SAJS
Case Report

Mucinous cystadenoma arising in a completely isolated infected ileal duplication cyst

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Summary
Gastrointestinal duplications are uncommon congenital lesions that can occur anywhere along the alimentary tract, and the symptoms of which generally develop during infancy or childhood. Completely isolated duplication cysts are an extremely rare variant of duplication, where no communication between the cyst and the adjacent bowel segment is present. We report the unique case of an adult who presented with right lower abdominal pain and systemic signs of inflammation caused by infection of a completely isolated ileal duplication cyst. Histological examination of the cyst additionally revealed a low-grade mucinous cystadenoma. We discuss the clinical presentations, diagnosis and treatment of this rare entity.

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Case report
A 56-year-old man presented at our emergency department with a 3-day history of pain in the right lower abdomen associated with tiredness, arthralgia, headache and shivering. Nausea was present without vomiting. On examination he was mildly pyrexial, and there was a tender mass as well as signs of peritoneal irritation in the right lower abdomen.

The positive laboratory findings were a white cell count of 18.03×10^9/l and a C-reactive protein level of 225 mg/l. A triple-contrast computed tomography (CT) scan of the abdomen revealed an inflammatory tumour measuring about 15 cm in diameter in the right lower abdomen, with enlarged mesenteric lymph nodes (Fig. 1). Laparoscopic exploration of the abdomen showed a whitish saucer-shaped ileal mass located close to Bauhin's valve and capping the inflammatory ileo-caecum (Fig. 2). After conversion to a low-middle laparotomy, ileo-caecal resection and selective lymphadenectomy were performed. No other tumour was noted in the abdominal or pelvic cavity. The patient made an uneventful recovery, with rapid regression of systemic inflammatory signs. He was discharged on the 5th postoperative day.

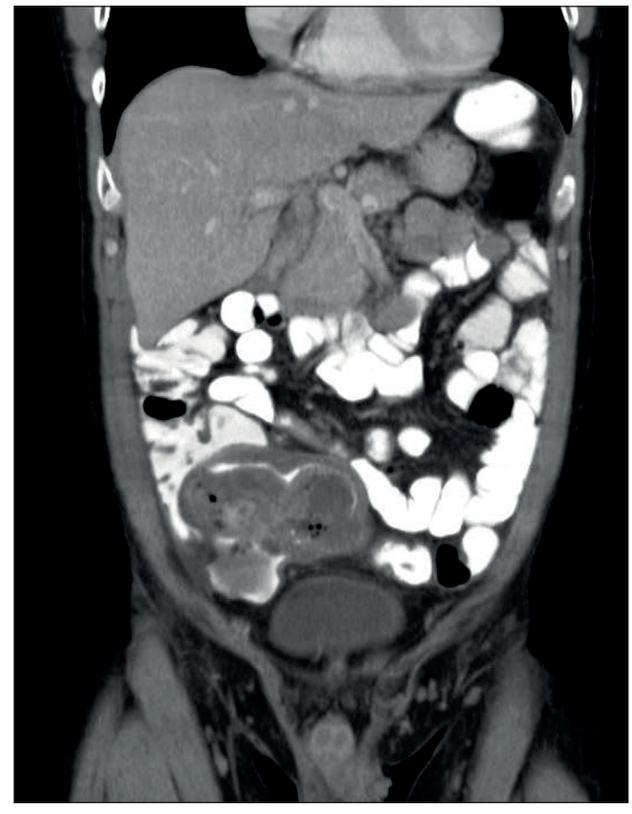

Fig. 1. Coronal view of the tumour in the right lower abdomen on computed tomography scan.

Macroscopic pathological examination revealed a spherical mucus-filled cyst of about 7 cm in diameter, located on the mesenteric aspect of the ileum without communication with the intestinal lumen (Fig. 3). Microscopically the cyst wall included small-bowel epithelium with cell atypia, underlined by a double-layered smooth-muscle coat and inflammatory serosa. Mesenteric lymph node sections showed reactive hyperplasia without malignant change. No parasite or bacteria could be

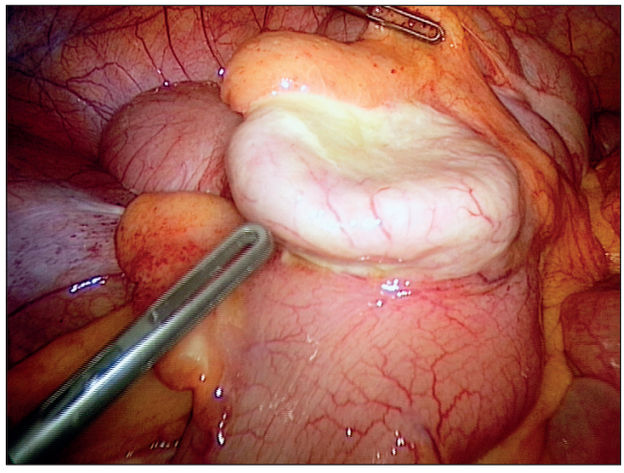

Fig. 2. Laparoscopic view of a white saucer-shaped ileal tumour located close to Bauhin's valve and capping the inflammatory ileo-caecum.

