



소장의 다발장중첩증을 동반한 라푼젤증후군의 드문 발현

민경진 · 차 한¹ · 김성민² · 최재연

가천대학교 의과대학 길병원 응급의학교실, ¹소아청소년과학교실, ²외과학교실

A rare presentation of Rapunzel syndrome with multiple small bowel intussusceptions

Kyoung Jeon Min, Hann Tchah¹, Seong Min Kim², Jea Yeon Choi

Departments of Emergency Medicine, ¹Pediatrics, and ²General Surgery, Gachon University Gil Medical Center, Gachon University College of Medicine, Incheon, Korea

Rapunzel syndrome is caused by gastric trichobezoar with extended tail and small bowel obstruction. Patients with gastric trichobezoar can be asymptomatic until the bezoar increases in size. We report a case of a girl who visited the emergency department with abdominal pain. She was finally diagnosed with Rapunzel syndrome that causes multiple small bowel intussusceptions associated with trichophagia. Surgery was needed to reduce the multiple intussusceptions, and to remove the large trichobezoar. This case highlights to consider the possibility of Rapunzel syndrome when diagnosing the main cause of intussusceptions.

Key words: Bezoars; Intestinal Obstruction; Intussusception; Pica; Trichotillomania

Introduction

Small bowel intussusception is unusual, representing 1%–10% of intussusceptions¹. It can be caused by infection, Meckel's diverticulum, polyp, duplication cyst, tumor, hematoma or vascular malformation. Multiple, simultaneous small bowel intussusceptions are rare.

Bezoars are accumulations of undigested materials in the gastrointestinal tract. In a trichobezoar, the main component is a hair bundle, and gastric bezoar can lead to small bowel obstruction. Rapunzel syndrome is defined as gastric trichobezoar with extended tail into the small bowel causing its obstruction². This entity presents with abdominal pain, nausea, vomiting, bowel obstruction, weight loss, hematemesis, peritonitis, and intussusception³.

Small bowel intussusception due to Rapunzel syndrome is clinically important because it is likely to need surgical intervention⁴. Several cases of Rapunzel syndrome presenting with small bowel obstruction containing intussusception and bowel obstruction have been reported. However, cases with multiple small bowel intussusceptions are rare^{5,6}. We present a case of a 10-year-old girl with

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Corresponding author

Jea Yeon Choi (ORCID 0000-0002-9326-5897)

Departments of Emergency Medicine, Gachon University Gil Medical Center, Gachon University College of Medicine, 21 Namdong-daero 774beon-gil, Namdong-gu, Incheon 21565, Korea

Tel: +82-32-460-3901 Fax: +82-32-460-3019

E-mail: g3marine@naver.com

large trichobezoar causing multiple small bowel intussusceptions.

Case

A 10-year-old girl visited the emergency department with 4-day history of abdominal pain, nausea, and vomiting. She had about 12-month history of chronic

abdominal pain, but did not visit a hospital because the pain was mild and intermittent. She was developmentally normal, and previously healthy.

On physical examination, the abdomen was distended and tender with a palpable mass in the left upper quadrant with normal bowel sounds. Laboratory values revealed a white blood cell count of 8,610/mm³, hemoglobin concentration of 14.5 g/dL, and C-reactive protein of 1.63 mg/dL. Other laboratory findings were normal.

Plain abdominal radiograph showed a large mass-like lesion within the stomach (Fig. 1). Abdominal ultrasonography showed multiple target lesions at 4 points in the small bowel (Fig. 2A), and a 5 cm-long echogenic lesion with posterior shadowing in the stomach (Fig. 2B). We subsequently performed computed tomographic scan, showing a large mass in the stomach with a long segmental intussusception in the proximal jejunum (Fig. 3A). Other intussusceptions in the jejunum were also detected (Fig. 3B).

In the laparoscopic exploration, 3 segmental small bowel intussusceptions were identified with the maximum length of 15 cm; the stomach was distended by a hard trichobezoar. The multiple intussusceptions in the jejunum were manually reduced, and an 8 cm-long gastrotomy was performed at the antrum. The huge, bile-stained trichobezoar was removed (Fig. 3C). The length of the trichobezoar was 13 cm, and the tail was extended into the proximal

