable.

The case of an adolescent presenting with small bowel obstruction is presented and the management of duplication cysts is discussed.

CASE REPORT

A 12-year-old boy with a longstanding ventriculo-peritoneal (VP) shunt was referred to the surgical service at the University Hospital of the West Indies (UHWI) with an 18-month history of recurrent episodes of abdominal pain associated with headache, vomiting and diarrhoea. The cycles of symptoms occurred every two weeks, with vomiting not always a constant feature. Physical examination during these episodes had revealed no significant findings apart from ascites that was thought to be due to the VP shunt, and investigations performed at various times, including stool cultures, barium meals, and abdominal ultrasounds were all non-diagnostic. Several times during this eighteen-month period, he was admitted, had ‘negative’ investigations, settled spontaneously and was discharged from hospital.

Examination on presentation revealed a thin, wasted young man in no painful distress. His abdomen was distended, with an obvious right upper quadrant scar consistent with the VP shunt. Peristalsis of the bowel was seen through his thin abdomen, despite the presence of ascites. There were no masses, and the abdomen was non-tender. Barium meal and follow through done showed marked dilation of the jejunum and ileum, with a contracted terminal ileum of
irregular outline, and filling distally only after several hours. The clinical impression at this admission was that of possible inflammatory bowel disease (Crohn’s) with intermittent small bowel obstruction. Colonoscopy was only possible to the ascending colon. There were no mucosal abnormalities seen grossly or on histologic assessment of random biopsy specimens.

With a long history of recurrent episodes of partial mechanical small bowel obstruction, the decision was made to proceed to exploration after the VP shunt was converted to a ventriculo-atrial (VA) shunt to minimize the risks of shunt contamination. Five days later, he underwent exploratory laparotomy under the cover of systemic broad spectrum prophylactic antibiotics. The jejunum and ileum were massively distended proximal to a large, cystic mass located just proximal to the ileocaecal valve (Figure). The caecum and overall histologic features were consistent with small intestine duplication.

DISCUSSION

Congenital malformations of the intestinal tract include obstructive defects, anomalies of fixation and duplication cysts, and usually present in infancy or early childhood. Various terminologies had been used to describe this anomaly until Gross and Holcomb suggested the term duplication be used for all such anomalies, irrespective of their site, morphology or embryologic derivations, in order to simplify the nomenclature (2, 3). Duplications arise from disturbances in embryonic development of the gut and, unlike a Meckel’s diverticulum, are located along the mesenteric border of the involved intestine with which they are intimately related. There are two main types, spherical and tubular (4, 5). The spherical or cystic duplications are usually separate from the lumen of the bowel, while the tubular duplications often communicate with the lumen. Both types have walls of smooth muscle and a mucosal lining of some part of the alimentary tract, all features that help to differentiate duplications from diverticula and mesenteric cysts.

Duplication cysts may be found anywhere between the mouth and the anus, but are commonly located along the small intestine, with the ileum being the most frequent site (6). They rarely present in persons over 15 years of age (1, 3, 7). Ileocaecal duplication cysts are a subgroup which are situated in the terminal ileum and may involve the ileocaecal valve (8). Clinical presentation, as in our case, is usually as a result of obstruction due to pressure on the adjacent bowel or occasionally from an intussusception or volvulus (9). Haemorrhage may result from erosion of the cyst wall into the surrounding vasculature, volvulus of the cyst with associated venous engorgement, or from the gastric mucosa present in some of these.

Despite the use of imaging techniques and laparoscopy, preoperative diagnosis of duplication cysts is uncommon (7, 10–13). It must be mentioned that in many centres, this patient would have been admitted and managed by paediatric surgeons who might have suspected the diagnosis.

Cystic duplications can usually be completely excised by enucleation or resection when a non-contiguous cyst with separate blood supply is found (10, 11). Others require resection of a small portion of bowel. The tubular types may represent a more significant challenge. For an extensive tubular duplication, bowel resection may result in short-gut syndrome. To avoid this complication, long duplications can be treated by marsupialization of the duplication, followed by excision of most of the wall of the duplication, then stripping the mucosa from the common wall. For non-obstructed bowel, some advocate mucosal resection only (1, 7), but there is little evidence to suggest that this is a practical procedure (11). When discovered incidentally, resection is
advised as the statistical risk for complications is unknown and malignancy, though rare, has been reported (1, 12, 13). Resection of the terminal ileum, cystic duplication and the caecum was performed in this case with good results.

REFERENCES