

Rapunzel syndrome - a case report

Case Report

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Abstract: Bezoars are collections of indigestible materials found in the gastrointestinal tract. Rapunzel syndrome is a rare complication of a gastric trichobezoar in which the mass of hair extends through the pylorus into the small bowel and can even reach the colon. A 12-year-old girl with severe pain and a feeling of "fullness" in the upper abdomen was admitted to Pediatric Surgery. Two days before admission, the patient presented with vague abdominal pain, vomiting with a little blood, and black stool. On physical examination she was pale and listless with patchy alopecia. Abdominal examination showed a mobile, palpable mass in the epigastrium. The initial hematocrit was 7,1%, and the erythrocyte count was $1,12 \times 10^{12}$ cells per liter. Abdominal ultrasound and computed tomographic (CT) examinations confirmed the presence of a tumor-like formation in stomach. A giant bezoar with ulceration on the back wall of the stomach was verified by upper gastrointestinal endoscopy. Endoscopic extraction was not possible because of the size of the bezoar. Gastrotomy was performed to remove the bezoar that occupied most of the stomach, with a long tail that extended deeply into the duodenum (Rapunzel syndrome). The bezoar, 35 by 10 cm in diameter, was black and consisted of hair and synthetic fibers. Prompt diagnosis and treatment of trichobezoars are important to avoid a possible fatal outcome.

Keywords: *Trichobezoar • Rapunzel syndrome • Laparotomy*

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1. Introduction

Bezoars are an agglomeration of indigestible organic or inorganic foreign materials found in the gastrointestinal tract [1]. Bezoars are classified according to the content of the bezoar, such as trichobezoars (composed of hairs and synthetic fibers), phytobezoars (vegetable fibers, seeds, and skin of vegetables), trichophytobezoar (a combination of both), lactobezoars (milk products), lithobezoars (concretion of mud and stone), and pharmacobezoars (medication impaction) [2]. Trichobezoars are the most common form of bezoars (55% of all bezoars) [3,4].

Rapunzel syndrome is a very rare complication of a stomach trichobezoar, in which the mass of hair extends through the pylorus into the small bowel and can even reach the colon [1]. This syndrome was originally described by Vaughan et al. in 1968. It is named after a fairy tale written in 1812 by the Brothers Grimm [5]. In the story, Rapunzel is a long-haired girl imprisoned at the top of the tower whom a prince attempts to rescue by making a ladder of her hair [1].

2. Case Report

A 12-year-old girl with severe pain and a feeling of "fullness" in the upper abdomen was admitted to Pediatric Surgery. Two days before admission, the patient presented with vague abdominal pain, 3 to 4 episodes of vomiting with a little blood, and black stool. Mild abdominal pain, irregular bowel habits, a little blood in the vomitus, and loss of weight and appetite had been noted during the previous 6 months. Her mother claimed that she refused contact with other children, and she did not attend school regularly.

On physical examination, the patient was thin, very pale, uncooperative, and listless, with an axillary temperature of $37,8^{\circ}\text{C}$, blood pressure 97/46 mm Hg, pulse 150 beats per minute, and respiratory rate 100 per minute. Her weight was 30 kg (3rd percentile), height 144 cm (18th percentile), and body mass index 14,5 kg per square meter (3rd percentile). Her skin and visible mucosa were pale; there was no evidence of icterus. Examination of her head and neck revealed patchy alopecia; visible generalised lymphadenopathy was

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Figure 1. Extraction of a trichobezoar through gastrotomy.

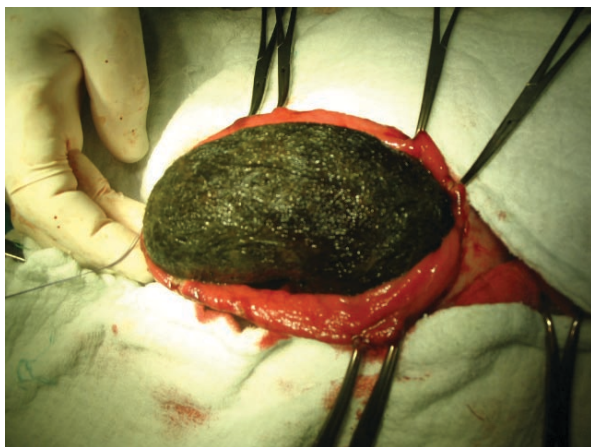


Figure 2. Ulcerous change on the back wall of the stomach caused by a trichobezoar.

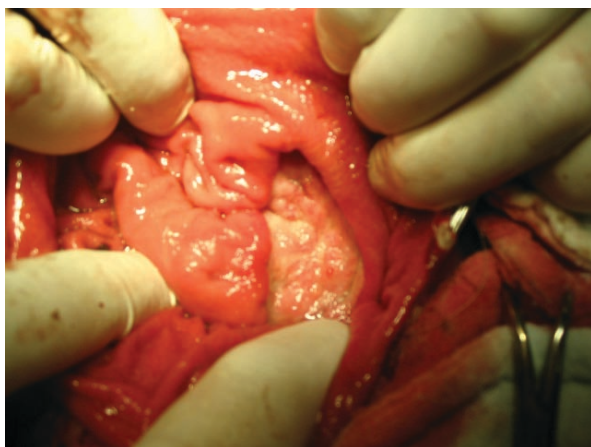


Figure 3. Specimen of extracted gastric trichobezoar with a long tail.



not present, and the supraclavicular lymph nodes were not enlarged. Examination of the chest and heart was normal. The patient had a leg prosthesis because of

the congenital absence of her left foot and distal lower leg. The abdomen was soft and nontender, with a firm, mobile, palpable mass in the epigastrium.

