Case report
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Rapunzel syndrome, a rare hairy tale: a case report

Nevine Macalintal[1], Shoshana Taube[2], Vinci Jones[3], Cornelia Muntean[1]

[1]Departments of Pediatric Emergency Medicine, [2]Emergency Medicine, and [3]Surgery, Good Samaritan University Hospital, West Islip, NY, USA

Abstract

Rapunzel syndrome is a rare condition that typically occurs in young females with trichotillomania or trichophagia. The name "Rapunzel" was given due to the presence of trichobezoars extending from the stomach down to various parts of the gastrointestinal tract. In this case, we discuss a 6-year-old female patient with a large trichobezoar that was further complicated by the occurrence of gastric perforation. This case emphasizes not only the presentation of a rare pathology with a rare complication but also the importance of meticulous history taking and physical examination for the approach to an acute abdominal emergency in the pediatric emergency setting.

Key words: Abdomen, Acute; Abdominal Pain; Bezoars; Intestinal Perforation; Trichotillomania

Introduction

Rapunzel syndrome is a rare condition that typically occurs in young females with underlying psychiatric disorders, and is a result of compulsively pulling out hair (i.e., trichotillomania) and subsequent swallowing it (trichophagia) (1-3). The syndrome was named due to the presence of trichobezoars, accumulated hair in the gastrointestinal tract, extending from the stomach down to the various parts of the gastrointestinal tract (2). This is a case of a 6-year-old female with a large trichobezoar that was further complicated by the occurrence of gastric perforation.

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Corresponding author
Nevine Macalintal
Department of Pediatric Emergency Medicine, Good Samaritan University Hospital, 1000 Montauk Highway West Islip, NY 11795, USA
Tel: +1-571-594-7224
E-mail: nevine.macalintal@gmail.com

Case

A 6-year-old female with no significant past medical or psychiatric history presented to the emergency department with sudden onset abdominal pain, nausea, and vomiting. Upon arrival, the initial vital signs were as follows: blood pressure, 103/57 mmHg; heart rate, 117 beats/minute; respiratory rate, 16 breaths/minute; temperature, 36.7 ˚C; and oxygen saturation, 97% on 2 L/minute nasal cannula. She appeared ill, lethargic, and falling asleep unless actively stimulated by tactile or verbal stimuli. Due to her acute distress noticed by a triage nurse, a pediatric rapid response was promptly activated. She was immediately taken to the resuscitation room for an urgent evaluation. A point-of-care glucose test showed 262 mg/dL. The head and neck examination was remarkable for the dry mucous membranes. She was found to have abdominal distension, absent bowel sounds, and generalized abdominal tenderness on palpation, as well as rebound tenderness with rigidity. These findings suggest the presence of an acute abdominal emergency requiring immediate surgical intervention.

With concerns for a possible new-onset diabetic ketoacidosis, the clinical team performed a venous blood gas, showing respiratory acidosis with pH of

7.29, bicarbonate of 22.6 mmol/L, and CO_2 of 47 mmHg. Complete blood count and metabolic panel were otherwise unremarkable. She was administered lactated ringers and piperacillin-tazobactam to manage her acute symptoms. A bedside chest radiograph showed free air under the diaphragm (Fig. 1), raising concerns about a perforated hollow viscus. Immediately, a computed tomography scan was done showing a large mass in the stomach and duodenum, along with the duodenum dilated with air-fluid levels (Fig. 2). There was a large amount of intraperitoneal free air, particularly in the region of the porta hepatis, indicating a duodenal perforation. Furthermore, we observed a dilation of the duodenal third portion and the proximal jejunum filled with material. These findings indicate the presence of a bezoar and a perforated hollow viscus.

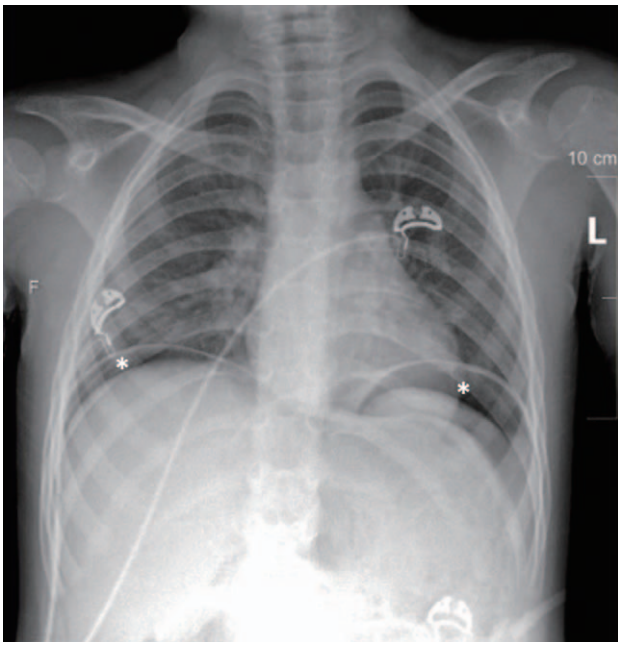

Fig. 1. Plain supine radiograph showing free air (asterisks) under the diaphragm.

