A rare presentation of Rapunzel syndrome with multiple small bowel intussusceptions

Kyoung Jeen 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 intussusceptions⁴. 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 obstruction⁶. This entity presents with abdominal pain, nausea, vomiting, bowel obstruction, weight loss, hematemesis, peritonitis, and intussusception⁶. Small bowel intussusception due to Rapunzel syndrome is clinically important because it is likely to need surgical intervention⁶. 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 rare⁶. We present a case of a 10-year-old girl with...
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).](image1)

![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).](image2)
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 1779\(^7\). Bezoars are classified according to their main components\(^4\): 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 stomach\(^4\).

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 malnutrition\(^8\). 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 wit 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 endoscopically, but there is no report of complete endoscopic removal of Rapunzel syndrome-associated bezoars. Kim et al. 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.

Acknowledgements

No funding source relevant to this article was reported.

References

1. Pandey A, Rawat JD, Wakhlu A, Kureel SN, Gopal SC. Simultaneous occurrence of jejun-jejunal and ileo-ileal intussusception in a child: a rare occurrence. BMJ Case Rep 2011;2011:bcr0820103294.
2. Vaughan ED Jr, Sawyers JL, Scott HW Jr. The Rapunzel syndrome. An unusual complication of intestinal bezoar. Surgery 1968;63:339-43.
3. Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P, et al. Rapunzel syndrome reviewed and redefined. Dig Surg 2007;24:157-61.
4. Gupta R, Prabhakar G, Mathur P, Goyal RB, Sharma C, Ali MA. Rapunzel syndrome and its variants in pediatric patients: our experience. Arch Int Surg 2014;4:152-7.
5. Prasanna BK, Sasikumar K, Gurunandan U, Sreenath GS, Kate V. Rapunzel syndrome: a rare presentation with multiple small intestinal intussusceptions. World J Gastrointest Surg 2013;5:282-4.
6. Ha SC, Koh CC, Lee CH. A rare case of Rapunzel syndrome with multiple small bowel intussusceptions and bowel obstruction. Formos J Surg 2016;49:67-9.
7. Faria AP, Silva IZ, Santos A, Avilla SG, Silveira AE. The Rapunzel syndrome: a case report: trichobezoar as a cause of intestinal perforation. J Pediatr (Rio J) 2000;76:83-6.
8. Anantha RV, Chadi SA, Merritt N. Trichobezoar causing intussusception: youngest case of Rapunzel syndrome in a boy in North America. J Pediatr Surg Case Rep 2013;1:e11-3.
9. Benatta MA. Endoscopic retrieval of gastric trichobezoar after fragmentation with electrocautery using polypectomy snare and argon plasma coagulation in a pediatric patient. Gastroenterol Rep (Oxf) 2016;4:251-3.
10. Kim SC, Kim SH, Kim SJ. A case report: large trichobezoar causing Rapunzel syndrome. Medicine (Baltimore) 2016;95:e3745.