Introduction

Duplications of the gastrointestinal tract are rare congenital anomalies that are often found early in life (more than 80% are diagnosed in children under 2 years of age, and rarely in adults) [1,2]. The preoperative diagnosis of duplication cyst is difficult because the clinical manifestations are highly variable, especially in adults. Moreover, duplication cysts sometimes cause obscure gastrointestinal bleeding.

Double-balloon enteroscopy has a prominent role in examination of the small intestine, especially in patients with obscure gastrointestinal bleeding. Double-balloon enteroscopy makes it possible to visualize, sample, and endoscopically treat (using clipping, argon plasma hemostasis, polypectomy, etc.) lesions.

In this case, we performed double-balloon enteroscopy in a patient with severe iron deficiency anemia, and were able to diagnose a duplication cyst preoperatively. We present the patient’s unique clinical course, enteroscopy images, and surgical pathology.

Case report

A 19-year-old man with severe iron deficiency anemia was admitted to our hospital. At age 15, he was diagnosed with anemia during a medical checkup, but was not treated. Three months before admission, he developed abdominal pain and exertional dyspnea. In January 2016, he visited our hospital due to sudden onset of painful paraphimosis. The urologist treated this with manual repositioning and circumcision was...
scheduled. Preoperative blood testing revealed severe iron deficiency anemia (red blood cells $2.53 \times 10^6/dL$, hemoglobin 4.3 g/dL, hematocrit 17.5%, mean corpuscular volume 69.2 fl, mean corpuscular hemoglobin 17.0 pg, mean corpuscular hemoglobin concentration 24.6 g/dL). The cause of bleeding was not evident on upper gastrointestinal endoscopy or colonoscopy. Abdominal computed tomography was performed and a cystic structure resembling Meckel’s diverticulum was found in the ileum. Double-balloon enteroscopy via the anal route was performed and revealed a narrowed distal ileal lumen, 50 cm from the ileocecal valve. After passage through the narrowed lumen, the intestinal tract was bifurcated (Fig. 1a), with one segment connected to a blind sac (white triangles). A large, shallow, and irregular ulcer near the bifurcation (white arrow) is shown. The ulcer bled easily from contact during endoscopy but there were no exposed vessels at its base. The mucosal surface of the blind sac was covered with normal villi. 

Fig. 1 Images on double-balloon enteroscopy. a The intestinal tract was bifurcated, with one segment connected to a blind sac (white triangles). A large, shallow, and irregular ulcer near the bifurcation (white arrow) is shown. The ulcer bled easily from contact during endoscopy but there were no exposed vessels at its base. b The mucosal surface of the blind sac was covered with normal villi.

Discussion

Duplication cysts are rare congenital malformations (1/10,000 live birth) that can appear anywhere in the gastrointestinal tract from the oral cavity to the anus [2–4]. Eighty percent are diagnosed in children below the age of 2 years, with a male predominance [1, 2]. The ileum is the most frequently affected site [2].
Symptoms include abdominal pain, vomiting, melena, constipation, intussusception, and ileus [1–8]. They can be easily misdiagnosed as other disorders, including appendicitis, Crohn’s disease, and Meckel’s diverticulum. For this reason, the preoperative diagnosis of a duplication cyst is difficult. Hoshi et al. reported that correct preoperative diagnostic yield for a duplication cyst was 11.2% in Japan [7].

Duplication cyst of the ileum must be differentiated from Meckel’s diverticulum. Meckel’s diverticulum is a true congenital diverticulum derived from the remnant omphalomesenteric duct during development of the terminal ileum, while a duplication cyst can occur anywhere in the gastrointestinal tract, most commonly in the ileum. Meckel’s diverticulum is normally located on the antimesenteric aspect, while ileal duplication cysts normally appear on the mesenteric aspect. When a duplication cyst is detected, resection and anastomosis, including that of adjacent normal intestine, is required in all cases, because of the shared common bowel wall and blood supply [4, 9]. All duplications should be surgically treated at the time of diagnosis owing to possible complications including bowel perforation, bleeding, obstruction, and malignant changes. Notably, 23% of intestinal duplication cysts in adults were affected by ileal cancer [3, 4, 9, 10]. In contrast, Meckel’s diverticulum has a vitelline artery or a clearly independent blood supply, and most cases require cystectomy alone.

Duplication cysts may cause severe anemia in some cases. Melena was the presenting symptom in 10.5% of patients with a duplication cyst, and ectopic mucosal tissue was detected in 54.5% [6, 7]. Tc-99m pertechnetate scintigraphy is helpful for diagnosis when duplication cysts have ectopic gastric mucosa. In our case, an ulcerated area was present in the duplication cyst, but no ectopic mucosa was detected. Therefore, the ulcer may have been caused by repeated transient intussusception and inflammation around the bifurcation of the sac. This might have explained to long-standing severe anemia and narrowing of the small intestine.
Conclusion

Duplication cysts are rare in adults, and there are few reports of diagnosis before surgery. We report a case of duplication cyst of the ileum, presenting with severe anemia, diagnosed with double-balloon enteroscopy, and treated without incident. Therefore, the differential diagnosis in a patient with unexplained anemia and abdominal pain should include a possible duplication cyst. Additionally, endoscopic evaluation should be performed.

Competing interests

None

References
