Duplication Cyst of The Caecum - A Rare Case Report with Review of Literature

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Abstract

Duplications of the alimentary tract encompasses a wide variety of mass lesions throughout the gastrointestinal tract that are either tubular or cystic and can occur anywhere from oropharynx to anus with predominance in males. They are more common in ileum but rare in the caecum. Eventhough several theories have been postulated, the true aetiology is not known. We report one such rare case of duplication cyst of caecum in a six year old female child, with review of literature.

Introduction

Gastrointestinal duplications are rare congenital malformations that may vary greatly in presentation, size, location, and symptoms. This term is applied to congenital lesions having three characterstics - (1) The presence of a well-developed coat of smooth muscle, (2) An epithelial lining representing some portion of intestinal tract mucosa, and (3) Intimate anatomic association with some portion of the gastrointestinal tract.[1] Furthermore, cystic duplication of caecum is especially rare with only 19 cases reported so far in English literature.[2]

Majority of duplications are diagnosed in first two years of life and usually made by prenatal ultrasonography. Abdominal pain, vomiting and abdominal mass are the most common signs and symptoms. Ileal duplications are often confused as Intussusception, Appendicitis or Meckel's diverticulum. Therefore it should be included in the differential diagnosis for paediatric surgical abdominal emergencies.

Case Report

Six year old female child presented with abdominal pain, vomiting and passage of blood in stools for the past three days. There was diffuse and severe colicky pain in abdomen associated with two to three episodes of vomiting per day, non-bilious with no history of haematemesis. History of passage of fresh blood in stools, not associated with mucus or melaena was also present. No history of fever or diarrhoea noted.

On examination, tenderness all over the abdomen was present and a palpable, firm, non-pulsatile, well defined, oval mass, 4 x 3 cm, not moving with respiration, was felt in infraumbilical region. Clinical diagnosis of acute intestinal obstruction with a possibility of intussusception was made. Ultrasonography revealed distended small bowel loops with absence of peristaltic movement which was traced upto the ileocaecal junction. Right iliac fossa showed evidence of bowel loops surrounded by other bowel loop, suggesting the possibility of intussusception. Intraoperatively, a cyst was noted at caecum stretching and obstructing it.

We received a segment of ileum, caecum, with attached appendix and a cyst at the caecum, altogether measuring 10 x 10 x 6 cms. Cyst measured 10 x 6 x 5 cms. External surface of the cyst showed congestion. There was no perforation or gangrenous change(Fig. 1). On cut section, unilocular cyst oozed out clear serous fluid approximately 40 ml with uniform wall thickness of two mm. There were no solid areas or papillary excrescenses.

Microscopic examination of the caecal cyst showed ulcerated lining and presence of intestinal type of glands in the mucosa, similar to the adjacent caecum (Fig. 2). The ulcerated mucosa is covered with dense chronic inflammatory cell infiltrate extending into the muscle layer and serosa. Muscularis propria was thinned out and showed congested and dilated blood vessels (Fig. 3). Sections from appendix showed features of chronic

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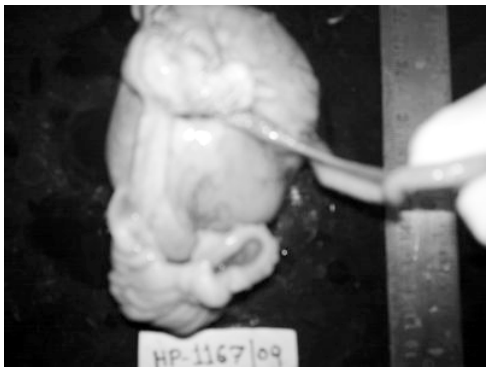

Fig. 1 Gross photograph of the caecal cyst with attached appendix.

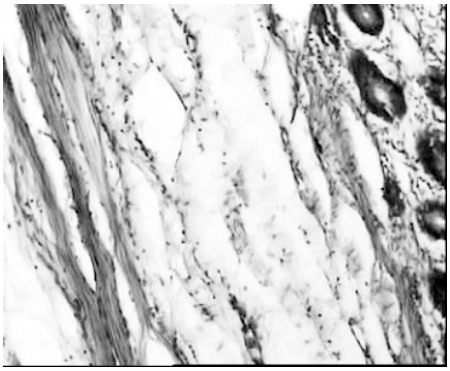

Fig. 2 H and E, 4x showing cyst wall lined by mucosa similar to the adjacent caecum.