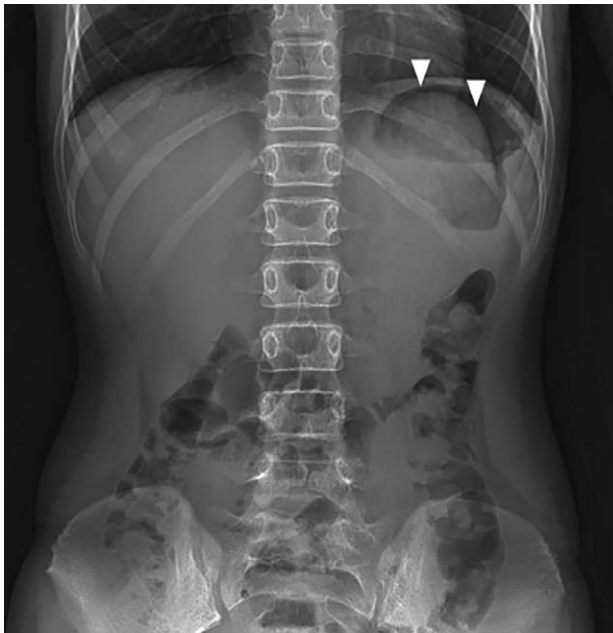


Fig. 1. Plain radiograph of the abdomen showing a large mass-like lesion within the stomach (arrowheads).

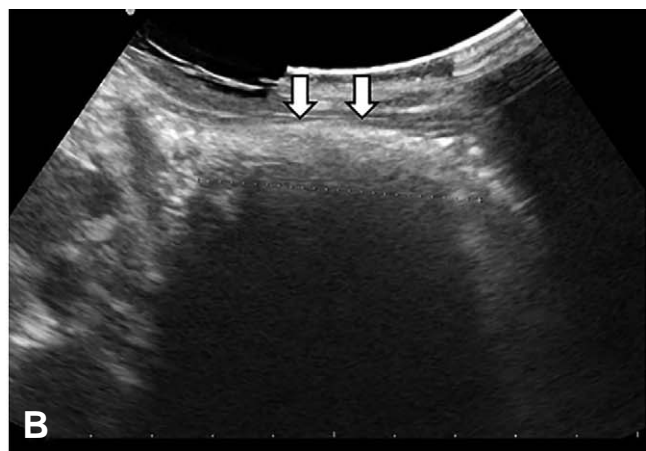
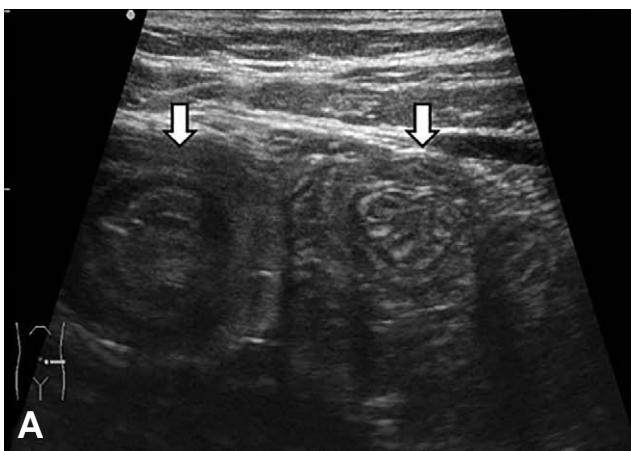


Fig. 2. Ultrasonography of the abdomen. Multiple target lesions are detected in the small bowel (arrows, **A**). There is a 5 cm-long echogenic lesion with posterior shadowing in the stomach, suggesting the trichobezoar (arrows, **B**).

jejunum. No necrosis or perforation of the bowel was observed.

Psychiatric consult showed that she had a habit of pulling out and eating her hair since the preschool age. The girl was discharged 4 days after the surgery without complication. At outpatient department follow-up, she was free of abdominal symptoms.

Discussion

A bezoar was first reported during autopsy on a patient who died of gastric perforation with peritonitis by Baudamant in 1779⁷. Bezoars are classified according to their main components⁴: plant and vegetable fibers (phyto-bezoars), hair (tricho-), milk protein or formula (lacto-), medicines (pharmaco-), or plastic materials (plasto-). Trichobezoar is a hair bundle in the stomach or small bowel. Hair strands are lodged in the gastric mucosal folds, and the slippery surface prevents moving by peristalsis. As hair strands accumulate,

a hair ball becomes too large to pass the stomach. This large bundle of hair takes on the shape of the stomach⁴.

Rapunzel syndrome is an unusual condition of trichobezoar in children with approximately 50 cases reported in the literature. Although there has been a little difference in definition in the published cases, Rapunzel syndrome is generally characterized by a trichobezoar with a tail-like extension and symptoms of small bowel obstruction^{2,3}. Most patients with trichobezoars are teenaged females, and a half of them have psychiatric disorders, such as trichotillomania or trichophagia.

