CASE REPORT

A rare case of Rapunzel syndrome with multiple small bowel intussusceptions and bowel obstruction

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Abstract
Rapunzel syndrome is a rare condition that is characterized by a gastric trichobezoar with a long tail extending from the stomach to the small bowel. Patients can be asymptomatic for a long period, and symptoms develop later when the bezoar enlarges in size. The most common presentations include chronic abdominal pain, malabsorption, gastrointestinal tract obstruction, gastrointestinal bleeding, and intussusceptions. We report a case of a 5-year-old girl with Rapunzel syndrome causing multiple small bowel intussusceptions. We were unable to identify the main cause of this condition during laparoscopic reduction. Definite diagnosis was made after the development of gastrointestinal tract obstruction. Surgeons should consider the possibility of Rapunzel syndrome when diagnosing the main cause of intussusceptions.

1. Introduction
A small bowel intussusception is rare and can be caused by infection, polyp, lymphoma, vasculitis, Meckel’s diverticulum, intestinal duplication, cystic fibrosis, and intramural hematoma. Multiple simultaneous small bowel intussusceptions are rarer. Rapunzel syndrome is an extremely rare intestinal condition resulting from a trichobezoar with a tail-like extension from the stomach to the small bowel. Rapunzel syndrome may present as chronic abdominal pain, malabsorption, gastrointestinal

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tract obstruction, gastrointestinal bleeding, and intussusceptions. Few cases of Rapunzel syndrome in patients presenting with intussusceptions have been reported. We report a case of a 5-year-old girl with Rapunzel syndrome causing intussusceptions and bowel obstruction.

2. Case Report

A 5-year-old girl presented with severe abdominal pain and nausea that started 2 days before presentation. She experienced cramping pain in the left abdomen for three weeks without other gastrointestinal symptoms. On physical examination, a left palpable abdominal mass was noted. Abdominal echo revealed typical imaging findings but an unusual location for intussusceptions. Abdominal computerized tomography (CT) revealed multiple target signs (Figure 1) over the jejunum. Radiological reduction was unsuitable for the multiple jejunal intussusceptions; therefore, laparoscopic exploration was performed. During operation, 8 segmental jejunal intussusceptions ranging from 2 cm to 8 cm in length were observed at a distance of 20–100 cm from the ligament of Treitz. All intussusceptions were laparoscopically reduced. A mass or diverticulum was not observed. The patient had a smooth recovery and was discharged on the 5th day after operation.

Six weeks after the discharge, the patient returned with severe pain in the upper abdomen. In addition, she complained of vomiting soon after a meal and experienced less pain after vomiting. Abdominal CT revealed a long bezoar impacted in the junction of the duodenum and jejunum with proximal gastrointestinal tract obstruction, which was not observed in the previous CT (Figure 2). Esophagogastroduodenoscopy was arranged for evaluating any other possible bezoars. During examination, a gastric bezoar consisting of long hairs was revealed; however, we were unable to extract it endoscopically. Midline laparotomy with gastrotomy was then performed. After exploration, a gastric bezoar approximately 4 cm in length with a connection stalk to another larger duodenal bezoar approximately 6 cm in length was removed through the gastrotomy wound. The total length of both bezoars including the long tail was approximately 30 cm (Figure 3). The gastrointestinal tract was examined by palpation for other residual bezoars; neither a mass nor an

Figure 1  Multiple target signs (red arrows) are revealed through computerized tomography.

Figure 2  A bezoar located at the junction of the duodenum and jejunum with distended stomach (red arrows) is revealed through computerized tomography.

Figure 3  A gastric bezoar with a connection stalk to another larger duodenal bezoar is removed through gastrotomy. D = duodenal bezoar; G = gastric bezoar; T = long tail of the bezoar extracted from the proximal jejunum.
intussusception was observed. The patient recovered smoothly. During history taking, her parents revealed that until recently, she had pulled and eaten her own hair for 3 months. She was then discharged and followed up in the psychiatry department.

3. Discussion

Bezoars include trichobezoar (hair), phytobezoar (vegetable material), lactobezoar (milk products), pharmacobezoar (medicines, particularly with cellulose residues), and plastobezoar (plastic materials). Rapunzel syndrome is an unusual form of trichobezoar. It is defined as a trichobezoar with a tail-like extension from the stomach to the small bowel. Clinical presentation includes the presence of a mass in the abdomen, abdominal pain, nausea, vomiting, weakness, weight loss, constipation, diarrhea, and malnutrition. The bezoar itself can have a mass effect on the gastrointestinal tract, causing intestinal obstruction or gastric ulceration. The tail of the trichobezoar can interfere with the peristalsis of the intestine and lead to intussusceptions.

Most cases of Rapunzel syndrome are reported in teenage females with long hair. Surgical exploration remains the main treatment option for removing the trichobezoar. Enterotomy is advised for removing bezoars that extend into the intestine. After removal, the remaining bowel should be distally examined for any perforation or synchronous bezoar.

In our case, the patient experienced two complications of Rapunzel syndrome. One was multiple intussusceptions at an unusual location resulting from the tail of the trichobezoar, and the other was gastrointestinal obstruction caused by the mass effect of the body of the trichobezoar. We were unable to identify the main cause during the first operation because we did not consider the possibility of Rapunzel syndrome. Moreover, using laparoscopic instruments for palpating small bundles of hair within the bowel was difficult because the bowel had swollen after the reduction. In addition, during the first operation, the patient probably had a gastric bezoar. We could not diagnose the patient’s condition earlier through initial CT because of the difficulty in distinguishing between a gastric bezoar and food content retention. Before performing laparotomy, we planned to perform both gastrostomy and enterostomy. However, because both the gastric and duodenal bezoars had a strong connection, we could remove both bezoars only through gastrostomy.

In conclusion, we report a case of a 5-year-old girl with Rapunzel syndrome who had multiple intussusceptions and bowel obstruction caused by a trichobezoar with a long tail. Multiple intussusceptions at unusual locations are extremely rare. Surgeons should consider the possibility of Rapunzel syndrome when diagnosing the cause of intussusceptions.

References