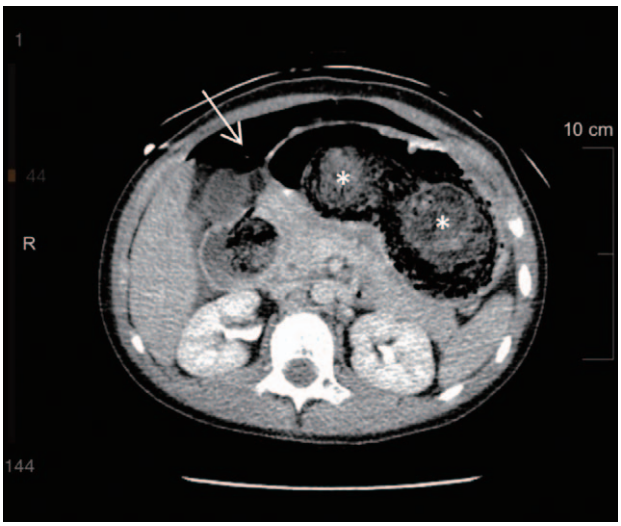

Fig. 2. Axial computed tomography scan showing a large amount of material in the stomach (asterisks) with a significant amount of free air detected in the peritoneum (arrow).

A pediatric surgeon immediately performed an exploratory laparotomy with foreign body extraction and repair of viscus perforation. Intraoperatively, the girl was found to have a gastro-duodeno-jejunal trichobezoar with gastric perforation (Fig. 3). On day 5, she was discharged uneventfully and fully recovered during surgical follow-up after the discharge. During the hospital stay, psychiatry and social evaluation found that she had used to consume the strands of hair of her and her teddy bear and had never sought healthcare regarding the issue. She was diagnosed with trichotillomania as well as anxiety, and prescribed fluoxetine.

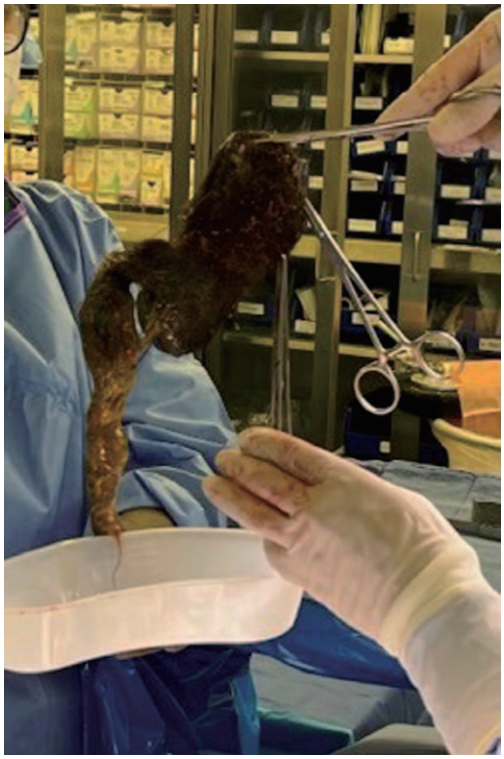

Fig. 3. Gastro-duodeno-jejunal trichobezoar retrieved intraoperatively.

Discussion

Bezoars are indigestible foreign materials found in the gastrointestinal tract, most commonly in the stomach (3). They can be classified into trichobezoars (hair particles), phytobezoars (vegetable fibers), and lithobezoars (stones). The incidence is approximately 0.4%-1% of the general population (1). Trichobezoars are formed when the hair strands become retained in the gastric mucosal folds and entangle over time. This is due to the hair being too resistant to the digestive enzymes and being able to evade gastric peristalsis with its slippery nature (2). The dark color and shiny appearance of trichobezoars stem from the denaturation of the hair protein by the gastric acid and the coating with the gastric mucus, respectively (4).

Rapunzel syndrome is a rare form of trichobezoars that is defined as gastric trichobezoars with the tails extending down to the ileocecal junction, or any size of bezoars that cause an intestinal obstruction (4). It is estimated that about 1% of patients with trichophagia develop trichobezoars (3). Patients are typically asymptomatic for years, but as the trichobezoars increase in size, symptoms can develop, such as abdominal pain, nausea, vomiting, halitosis, early satiety, and weight loss. Complications include intestinal obstruction, gastrointestinal bleeding, pancreatitis, cholangitis, and perforation.

The management is focused on the removal of the mass, which can be achieved either by endoscopy, laparoscopy, or conventional laparotomy, depending on the consistency, size, and location of the bezoars (2). The endoscopic approach can be effective for phytobezoars, as they are usually smaller in size and this method aids in the diagnosis and differentiation of different intragastric bezoars (2,4). However, in cases of Rapunzel syndrome, the trichobezoars are less likely to be removed by endoscopy due to their size and extension. Surgery is also indicated when the bezoars cause perforation or hemorrhage (3).

Conventional laparotomy has been proven superior to the laparoscopic approach, particularly in a case of perforation, like in this case. Recurrence of bezoars has been reported after the initial removal. Given the association of the bezoars with psychiatric comorbidities, a psychiatric assessment and long-term follow-up is advised to prevent the recurrence (3).

This case underscores the importance of a thorough history and physical examination, as pediatric patients can also present with gastric perforation secondary to trichobezoar.

ORCIDs

Nevine Macalintal
(https://orcid.org/0009-0002-6356-6207)
Shoshana Taube
(https://orcid.org/0000-0001-7487-258X)
Vinci Jones
(https://orcid.org/0000-0003-0732-7615)
Cornelia Muntean
(https://orcid.org/0009-0003-9292-0815)

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