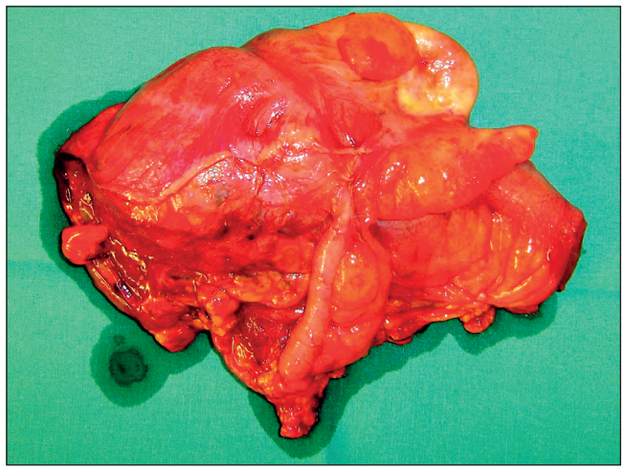

Fig. 3. Macroscopic view of the resection specimen. The saucer-shaped ileal cyst is located on the mesenteric aspect of the ileum.

identified. The proliferative activity of the tumour cells, measured immunohistochemically by expression of Ki-67 antigen, was low. A negative stain for Pan-Cytokeratin AE1/AE3 ruled out an invasive carcinoma. Macroscopic as well as microscopic pathological features, including the presence of cell atypia, established the diagnosis of low-grade mucinous cystadenoma arising in a completely isolated and infected ileal duplication cyst.

Discussion

Duplications of the alimentary tract are uncommon congenital lesions that can occur anywhere along the tract. They are most commonly found in the jejunum or ileum (30 - 50%). Less commonly, they may be oesophageal (17%), colonic (15%), gastric (8%), duodenal (6%), rectal (6%) or oral (1%).[1] These lesions are usually isolated spherical or tubular structures attached to the mesenteric side of the gastrointestinal tract, often sharing a common smooth-muscle wall and vascular supply with histological features of a layered smooth-muscle coat and gastrointestinal mucosal lining. Completely isolated duplication cysts are an extremely rare variant of gastrointestinal duplication cyst, where no communication between the cyst and the adjacent bowel segment is present.

Several theories have been proposed to explain the causation of duplication cysts, including aberrant recanalisation of the gut lumen, persistent embryonic diverticulum, hypoxic or vascular occlusive events, partial twinning, and the 'split notochord' theory.[1] Symptoms of alimentary tract duplications generally develop during infancy or childhood, and much more rarely during adult life. Common manifestations include gastrointestinal haemorrhage due to peptic ulceration of heterotopic gastric mucosa, gastrointestinal obstruction from mass effect or intussusceptions, and abdominal pain. Our patient had systemic signs of infection and abdominal pain caused by infection of the cyst and its surrounding tissue. To our knowledge, only one case of an infected ileal duplication cyst (with *Salmonella*) has been published, occurring in a 2-year-old boy.[2]

In the case of unrecognised asymptomatic duplication, neoplastic changes can occur. Malignant transformation in oesophageal, gastric, intestinal and colonic duplications have been reported.[3] To avoid gastrointestinal complications, including malignant transformation, surgery is advocated for the treatment of alimentary tract duplication even in asymptomatic patients. Complete resection, usually accomplished by resection of the adjacent bowel and mesentery with primary anastomosis, should be performed. On occasion, long tubular duplications will necessitate removal from within the mesentery by dividing vessels on one side of the mesenteric bilayer to preserve blood supply to normal bowel.[4] Alternatively, in the case of completely isolated duplication cysts, resection can be carried out safely without bowel resection as demonstrated by Srivastava *et al*.[5]

Conclusions

Although manifestations of alimentary tract duplications are most common in the paediatric age group, visceral surgeons who treat adults should be aware of this rare entity. Owing to potential complications, including malignant transformation, surgical treatment is advocated, even in the absence of symptoms.

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