Clinical manifestations include the presence of a mass in the abdomen, abdominal pain, nausea, vomiting, constipation, diarrhea, general weakness, weight loss, and malnutrition⁸. In our case, the patient had abdominal pain, nausea, vomiting, and a palpable mass in the left upper quadrant. The mass effect of a bezoar results in obstruction, ulceration, bleeding or perforation of the gastrointestinal tract. The extended tail of the trichobezoar may reduce

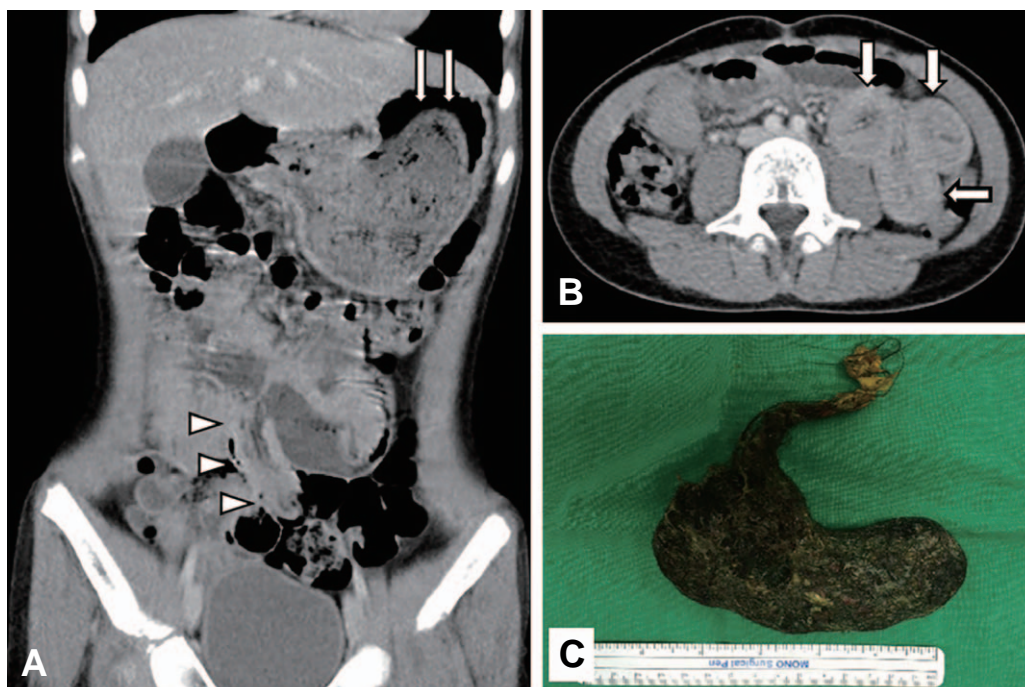


Fig. 3. Computed tomography and a gross specimen of the removed trichobezoar. Long arrows indicate the gastric trichobezoar, and arrowheads indicate the long segmental intussusception in proximal jejunum (A). Other multiple intussusceptions are shown in the jejunum (arrows, B). The maximum diameter of the trichobezoar body was 13 cm (C).

peristalsis, and cause intussusceptions. Definitive diagnosis is made with endoscopy. Ultrasonography shows echogenic lesion with posterior shadowing due to a mixture of hair, air, and food. Computed tomography shows a heterogeneous mass filled with air in the gastrointestinal tract.

The goal of treatment of bezoars is removal and prevention of recurrence. Surgical exploration remains the main treatment option in Rapunzel syndrome. Endoscopic removal of gastric bezoar can be less invasive and cost-effective. In a recent report, an 8×4 cm-sized trichobezoar was removed endoscopically⁹⁾, but there is no report of complete endoscopic removal of Rapunzel syndrome-associated bezoars. Kim et al.¹⁰⁾ reported surgical removal of a large bezoar after it was endoscopically cauterized with argon plasma. Psychiatric therapy for trichotillomania or trichophagia should be considered to prevent its recurrence.

In conclusion, we report a pediatric case of Rapunzel syndrome presenting with multiple small

bowel intussusceptions caused by a trichobezoar with an extended tail into the proximal jejunum. The bezoar was completely removed through gastrotomy after manual reduction of the multiple small bowel intussusceptions. This type of intussusception in Rapunzel syndrome are rare, but need timely diagnosis and surgical intervention to prevent fatality. This case highlights to consider the possibility of Rapunzel syndrome when diagnosing the cause of unusual intussusception.

Conflicts of interest

No potential conflicts of interest relevant to this article were reported.

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