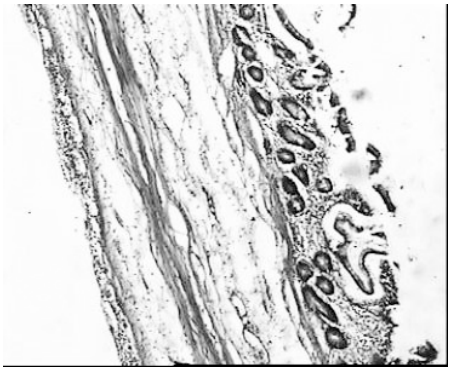

Fig. 3 H and E, 10x showing thinned out cyst wall with muscle coat.

appendicitis and that from the ileum was unremarkable.

Discussion

Duplications of the alimentary tract was first reported by Calder in 1733.[3] They have been variously referred to as giant diverticula, enterogenous cysts, ileal or jejunal duplex, giant thoracic cysts, duplications and reduplications and unusual Meckel's diverticula.

In 1937, William E-Ladd used the term 'Duplications of the Alimentary tract'. Ladd and Gross reported 18 such cases in 1941 and Gross reported 68 cases in 1953. A meta-analysis by Heiss of 12 large series of enteric duplications encompassing 580 patients demonstrated that 20% of these lesions occurred in the chest and remaining 80% in the abdomen with a small percentage being thoraco-abdominal. It is common to refer to duplications as foregut, midgut or hindgut derived, depending on their location. Enteric duplications frequently contain mucosa similar to that of their adjacent gastrointestinal location.[4]

In 10-20% of patients, enteric duplications are multiple and the presence of one such lesion should warrant a search for others.[5] Children with enteric duplications seem to have a high incidence of other associated anomalies, like spinal malformations, intestinal malrotation or atresias, urinary tract anomalies and skeletal anomalies.

The most frequent location for a duplication cyst of the intestinal tract is the region of terminal ileum and ileocaecal valve, 53%.[4] The next most common areas are oesophagus, stomach and duodenum. Duplication cysts of the caecum and colon are especially rare with reported incidence of 13%.[1]

Enteric duplications are generally cystic or tubular masses and present in a variety of ways depending on their size, location, adjacencies and whether they contain heterotopic gastric mucosa. Many duplications will have few or no symptoms and usually found incidentally. The

diagnosis may be made by prenatal ultrasonography. Cystic masses appear as black holes on ultrasonography.[4]

Because duplication is a cystic mass, acute distension with secretions or infection may cause severe pain in the abdomen. An acutely enlarging cystic mass may cause obstruction and result in nausea, vomiting and cramping. A large duplication may even cause localized volvulus. Mass is palpable per abdomen in upto 50% patients.

Duplications of the midgut or hindgut are more likely to cause abdominal pain, distension, melaena or perforation. Those arising in the ileum may be confused with acute appendicitis and it's difficult to diagnose preoperatively. Small bowel duplications may present as intussusceptions by acting as a lead point.

Radiologic studies such as USG, plain radiographs, gastrointestinal contrast studies, CT or MRI may assist in the diagnosis.

Midgut and hindgut duplications may be more difficult to diagnose, and their diagnosis is often made at operation. These lesions may present as Meckel's diverticulum or volvulus or intussusception. Therefore, duplication cysts should always be included in the differential diagnosis when evaluating abdominal conditions in paediatric age group.

Duplications of small intestine are the most common enteric duplications encountered, and the majority of these occur in the ileum. They are either cystic or tubular and most are located on the mesenteric side. This is in contradistinction to Meckel's diverticula, which characteristically occur on the antimesenteric side. Communication with the lumen of the intestine may be variable.

Optimal treatment is removal. This is usually accompanied by excising the cystic duplication with its adjacent bowel with primary reanastomosis.[4]

Conclusion

Duplication cysts of the intestine may present in diverse ways and encompass a wide variety of lesions from neck to anus. They can be simple and cystic, complex, multiple or tubular. They may be associated with spinal and genitourinary anomalies. Optimal treatment is resection and patients have excellent long-term outcomes and quality of life.

References

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EARLY TIPS IN CIRRHOSIS WITH VARICEAL BLEEDING

In this randomized trial involving patients with cirrhosis and acute variceal bleeding who were at high risk for treatment failure, control of bleeding was more common and mortality was lower among patients as-signed to early treatment with a transjugular intrahepatic portosystemic shunt (TIPS) than among those assigned to standard treatment with rescue TIPS, if needed.

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