Laboratory test results showed a hemoglobin concentration of 2,5 g per deciliter; the hematocrit was 7,1%, erythrocyte count $1,12 \times 10^{12}$ cells per liter, leukocyte count $6,43 \times 10^9$ cells per liter, and platelet count 579×10^9 cells per liter. Other laboratory tests showed a serum protein of 45 g per liter and serum albumin of 22 g per liter. There was no electrolyte disturbance, and results of liver function tests and levels of serum amylase and lipase were normal.

Abdominal radiographs demonstrated a mass of opaque soft tissue in a swollen stomach. The bowel-gas pattern was normal. A CT scan of the abdomen revealed a large solid mass in a distended stomach. A pediatric endoscopist was consulted and confirmed the presence of a tumor-like formation in the stomach. It was vertically mobile, with a smooth surface, and occupied most of the stomach. An ulceration seen in the upper portion of the posterior surface of the body of the stomach had a smooth, regular, rounded edge, with a flat, smooth base. No evidence of bleeding from the ulceration was found.

Endoscopic extraction was not possible because of the size and hardness of the bezoar, and the patient was referred for surgical evaluation.

After preoperative preparation, gastrotomy was performed to remove the bezoar. At operation, an intraluminal mass was found to occupy most of the stomach, with a tail-like formation that extended deeply in duodenum (Rapunzel syndrome) (Figure 1). An ulceration, 3 by 3 cm in diameter, was found on the posterior surface of the body of the stomach (Figure 2). Pathological examination revealed a stomach ulcer. The bezoar was 35 by 10 cm in diameter, black, and J-shaped and consisted of hair and synthetic fibers (Figure 3). The length of the tail extension was 20 cm.

The postoperative course was uncomplicated. On retrospective questioning, the girl denied ingestion of hair or synthetic fibers. The parents were advised to consult pediatric psychiatry and her school psychologist for follow-up. It was recommended that social services be involved to help the family manage the child's psychiatric disorder.

3. Discussion

Rapunzel syndrome is a rare form of trichobezoar [5]. Up to 2008, 28 cases had been reported in the literature, with variable clinical features [6]. We report Rapunzel syndrome in a 12-year-old girl, with a tail of hair that extended deeply into the duodenum. All the cases

reported in the literature involve females, except one, which was that of a 6-year-old male who ate the fallen hair of his older sister [7].

Trichobezoars are commonly found in girls with underlying psychiatric disorders. They may occur as a result of habitual hair plucking (trichotillomania) and long-standing ingestion of hair (trichophagia) [8]. Only 50% of patients have a history of trichophagia, but most of them deny it, even when specifically asked [4]. Besides hair, the trichobezoars may consist of other undigestible fibers (nylon, wool, and bristle) [8]. In this case, the patient denied ingestion of hair, although there were alopecia plaques and the bezoar was found to contain synthetic fibers and human hair.

Trichobezoars grow slowly over many years until they form a mass in the shape of the stomach as in our case. The acidic contents of the stomach denature the hair protein resulting in a black colour. Decomposition and fermentation of fats give it a putrid smell. Mucus covering the bezoar provides a glistening shiny surface that prevents propulsion by peristalsis through the duodenum [5]. The bezoar becomes entrapped within the mucosal folds, where it then becomes enmeshed [9].

Patients are usually asymptomatic until trichobezoars reach a critical size [2]. The clinical picture includes abdominal pain, nausea, vomiting, early satiety, and weight loss. In cases of the Rapunzel syndrome, a tail in the intestines can trigger peristaltic movements, resulting in colicky abdominal pain [2]. Naik et al. in their review presented the most common features of Rapunzel syndrome: abdominal pain (37%); nausea and vomiting (33,3%); obstruction (25,9%); and peritonitis (18,3%). Patients have also presented with weight loss (7,4%), anorexia, hematemesis, and intussusception (7,4%) [5].

More severe complications of Rapunzel syndrome are gastrointestinal ulceration; gastrointestinal bleeding; intestinal, pancreatic, or biliary obstruction; perforation; and peritonitis. Jaundice and acute pancreatitis due to obstruction of the ampulla of Vater have also been reported [10]. Recurrent trichobezoars are extremely rare [8]. In our case, it was assumed that there had been occult bleeding from the stomach ulceration during the few months before hospitalization.

Diagnostic modalities include ultrasound study, CT scanning, and upper endoscopy [9]. On conventional radiography, a gastric bezoar is seen as a mass of opaque soft tissue in a swollen stomach. Ultrasonography of the trichobezoars reveals the high echogenicity of hair and the presence of multiple acoustic interfaces created by trapped air and food. CT reveals a well-defined intraluminal mass, with interspersed gas and localization of the bowel obstruction, and it has a high accuracy rate in patients with bezoars [3].

The differential diagnosis of a patient presenting with an epigastric mass includes a tumour of the left lobe of the liver, splenic enlargement due to lymphoma (e.g., Burkitt's or non-Hodgkin's lymphoma), a neuroblastoma, and a rare carcinoma of the stomach [11].

The treatment of large bezoars is laparotomy, with gastrotomy or enterotomy. If bezoars are small, they may be removed endoscopically [12]. Exploration of the rest of the stomach and the small intestine is recommended to look for retained bezoars [13].

In conclusion, the diagnosis of a trichobezoar should be considered in young females in association with emotional disorders, epigastric pain, a palpable, mobile abdominal mass, or alopecia. Prompt diagnosis and treatment are important to avoid a possible fatal outcome [9].

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