Case Report

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Rapunzel Syndrome in A 13-Year-Old Girl with Abdominal Pain Presenting to The Pediatric Emergency Department: A Case Report

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ABSTRACT

Trichobezoar is a rare form of bezoar characterized by the accumulation of swallowed hair in the stomach. An unusual form of bezoar, in which the bezoar spreads from the stomach to the small intestine or beyond, is called Rapunzel syndrome. This syndrome is mainly seen in females and is often associated with psychiatric disorders. Trichobezoar usually presents with abdominal pain, early satiety, and nausea. However, it may also present as an asymptomatic abdominal mass or obstruction and perforation of the gastrointestinal tract. This diagnosis should be considered in young women with abdominal pain, an epigastric mass, and malnutrition, especially in patients with a history of trichophagy or psychiatric disorder.

The case of a 13-year-old girl with Rapunzel syndrome is described. She presented to the emergency ward with a history of abdominal pain, non-bilious vomiting after meal and fluid intake, halitosis, decreased appetite, and weight loss. The patient was treated successfully by laparotomy.

Introduction

The term “bezoar” is derived from the Persian word badzehr, meaning antidote. Some types of bezoar can precipitate or bind to arsenic compounds (commonly used in toxin compounds), so these act as antidotes [1]. Bezoars are defined as masses of indigestible material that accumulate and harden in the stomach and intestines after ingestion. Bezoars are classified by content. For example, persistent consumption of plastics (plastobezoar), antacid drugs (pharmobezoar), vegetables and fruit-based ingredients (phytobezoar), milk and dairy products (lactobezoar), and hair (Trichobezoar) are called [2]. Phytobezoars are usually the most known form of bezoars and account for approximately 40% of all cases [3].

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Trichobezoar is composed of hair, undigested fat, and mucus. The source of hair may be from the patient, other humans, animals, carpets or blankets [1]. Hair protein is denatured by gastric acid, which causes the ball to be blackened; fat is trapped and fermented in the hair fibers, causing a foul odor [1].

Most cases of Trichobezoar have been reported in women between the ages of 13 to 20 years [4]. In young women, Trichobezoar is associated with psychiatric disorders such as trichotillomania and pica trichophagy [5, 6].

Trichobezoars usually form in the body of the stomach. However, some trichobezoars may spread through the pylorus to the duodenum, jejunum, ileum, and even the colon, called Rapunzel syndrome. The name of this syndrome is derived from the legend of the German princess with long hair [7]. Rapunzel syndrome may be asymptomatic for a long time and is associated with varying degrees of obstruction due to the gradual enlargement of the bezoar. The most common symptoms include abdominal pain, nausea and vomiting, intestinal obstruction, and peritonitis. Other less common symptoms include weight loss, anorexia, hematemesis, protein-losing enteropathy, intussusception, iron deficiency, gastric ulcer, megaloblastic anemia, obstructive jaundice, acute pancreatitis, and gastric emphysema; it has been reported following large bezoars [8].

Small gastric bezoars can be removed by less invasive procedures such as endoscopy, but large trichobezoars, often non-biodegradable, can only be removed by open surgical intervention [9].

Case presentation

A 13-year-old girl was referred to the pediatric emergency unit with a history of abdominal pain, non-bilious vomiting after meals and fluid intake, halitosis, decreased appetite, and weight loss. Treatment in various outpatient clinics was pursued under the assumption of gastroenteritis. The patient had no past medical history or drug history.

Upon physical examination, stable vital signs were observed, and the general health state was good. The patient's height was 135 cm (< 3 %), and weight was 31 kg (<3 %). During the abdominal examination, a solid, mobile mass with crepitation was palpable in the epigastric region. The mass was firm in consistency and was not tender.

Laboratory evaluations revealed a WBC count of 9800 / mL (Neut:70%, Lymph:22%, Mono:5%, Eos:3%), hemoglobin of 14.6 g/dL, platelets of 320,000/mL, and CRP 0.05mg/L.

An abdominal computed tomography (CT) scan showed a large heterogeneous intraluminal mass with a mottled gas pattern in the stomach (Fig.1) and proximal Jejunojejunal and ileoileal intussusception (Fig.2) in favor of Trichobezoar.

Fig. 1. Abdominal computed tomography.

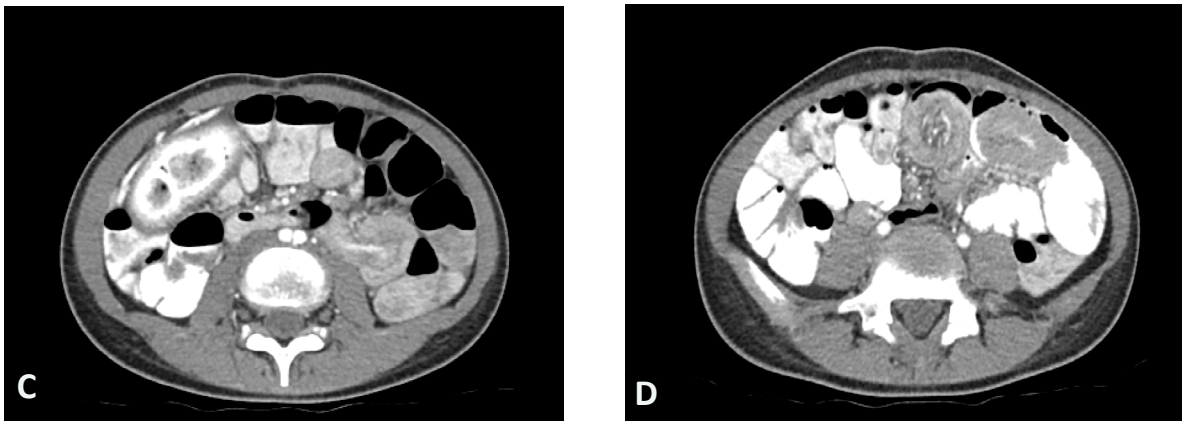

Fig. 2. Abdominal computed tomography.

Fig. 3. Intraoperative image.

Treatment

After the correction of fluid and electrolyte deficit, the patient was referred to surgery. Under general anesthesia administered by a pediatric surgeon, a supraumbilical midline laparotomy and anterior gastrotomy were performed. Upon exploration, a huge trichobezoar that had taken the shape of the stomach was identified (Figure 3).

Following the removal of the stomach-shaped trichobezoar, gastrography was performed with continuous Vicryl (Figure 4). The patient was discharged after five days. The parents were advised to schedule a follow-up visit with pediatric psychiatry.

Discussion

Gastric Trichobezoar is caused by continuous ingestion of hair. Due to the indigestion of hair in the stomach, a mass of hair forms that often fills the stomach and sometimes extends to the duodenum and beyond, a condition known as Rapunzel syndrome [10]. Rapunzel syndrome was first described by Vaughan ED et al. [11]. This name is derived from a legend written by the Grimm brothers in 1812 about a young prince who was imprisoned in a castle and had his long hair hung from a window [12].

However, most patients with Trichobezoar have psychiatric disorders, including trichotillomania

Fig. 4. Trichobezoar after the extraction.

and trichophagy. Trichobezoar occurs in only 1% of patients with trichophagia. More than 95% of cases of Trichobezoar occur in females [13]. The age of onset ranges from 1 year two months to 22 years, with 68.9% of cases in the age group of 5 to 15 years [12]. The prevalence of trichobezoar increases between 7-8 and 11-12.5 years [14]. Predisposing factors include psychiatric problems, including underlying depression, anxiety, obsessive-compulsive disorder, emotional factors such as stress, troubled family environment, loss of mother or father, or mental retardation [2]. The patient also lost his mother about a year ago.

Patients rarely remain asymptomatic for several years. Symptoms begin when the size of the bezoar increases to the point of obstruction. Patients with gastric Trichobezoar usually present with nonspecific symptoms, including abdominal pain [70], nausea and vomiting (64%), gastrointestinal bleeding (61%), epigastric discomfort, early satiety dyspepsia, weight loss (38%), diarrhea or constipation (32%). (5) Complications of a major bezoar include obstructive jaundice, severe anemia due to malabsorption or gastrointestinal bleeding, acute pancreatitis, and gastric emphysema, a rare complication that can increase mortality by up to 30%. [15] Treatment varies depending on the size and location of the bezoar. Management includes the complete elimination of bezoar and the prevention of recurrence. Due to the risk of perforation, only small bezoars can be removed with an endoscope. Data from case reports showed that out of 40 cases in which attempts were made to remove endoscopically, only two (5%) were successful. [16] For large bezoars extending to the duodenum, gastrotomy is the first-line treatment due to low complications and low recurrence, as well as the possibility of assessing satellite lesions. [17, 18] In the patient, endoscopic extraction was challenging and associated with potential risks due to the large size of the bezoar, so the removal was performed surgically.

Conclusion

This diagnosis should be considered in children with a long history of gastrointestinal problems, including abdominal pain, epigastric mass, halitosis, and malnutrition, especially in patients with a history of trichophagy or psychiatric disorders. All patients with Trichobezoar should be referred for psychiatric evaluation after treatment to prevent recurrence.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

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